abbvie

Galapagos Biotech Ltd

An Alfasigma company







Irish Society for Rheumatology HOTEL TILLE TILLE **leenn**e April 2024 Κt







Welcome Message from the ISR President Dr John Ryan



Dear Guests, Colleagues and Friends,

A very warm welcome to the ISR Spring Meeting, here at The Grand Hotel in Malahide, County Dublin. We are thrilled to have you all here for what promises to be an enriching and informative event.

Our agenda for the next two days is packed with insightful sessions, with a particular focus on maternal health. Prof. McAuliffe will open the meeting with the obstetrics perspective on Rheumatologic diseases, with presentations from Dr Sinead Maguire on pregnancy in AxSpA, Dr Gorman on her experience in combined Rheumatology/Obstetrics clinics and the development of guidelines a national implementation of such services. Prof. Ni Ainle will discuss obstetric APS.

We'll also delve into fascinating areas with our international speakers- including the advancements in psoriatic arthritis management delivered by Prof. Behrens, and the application of artificial intelligence in healthcare by Prof. Loh. Dr Belviso who has travelled from medicine to mindfulness will tell us how to incorporate mindfulness into our daily routines in medicine.

Amidst our engaging sessions, I encourage you to take advantage of networking opportunities and visit our industry partners during coffee breaks.

On Thursday, we'll come together for a reception and dinner, offering a chance to unwind and connect with fellow attendees in a relaxed atmosphere.

As we embark on this journey of learning and collaboration, I extend my deepest gratitude to our speakers, sponsors, our local organisers- Prof. Ronan Mullan and Dr Shawn Chavrimootoo, and of course Michael Dineen and colleagues for making this event possible.

I want to express my gratitude to Prof. Geraldine McCarthy for her exceptional leadership of the ISR. Her unwavering optimism, even amidst the toughest days of the COVID pandemic when the prospect of in-person meetings seemed bleak, was truly commendable. While the days of quarantining feel distant now, it was always a pleasure to attend an ISR board meeting with Geraldine leading the way!

Thank you all for being here, and I wish you a rewarding and enjoyable experience ahead.

Dr John Ryan President ISR



ISR Spring Meeting 11-12 April 2024

Programme

Thursday, 11 April

- 12.45-13.45: Light lunch
- 13.55-14.00: Opening Address: Dr John Ryan, ISR President
- 14.00-14.45: **Obstetric care for women with Rheumatological disease Professor Fionnuala McAuliffe,** Consultant Obstetrician and Gynaecologist, UCD/National Maternity Hospital, Dublin.
- 14.45-15.30: **Pregnancy in Axial Spondyloarthritis Dr Sinead Maguire,** Toronto Western Hospital, Canada.
- 15.30-16.00: Coffee Break and Visit Industry
- 16.00-16.45: From Medicine to Mindfulness Dr Ceara Belviso, Rheumatologist and Mindful Living Coach
- 16.45-17.45: **Advancements in Psoriatic Arthritis Management Professor Frank Behrens**, Geothe-University & Fraunhofer ITMP Frankfurt am Main, Germany – UCB sponsored.
- 19.30: Reception/Dinner

Friday, 12 April

- 09.00-10.00: Clinical case presentations
- 10.00-10.45: Artificial Intelligence in medicine Professor Erwin Loh, President Elect, Royal Australasian College of Medical Administrators; University of Melbourne/Monash University/ Macquarie University
- 10.45-11.15: Coffee Break and Visit Industry
- 11.15-12.00 National Rheumatology Obstetrics Service (ROSE) Dr Áine Gorman, Consultant Rheumatologist, Midland Hospital Group, Tullamore, Co Offaly.
- 12.00-12.45: **Obstetric antiphospholipid syndrome Professor Fionnuala Ní Áinle,** Consultant Haematologist, Mater Hospital and Rotunda Hospital, Dublin.
- 12.45: **Prize-giving and close of meeting followed by lunch**



Biographical Sketches

Speakers

Professor Frank Behrens

Geothe-University & Fraunhofer ITMP Frankfurt am Main, Germany

Frank Behrens first studied biology at the Johannes Gutenberg University in Mainz and later medicine. He graduated



from the Johann Wolfgang Goethe University Frankfurt am Main, specializing in internal medicine and rheumatology. Following a Fellowship at the Pharmazentrum Frankfurt (Immunopharmacology) researching the regulation of T-cells, Dr Behrens returned to the field of Rheumatology. He has been Director of the Rheumatology Clinic of Goethe University since 2007 where he founded the interdisciplinary Centre for drug research, development, and security (ZAFES). In 2011, Dr Behrens founded the Centre for innovative diagnosis and therapy in rheumatology/immunology (CIRI) in partnership with Goethe University.Dr Behrens is a founding member of various international committees and working groups, such as GRAPPA (Group of research and assessment of psoriasis and psoriatic arthritis), EPOSS (Expert Panel on outcome of systemic sclerosis), PAGE (psoriatic arthritis genetic European Consortium), etc. He is involved in clinical trials as a home inspector, investigator and senior examiner for Germany since 2001. He is also responsible for one of the largest investigator-led studies (IIT) in rheumatology in Germany and set-up of the German study network NESTRA.

Dr Ceara Belviso

Rheumatologist and Mindful Living Coach

Dr Ceara Belviso is a wife and mum living in Dublin. Graduating from UCD in 1999, she spent 13 years in clinical

practice, followed by 11 years in the corporate world of pharmaceutical medicine.

During her time in practice, Ceara witnessed the growing prevalence of stress, burnout and chronic illnesses on society. After starting her family and struggling with some personal health issues, Ceara experienced first-hand the effects that chronic low-grade stress can take on your health. At this time she discovered mindfulness and selfcompassion, eventually undertaking teacher training in mindfulness and positive psychology.

In 2023, Ceara stepped back from medical practice to focus on sharing the transformative power of mindful living with others. Through bespoke offerings and her membership 'CAILM', she works with diverse audiences, including healthcare professionals, corporate professionals, and individuals seeking greater balance and fulfilment in their lives.

Ceara firmly believes that true health comes from a baseline of inner peace, which encompasses not only physical health but also mental and emotional wellbeing, and she is passionate to share the benefits of mindfulness with the purpose of creating a healthier and happier world. **A** work resulted in a PhD from Trinity College Dublin focused

Dr Áine Gorman

Consultant Rheumatologist, Midland Hospital Group, Tullamore, Co Offaly

Dr Áine Gorman has a degree in Medicine from NUI Galway. She completed her

rheumatology training in Ireland in 2021. She developed an interest in pregnancy in patients with rheumatic disease while working as a research fellow at St Vincent's University Hospital. Dr Gorman subsequently worked as a clinical research fellow at the Royal National Hospital for Rheumatic Diseases (RNHRD) in Bath. She also holds a master's degree in clinical education. Dr Gorman works as a locum consultant rheumatologist at Midlands Regional Hospital Tullamore.

Professor Erwin Loh

President Elect, Royal Australasian College of Medical Administrators; University of Melbourne/Monash University/Macquarie University



Professor Erwin Loh is President Elect of

the Royal Australasian College of Medical Administrators and Chief Medical Officer at Goulburn Valley Health. Prior to that, he was national Chief Medical Officer and Group General Manager Clinical Governance for St Vincent's Health Australia, the nation's largest not-for-profit health and aged care provider.

He is Professor at Monash University, where he leads the Clinical Leadership and Management Unit at the Monash Centre for Health Research and Implementation. He is Honorary Clinical Professor with the title of Professor at the Department of Medical Education, University of Melbourne. He is Honorary Professor at Macquarie University at the Centre for Health Systems and Safety Research.

He has been an invited speaker at local and international conferences, and is a member of the Association of Professional Futurists, with an interest in medical futurology. He is co-editor of the textbook "Artificial Intelligence in Medicine - Applications, Limitations and Future Directions", and has published chapters and journal articles on the use of artificial intelligence in health.

He received the Distinguished Fellow Award from RACMA in 2017 for "commitment to governance, research and publication".

Dr Sinead Maguire

Toronto Western Hospital, Canada

Sinead is currently working as a Clinical and Research Fellow in the Spondylitis Program at Toronto Western Hospital in Canada. She attended medical school



at the Royal College of Surgeons in Ireland graduating in 2012, and then completed her higher specialist training in General Internal Medicine and Rheumatology. During her HST she undertook a period of research based out of St James Hospital working with Professor Barry O'Shea and the Ankylosing Spondylitis Registry of Ireland (ASRI). This





on women's health in axial spondyloarthritis and led to the development of her interest in pregnancy in women with axSpA. Her work has been featured in peer reviewed Rheumatology journals, podcasts, patient advocacy groups, national and international meetings. She is very excited to be returning to Ireland to join the Rheumatology Department at Our Lady's Hospital Navan.

Professor Fionnuala McAuliffe

Consultant Obstetrician and Gynaecologist, UCD/National Maternity Hospital, Dublin



Fionnuala McAuliffe is a Full Academic Professor of Obstetrics and Gynaecology

at National Maternity Hospital Dublin, Head of Women's and Child's Health at University College Dublin. She is Director of the UCD Perinatal Research Centre, a multidisciplinary research centre aiming to improve outcome for mother and baby though clinically relevant pregnancy research. She is a maternal and fetal medicine specialist and Director of the MFM training programme at National Maternity Hospital and lead clinician for maternal medicine services.

She has received over euro 50 million as CoPi/PI. has over 400 peer reviewed publications including national and international pregnancy care guidelines. She is Past Chair of International Federation of Gynaecology and Obstetrics (FIGO) committee on Impact of Pregnancy on Long- term Health and is a FIGO executive council member.

Professor Fionnuala Ní Áinle

Consultant Haematologist, Mater Hospital and Rotunda Hospital, Dublin



Professor Ní Áinle is a Consultant Haematologist at the Mater Misericordiae

University Hospital (MMUH) and Rotunda Maternity Hospital, Dublin, Ireland. She is a Full Clinical Professor at University College Dublin (UCD) School of Medicine. She is privileged to treat patients affected with Venous Thromboembolism (VTE) and has a strong interest in women's issues in thrombosis and haemostasis. Prof. Ní Áinle serves as Director of the Irish Network for VTE Research and co-directs the SPHERE Research Group, UCD Conway Institute.

Prof. Ní Áinle was elected to the Council of the International Society on Thrombosis and Haemostasis (ISTH) in 2022 (Class of 2028) and acts as vice-chair of the ISTH Guidelines Committee. Prof. Ní Ainle was recently appointed National Lead for VTE by the Irish Health Services Executive (Term commencing in Q4 2022).

ISR Board members

Dr John Ryan

President Consultant Rheumatologist, Cork University Hospital, Cork

Dr John Ryan is a graduate of the Royal College of Surgeons in Ireland, he completed his higher medical training in



rheumatology and general internal medicine in Ireland. He undertook a fellowship at the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS) in Bethesda, Maryland. During this time he undertook translational research into disordered innate immunity manifesting as recurrent fever syndromes. He joined Dr Sinead Harney in the Rheumatology service at Cork University Hospital in 2010. The Rheumatology department has since expanded to include Dr Grainne Murphy. In July 2017 he took up the post of National Specialty Director for Rheumatology.

Dr Claire Sheehy

Honorary Secretary Consultant Rheumatologist University Hospital Waterford



Dr Claire Sheehy is a Consultant Rheumatologist in University Hospital Waterford. A graduate of Trinity College

Dublin, she completed the higher specialist training in rheumatology and general medicine, and was awarded an MD for work exploring the role of anti TNF therapy in early rheumatoid arthritis. She undertook a fellowship in connective tissue disease and vasculitis between Norfolk and Norwich University Hospital, and Addenbrookes Hospital. She took up her post in 2012; her current clinical interests include early inflammatory arthritis and connective tissue disease.

Dr Shawn Chavrimootoo

Honorary Treasurer Consultant Rheumatologist, Our Lady's Hospital, Navan, Co Meath.



Shawn Chavrimootoo is a Consultant Rheumatologist at Our Lady's Hospital,

Navan, Co. Meath. He graduated in Medicine from RCSI, Dublin in 2002 and developed an interest in Rheumatology during his Senior House Officer years in Connolly Hospital, Blanchardstown. Following this, he completed higher specialist training in Cork University Hospital, Kerry General Hospital, Connolly Hospital and St Vincent's University Hospital in Dublin. He was appointed to his Consultant Rheumatologist post in 2013 when he joined Dr Ramakrishnan at Our Lady's Hospital, Navan, from where they currently provide a regional Rheumatology service for the North East of Ireland. His clinical interests include osteoporosis as well as gout, inflammatory arthritis, spondyloarthritis, connective tissue disease and vasculitis.



Spring Meeting 2<u>024</u>

Dr Nicola Ambrose

Consultant Rheumatologist, Blackrock Clinic, Co Dublin

Dr Nicola Ambrose is a graduate of Trinity College Dublin. She completed her specialist training in rheumatology and general internal medicine in Ireland,



before obtaining an Arthritis Research UK (ARUK) fellowship to undertake a PhD at Imperial College London, studying inflammation in Behçet's Syndrome. She then obtained a Richard Steeven Fellowship from the HSE to undertake a Clinical Fellowship at the ARUK Adolescent Rheumatology Centre of Excellence at University College London Hospital (UCLH). She stayed at UCLH as an Adolescent and Adult consultant rheumatologist, and was the Clinical Lead for Adolescent Rheumatology. Special interests: Adolescent and Young Adult Rheumatology including JIA; Behçet's Syndrome; SLE and dermatomyositis; Gout Osteoporosis and fracture secondary prevention; Inflammatory arthritis. She has published 23 peer review papers as well as 6 book chapters.

Dr Elizabeth Ball

Consultant Rheumatologist Musgrave Park Hospital/ Belfast City Hospital



Dr Liz Ball is a graduate of Queen's University Belfast and was appointed as a Consultant Rheumatologist at Musgrave

Park Hospital/ Belfast City Hospital in 2014. She is also an Honorary Lecturer at Queen's University Belfast. She has a special interest in autoimmune disease and lupus and was awarded an MD entitled 'A study of hand arthritis in Systemic Lupus Erythematosus from a Clinical, Imaging and Cytokine Perspective' from Queen's University in 2013. She is involved in postgraduate medical education and holds a Training Programme Director role within the Northern Ireland Deanery and is currently completing a Masters in Clinical Education. She is a musculoskeletal ultrasound tutor and regularly teaches regionally and nationally.

Dr Andrew Cairns

Consultant Rheumatologist, Musgrave Park Hospital, Belfast

Dr Andrew Cairns graduated in Medicine from Queen's University Belfast in 1995. He completed specialist training in Belfast and also at the Rheumatic Diseases Unit



Dr Michele Doran

Consultant Rheumatologist and General Physician, St James's Hospital Dublin

Dr. Michele Doran has been working as a Consultant Rheumatologist and General Physician at St. James's Hospital, Dublin since 2003. She graduated in Medicine from UCD in 1993, and completed her



clinical training in General medicine and Rheumatology in Dublin and Bath, UK. She undertook a 2 year Research Fellowship at Mayo Clinic, Rochester, USA, where she completed an MD degree with research relating to the Epidemiology of Rheumatoid Arthritis. During this time she completed a Master's Degree in Biomedical Sciences, Clinical Research, in the Mayo Clinic Graduate School. She was involved with the establishment of and is on the steering committee for the Rheumatoid Arthritis Biologics Registry of Ireland (RABRI).

Professor Ursula Fearon

Head of Molecular Rheumatology, School of Medicine, Trinity Biomedical Sciences Institute, Trinity College Dublin.



Professor Ursula Fearon is head of Molecular Rheumatology, School of Modicine Trinity Piomodical Sciences I

Medicine, Trinity Biomedical Sciences Institute, Trinity College Dublin. Professor Fearon's research is a bench-tobeside translational approach, focusing on understanding underlying mechanisms that the drive disease pathogenesis; her team specifically examine components of joint inflammation at a cellular and molecular level to dissect out the signalling and gene pathways that are involved in the pathogenesis of inflammatory arthritis and rheumatic diseases. She has established strong collaborative research networks across Europe, USA and Singapore. Professor Fearon, has been awarded significant research funding from Arthritis-Ireland, Health Research Board, Science Foundation Ireland, IRCSET, European-ASPIRE, JU Innovative Medicines Initiative (IMI) and Maeve Binchy Funding for Arthritis Research, in addition to industry collaborative partnerships. She has published extensively in high impact peer-reviewed journals, and her research has been awarded several National/International awards.

Dr Natasha Jordan

Consultant Adult and Adolescent Rheumatologist St James's Hospital and Children's Health Ireland



Dr Natasha Jordan is a Consultant Adult and Adolescent Rheumatologist, currently

based at St James's Hospital and Children's Health Ireland, Crumlin. Her major clinical and research interest is SLE. Dr Jordan studied Medicine at University College Dublin

followed by Higher Specialist Training in General Internal Medicine and Rheumatology. She subsequently worked for 5 years at the Louise Coote Lupus Unit at St Thomas' Hospital in London, where she was the recipient of both an Arthritis Research UK Fellowship and a Graham Hughes Clinical Research Fellowship. She obtained her PhD from King's College London investigating the genetics of



lupus nephritis. She then took up a substantive post as a Consultant Rheumatologist for 7 years at Addenbrooke's Hospital in Cambridge. While in Cambridge, she was Deputy Director of the Rheumatology Clinical Research Unit and ran phase II and III trials in SLE. In 2018, she led the bid for Addenbrooke's to become a Lupus UK Centre of Excellence, recognising the unit's commitment to providing high quality care for patients with SLE.

Dr Jordan's interest in pregnancy management in SLE began during her time at St Thomas' and continued while in Cambridge where she worked collaboratively with the departments of Nephology and Obstetrics in providing combined care clinics for pregnant patients with complex autoimmune rheumatologic conditions.

Professor David Kane

National Lead for Rheumatology HSE Clinical Programme Consultant Rheumatologist, Tallaght University Hospital, Dublin



Prof. David Kane attended medical school at Trinity College, Dublin, Ireland and was conferred MB BCh BAO BA in 1991, PhD

in 2002 and FRCPI in 2006. He has trained in rheumatology with Prof. Barry Bresnihan and Prof. Oliver FitzGerald at St. Vincent's University Hospital, Dublin, Ireland and with Prof. Roger Sturrock, Prof. lain McInnes and Dr Peter Balint at Glasgow Royal Infirmary, Glasgow, United Kingdom. He was appointed as Senior Lecturer in Rheumatology at the University of Newcastle (2003-2005) and is currently working as Consultant Rheumatologist at the Adelaide and Meath Hospital and Clinical Professor in Rheumatology at Trinity College Dublin. His special interests are musculoskeletal spondyloarthopathy and ultrasound, synovial inflammation. He is a member of the European Working Party on Musculoskeletal Ultrasound and the OMERACT special interest group on musculoskeletal ultrasound, previous organiser of the BSR Musculoskeletal Ultrasound course and is Faculty member of the EULAR Musculoskeletal ultrasound course. He has served as a Board member of the Irish Osteoporosis Society, as President and Treasurer of the Irish Society for Rheumatology and is currently a Board member of Arthritis Ireland.

Dr Emma Jane MacDermott

Consultant Paediatric Rheumatologist, CHI Crumlin

Emma Jane MacDermott, is a Consultant Paediatric Rheumatologist in CHI Crumlin where she joined the team in 2012 and has helped oversee the ongoing growth and development of the paediatric



rheumatology department into a dynamic national service, now including a growing research and education component. With a special interest in education she enjoys working with patients, parents and medical providers to raise the profile and understanding of rheumatologic disease. She works with the national advocacy groups continuing to raise the profile for Irish paediatric rheumatology patients. Areas of interest include Juvenile arthritis, Paediatric Lupus and autoinflammatory disease. A graduate of University of Dublin, Trinity College Medical School she pursued her post graduate training in paediatrics, becoming a member of the Royal College of Physicians in 2001. She subsequently moved to New York, where she completed a fellowship in Paediatric Rheumatology, from Weill Cornell Medical School, working at Hospital for Special Surgery and the Cornell Campus of New York Presbyterian Hospital as Assistant Attending in Paediatric Rheumatology at Hospital for Special Surgery and Assistant Professor of Paediatrics at Weill Cornell Medical School until her return to Ireland in 2012.

Emma is a member of the Royal College of Physicians of Ireland, the American College of Rheumatology, the Irish Rheumatology Society, the British Society of Adolescent and Pediatric rheumatology.

Professor Geraldine McCarthy

Consultant Rheumatologist Mater Misericordiae University Hospital Dublin and Full Clinical Professor of Medicine University College Dublin



Geraldine McCarthy graduated in Medicine from NUI. She received her

Fellowship in Rheumatology at the Medical College of Wisconsin. Her research has focused on the biological effects of calcium-containing crystals in degenerative joint disease as well as in atherosclerosis and breast cancer. Promoted to Associate Professor of Medicine at the Medical College of Wisconsin in 1996 where she remained until her return to Dublin.

Prof. McCarthy was appointed Consultant in Rheumatology at the MMUH and Cappagh National Orthopedic Hospital Dublin in 1999 where she continues to run a busy clinical practice. She teaches as part of the University College Dublin Faculty of Medicine where she was the first clinician to be appointed Full Clinical Professor of Medicine through the Clinical Pathways in 2009.

Geraldine has current international collaborations in the UK, USA, Europe, Australia, New Zealand and Canada, particularly in relation to calcium crystal deposition diseases as well as gout. She continues her involvement in bench research related to the pathogenesis of basic calcium phosphate crystal-induced joint disease and participates in and contributes to numerous international collaborations related to gout. Other research interests include platelet activation in inflammatory arthritis and its role in enhanced cardiovascular risk. She also participates in collaborative studies of the pathogenesis of giant cell arteritis and HIVassociated bone pathology.

Author of over 130 publications, including original manuscripts, editorials, reviews and book chapters and has spoken at many national and international meetings. She has been winner of several research and teaching awards and has mentored many medicine and science graduates in clinical practice and in research.



Dr Wan Lin Ng

SpR Rep on ISR Board

Dr Wan Lin Ng is a medical graduate from the Royal College of Surgeons in Ireland (RCSI). She has completed her basic specialist training in Ireland and is currently in the higher specialist training

programme in Rheumatology. She is a recipient of the StAR MD scholarship from RCSI and the ISR Rheumatology Patient Initiative Fund. With a keen passion in teaching and education, Dr Ng was previously an affiliate tutor with University of Limerick and RCSI. Her clinical interests include connective tissue disease related interstitial lung disease and musculoskeletal ultrasound.

Irish Society

for Rheumatology

Professor Barry O'Shea

Consultant Rheumatologist, St James's Hospital, Dublin

Barry O'Shea is a Consultant Rheumatologist in St James's Hospital, and a Clinical Associate Professor in the School of Medicine in Trinity College Dublin.



He took up his position in St James's in 2010. During his specialist training in Rheumatology he worked in St Vincent's Hospital, Waterford Regional Hospital, St James's Hospital and the Mater Hospital. He was the inaugural recipient of the Irish Society for Rheumatology / Wyeth Travelling Fellowship award. This facilitated the completion of his training in the University of Toronto and Toronto Western Hospital, Canada. He went on to undertake a Research Fellowship in Toronto with Dr Robert Inman with a focus on patients with Ankylosing Spondylitis and Psoriatic Arthritis. He has presented at the American College of Rheumatology Annual Meeting on this work. He is an active member of ASAS (Assessment of SpondyloArthritis international Society), an international group of experts in the field of Ankylosing Spondylitis. He is a co-founder and principal investigator of ASRI – the Ankylosing Spondylitis Registry of Ireland, a national database of patients with AS from across Ireland. He is the Clinical Lead for Rheumatology in St James's Hospital. In 2022 he was appointed the National Speciality Director (NSD) for Rheumatology training in the Royal College of Physicians of Ireland.

Dr Bryan Whelan

Consultant Rheumatologist Our Lady's Hospital, Manorhmailton, Co Leitrim

Dr Bryan Whelan is a Consultant Rheumatologist in Our Lady's Hospital in Manorhmailton, Co Leitrim and an



Honourary Senior Lecturer in Medicine in NUIG. He qualified from UCD in 2000 and completed BST in the Mater Hospital in Dublin. He completed SpR training in Rheumatology in CUH, the Mater Hospital and University College London. He has an MD and Masters Sports and Exercise Medicine from UCC and an MSc in Epidemiology from the London School of Hygiene and Tropical Medicine. He is currently a board member of Arthritis Ireland, the SUH Research and Education Foundation, a member of the Academic Committee of the FSEM and a member of the Advisory Committee for Human Medicines Clinical Trials Subcommittee of the HPRA. His current research interests include muscle disease, exercise in rheumatology and osteoarthritis.

Dr Maria Wray

Consultant Rheumatologist Antrim Hospital, Northern Ireland.

Dr Maria Wray is a consultant rheumatologist in Antrim Hospital, Northern Ireland. She graduated from Queens University Belfast in 2000 and



began rheumatology training in Northern Ireland. She then undertook long term specialty doctor roles in rheumatology firstly in Musgrave Park Hospital and then the South East Health Trust where she developed particular expertise in musculoskeletal and vascular ultrasound. She was awarded specialist registration in rheumatology and joined the team in Antrim hospital in the Northern Trust as a consultant in 2018. Her specialist interests include PMR/GCA and "fast track" diagnostic imaging.





Spring Meeting 2024







Spring Meeting 2024







ISR Meeting Spring, 2024 Sponsors

The Irish Society for Rheumatology wishes to express its gratitude to all its sponsors and in particular to the following 'Major Sponsors'

AbbVie Ltd Galapagos Biotech Ltd an Alfasigma company Novartis Ireland Ltd Pfizer Healthcare Ireland UCB (Pharma) Ireland Ltd

Accord Healthcare Ltd Amgen Ireland AstraZeneca Pharmaceuticals (Ireland) DAC Athena Pharmaceuticals Ltd Biogen Idec Ireland Ltd Bristol-Myers Squibb Pharmaceuticals Celltrion Healthcare Ireland Ltd Clonmel Healthcare Eli Lilly & Company (Ireland) Limited Fannin Ltd Fresenius-Kabi Janssen Sciences Ireland UC MSD Ireland Ltd Nordic Pharma Ireland

The Pharmas listed above have all supported this meeting through a payment to exhibit a stand. They have had no involvement in any other aspect of this meeting.



Spring Meeting 2024

ISR Presidents

Dr John Ryan 2023 -Cork

Prof. Geraldine McCarthy 2020 - 2023 Dublin

> Dr Sinéad Harney 2018 - 2020 Cork

Dr Sandy Fraser 2016 - 2018 Limerick

Prof. D. Kane 2014 - 2016 Dublin

Dr G.Wright 2012 - 2014 Belfast

Pro.f Gaye Cunnane 2010 - 2012 Dublin

Dr R. Kavanagh 2008 - 2010 Galway

> **Dr J. Lee** 2006 - 2008 Craigavon

Dr P. O'Connell 2004 - 2006 Dublin

Prof. O. FitzGerald 2002 - 2004 Dublin

Dr A. Taggart 2000 - 2002 Belfast

Dr D. Raman 1998 - 2000 Sligo

Dr A. Bell 1996 - 1998 Belfast

Prof. B. Bresnihan 1994 - 1996 Dublin

Prof. M. Molloy 1992 - 1994 Dublin

Dr E. Casey 1990 - 1992 Dublin

Dr S. Roberts 1988 - 1990 Belfast

Dr C. Barry 1985 - 1987 Dublin

Dr D. Roden 1983 - 1985 Dublin

Dr W. Boyd 1981 - 1983 Belfast

Dr T. Gregg 1979 - 1981 Dublin

Dr J. Molony 1977 - 1979 Dublin

Dr M .McMahon 1975 - 1977 Cork Irish Society for Rheumatology Board Members

PRESIDENT

Dr John Ryan Consultant Rheumatologist, Cork University Hospital, Cork

HONORARY SECRETARY Dr Claire Sheehy Consultant Rheumatologist University Hospital Waterford

HONORARY TREASURER

Dr Shawn Chavrimootoo Consultant Rheumatologist, Our Lady's Hospital, Navan, Co Meath.

BOARD MEMBERS

Dr Nicola Ambrose Consultant Rheumatologist Blackrock Clinic, Dublin

Dr Elizabeth Ball Consultant Rheumatologist Musgrave Park Hospital/ Belfast City Hospital

> **Dr Andrew Cairns** Consultant Rheumatologist Musgrave Park Hospital, Belfast

Dr Michele Doran Consultant Rheumatologist and General Physician St. James's Hospital, Dublin

> Professor Ursula Fearon Head of Molecular Rheumatology, School of Medicine, Trinity Biomedical Sciences Institute, Trinity College Dublin.

Dr Natasha Jordan Consultant Adult and Adolescent Rheumatologist St James's Hospital and Children's Health Ireland

> Professor David Kane National Lead for Rheumatology HSE Clinical Programme Consultant Rheumatologist, Tallaght University Hospital, Dublin

Dr Emma MacDermott Consultant Paediatric Rheumatologist, CHI Crumlin

Professor Geraldine McCarthy Consultant Rheumatologist Mater Misericordiae University Hospital Dublin and Full Clinical Professor of Medicine University College Dublin

> **Dr Wan Lin Ng** SpR Beaumont Hospital Dublin

Professor Barry O'Shea Consultant Rheumatologist, St James's Hospital, Dublin.

Dr Bryan Whelan Consultant Rheumatologist Our Lady's Hospital, Manorhmailton, Co Leitrim

Dr T.O'Reilly 1973 - 1975 Dublin

12

Dr Maria Wray Consultant Rheumatologist Antrim Hospital, Northern Ireland



Message from Michael Dineen



Dear Friends

This year for our Spring Meeting we head for beautiful Malahide in the hope of getting an early summer. We have been giving some serious thought to our Spring Meeting since the pandemic. It was always a one-day meeting which limited the areas which we could select venues from. It also removed the opportunity for any worthwhile networking which is so valued by many of our ISR membership. The two half days with an evening dinner seem to tick most of the boxes for the majority. I feel that it provides adequate time for learning, acquiring new technology and networking.

A vote of thanks to Shawn and Ronan in Navan for putting together the academic programme for this meeting. It is never an easy task to please everybody when putting an agenda together for any meeting, so well done to all concerned.

When talking about meetings I must make a brief reference to last year's Autumn Meeting in September at Killiney. This was certainly the most memorable and nostalgic meeting that I have ever been involved with. The meeting presentations and speakers were outstanding with a hugely responsive attendance. The dinner and Lifetime Achievement Award took pride of place. A brief report on Linkedin is still attracting positive comment. It has presently received in the region of 4500 hits. Our continued best wishes to Doug, Ursula and family.

Great to see the Bernard Connor Medal regaining some of its former glory. I hope that this important award will go from strength to strength. We would ask that you encourage your trainees to participate in this educational process which honours a great scientist.

For the past two years ISR has benefited from employment of Dr Mythri Shaji as a Clinical Research Associate in the Biologics field. She will be leaving us at the end of May to join her husband in Canada, having recently married. We wish them both well in their lives together. We will indeed miss her.

I hope you all will have a very enjoyable and educational meeting.

Michael Dineen Chief Executive ISR



ISR Bernard Connor Medal

The Irish Society for Rheumatology (ISR) has established the Bernard Connor Medal to promote interest in the field of rheumatology among BST trainees in medicine, and to support engagement with the Irish Society for Rheumatology.

The award is open to BST trainees in medicine who *fulfil the eligibility criteria below*. In addition to receiving the Connor Medal, the winner will be invited to attend the annual scientific meeting of the ISR to present their work to the membership, as a guest of the society. *Additionally, and*



at the discretion of the judging panel, up to two runners-up may be awarded full registration to attend the ISR annual scientific meeting.

Bernard Connor

The Connor Medal is named in honour of Bernard Connor, an Irish physician who observed and described the characteristic skeletal and clinical features of ankylosing spondylitis in 1693, while a medical student in Paris. This award will be made annually on the basis of competitive submission.

Theme

The theme chosen by the ISR is a **Case Report with relevance to rheumatology**. The submitted work should highlight your observations/insight in relation to rheumatology as a BST trainee.

Submission Only one submission per trainee will be accepted.

Case Report

The case report should be submitted in full form and present the details of an interesting case followed by a discussion on your observations of the key points of interest, and relevant literature review. These should be submitted in full [**max 800 words**, concluding with summary key message and references (up to 5 in total)].



Bernard Connor Medal contd...

Eligibility

- Applicants must be registered RCPI BST trainees by 2 July 2024.
- Applicants must submit completed entries to the ISR by the 12 August 2024.
- It is hoped that the re-launch of this competition in its new format will not only honor a great clinician but also draw the specialty of Rheumatology to the attention of BST trainees. The ultimate aim is to attract more trainees to HST in Rheumatology.

How to Apply

Application forms for the Connor Medal will be available for download from the ISR website www.isr.ie. These must be completed in full and returned together with your submission to info@isr.ie

Closing Date: 12 August 2024.

Judging Criteria

The Medal will be awarded according to the criteria below which will be applied to all submissions

- Relevance of the submitted work to rheumatology
- Originality and Merit of the work

The Board of the Irish Society for Rheumatology shall convene the judging panel. The decision of the panel shall be final and no correspondence on submissions or final outcome can be entered into.





CLINICAL PRESENTATIONS

Friday morning, 12 April

Abstract No.	Time
24S110	09.00
24S116	09.15
24S122	09.30
245124	09.45

CLINICAL CASES

24S101

Beyond Lipid-Lowering

Author(s)

Eva McCabe, Abdelrahman Elsheikh, Carl Orr

Department(s)/Institutions

Rheumatology Department, St Vincent's University Hospital

Introduction

Anti-HMG Co-enzyme reductase necrotising myositis is a rare subtype of immune-mediated myositis. Increased availability of antibody testing has been helpful to confirm suspected diagnoses in an era where statin prescribing is highly prevalent.

Aims/Background

A 62-year-old man presented to the Rheumatology clinic with a 10-week history of progressive lower limb weakness, fatiguability and lethargy. He had a notably high CK > 16,000 and a marginally elevated CRP 9. He was admitted for further workup. Initial serology was negative for ANA, ENA and ANCA. Complements were normal. His troponin was 400, though ECGs and echocardiogram were normal. MRI of the thighs confirmed florid myositis. Rectus femoris muscle biopsy identified profoundly abnormal tissue with areas of necrotisation, though there was not a significant inflammatory infiltrate. His EMG was consistent with a severe active necrotising myopathy process affecting proximal and distal muscles in the upper and lower limbs. An extended myositis panel confirmed low level positivity for anti-Mi2 beta antibodies. Anti-HMG Co-enzyme A reductase antibodies subsequently returned positive at a high titre: 363 units (ULN 20 units).

The antibody, biopsy and EMG results favoured an underlying diagnosis of immune-mediated necrotising myositis. By way of background, this gentleman had been treated for a STEMI 18 months prior to this presentation and had been commenced on Atorvastatin 80mg, which was felt to be the precipitant.

To screen for secondary causes of myositis, a CT TAP was also performed. This identified a 3cm heterogeneous mass in the left adrenal gland, later determined to be functioning, secreting adrenaline metabolites. He underwent adrenalectomy with histology confirming a diagnosis of phaeochromocytoma.

High dose prednisolone 60mg and mycophenolate mofetil 500mg bd was initiated on the day of muscle biopsy. MMF was uptitrated to 1g bd one week later. Unfortunately, his myopathy progressed and he required IVIG and prolonged rehabilitation to stabilise his disease.

Conclusions

Learning points

- HMGCR-associated IIM is a rare but debilitating complication of statin use
- It is often diagnostically and therapeutically challenging with persistent disease despite cessation of the drug
- IIM may be precipitated by more than one cause

24S104

Macrotroponin as a cause of a falsely elevated cardiac troponin in Systemic Sclerosis

Author(s)

Sherdya W Tio (1), Caroline M Joyce (2), David Halsall (3), John Ryan (1)

Department(s)/Institutions

1. Cork University Hospital, Rheumatology, Cork, Ireland 2. Cork University Hospital, Biochemistry, Cork, Ireland 3. Cambridge University Hospitals Trust, Addenbrookes Hospital, Clinical Biochemistry, Cambridge, United Kingdom

Introduction

The prevalence of cardiac involvement in systemic sclerosis (SSc) is high but the finding of a raised troponin level is concerning, especially in younger patients.

Aims/Background

A persistently raised cardiac troponin in the absence of diagnostic clinical evidence should raise the suspicion of a false positive result. Macrotroponin is a high molecular weight complex of immunoglobulin and troponin. It is not biologically active but it's presence interferes with measurement of cardiac troponin, causing falsely elevated result.

Method

We present the case of a 37-year old Caucasian male who had a recent diagnosis of anti-Scl70 positive antibody SSc with persistently elevated cardiac troponin-I (cTnI) 500-800ng/L (normal<34ng/L) and cardiac troponin-T (cTnT) 169ng/L (normal <5ng/L). He had progressively worsening skin tightness, Raynaud's phenomenon and markedly reduced exercise tolerance despite previous high performance status. Thus, the elevated cTnI raised concern for cardiac involvement.

Results

Resonance Imaging were all normal.

Given the discordance of clinical and laboratory results, the presence of an analytical inference was suspected. Biochemistry laboratory investigations included dilution studies which were non-linear and Polyethelene glycol (PEG) precipitation which yielded maximum troponin recovery of 3%, suggesting the presence of interferent.

Gel filtration chromatography (GFC) confirmed the presence of approximately 65% macrotroponin in the patient's sample (Graph 1).

Conclusions

This case highlights the importance of interpreting cardiac troponin results with caution when they don't fit the clinical picture and communicating with laboratory experts to initiate further investigations.

The exact prevalence of macrotroponin is still unknown but this case intends to raise awareness of the possibility of a macrotroponin when unexpected raised troponin results are obtained.

Graph 1 Gel Filtration Chromatography





24S105

Challenging case of Eosinophilic Fasciitis (Shulman's disease)

Author(s)

Sumreen Sarfaraz, John Ryan

Department(s)/Institutions

Rheumatology Department, Cork University Hospital, Wilton Co. Cork

Introduction

Eosinophilic fasciitis is rare disease characterized by subacute onset of erythema, edema, induration of the skin and soft tissues of the limbs and trunk followed by fascial fibrosis, leading to a cobblestone appearance or peau'd orange.

Aims/Background

We are presenting a case of 61 year old lady who was attending rheumatology outpatient following recent hospitalization with lower respiratory tract infection and polyarthralgia. She had medical history of Hypothyroidism on Eltroxin, Eczema, Rectocele repair, Long QT syndrome and Vaginal prolapse. Her initial joints symptoms included swelling and tenderness affecting wrists, MCPs and PIP. She received a taper course of steroids following treatment of her chest infection. Her arthritis recurred on weaning steroids, leflunomide and subsequently anti TNF was added. She reported a new onset rash and was found to have diffuse thickening and nodular appearance of skin of forearm1. Biopsy showed subcutaneous and fascial inflammatory process with epicenter in deeper facial tissue extending into contiguous epimysium, perimysium and inflammation of adjacent muscle. Components of inflammatory process include lymphocytes, plasma cell with infrequent eosinophils. MRI of wrists also were consistent with an eventual diagnosis of Eosinophilic Fasciitis. She was treated with 3 courses of intravenous immunoglobulins with minimal improvement. The patient was reluctant to start methotrexate but did so with encouragement . She continued to feel tightened skin despite the addition of JAK inhibitor. She required ongoing steroids (prednisolone 10mg), in combination with methotrexate and intravenous infliximab. Mepolizumab was commenced and the effect of treatment is awaited.

Method

Case Report

Results

Currently there is lack of RCT for treatment of EF. Steroids are considered as first line treatment followed by addition of methotrexate. Other agents include IVIG, cyclosporine, rituximab and infliximab. Refractory cases have been reported responding to interleukin 5 blockade which has been commenced

Conclusions

In conclusion, steroids are considered as first line of treatment with 75% response rate. Refractory or relapsed cases requires additional therapy. The presence of peau d'orange and skin disease affecting the trunk is associated with an incomplete response to therapy.



24S106

A case report of myelodysplastic syndrome and VEXAS.

Author(s)

Connerton Á, Murphy E, Duffy T, Murphy CL.

Department(s)/Institutions

Rheumatology Department, Connolly Hospital Blanchardstown.

Introduction

A 65-year-old gentleman presented with lethargy, weight loss, dyspnoea, cough and fatigue. His background history was significant for newly diagnosed myelodysplastic syndrome (MDS), exsmoker, and chronic renal insufficiency. He underwent extensive investigations including endoscopy, CT Thorax Abdomen Pelvis, ECHO, bronchoscopy, and autoimmune serology which were all normal. On discharge, his symptoms were thought to be explained by MDS.

Aims/Background

His condition progressed and he reported worsening dyspnoea and skin lesions. He had recurrent episodes of painful subcutaneous erythematous nodules involving his lower legs, trunk, and face, usually lasting one week. CT thorax was repeated, showing multifocal ground-glass opacities as well as bibasal consolidations and pleural effusions. See Fig.1. Skin biopsy revealed neutrophilic inflammatory infiltrate with eosinophils and light leucocytes, consistent with Sweet's syndrome. His rash responded to short courses of steroids.

7 months later he presented to the hospital with periorbital oedema, dyspnoea (modified Medical Research Council dyspnoea scale 1), migratory arthralgia, and an acute kidney injury. Our concern at this point was an evolving vasculitis or an autoinflammatory disorder. His symptoms again rapidly resolved with a short course of steroids.

Method

He was followed regularly in Rheumatology clinic and the main issues were recurrence of subcutaneous nodules, ear chondritis, periorbital oedema, dyspnoea, and worsening renal function. His original diagnosis was reviewed and given the predominant chondritis involvement he was sent for genetic tests to the UK looking for an underlying UBA1 abnormality or cryopyrin-associated periodic syndromes (CAPS). The results confirmed a diagnosis of vacuoles, E1 enzyme, X-linked, autoinflammatory, somatic (VEXAS) syndrome which can be seen in combination with MDS.

Results

Like this case, rashes that are typically seen in Sweet's syndrome can be part of VEXAS. CT Thorax abnormalities are present in 91% of men at diagnosis. Pulmonary manifestations are non-specific but can include ground-glass opacities, along with mediastinal adenopathy.

Conclusions

VEXAS is a recently recognized adult-onset autoinflammatory syndrome primarily affecting men. This case highlights the importance of considering this diagnosis in patients seen in rheumatology clinic, particularly if they have an underlying haematological disorder.



17



Spring Meeting 2024







24S107

A Traumatic Consequence of Deep Sea Diving

Author(s)

Rebecca O'Farrell, Sara Treacy, Paul O'Grady, Carmel Silke.

Department(s)/Institutions

Department of Rheumatology, Our Lady's Hospital Manorhamilton, Department of Orthopaedics, Mayo University Hospital.

Introduction

We describe the case of a subacute un-displaced left neck of femur stress fracture in a 47 year old gentlemen with a significant history of occupational deep sea diving in the Navy.

Aims/Background

Femoral neck stress fractures are a relatively rare injury and diagnosis highly depends on imaging techniques. These injuries most often effect military recruits and endurance athletes. It is well established that radiographs are incapable of diagnosing this injury in the early stages. This condition is often under recognised by clinicians.

Method

Our patient described the insidious onset of atraumatic left hip pain whilst running, relieved by rest. His last sea dive was 8 years ago and he experienced no prior pain in his left hip. Initial plain film radiographs of both hips were normal. Subsequent Magnetic resonance imaging demonstrated a subacute un-displaced femoral fracture with associated oedema. This patient had no additional risk factors for osteoporosis or medical comorbidities. The patient's history was significant for prolonged exposure to hyperbaric environment. Our patient spent cumulatively 18 months deep sea diving at a depth of 150 m over a 6 year period. Each exposure was a period of 25 days spent in a small hyperbaric chamber resting in between periods of deep sea diving, followed by 5 consecutive days in a decompression chamber. His last hyperbaric exposure was 8 years prior to this diagnosis. He ran approximately 20km per week and cycled 100km weekly. Given his diving history, we hypothesise he experienced a stress fracture following a combination of prolonged decompression periods and immobility. This gentlemen may have suffered subclinical or latent osteonecrosis leading to compromised bone health whilst engaging in moderate running.

Results

This patient was treated surgically with a dynamic hip screw with an uncomplicated perioperative course.

Conclusions

This case represents an easily missed clinical example of a femoral stress fracture after prolonged exposure to a hyperbaric environment. Additionally this case highlights the importance of early recognition and initiation of treatment. If left untreated patients may progress to a complete displaced fracture. Lifestyle advice should be given to those individuals with occupational hyperbaric exposure.





24S108

A Case of Unusual Presentation of Behcet-like Paraneoplastic Syndrome

Author(s)

H. Tan, S. McConkey, G. Wright and C. Donaghy

Department(s)/Institutions

Rheumatology Department, Musgrave Park Hospital

Introduction

Paraneoplastic syndrome is a rare disorder which occurs when immune system has reaction to cancerous tumour. [1] We present an unusual case of paraneoplastic syndrome presented with Behcet's related symptoms.

Aims/Background

A 69-year-old man presented with 3 weeks history of erythema nodosum and pyrexia following a recent admission with orbital cellulitis. This was preceded by recent diagnosis of right epididymoorchitis and left calf DVT. He had widespread torso rash, panniculitis and thrombophlebitis on his upper limbs, which were swollen with pitting oedema. Blood test showed elevated ESR (124) and CRP (265). He was treated with broad spectrum antibiotics. Ultrasound Doppler upper limbs showed right subclavian and left cephalic vein thrombus. There was subtle hilar lymphadenopathy and marked pulmonary emphysema on CTPA. No malignancy on imaging. Autoimmune screen, tumour markers, and infection screen were negative. Pathergy test was negative. He was discharged with outpatient follow-up.

Unfortunately, he had relapse of swelling in all limbs, livedo reticularis, further weight loss and shortness of breath on follow-up review after few weeks. There was no history of recurrent mucosa or genital ulceration. Repeat imaging confirmed new bilaterally axillary vein thrombus, potential internal jugular vein thrombus and left subclavian artery thrombus. IgG4 and antiphospholipid screen are negative. Cardiac MRI showed non-ischaemic undilated cardiomyopathy possibly secondary to recent myocarditis. Due to persistent high inflammatory markers, multiple thrombosis and lack of improvement, he was commenced on 40mg Prednisolone. His anticoagulation was optimised. His inflammatory markers and symptoms all improved.

Method

A Case report

Results

CTPET on last admission had shown evidence of uptake query mucous plugging, which was treated with antibiotics. Chest X-ray showed right middle lobe collapse. Red flag bronchoscopy was performed due to these findings. Cytology on bronchial washing showed squamous cell carcinoma. He was referred to respiratory team for further management.

Spring Meeting 2024



Spring Meeting 2024





Spring Meeting 2024







Conclusions

This case highlights multisystem inflammatory response which mimics Behcet's disease. There are 95% of Behcet's patients have oral ulcers and 80-80% have genital ulcers. [2] Clinicians need to be aware that lack of hallmark features should prompt for further investigations. To date there is no report of similar paraneoplastic features related to squamous cell lung cancer.

24S109

A case of rapidly progressive interstitial pulmonary fibrosis refractory to treatment in the setting of Anti-MDA-5 Dermatomyositis (MDA5-DM)

Author(s)

Dr J Sin, Dr A Elliott, Dr G Wright

Department(s)/Institutions

Rheumatology Department, Belfast Health and Social Care Trust

Introduction

60 year old male admitted to the Intensive Care Unit (ICU) with progressive hypoxic failure secondary to rapidly progressing interstitial lung disease. The diagnosis of MDA5-DM was made from clinical history alone whilst antibody results were awaited in an emergency setting.

Aims/Background

A 60 year-old gentleman was admitted to hospital with hypoxic respiratory failure. He was diagnosed with mild-moderate interstitial lung two months prior, this was his second admission to hospital for lower respiratory tract symptoms. Despite antibiotic and oxygen therapy at ward level, he required escalation to the ICU for endotracheal intubation. A CT-scan of the chest revealed widespread progressive interstitial lung disease– peripheral, ill-defined interstitial opacification, and he had developed a pneumomediastinum post-intubation. Despite escalated antibiotic treatment and oxygen support, he deteriorated, requiring proning, IV methylprednisolone and concurrent nitric oxide to help with oxygenation.

The patient was referred to rheumatology as the ICU team had noticed bilateral ulcerated lesions across his metacarpophalangeal joints (MCPs). After a detailed collateral history and examination, this gentleman was found to have ulcerative lesions at the site of Gottron's papules and along the ears, joint synovitis at the knees and MCPS along with subungal erythema at the nail bed. These findings along with the history of pneumomediastinum and no definite history of muscle weakness or myopathy with a creatinine kinase was 41U/L-lead to a suspicion of amyopathic MDA5-DM. A myositis panel was sent off but treatment was started based on the clinical findings and the seriousness of his clinical condition.

Method

Case-report

Results

Despite treatment with further pulses of IV methylprednisolone and later prednisolone, plasma exchange, cyclophosphamide and tacrolimus; the patient's condition continued to deteriorate. The patient passed away due to worsening hypoxic respiratory failure before the myositis panel results returned- testing positive for Anti-MDA-5 antibodies.

Conclusions

MDA5-DM is a rare but serious systemic inflammatory condition which is clinically challenging to diagnose with a lack of myopathic features and a potentially life threatening course as seen in this case. Rapidly progressing interstitial lung disease and hypoxic respiratory failure is often associated with this sub-type of dermatomyositis and is associated with worse outcomes and high mortality rates.



24S110

Paraneoplastic Linear IGA bullous disease as a Behcet's mimic

Author(s)

Samreen Tariq1, Christina Grechin2, Muhammad Jehangir1, Malaz Ahmed1, Martina Carolan1, Catriona Stafford1, Omer Hussein1, Cliona Feighery2, Shawn Chavrimootoo1

Department(s)/Institutions

1)Rheumatology Department, Our Lady's Hospital, Navan 2) The Department of Dermatology, Our Lady of Lourdes Hospital, Drogheda.

Introduction

Behcet's disease (BD) is a relapsing multisystem vasculitis which commonly presents with recurrent oral and genital ulceration. The diagnosis is purely based on the mucocutaneous manifestations and the application of the International Criteria for Behcet's Disease (ICBD) may result in overdiagnosis of the disease. We report a rare case of a Linear IgA bullous Dermatosis (LABD), likely paraneoplastic in origin, as a mimicker of Behcet's disease (BD).

Aims/Background

Case report

Method

A 63 years old lady was seen in the Rheumatology clinic with 4 months history of recurrent oral and genital ulceration and new onset ocular pain and conjunctival redness. Over the last 1 month, she had developed ulcerating skin lesions in the axillae and over the eyelids. Poor response to colchicine prescribed by the GP.

Initial clinical examination confirmed two small oral ulcers and multiple round erythematous skin lesions with blistering borders in the axillae and on the eyelids. Laboratory workup revealed a very high ESR of 100 and a CRP of 86. She was clinically diagnosed with BD, meeting the ICBD criteria and treatment with tapering steroids and azathioprine (AZA) was started and the dermatology review was requested.



Spring Meeting 2024







Despite initial response to the steroids, the patient clinically worsened with increasing genital and skin lesions. AZA was switched to adalimumab for a short period. It was stopped when she developed constitutional symptoms of anorexia, weight loss and pelvic pain. An Ultrasound of the pelvis requested by the GP raised a suspicion of an ovarian mass.

The dermatologist performed a biopsy of one of the genital lesions, and immune-florescent studies revealed a linear deposition of IgA antigen at the basement membrane, thus directing the diagnosis towards LABD. This is a rare auto-immune blistering disease mostly idiopathic or drug induced but has been loosely linked to various malignancies.

Results

Our patient was subsequently diagnosed with high grade serous cell ovarian carcinoma. She underwent hysterectomy, oophorectomy and is currently receiving adjuvant chemotherapy with carboplatin/ paclitaxel. Her ocular disease remains active and responds to the rounds of chemotherapy, indicating the paraneoplastic aetiology of the LABD.

Conclusions

Behcet's can mimic various dermatological conditions. This case highlights the importance of confirming the diagnosis prior to initiating immunotherapy.





245111

Reactive arthritis following local intravesical Bacillus Calmette Guerin (iBCG) immunotherapy for high-grade papillary urothelial carcinoma.

Author(s)

Connolly E, Abdallah I, Mohamed H, Shinners E, Camon A, Gorman A, O'Rourke K, Mohammad A.

Department(s)/Institutions

Department of Rheumatology, Midland Regional Hospital, Tullamore

Introduction

Bladder cancer is the 9th most frequently diagnosed cancer, with the most common being superficial transitional cell carcinoma. Patients with high-grade non-invasive carcinoma are treated with transurethral resection of bladder tumour (TURBT) followed by local intravesical Bacillus Calmette Guerin (iBCG) immunotherapy. Reactive arthritis (ReA) is estimated to occur in 0.5-1% of cases of iBCG therapy.

Aims/Background

We examine a case of a 49-year-old male with a three-day history of polyarthralgia, painless red eyes and fevers on a background of high-grade papillary urothelial carcinoma. He was treated with TURBT followed by iBCG immunotherapy. He presented four weeks after his fourth iBCG. He had evidence of synovitis at his bilateral metacarpophalangeal joints, proximal interphalangeal joints and knees with bilateral conjunctival hyperaemia.

Method

Initial laboratory evaluation showed C reactive protein 390 mg/ dl(normal 0-5mg/dl), white blood count was $18.8 \ 10^{-9}\text{/L}$ (normal 3.8-9.5 10^{-9}/L) and neutrophilia of $16.12 \ 10^{-9}\text{/L}$ (normal 2-6 10^{-9}/L). His x-ray images of bilateral knees were normal.

Rheumatoid factor, Anti-cyclic citrullinated peptide and human leukocyte antigen B27, anti-nuclear antibodies and anti-neutrophil cytoplasmic antibodies, blood cultures, urine cultures and QuantiFERON were all negative.

He had multiple diagnostic joint aspirations taken of synovial fluid with no growth or crystals found. A sample was sent for GeneXpert assay, which did not detect mycobacterium tuberculosis complex.

Results

He was initially commenced on Vancomycin and Ceftriaxone for five days, which was then stopped. He was then commenced on intravenous methylprednisolone for three days and was de-escalated to prednisolone thirty milligrams daily. He was started on isoniazid, azithromycin and rifampicin. His stay was complicated by worsening liver function tests due to drug-induced liver injury (DILI). The DILI meant he was unable to be treated with a drug-modifying agent (DMARDS), and resulted in the cessation of rifampicin. He had minimal improvement, and treatment was escalated to Tocilizumab weekly. He responded well and was discharged with tapering prednisolone, Tocilizumab, and isoniazid.

Conclusions

ReA should be considered in patients with inflammatory arthropathy with a history of iBCG. First line treatments include discontinuation of iBCG therapy and non-steroidal anti-inflammatory drugs. Secondline treatments include steroids, anti-tubercular treatment and DMARDS, with Tocilizumab used only in refractory cases.

245112

Rice body tenosynovitis! Rare but exists !!

Author(s)

M Ahmed 1, S Tariq 1, S Chavrirmootoo 1, R Mullen 1, P Harrington 2

Department(s)/Institutions

Rheumatology department 1, Orthopaedics department 2, Our Lady's hospital Navan

Introduction

Rice-body tenosynovitis is rare and it's association with Rheumatoid arthritis,tuberculosis,infections or trauma is reported only in a few cases in the literature.In almost all cases the recovery is complete after synovectomy and treatment of the underlying cause if it is found



Aims/Background

Mr.BB 70 YO M farmer, referred from his GP with Left hand pain and swelling. On reviewing him he mentioned 5 years gradual swelling in the left wrist and little finger with intermittent pain and intermittent yellow fluid discharge. He cannot fully flex his little finger or his wrist. No trauma. No other joints symptoms. On examination there is a swelling on the ulnar side of his wrist and little finger, no tenderness. Investigations showed normal inflammatory markers, routine bloods, bone profile, CCP and Rheumatoid factor. XR of left hand and wrist showed OA changes and no erosions

Method

MRI of the Left hand and wrist(figure1)showed OA changes with full thickness cartilage defects and subchondral cystic lesion, and evidence of extensor tenosynovitis with rice-bodies suggestive of inflammatory arthritis. The MRI was discussed in our radiology conference with conclusion this is unlikely infection or tumor. The patient was contacted and a trial of oral steroids with clinic follow up was agreed. Unfortunately no response to steroids, and the swelling hadn't unchanged.Orthopaedics were contacted for an exploration of his tenosynovitis. Three months later he had tendon sheath exploration which showed complete replacement of the sheath by soft tissue growth with large amount of yellow mucinous material(figure 2).Specimens were sent for microbiology with negative result for TB and other infections.Histology showed fibroconnective tissue showing non-caseating granulomatous infiltration, with areas of cystic-spaces filled with mucous. The differential included: infection ,sarcoidosis or ruptured mucoid cyst

Results

His history didn't suggest any systemic symptoms of sarcoidosis as well as his CXR and his MRI hand and wrist. The diagnosis was concluded to be a ruptured mucoid cyst

We continued to see Mr.BB regularly until 1 year after his surgical intervention and he remained well with no recurrence

Conclusions

Tenosynovitis is not always inflammatory. History taking remains a key to the correct diagnosis and it's certainly advisable not to start immunosuppression treatment without solid evidence.

Multidisplinary approach is important to treat complex patients as our orthopaedics colleagues helped us narrow down the differential here



24S113

A case of rapid complete recovery of visual loss in GCA with intravenous tocilizumab

Author(s)

A Farouk, K Murray, J Devlin, A Fraser, A Sebastian

Department(s)/Institutions

Rheumatology Department, University Hospital Limerick.

Introduction

A major and dreaded complication of cranial giant cell arteritis (GCA) is well known to be anirreversible permanent visual loss (PVL) and infact an incidence of almost 30% has been reported.1 Only a small number of studies have demonstrated recovery of vision following high dose glucocorticoids and more recently, tocilizumab, a recombinant humanised anti-interleukin-6 receptor monoclonal antibody has been used with more success.2

Aims/Background

We report a case of a 69-year-old female, presented with sudden onset left-sided visual loss with no light perception occurred hours before presenting to hospital. She had no associated symptoms of headache, scalp tenderness or jaw claudication. She was admitted for transient ischaemic attack (TIA) assessment but was noted to have elevated inflammatory markers with a C-reactive Protein (CRP) of 27 mg/L and Erythrocyte Sedimentations Rate (ESR) of 65 mm/ hr. Her Ocular coherence tomography (OCT) confirmed left central retinal artery occlusion (CRAO). Southend GCA pre-test probability score (SGCAPS)3 was 17. Temporal and axillary arteries ultrasound (US) showed bilateral inflammatory changes (halo sign) of the vessel walls in the common temporal, frontal and parietal branches (figures 1 and 2) consistent with diagnosis of GCA.

Method

Case report

Results

She was initiated on intravenous (IV) methylprednisolone at presentation followed subsequently by IV tocilizumab infusions following US results. Significant clinical and serological improvement was noted with near complete resolution of vision loss within a few days (80%) and full resolution within 6 months as well as sustained GCA remission.

She had repeat ultrasound of the temporal arteries two months after initial presentation which confirmed improvement in her vessel wall thickening. Ophthalmology also reported full resolution of left visual defect.

She remains on IV monthly tocilizumab infusions with a view to change her to subcutaneous in the near future.

Conclusions

Tocilizumab is approved in the treatment of refractory GCA and has been shown to have an excellent efficacy in sustaining remission. Our case demonstrates not only the efficacy of tocilizumab in the treatment of GCA but in our case, IV administration had significant impact on rapid recovery of visual loss related to cranial GCA.







Figure 2

Temporal artery ultrasound. The non-compressible, hypoechoic 'halo sign' is seen in longitudinal of the (D) frontal branch of the left temporal artery and (E) parietal branch of the left temporal artery.

24S114

Scleroderma myositis overlap: 2 rheumatological cases are not better than one

Author(s)

Conall Mac Gearailt, Mohamed Eltahir, Gillian Fitzgerald, Bernadette Lynch

Department(s)/Institutions

Galway University Hospital

Introduction

Overlap conditions are common in Rheumatology. This clinical case discusses a complex presentation of Scleroderma myositis overlap complicated by Interstitial Lung Disease (ILD) as well as active Cytomegalovirus (CMV) infection. The delicate balance between immunosuppression and treatment of infection is addressed as is the relevance of cardiac markers in patients with minimal symptoms.

Aims/Background

A case report

Method

A 73 year old lady presented electively to Galway University Hospital for muscle biopsy following discussion between Respiratory and Rheumatology. She was initially managed for interstitial lung disease with mycophenolate mofetil as well as cytomegalovirus infection with valganciclovir. Incidental significantly raised creatine kinase was noted and the patient was subsequently referred to GUH. Her symptoms were of significant fatigue, dry eyes and skin tightening. Examination revealed abnormal cuticles, skin tightening to MCPs, facial telangiectasia and moderate fatiguability of large muscles.

Results

CK was 2000U/L and troponin was 1000ng/L. PM SCL 75/100 antibodies were noted. Thigh MRI showed extensive multifocal hyperintensity suggesting myositis. Biopsy revealed multifocal endomysial inflammatory infiltrates in keeping with myositis. Cardiac MRI showed global myocardial oedema with no fibrosis. She was treated with Methylprednisolone, Intravenous Immunoglobulin (IVIG) and Rituximab despite concerns regarding immunosuppression with good effect.

Conclusions

Scleroderma myositis overlap is uncommon. Myocardial involvement has an increased prevalence in Scleroderma myositis overlap compared to Scleroderma alone. The absence of overt cardiac symptoms does not preclude patients from having cardiac involvement. These patients must be closely monitored and treated promptly in order to ensure the best outcomes. This case demonstrates with complexity of some rheumatological conditions and the importance of multidisciplinary care.

Spring Meeting 2024





24S115

An Eye catching Rheumatological case

Author(s)

Conall Mac Gearailt, Gillian Fitzgerald, Bernadette Lynch

Department(s)/Institutions

Galway University Hospital

Introduction

IgG4-Related disease should be considered in cases of bilateral eye swelling. Normal IgG4 levels do not exclude this diagnosis. B cell depleting therapy is highly effective treatment option. IgG4related disease should also be considered in cases with widespread lymphadenopathy with evidence of malignancy. We describe a clinical case which involved a high amount of clinical uncertainty.

Aims/Background

A case report

Method

A 28 year old man previously well presented with bilateral eye swelling and feeling generally unwell. He subsequently developed daily fevers, night sweats and reduced appetite. He had no relevant medical history with no regular medications. On exam, he had proptosis of his left more than right eye. Normal eye movements. Palpable lymphadenopathy in cervical chain. No organomegaly.

Results

His bloods revealed White Cells 5.4x10^9/L, Haemoglobin 12.7g/L, Eosinophils 1.35x10^9/L, CRP 76, U+E Normal, ALP 174U/L, ALT 19U/L, GGT 119U/L. ANA, Anti CCP, Rheumatoid factor normal. p ANCA was positive but PR3 and MPO were both negative. IgG was mildly elevated at 18.01g/L. IgG4 level was 0.58g/L (0-1.29). Normal CXR. CT TAP showed multiple bilateral pulmonary nodules and peribronchial soft tissue in the right upper lobe and middle lobe. Extensive retroperitoneal and pelvis adenopathy. Confluent adenopathy demonstrated in the para-aortic and aortocaval iliac



regions. Hepatosplenomegaly was shown. Para-aortic biopsy showed no evidence of malignancy. Increased eosinophils were noted in the peripheral fibroconnective tissue away from the lymphoid tissue. Lacrimal gland biopsy showed perivascular fibrosis.

Conclusions

Overall, the history, clinical exam and results were most in keeping with IgG4-related disease. He was treated with Prednisolone tapering from 60mg orally. Rituximab was subsequently introduced to good effect following a negative biologic screen. He was symptom free on low dose of steroids on his most recent assessment.



24S116

Cerebral Malakoplakia: A Rare Complication in an Immunosuppressed Patient

Author(s)

Musab Suliman, Mohanad Abdulrahman, Jordan Grant, Anwar Al Albri, Alwin Sebastian

Department(s)/Institutions

Rheumatology Department, Limerick Regional Hospital

Introduction

Cerebral Malakoplakia represents a rare chronic inflammatory condition characterized by distinct pathological features, including Michaelis-Gutmann bodies, which are indicative of abnormal lysosomal function within macrophages in response to infectious agents. Predominantly affecting the genitourinary system, cerebral involvement is exceedingly rare, often presenting diagnostic challenges due to its tumor-mimicking symptoms. Patients with a history of immunosuppression, whether from medications or underlying diseases such as HIV, post-transplant status, or inflammatory bowel disease, are at increased risk.

We report a case involving a 20-year-old male with a history of Common Variable Immune Deficiency (CVID), epilepsy, and Evans Syndrome, who has been receiving monthly IVIG treatments alongside rituximab for recurrent thrombocytopenia. The patient's initial presentation to the hospital was characterized by sudden severe occipital headache, neck pain, and vomiting. Following brain biopsy and histopathology, a diagnosis of cerebral Malakoplakia was confirmed.

Aims/Background

This case aims to underscore the potential for Malakoplakia to occur as a complication in immunosuppressed patients, particularly those on immunosuppressive therapies, highlighting the need for heightened awareness among clinicians.

Method

Case report

Results

Initial treatments involving various antibiotics were ceased due to adverse reactions or insufficient efficacy, ultimately leading to the selection of ceftazidime as the primary treatment. Upon discharge, long-term prophylactic oral co-trimoxazole was prescribed. Despite the patient's intricate medical history, characterized by CVID and rituximab use, prompt recognition and assertive management of cerebral Malakoplakia resulted in positive outcomes. Upon discharge, the patient showed a remarkable response to the antibiotic regimen, evidenced by serial brain imaging that revealed substantial improvement in the cerebellar lesion (Images 1-4), alongside the resolution of symptoms.

Conclusions

Malakoplakia, affecting organs such as the brain, genitourinary system, gastrointestinal tract, skin, lungs, bones, or endometrium, should be included in the differential diagnosis for patients receiving immunosuppressive treatment who present with symptoms indicative of involvement of these organs.





First CT brain without contast on admission: Arrow Showing Let: cerebellar hemisphere edema with focal hemorrhagic component, potential underlying mass or infarct. There is evidence of raised intracranial pressure and hydrocephalus.

and previous and admassive crucicle or emailing resort in the encoded and approximate with marked perilesional edema with associated mass effect, effacement of the burth ventricle with acute hydroceptalus.





Repeases Mix brain after in antibiotics: interval decrease in extent of abnormal 12/VAW hyperintensity in the left cerebellar hemisphere. Re-identified enhancing lesion. Demonstrating interval decrease in the extent of enhancement.

 suppose that approximately two months on iv antibuotos: (nor a sciencially to interval change with post-operative changes in the left posterior fossa with minor high signal within the left cerebellum.

24S117

Complex Presentation of Systemic Autoimmune Disease with Neurological, Hematological, Cardiac, Respiratory, and Renal Manifestations: A Diagnostic Challenge

Author(s)

Dr. Jordan Grant, Dr. Musab Suliman, Dr. Anisah Farouk, Dr. Yaseen Yacoob, Dr. Liam Casserly, Dr Kieran Murray, Dr Joe Devlin, Professor Alexander Fraser, Dr. Alwin Sebastian

Department(s)/Institutions

Rheumatology Department, University Hospital Limerick

Introduction

Systemic autoimmune diseases can present with a broad spectrum of symptoms across multiple organ systems, posing significant diagnostic challenges. We report a case of a 65-year-old female with a history of hypothyroidism, hypercholesterolemia, and hypertension, presenting with progressive multi-system involvement, including neurological symptoms, haematological abnormalities, and renal impairment, indicative of an underlying systemic autoimmune disorder.



Spring Meeting 2024







Aims/Background

The patient, initially well and active until 2018, presented with imbalance, frequent falls, ataxia, myoclonic jerks, chronic hypothermia, recurrent infections, and intermittent pancytopenia. Later, she developed pneumonia with neutropenic sepsis, worsening mobility with dysarthria, peripheral oedema, hypertensive urgency, severe abdominal pain, hearing impairment, progressive dysphagia and seizures. Notably, investigations revealed intermittent thrombocytopenia, neutropenia, chronic deranged liver function tests, hypoalbuminemia, elevated serum amyloid A, and evidence of minimal change disease on renal biopsy without immune complex deposition. Neurological evaluations demonstrated encephalopathy, as well as an episode of subclinical status epilepticus; and nerve conduction studies demonstrated evidence of low-grade neuropathy. Imaging studies highlighted bilateral mastoid effusions, pleural effusion, reduced cortical medullary differentiation in kidneys, small pericardial effusion and progressive heart failure with severe mitral regurgitation. Extensive serological, genetic and immunological testing for autoimmune and paraneoplastic markers yielded largely negative results; though, HLA DRB1*03:01 was positive, and direct antiglobulin test confirmed warm autoimmune hemolytic anaemia. Bone marrow biopsy suggested possible myelodysplastic syndromes versus reactive changes.

Method

Case report

Results

This case underscores the complexity of diagnosing systemic autoimmune diseases with multi-system presentations and inconclusive diagnostic markers. The differential diagnosis remains broad, including systemic lupus erythematosus, vasculitis, paraneoplastic syndromes, multisystem mobility disorder, and myelodysplastic syndromes. The multidisciplinary management approach focuses on symptomatic treatment, immunosuppression, and close monitoring for disease progression and treatment response. Notably, maintenance therapy with hydroxychloroquine, steroids, and monthly immunoglobulins has effectively improved both the encephalopathy and thrombocytopenia (figure 1-2), and has reversed the patient's hearing impairment.

Conclusions

Systemic autoimmune diseases can present with a wide array of challenging and overlapping symptoms. A comprehensive and iterative diagnostic process, involving multiple specialties, is crucial for such complex cases. This case highlights the need for awareness of the potential for seronegative systemic autoimmune diseases to present with non-specific multi-system involvement.



24S119

Atypical Presentation of Myositis with Giant Cell Arteritis.

Author(s)

Dr. MohamedElmansour Muhieldin, Dr. Muhammad Daniyal Memon, Dr. Eleanor Connolly and Dr.Linda Oshea

Department(s)/Institutions

Internal Medicine of Midland Regional Hospital Tullamore

Introduction

Giant cell arteritis (GCA) is a type of vasculitis, a group of diseases whose main feature is inflammation of blood vessels. GCA can overlap with other rheumatological disorder however it rarely presents with inflammatory conditions such as myositis. This paper reports the presents of Myositis with GCA.

Aims/Background

Case Report

Method

A 71-year-old male with a background of dilated cardiomyopathy with Heart failure, hypertension and Type 2 diabetes mellitus, presented with joint and muscle pain and fatigue-limiting mobility for weeks. Initial systemic review and examination were unremarkable. Preliminary investigations noted elevated CRP of 214 and WCC of 10.6 but no obvious source of infection. The patient was admitted with an infection of unknown origin and started on intravenous Piperacillin/tazobactam 4.5g, with blood and urine cultures sent for culture and sensitivity. However, the patients showed no improvement, with no recorded temperature spikes over four days. Follow up examination had noted occipital headaches with temporal tenderness. Along with shoulder and pelvic joint synovitis and tenderness. In addition to proximal muscle weakness that was unexplained. An autoimmune screen was sent off, which noted elevated ESR and CK levels but antineutrophil antibodies and Myositis screens were negative.

He underwent MRI scan of the whole body which noted the presence of polymyositis and vasculitis. Thus follow up Temporal artery and Muscle biopsies were done which noted presence of GCA.

Results

He was started on prednisolone 60mg once a day ongoing and Privigen 30g once a day for five-days prior to biopsy results. He noted significant improvement with the regiment and was then discharged post biopsy on steroid regiment. During the outpatient appointment he was started on a course of Rituximab infusion.

Conclusions

The presence of Myositis and GCA is a quite rare presentation with a limited number of reported cases in literature. As such it is imperative to remember rheumatological disorders such as myoscitis and GCA while unrelated in common can be present concurrently and even remit presentation up to two years from symptoms onset, thus regular follow ups are imperative.







Spring Meeting 2024







24S120

Clinically suspected arthralgia with immediate emergent of what seems like classifiable RA after Pembrolizumab

Author(s)

Abdelrahman Elshiekh 1 Dr Carl Orr 1 Dr David O'Sullivan 1

Department(s)/Institutions

1 Rheumatology department, St Vincent's Unversity Hospital

Introduction

Pembrolizumab is an immune checkpoint inhibitor that is used in the treatment of a variety of cancers in the adjuvant or metastatic setting. Adverse effects include non-specific activation of T cells, leading to immune-related adverse events in downstream organs.

Aims/Background

72-year-old man.presented in July of last year, and at that stage he had minimal symptoms, but some pain and stiffness, it seems, in his right index finger. He is left hand dominant. It does not seem he had any definitive findings on clinical examination at that time ,. His symptoms remained relatively subtle until such time, it seems, as he started Pembrolizumab, he thinks about 3 months ago. He developed, pretty soon after his first infusion, pain and stiffness in his hands, such that he could not make a fist, and debilitating shoulder symptoms, which limited range of motion. No other joints have been involved. His Pembrolizumab is following radiation therapy in September of last year to treat metastatic lung cancer, adenocarcinoma on histology in August 2023. Other than his oncology background, he has a history of hypertension and he has been taking Aspirin and statins.

Method

Prior initiation of Pembrolizumab :his rheumatoid factor was borderline positive at 9.7 and ACPA was positive at 13. ANA positive at 1:400 and anti-centromere 14 units

Notably plain film radiographs of his hands and feet at that time only identified degenerative changes and did not identify features that would be compatible with rheumatoid arthritis.

Results

After Prednisolone was started at a dose of 20mg tapered over the course of 4 weeks, his symptoms improved within 48 hours of commencing steroids. He is essentially asymptomatic afterwards.

Conclusions

Approximately one-third or more of patients with preexisting rheumatic or other systemic or autoimmune disease have experienced flares of their prior disorder in association with treatment using immune checkpoint inhibitors (ICIs) for malignancy. Many have been successfully managed with glucocorticoids or other treatments, but some have required discontinuation, usually temporarily, of their ICI.

24S121

Unusual Presentation of Cord Compression with Incidental Diagnosis of Scleroderma and CPPD

Author(s)

Dr Shagufta Kiran¹, Dr. Patricia Cunningham³, Dr Malik Maqsood Anwar 2, Professor Ronan Mullan 1, Dr Shawn Chavrimootoo 1

Department(s)/Institutions

1. Rheumatology Department, Our Lady's Hospital, Navan 2. Medical Department, Our Lady's Hospital, Navan 3. Radiology Department, Our Lady's Hospital, Navan

Introduction

This is a case report describing the most unsuspecting cause to patient's symptoms. A 59 year old Estonian male presented to ED with new gait disturbance, fall, and 3-day preceding history of central chest pain radiating to the back. He only had a background of Duodenal Ulcer. Examination revealed truncal and lower limb ataxia with reduced ankle reflexes bilaterally. Distal proprioception was reduced in toes with normal vibrioception and fine touch. He had no urinary retention, anal sphincter tone was normal with no saddle paraesthesia. There was no axial weakness and cranial nerve examination was normal.

Aims/Background

CT brain, CT Aorta and MRI Lumbosacral Spine failed to reveal the cause of his symptoms, with no dissection seen. His symptoms improved with physiotherapy. An incidental finding of sclerodactyly to mid forearms was noted which the patient reported had appeared along with symptoms consistent with Raynaud's phenomenon 1-year previously.

Method

Subsequent connective tissue and paraneoplastic screens documented positive ANA but negative ENA and myositis panel. Capillaroscopy showed dilated capillary loops. Hand X-rays did not show calcinosis or tuft resorption. CT TAP was unremarkable.

Results

A subsequent CT angiogram of the aortic arch, MRI cervical and thoracic spines identified extra axial spinal lesions at T1 bilaterally with a large 30 mm lesion on the right and a smaller shallower 7 mm lesion on the left. These were of low signal intensity on T1 & T2 and did not enhance following contrast. They showed significant impingement on the cord with no abnormal signal present within the cord itself. On CT cervical spine they looked heavily calcified. He was transferred to Neurosurgery for a T1 Laminectomy for excision of the extradural lesion, which consisted of multiple pieces of opaque white/tan tissue.

Conclusions

The biopsy showed fibrocollagenous connective tissue associated with numerous large deposits of coarse basophilic granular material felt to be histologically consistent with Calcium Pyrophosphate Disease. He attended our Rheumatology clinic 2 months following surgery with no further ataxia. Our plan is to investigate for CPPD and start immunosuppression therapy for his underlying Scleroderma.





24S122

VEXing but not VEXAS

Author(s)

Dr. Michelle Colfer, Dr Patrick Mulkerin, Dr John Quinn, Dr. Laura Durcan, Dr. Eoghan McCarthy

Department(s)/Institutions

Department of Rheumatology, Beaumont Hospital

Introduction

Below

Aims/Background

WM, a 66-year-old male, presented with a five-month history of intermittent temperatures, mouth ulcers, generalised arthralgia and weight loss. Background history was significant for Prostate Cancer, Type 2 Diabetes and COPD. He was admitted under the Infectious Diseases team for workup of pyrexia of unknown origin (PUO).

Method

Examination revealed mouth ulcers and a diffuse salmon pink rash. Bloods identified a normocytic, normochromic anaemia with high CRP. Extensive screening for infectious causes were negative as was screening for connective tissue disease. A CT-PET scan showed a hyperactive bone marrow. Subsequent bone marrow showed a hypercellular marrow with myeloid hyperplasia consistent with myelodysplastic syndrome (MDS).

Rheumatology were asked to review WM during a subsequent admission. He was commenced on Prednisolone for a presumed diagnosis of Adult Onset Stills with improvement in symptoms and inflammatory markers. Subsequent attempts to taper to below 15mg of Prednisolone resulted in recurrence of symptoms and Tocilizumab was introduced. This again resulted in a dramatic improvement in both inflammatory markers and WM's symptoms initially however he remained refractory to attempts to wean Prednisolone below 10mg.

Results

Detailed review of notes and investigations at that point identified the presence of vacuoles on bone marrow and a neutrophilic dermatosis on skin biopsy which in combination with the apparent refractory nature of his autoinflammatory disease lead to a provisional diagnosis of VEXAS Syndrome. Testing for the UBA1 gene to confirm diagnosis was negative however analysis identified somatic mutations in the ZRSR2 gene located on the X chromosome and EZH2 gene on Chr 7 both of which are associated with MDS and autoinflammation. On this basis his Tocilizumab was switched to Ruxolitinib and since then he has been maintained on 5mg Prednisolone and remains symptom free.

Conclusions

Despite our patient having almost all the clinical features of VEXAS, the mutations in the ZRSR2 and EZH2 genes made this the incorrect diagnosis. UBA 1 testing remains mandatory to confirm a VEXAS diagnosis. Almost 50% of patients with biopsy-proven MDS can have an auto-inflammatory phenotype mimicking Stills Disease. Detailed genetic testing in such patients can confirm diagnosis, alter treatment and inform prognosis.

24S123

Diagnostic and treatment dilemma of a suspected case of Primary Angiitis of the Central Nervous System, in a patient with posttransplant lymphoproliferative disease.

Author(s)

Karolina Warciak, Marah Shaikh Yousef, Kathy Gallagher, Orla G Killeen, Emma Jane MacDermott, Peter McCarthy

Department(s)/Institutions

National Centre for Paediatric Rheumatology, CHI- Crumlin, Ireland, Department of Radiology, CHI- Crumlin, Ireland, Department of Haematology and Oncology, CHI- Crumlin, Ireland

Introduction

We present a case of suspected Primary Angiitis of the Central Nervous system (PACNS), in a patient with Ebstein Barr Virus (EBV) driven Post Transplant Lymphoproliferative Disease (PTLD). We aim to highlight the importance of multi-disciplinary (MDT) approach to diagnosis and management.

Aims/Background

PACNS is a rare paediatric disease, now better recognised as a cause of cerebrovascular accident in childhood1,2. There are no standardised treatment protocols. Many reports and studies show a better response with Mycophenolate Mofetil (MMF) maintenance therapy, after an initial cyclophosphamide and glucocorticoid therapy2,3,4.

Method

15-year-old female presented with left-sided limp and facial droop and weakness of left upper limbs eight days post commencement of PTLD treatment. This is on a complex background of renal failure secondary to congenital urogenital malformation, peritoneal dialysis in 2020 and haemodialysis until 2022. First renal transplant in 2021 failed immediately intraoperatively. Second renal transplant was successful in 2022, with MMF and Tacrolimus maintenance. MMF was discontinued in October 2023 due to PTLD diagnosis.

Magnetic Resonance Imaging (MRI) showed multifocal acute infarcts in the right middle cerebral artery territory. Patient was treated with high dose intravenous Methylprednisolone followed by oral Prednisolone. Over the following two weeks patient had two more left-sided neurological events. Repeat imaging showed new areas of infarction relying on blood supply from collateral vessels consistent with PACNS, with no evidence of systemic vasculitis. A brain biopsy was deemed too high risk at present.

Results

As a result of increasing concern for further cerebrovascular injury, she underwent cerebral by-pass surgery. Six weeks post-surgery, patient had left-sided hand tremor, with new areas of acute ischaemia on MRI of brain with contrast.

She has now completed her full treatment, of two doses of Rituximab and six doses of Cyclophosphamide. Remains on oral Prednisolone. We now face a treatment dilemma, as maintenance options are limited due to the risk of EBV reactivation and PTLD recurrence with additional immunosuppression, especially with MMF.

Conclusions

PACNS is an exceptionally rare condition that can lead to permanent neurological damage and often proves fatal. Patients with comorbidities pose greater challenges. Prompt diagnosis, aggressive treatment and comprehensive MDT approach is paramount.



24S124

Not so "SAVI" after all! A diagnostic and therapeutic conundrum

Author(s)

Author: Anitha Sokay, Kathy Gallagher, Emma Mac Dermott, Ronan Leahy, Orla G Killeen

Department(s)/Institutions

National Centre for Paediatric Rheumatology, CHI at crumlin

Introduction

Interferonopathies are a constellation of diseases that cause recurrent pathogenic inflammation, arising primarily through antigenindependent hyperactivation of immune pathways. It is primarily due to abnormal production or signalling of type I IFNs. A disorder of both the innate and adaptive immune system, this condition is relatively new with the first case only recognised in 2011. Currently there are more than 20 interferonopathies recognised with new interferonopathies are with SAVI.

Aims/Background

We present a case of a 11-month-old female with a SAVI like phenotype but unsupportive genetics.

Method

Clinical data was obtained from patient files and collaboration with subspecialities

Results

Our patient first presented with a severe panniculitis rash, swelling of her limbs and hepatosplenomegaly at 5 weeks of age. She developed significant respiratory distress requiring non-invasive ventilation. A CT Thorax revealed diffuse groundglass opacification suggestive of interstitial lung disease.She had elevated inflammatory markers, a transaminitis and positive cardiolipin (low positive) and B2 glycoprotein antibodies (74 U/ml). Clinically SAVI was suspected as the most likely diagnosis.Her interferon signature gene assay demonstrated exaggerated IFI 27 and IFI44L levels. A Trio exome sequencing and IKBKG sequencing to dateh however have not been indicative of any currently recognised interferonopathy. Further genetic analysis has been requested to explore possible novel pathogenic mutations. A reasonable clinical response was seen with corticosteroid therapy in combination with Jak Inhibition. With worsening of her cutaneous disease and an increasing need for corticosteroid use a trial of anti IL-6 therapy was initiated. This was subsequently discontinue and escalating doses of ruxolitinib, pulse high dose corticosteroids and epoprostenol infusion instituted when she recently presented with severe acute digital ischemia.

Conclusions

This case enhances our current understanding of interferonopathies. Although the phenotype and course of the disease is compatible with SAVI, genetic testing is negative to date. Presently there are eleven heterozygous gain-of-function (GoF) variants for SAVI, with JAK inhibition as the most successful therapeutic option to date. Our case possibly represents an as yet unreported genetic variant, further studies are awaited.

24S125

The Boy who Cryo'd Wolf

Author(s)

Dr. Shalen Naidoo, Prof Ronan Mullan

Department(s)/Institutions Tallaght University Hospital

Introduction

Type II cryoglobulinemia is a mixture of monoclonal IgM (or IgG or IgA) with RF activity and polyclonal Immunoglobulins. It is associated with Hepatitis C (less frequently with Hepatitis B and HIV) and autoimmune diseases (SLE, Sjogrens, RA) as well as Lymphoproliferative disorders and 10% no cause is identified, "essential"(2)

Often constitutional symptoms, fever, fatigue, myalgia/arthralgia (Meltzers triad –Purpura, arthralgia, weakness)

It is a difficult diagnosis to make as collection of samples to isolate cryoglobulins require optimal conditions on collection and analysis.

Aims/Background

A 69 year old female presented to the Emergency Department having been, febrile at home,38°C and unwell for a week and being fatigued. As it was during the pandemic she was swabbed for Covid-19 and found to be antigen positive a day prior to presenting. She had rashes to bilateral lower legs, non-blanching, purpuric in nature that were associated with numbness to both hands and wrists.

She also complained of arthralgia to both hands and feet associated with swelling, diffuse large joint arthralgia, to her shoulders and hips and difficulty mobilizing secondary to pain, swelling and numbness to feet.Her extremities were cold. She had a dry cough.

She noted that the lower limb rash relapsing/remitting for months.

Seen in Rheumatology outpatient clinic on 4 other occasions with a rash associated with arthralgia.

Complements specifically C4 was found to be <0.02 with a normal C3. Her Rheumatoid Factor was elevated at 293.

Her complements were further studied noting absent C1q with negative C1q antibodies. Small monoclonal IgG band with a likely mixed cryoglobulinemia type 2.

Nerve conduction studies showed, abnormal findings supporting a mild-moderate length dependant large fibre sensory-motor axonal neuropathy, reassuringly motor and sensory nerve conduction data from both upper limbs were normal.

She was treated with high dose prednisolone 60mg OD, which was tapered over 6 weeks. Given her prolonged sensory deficits she received 2 doses of rituximab 1g. Her rash has resolved but sensory symptoms persist.

Conclusions

Mixed cryoglobulinemia is a challenging diagnosis with varied treatment options. Isolating the correct subtype of cryoglobulin can lead to a very differing diagnosis and treatment plan as well as varying outcomes





24S127

PET Project

Author(s)

Dr David O'Sullivan (1) Dr Carl Orr (1) Professor Eamonn Molloy (1) Dr Abdul Elshiekh (1) Dr Stephen Skehan (2)

Department(s)/Institutions

1. Department of Rheumatology, St Vincents Hospital 2. Department of Radiology, St Vincents Hospital

Aims/Background

Case

A 63-year-old female presented to the ED describing a 3-month history of abdominal pain, arthralgia affecting the large joints of the upper and lower limbs as well as her spine, 10kg weight loss, fatigue and generally feeling unwell. On a background of fibromyalgia, as appraised by a rheumatologist years previously. She denied any fevers, rashes, hair-loss or mouth ulcers. Her symptoms onset immediately followed an emergent sigmoid colectomy.

Her symptoms had improved somewhat a month prior with a oneweek trial of prednisolone 20mg, initiated by her GP, considering a working diagnosis of PMR.

Method

Clinical examination identified mild discomfort on internal and external rotation of the right hip, but it was otherwise unremarkable.

Results

Laboratory indices identified a CRP level of 163mg/l, normal WBCs, moderately abnormal LFTs, lactate 4.1 mmol/L, fibrinogen 7.04g/L and normal complement levels. Serology on ELISA/IIF was negative for RF, ACPA, ANA and ANCA. LDH and ferritin were >1800IU/L and 3519 μ g/L respectively.

Plain film radiographs of the large joints as well as a temporal artery ultrasound were normal. Contrast enhanced CT-TAP identified low volume retroperitoneal, cervical and supraclavicular lymphadenopathy.

The patient subsequently had a PET-CT, which identified extensive multifocal uptake in the bone marrow with multiple avid lymph nodes.

She underwent a bone marrow biopsy and was diagnosed with diffuse large B cell lymphoma. She is now in remission having received appropriate therapy.

Conclusions

Learning-points:

- Whole body PET-CT imaging is becoming increasingly important in identifying pathology presenting apparently with rheumatologic symptoms. It is superior to contrast enhanced CT and assists in identifying a relevant target tissue to biopsy.
- This patient's heterogeneous symptoms could have been attributed to her diagnosis of fibromyalgia, but for her abnormal acute phase reactants. However, not all patients will mount an acute phase in this manner, and this would contribute to diagnostic delay or inaccuracy.
- Destination is Destiny? This lady was referred to rheumatology due to the nature of her musculoskeletal symptoms, her fibromyalgia diagnosis, the attribution of her symptoms to PMR by her GP and her partial response to corticosteroids (a common feature of lymphoma), however, it is important to vigilant and remain openminded.





Spring Meeting

2024

24S128

A challenging case of Checkpoint Inhibitor induced autoimmunity

A

Author(s)

Dr. Matthew Kelly, Prof. Jarushka Naidoo, Dr. Laura Durcan, Dr. Eoghan McCarthy

Department(s)/Institutions

Beaumont Hospital Rheumatology Department

Introduction

Immune Checkpoint Inhibitors (ICIs) are the most commonly used type of cancer immunotherapy but can cause a number of immunerelated adverse events due to immune activation and inflammation.

Aims/Background

To describe a case of ICI induced autoimmunity.

Method

Review of patient chart, labs and imaging.

Results

MH, a 57 year old female, was diagnosed with metastatic lung carcinoma following left adrenalectomy for left adrenal mass in December 2020. Past medical history included HTN and COPD (ex-smoker with 40 pack year history). Following surgery she commenced treatment with Carboplatin/Pemetrexed in conjunction with Pembrolizumab - anti programmed cell death-1 (PD-1) antibody.

In September 2021 she developed PR bleeding with sigmoidoscopy revealing colitis. She was treated with IV Methylprednisalone and tapering Prednisalone on discharge to good effect. Following her next course of immunotherapy she developed periorbital oedema and exophthalmos. She was again admitted for IV Methylprednisalone and tapering Prednisolone but noted worsening visual acuity (6/60) on tapering in the community and was admitted for urgent ophthalmology review. She was treated with pulse methylprednisolone to good effect. Lacrimal Gland Biopsy showed acute and chronic inflammation and interstitial fibrosis in keeping with dacryoadenitis. There was no evidence of malignancy (MRI Brain and PET CT). A diagnosis of checkpoint inhibitor induced dacryoadentitis was made. Rheumatology were consulted with Methotrexate introduced as a steroid sparing therapy. On reduction of Prednisolone to 15mg patient developed new onset shortness of breath. CT showed pneumonitis. Bronchoscopy was negative for infection and symptoms improved with increase in steroid. Methotrexate was changed to Azathioprine given the concern re pneumonitis and to facilitate use of Co-trimoxazole. Her eye disease gradually improved and she has successfully reduced prednisolone to 5mg and remains well on follow up with no cancer recurrence.

However the prolonged steroid therapy resulted in multiple complications including cushingoid appearance, exacerbation of anxiety, oral thrush, compression fractures, stress fracture of distal tibia and bilateral AVN of navicular bones.



Spring Meeting 2024







Conclusions

ICI have improved cancer outcomes but are associated with the development of autoimmune complications. Early multidisciplinary management can aid diagnosis and reduce complications associated with therapies used to manage ICI associated toxicity.





24S129

Not All "Fava Beans and a Nice Glass of Chianti"

Author(s)

Sarah Quidwai, Ronan Mullan, Patricia Cunnigham, Shawn Chavrimootoo

Department(s)/Institutions

Department of Rheumatology, Our Lady's Hospital, Navan, Co Meath.

Introduction

We present a 66 year-old male with ALT derangement, four years after a diagnosis of undifferentiated inflammatory arthritis treated with Methotrexate. Following an initial presentation with a left ankle monoarthritis in 2019, remission was rapidly achieved with Methotrexate 20mg + Folic Acid 5mg weekly. His medical history included hypercholesterolaemia, for which he was taking Atorvastatin 10mg nocte for 7 years.

Aims/Background

In February 2023, routine bloods revealed ALT 144U/L. Other LFTs and inflammatory markers were normal. Methotrexate was stopped but ALT continued to rise with ALT 631U/L in April 2023. There was no history of alcohol intake or new medications. Liver ultrasound was normal. Metabolic, immune and viral liver screens were normal. Atorvastatin was stopped. The patient was referred to hepatology while remaining clinically well. Method.

Results

In May 2023 the patient was admitted with proximal muscle weakness, dysphonia and dysphagia. Bloods showed ALT 772U/L, AST 567U/L and CK 9193U/L. FBC, renal, bone profile, CRP and immunoglobulins were normal. Myositis panel, ANA, ENAs and ANCA were negative. Liver biopsy was normal. CT TAP was unremarkable. MRI femurs showed symmetric muscle oedema in both thighs. Right rectus femoris biopsy was taken. Prednisolone 80mg was commenced.

On review three weeks later, the patient reported no improvement in symptoms and new upper limb weakness despite steroids. Biopsy results were consistent with an immune mediated necrotising myositis (IMNM). Further antibodies were checked and HMG CoA reductase inhibitor was positive. The patient was started on IVIG therapy and Methotrexate with a plan to taper steroids. After two months of treatment, the patient was asymptomatic. CK and ALT normalised.

Conclusions

An abnormal ALT is not always indicative of liver pathology, as demonstrated in this case. Anti HMGCR positive IMNM is rare. The

incidence is 2- 3 per 100,000 patients on statin treatment. Median duration for statin exposure before symptom onset is 32-38 months. Upto a third of patients do not develop muscle weakness until after statin discontinuation. Atorvastatin is the most commonly associated statin and doses as low as 10mg have been associated with IMNM. Clinicians should remain vigilant to this serious complication of a commonly prescribed medication.

	ALT (U/L)	CK (U/L)	Bili (umol/L)	ALP (U/L)	GGT (U/L)
18/02/19	20		8	92	18
16/02/23	144		11	65	21
02/03/23	173		10	78	19
14/03/23	214		11	74	21
27/03/23	278		11	75	18
11/04/23	383		10	77	17
27/04/23	631		10	76	16
08/05/23	772	9193	10	68	15
16/05/23	723	8780	14	78	16
15/06/23	630	3343	13	69	16
26/07/23	267	1854	7	50	15
02/08/23	191	1633	11	46	15
30/08/23	80	955	9	50	17
27/09/23	25	177	9	46	17





24S130

Case of Neuromyelitis Optica (NMO) in Systemic Lupus Erythematosus following SARS-CoV-2 vaccine

Author(s)

Anisah Farouk, Muhammad Hassan Malik, Yaseen Yacoob, Kieran Murray

Department of Rheumatology, University Hospital Limerick.

Department(s)/Institutions

Introduction

We report a case of Neuromyelitis optics (NMO) as a manifestation of SLE following SARS-CoV-2 vaccination.

Aims/Background

A 31-year-old female presented with eight-day history of worsening ascending bilateral lower limb weakness, inability to weight bear, neck pain associated with ascending altered sensation, facial dysesthesia and right sided blurred vision. Her symptoms commenced one day following third Pfizer-BioNTech mRNA SARS-CoV-2 vaccine. She had one year diagnosis of SLE following presentation with arthralgia, malar rash and positive anti-double stranded DNA (anti-dsDNA) which was treated with hydroxychloroquine. She was unfortunately lost to rheumatology follow up and discontinued her medications.

On examination, she had asymmetric spastic quadriparesis involving bilateral lower and upper limbs, worse distally with brisk lower limb reflexes and positive Babinski sign. Sensation was reduced bilaterally to a high cervical level worse on right side (C2-T12). She had right relative afferent pupillary defect (RAPD) with right optic nerve head swelling. She was clinically in urinary retention.

Method

Case report

Results

Creatinine Kinase (CK) was elevated, 233U/L, anti dsDNA positive with a titre of 350IU/ml and positive p-ANCA but negative myeloperoxidase (MPO) and anti-proteinase-3 (PR3) antibodies. Cerebrospinal fluid (CSF) pleocytosis was identified with mononuclear predominance (99%) but negative infectious screen. MRI brain, cervical and thoracic spine showed long segment intramedullary cervical spinal cord signal abnormality extending from the medulla to C7 level with cord oedema and heterogenous enhancement and involvement of most of the cross-sectional area of the cord, consistent with transverse myelitis. {Figure 1}

A diagnosis of SLE-induced transverse myelitis was made



and was initiated with intravenous (IV) methylprednisolone, IV immunoglobulins (IVIG), hydroxychloroquine and Cyclophosphamide. Her power improved but asymmetrical weakness with spasticity persisted. Aquaporin 4 antibodies (Anti-AQP4) returned positive, prompting the suspicion of Neuromyelitis Optica (NMO). Rituximab, the anti-CD20 chimeric monoclonal antibody, was commenced and significant improvement was noted.

Conclusions

There is an increased rate of demyelinating disease in patients with underlying auto-inflammatory conditions, including SLE.1 Prior research proposes vaccinations may trigger NMOSD. This patient with SLE presented with NMO following SARS-CoV-2 vaccination and we hypothesize patients with underlying autoimmune disease such as SLE are a cohort at particular risk of developing NMO following SARS-CoV-2 vaccination.



Figure 1; Magnetic Resonance Imaging of cervical spine, thoracic spine (A, C, D) and brain (B)

- A: Sagittal view T2 weighted image showing long segment intramedullary hyperintensity with cord oedema and
- expansion involving the cervical spinal cord extending from the lev of the lower medulia to C7
- B: Axial view T2/Flair weighted image showing hyperintensity in th right optic nerve.
- C: Sagittal view T1- weighted pre gadolinium image showing abnormality
- abhormainty D: Sagittal view T1- weighted post gadolinium showing 2 contri enhancing regions at lower medullary region and C3 to C6

24S131

A Myositis Mimic

Author(s)

Sumaia Abuelbasher, Peter Browne, Lorraine O'Neill

Department(s)/Institutions

Department of rheumatology, University Hospital Kerry

Introduction

Metastatic tumours presenting as a soft tissue masses are relatively rare and can be a source of diagnostic confusion both clinically and pathologically [1]. This case report describes a patient with primary adenocarcinoma of lung presenting with upper limb muscle metastasis.

Aims/Background

To investigate arm swelling presented with feature of infective myositis

Method

A 50-year-old woman presents with right upper limb pain and swelling for two months, worsening in the two weeks prior to admission. The pain was concentrated in the forearm and aggravated with movement. She had no history of trauma, cuts or bites, fever nor constitutional upset. Review of systems was otherwise negative.

Results

Swelling of right upper limb (UL) around elbow joint with extension distally and proximally with subcutaneous edema (Figure 1). There was no neurovascular compromise.

Doppler US RT upper limb:

Doppler US (Figure 2)–defined a heterogenous lesion seen in the subcutaneous tissue of the anterolateral aspect of the right mid forearm measuring about 4.9 cm x 1.6 cm.

Differential Diagnosis (DD) haematoma or beginning abscess formation, for clinical correlation and further MRI assessment MRI SCAN RT UPPER LIMB:

Multiple intramuscular abscesses (figure 3), the largest within the flexor carpi ulnaris and digitorum muscles.

Associated oedema throughout the muscles consistent with an infective myositis.

US guided aspiration is advised.

US GUIDED BIOPSY:

US guided aspirate failed – solid lesions, not amenable to aspirate. Open biopsy was done – C&S negative, AFB negative. Blood cultures negative and HIV negative as well.

Biopsy and histopathology:

Fibrous tissue infiltrate by poorly differentiated adenocarcinoma. Immunohistochemistry shows variable positive staining of neoplastic cells with TTF1 which raise possibility of lung as primary. CT TAP:

Multiple bilateral lung nodules involving all lung segments in keeping with metastases (Figure 4). Large right-sided pleural effusion.

Conclusions

Although skeletal muscle comprises nearly 50% of the total human body mass and are well vascularised, metastases in the musculature are rare [2]. The reported prevalence of skeletal muscle metastasis (SMM) in autopsy series of cancer patients ranges from 0.03% to 16% [2,3,4 and 5]. Soft tissue metastasis in lung cancer has been considered a devastating prognostic indicator with a median survival of less than 6 months even on chemotherapy [6].



Figure 1: Shows unilateral right UL Swelling

Doppler US RT upper limb



Figure 2: Doppler US



24S133

Granulomatosis with Polyangiitis Presenting as Isolated Cerebritis – A Case Report

Author(s)

Burce Isik1,2, Matthew G. Davey2, Alwin Sebastian1,2, Alexander Fraser1,2

Department(s)/Institutions

1University of Limerick School of Medicine, Co. Limerick, Republic of Ireland; 2University Hospitals Limerick, Co. Limerick, Republic of Ireland

Background:

Isolated central nervous system involvement in granulomatosis with polyangiitis is rare. There are limited accounts of such a presentation in the literature.

Case presentation:

A 61-year-old gentleman presented with right upper limb and lower limb weakness, slurred speech and confusion following two instances of mechanical falls while at home. A cerebrovascular event was suspected. He underwent computed tomography brain and magnetic resonance imaging brain which demonstrated leptomeningeal thickening of left frontal region with vasogenic oedema. The differential diagnosis included malignancy, infection and contusion. He initially commenced on Dexamethasone 8mg, Keppra 250 mg and a combination of Ceftriaxone, Linezolid, Metronidazole to treat vasogenic edema and a possible infectious cerebritis, respectively. His vasculitis screen demonstrated a positive p-ANCA, positive for high titer PR3. He was diagnosed with cerebral vasculitis secondary to granulomatosis with polyangiitis and commenced on immunosuppressant treatment.

Conclusion:

Granulomatosis with polyangiitis with isolated central nervous system involvement can mimic the presentations of acute stroke, cerebritis, malignancy, and traumatic brain injury due to ambiguous symptoms. In the absence of other symptoms of granulomatosis with polyangiitis, isolated cerebral vasculitis often leads to delayed presentation, diagnosis, and treatment of this rare disease.

Key Words: granulomatosis with polyangiitis, cerebral vasculitis, vasculitis, p-ANCA, PR3





Spring Meeting 2024







Spring Meeting 2024





Spring Meeting 2024







Spring Meeting 2024



