Irish Society for Rheumatology

Spring Meeting

13 May 2021

Welcome Message from the ISR President Prof Geraldine McCarthy

Dear Colleagues and Friends

Having had no option but to cancel our visit to Sligo in 2020, I am delighted to welcome you all to the first virtual ISR Spring meeting. We had all hoped for an in-person Spring event in 2021. Since that is still not possible, we are greatly compensated by the generosity of our wonderful guest speakers who have taken the time to participate. The extra effort it has taken for our speakers to prepare their presentations in advance is much appreciated. We will start with four clinical cases which will be an excellent warm-up for the meeting. We look forward to then hearing Dr Sarah Mackie from the University of Leeds who will bring us the most update information about polymyalgia rheumatic and giant cell arteritis. Dr Sanjeev Patel from Kings College Hospital London will tell us all we need to know about primary bone marrow oedema syndromes. We will hear about haemochromatosis arthropathy from Professor Patrick Kiely from St Georges University Hospital London, which is particularly relevant at present because of the current EULAR initiative to classify haemochromatosis arthropathy. Professor AV Ramanan will bring us up to date on the current status of tocilizumab use in COVID-19. Dr Duncan Rogers will explain how ophthalmic imaging can assist rheumatologic diagnosis. The last two talks will have a lupus focus with addresses by two key opinion leaders. Dr John Hanly will inform us about the current understanding of the effects of lupus on the nervous system and Dr Michelle Petri will bring us completely up to date about the management of lupus. I regret that we can not invite our international speakers to join us in person on this occasion, but they will all be prioritized in the future.

We hope that our Autumn meeting might be more sociable but, as we have learned, there are no guarantees where COVID-19 is concerned.

I would like to offer special thanks to Michael Dineen and Marie Caston who have worked extremely hard to make this meeting a success, despite all the challenges and changing landscape.

I am especially grateful to our colleagues in the Pharmaceutical Industry for the continued support of ISR and its members, especially during these unprecedented times.

Finally, my sincere thanks to the members of ISR board for all their continued support.

Enjoy the meeting

Prof Geraldine McCarthy
President ISR

ISR Spring Meeting

Thursday 13 May 2021

Virtual Programme

10.00 **Opening Address – Professor Geraldine McCarthy,** President, ISR

10.05 **Clinical Cases – Four** (4)

11.00 **Commercial video**

11.10 **Dr Sarah Mackie**
Consultant Rheumatologist and Associate Professor, University of Leeds
Topic: ***"Polymyalgia rheumatica and giant cell arteritis"***

11.35 **Dr Sanjeev Patel**
Consultant Rheumatologist, Kings College Hospital, London
Topic: ***"Primary Bone Marrow Oedema Syndromes"***

12.00 **Commercial video**

12.10 **Professor Patrick Kiely**
Consultant Rheumatologist, St George's University Hospital, London
Topic: ***"Haemochromatosis Arthropathy"***

14.00 **Professor A V Ramanan**
Consultant Paediatric Rheumatologist, Bristol Royal Hospital for Children and Royal National Hospital for Rheumatic Diseases
Topic: ***"Tocilizumab in COVID-19"***

14.35 **Dr Duncan Rogers**
Consultant Medical Ophthalmologist, Mater Misericordiae University Hospital, D7
Topic: ***"How Ophthalmic imaging can assist rheumatological diagnosis"***

15.00 **Commercial video**

15.10 **Dr John Hanly**
Professor of Medicine and Pathology at Dalhousie University and Attending staff physician at the Queen Elizabeth II Health Sciences Centre, Halifax, Nova Scotia
Topic: ***"Lupus and the Nervous system – is the fog lifting?"***

15.35 **Dr Michelle Petri**
Professor Division of Rheumatology, Dept of Medicine, Johns Hopkins University
Topic: ***"Modern Lupus Treatment - What's New"***

N.B. While all of the Clinical Cases received are included in this brochure the 4 selected for Oral Presentation will not be disclosed until the morning of the meeting when delegates will have the opportunity of voting for the presentation of their choice.

Biographical Sketches

Speakers

Athimalaipet V. Ramanan
FRCPCH, FRCP

Athimalaipet V. Ramanan is a consultant pediatric rheumatologist at the Bristol Royal Hospital for Children. He is the joint lead for research (Division of Women and Children) at the Bristol Royal Hospital for Children, and Professor of Paediatric Rheumatology at the University of Bristol.
Professor Ramanan is a medical advisor for Olivia's Vision and the National Ankylosing Spondylitis Society. He is also Chair for the National Institute for Health Research Clinical Research Network: Children/Arthritis Research UK (ARUK) Paediatric Rheumatology Clinical Studies Group, and Associate Director for the UK Experimental Arthritis Treatment Centre for Children (JIA-Uveitis and Industry work streams). He was awarded the British Society of Rheumatology's Innovation in Clinical Practice Award in 2010. He was also awarded the University of Bristol Vice Chancellor's Health Impact award in 2017 and Royal College of Physicians/NIHR CRN Award for outstanding contribution to research in 2018
Professor Ramanan's has published >190 articles and numerous book chapters covering a variety of topics in the field of rheumatology. He is the Co-Editor of Rheumatology and Associate Editor for the Archives of Diseases in Childhood. He is also leading trials of Tocilizumab in COVID-19 and part of the paediatric steering committee of the RECOVERY trial.

Dr Sarah Mackie
Consultant Rheumatologist and Associate Professor, University of Leeds.

Dr Mackie's research is focused around two linked conditions: giant cell arteritis and polymyalgia rheumatica. She is also interested in the effects of the glucocorticoid therapy used to treat them and interested in how new scientific insights may be used to improve clinical care and clinical outcomes.

Professor Patrick Kiely
PhD FRCP

Consultant Rheumatologist and Professor of Practice, Clinical Rheumatology at St George's University Hospitals NHS Foundation Trust and the Institute of Medical and Biomedical Education, St George's University of London, UK Prof. Kiely is a general adult rheumatologist. He holds specialist clinics in rheumatoid arthritis, inflammatory muscle disease (myositis), primary systemic vasculitis, interstitial lung disease associated with connective tissue diseases and the only UK clinic dedicated to patients with haemochromatosis arthropathy.
Prof. Kiely qualified from the University of London in 1988, and has been a consultant at St George's University Hospitals NHS Foundation Trust since 1999. He was a joint author of the NICE clinical guidelines (1999) and Quality Standards (2013) for RA. He is a member of the EULAR task force producing guidelines for therapeutic monitoring of biopharmaceuticals, the BSR task force for guidelines for management of myositis, and leads the EULAR task force to create classification criteria for haemochromatosis arthropathy. His research has resulted in over 100 papers published on aspects of rheumatoid arthritis outcomes optimisation, myositis and haemochromatosis arthropathy.
Member: British Society of Rheumatology
Chairman: South West Thames Regional Rheumatology Group
Medical Advisor: National Rheumatoid Arthritis Society, Haemochromatosis UK
Executive member: Early Rheumatoid Arthritis Network

John G Hanly M.D.

Dr. John Hanly is Professor of Medicine and Pathology at Dalhousie University and Attending staff physician at the Queen Elizabeth II Health Sciences Center, Halifax, Nova Scotia, Canada. He is the Director of the Dalhousie University Lupus Clinic in Halifax. He obtained his medical degree from the National University of Ireland in 1978 and trained in general internal medicine and clinical rheumatology in Ireland prior to immigrating to Canada in 1984. He undertook clinical fellowships in Rheumatology and Immunology at the University of Toronto and McMaster University before joining the Faculty of Medicine at Dalhousie University in 1987.
Dr. Hanly is a member of national and international research networks involved in clinical studies of systemic lupus erythematosus. He is the Past Chair of the Systemic Lupus International Collaborating Clinics (SLICC), a research network comprising 52 lupus investigators in 43 academic centers in 16 countries. He has published extensively and has received awards in recognition of his achievements in clinical research in lupus. Peer-review funding includes continuous operating grant support as Principal Investigator from CIHR since 2002.
Dr. Hanly's major research focus is the study of pathogenic mechanisms and clinical outcomes in systemic lupus erythematosus, with a particular emphasis on ways in which lupus may affect the brain and other parts of the nervous system.

Dr Duncan Rogers
Medical Ophthalmology Consultant

Dr Duncan Rogers is a Medical Ophthalmology Consultant at the Mater Misericordiae Hospital in Dublin. He previously practised as a Consultant in Uveitis at Moorfields Eye Hospital in London. He was the first Medical Ophthalmologist trained in London specialising in Rheumatology, Neurology and Ophthalmology. He has a research interest in Artificial Intelligence role in referral pathways and risk stratification and is currently in receipt of a £1.2 million grant from the National Institute of Healthcare Research and NHSx Digital. His area of clinical interest is the ocular manifestation of rheumatic and neurological disease within the eye.

Dr Sanjeev Patel

Sanjeev has been a consultant rheumatologist since 1996 and works at King's College Hospital in London. He qualified from the University of Southampton and did his training in the UK and New Zealand before settling in London. He is an experienced general physician and rheumatologist. He has carried out a wide portfolio of clinical activities including acute general medicine, all aspects of rheumatology and running a metabolic bone service. He sub-specialises in metabolic bone disease with a focus on secondary causes of osteoporosis caused by eating disorders, chronic kidney disease and steroids. He is also an acknowledged expert in the diagnosis and management of bone marrow oedema syndromes.
Sanjeev has held a variety of roles dedicated to education and training of medical students and junior doctors. He is Vice-Chair of a NICE Health Technology Appraisal Committee for NICE and is President of the British Society for Rheumatology.

Michelle Petri, M.D.
Professor of Medicine, Johns Hopkins University

Michelle Petri, M.D. M.P.H. is a Professor of Medicine at the Johns Hopkins University School of Medicine. She attended medical school at Harvard University and fulfilled her internal medicine residency at the Massachusetts General Hospital. In addition, she completed two fellowship programs at the University of California, San Francisco in allergy and immunology and rheumatology. Dr. Petri is the Director of the Hopkins Lupus Cohort, a longitudinal study of morbidity and mortality in systemic lupus erythematosus, and Co-Director of the Hopkins Lupus Pregnancy Center.

ISR Board members

Professor Geraldine McCarthy
President
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 HIV-associated 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 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.

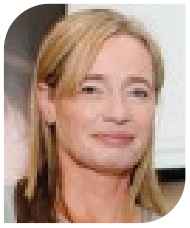
Dr Nicola Ambrose
Consultant Rheumatologist,
Blackrock Clinic, Co Dublin

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 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 in Edinburgh. He was awarded an MD by thesis entitled "Leucocyte surface receptor expression of relevance to apoptotic cell clearance in systemic lupus erythematosus" from Queen's University Belfast in 2001, and an MSc in Sport and Exercise Medicine from the University of Ulster in 2008. He is a Consultant Rheumatologist at Musgrave Park Hospital Belfast where he was appointed in 2004, and also provides rheumatology clinics at Belfast City Hospital and Lagan Valley Hospital Lisburn. He is a Fellow of the Royal Colleges of Physicians of Ireland, London and Edinburgh. He has published in a wide range of rheumatic diseases and is an enthusiastic proponent of musculoskeletal ultrasound.

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 Medicine, Trinity Biomedical Sciences Institute, Trinity College Dublin. Professor Fearon's research is a bench-to-beside translational approach, focusing on understanding the underlying mechanisms that 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.

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 Iain 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 ultrasound, spondyloarthopathy and 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 Colm Kirby
Cork University Hospital, Cork

Colm graduated with a degree in medicine from UCC in 2011. Following completion of CUH medical BST I commenced the Rheumatology HST in 2016. After spending some time University College Hospital Galway and Cork University Hospital, he now works in Tallaght University Hospital. Special interests include Musculoskeletal Ultrasound and premature atherosclerosis associated with systemic inflammatory diseases.

Dr Bernadette Lynch
Consultant Rheumatologist,
University Hospital, Galway.

Dr Bernadette Lynch graduated from the Royal College of Surgeons in Ireland in 2003. She completed her higher specialist training in Rheumatology and General Medicine in 2013 having worked and studied in Dublin, Galway and London. She was awarded an MD from University College Dublin in 2011 for work on IL-22 and musculoskeletal ultrasound in Inflammatory Arthritis. She undertook a fellowship in Scleroderma and Vasculitis at the Royal Free Hospital Hampstead under Professor Chris Denton and Dr Aine Burns. During this time, Bernadette was part of the UK Scleroderma Study Group (UKSSG) which developed the national guidelines on the management of complications of Scleroderma. She took up her current appointment as Consultant Rheumatologist and General Physician in University Hospital Galway in 2015. Her principal clinical and academic interests are Scleroderma and Inflammatory Arthritis.

Dr John Ryan
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 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.

Dr Clare Matthews
Consultant Rheumatologist
Ulster Hospital, Belfast

Consultant Rheumatologist, Ulster Hospital, Belfast Dr Clare Matthews graduated from Queens University Belfast in 1994. She completed registrar training with CCT in Rheumatology and general medicine in 2007. She completed an MD "Clinical, genetic and immunohistochemical findings of early inflammatory arthritis" from The Queen's University, Belfast in 2004. She trained in Belfast with a period of training in St Vincent's University Hospital Dublin through her research interest in synovial disease. Dr Matthews was first appointed as a consultant in Belfast City Hospital and moved to her current post in The South Eastern Trust in 2009.

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.

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Consultant Rheumatologist
Mater Misericordiae University Hospital Dublin
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Dr Bryan Whelan
Consultant Rheumatologist
Our Lady's Hospital, Manorhmailton,
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Rheumatoid Arthritis Biologics Registry of Ireland (RABRI)

Gerry Wilson UCD/MMUH/SVUH

Registries recording data on RA patients being treated with biological and JAK inhibitors are present in many countries. Although the efficacy of these agents is determined primarily in phase III studies, these are of relatively short duration (24-52 weeks) and involve several hundred highly selected RA patients. The potential long term efficacy and also the detection of side effects require post-authorisation phase IV or registry data involving one or two orders of magnitude more patients follow-up for 5 years or more. For example, the use of anti-TNF agents was initially thought to likely increase the risk of malignancy, however data from European and North American registries have not found evidence to support this concern. In addition, registries have confirmed beneficial effects of biological therapies on reducing the cardiovascular risk. Registries also give important data on the use of biological agents in each country, such as the disease activity score at initiation, the average treatment period of each agent, and uptake of new agents. Some of these data are now being collected automatically with recent development of central electronic prescribing platforms.

The Rheumatoid Arthritis Biologics Registry of Ireland (RABRI) was initiated with the aims of studying post-approval efficacy and side-effects of established and newly approved agents, including biosimilars. The design is similar to that of the British Society for Rheumatology Biologics Registry (BSRBR) involving 5 years of follow-up following treatment initiation; 6 monthly during years 1-3, and annually in years 4 and 5. However whilst the BSRBR data is collected using paper and post, RABRI has a bespoke online data capture interface. Data input at initiation takes around 20 minutes and subsequent visits requires less than 10 minutes. Centres contributing patients to RABRI are supported financially based on recruitment and follow-up visits.

In line with EULAR recommendations for drug registries the governance, including finances, is controlled by the national body, so in Ireland it is the Irish Society for Rheumatology. A steering committee meets regularly to review all aspects of the study including recruitment. All but one of the pharmaceutical companies have supported the creation and maintenance of RABRI, 6 monthly reports on any serious adverse events on individual drugs are sent to the relevant company, although these have been very rare. The major expenses have been the creation and maintenance of the electronic database, salaries for the study manager and coordinator (both 0.25FTE), and payments to contributing units.

Recruitment over the past two years has been adversely impacted initially by issues around the General Data Protection Guidelines and subsequently by the global pandemic. Hopefully, as the disruption from the latter recedes, recruitment will pick up again during 2021. We aim to submit an abstract on current RABRI data to the 2021 ISR AGM.

If you would like to recruit to RABRI or would like to undertake a study using the study data please email: registries615@gmail.com. We are keen for members of the ISR to join the steering committee.

RABRI Steering Committee

NAME	LOCATION
Gerry Wilson	Mater
Doug Veale	SVUH
Phil Gallagher	SVUH
Michele Doran	SJH
David Kane	TUH (ISR)

Speakers at the Autumn 2020

Professor David Kane

Dr Rachael Flood winner YIA 2020

(20S101) CLINICAL CASE 1

A Rugby Injury with a Difference

Author(s)
Ward. C, Kenny. G, Feeney. E
Department(s)/Institutions
St. Vincent's University Hospital, Ireland East Hospital Group, Dublin, Mid Leinster.

Introduction
A seventeen year old, previous healthy rugby player presented to a private Emergency Department with a two week history of groin pain, and leg stiffness following a rugby match.

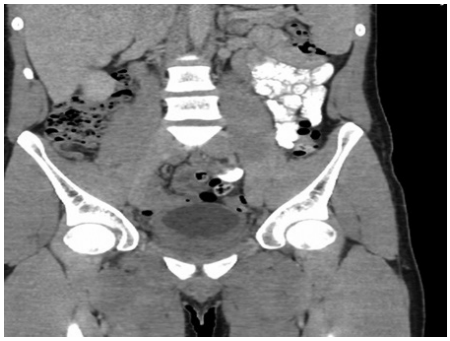
Aims/Background
He had no sexual partners and was fully vaccinated. His brother had been diagnosed with mumps eight months previous. His CRP was elevated and he was febrile. Mumps Ig G was positive but Ig M was negative. He was diagnosed with mumps orchitis and discharged home on seven days of amoxicillin.

Method
Ten days later he presented to the Emergency Department of SVUH with a worsening limp and fevers of 38.6°C. He was tender suprapubically and in the right iliac fossa. Testicular examination was normal. His CRP was elevated at 190mg/l and an STI screen (including HIV and syphilis), blood and urine cultures were negative. Mumps serology was consistent with previous infection or vaccination. CT abdomen revealed thickening of the urinary bladder wall and surrounding infiltration of the perivesical fat. MRI demonstrated a small joint effusion and florid oedema surrounding the pubis symphysis. There was an associated erosion of the cortex of both pubic bones, more marked on the right side equating to a diagnosis of septic arthritis of the symphysis pubis.

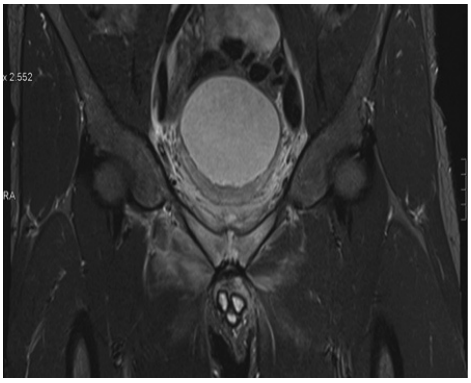
Results
Due to the location of the process and the absence of a drainable collection no sample was taken. He was commenced on empiric ceftriaxone 2g IV daily and had a clinical and biochemical response with falling CRP. He was discharged on Outpatient Antibiotic Therapy to complete a six week regimen. A repeat MRI performed at three months revealed complete resolution.

Conclusions
Pubic symphysis septic arthritis is rare. Although cultures were negative, Staphylococcus aureus is the main causative organism, presumably from haematogenous spread. It is most common in young athletes particularly football and rugby players owing to micro-trauma caused by repetitive over adduction and twisting. (1) It is likely that the previous prescription of amoxicillin prevented the bacteria growing in blood cultures. Owing to its rarity and variable presentation; diagnosis requires a high index of suspicion as symptoms can mimic other conditions which occurred in this case.

Figure



Figure



(20S102) CLINICAL CASE 2

HIV infection masquerading as connective tissue disease.

Author(s)
Z Javaid 1,2, M Suhaib 1,2, B Mekkayil 2, A Paul 2
Department(s)/Institutions
1 Acute Medical Unit, Darlington Memorial Hospital, County Durham & Darlington NHS Foundation Trust, DL3 6HX 2 Rheumatology Unit, The James Cook University Hospital, South Tees Hospitals NHS Foundation Trust, TS4 3BW

Introduction
We describe a case of 58 years old non-smoker lady,who presented with acute onset livedo reticularis rash on lower limbs.

Aims/Background
She had background of sicca symptoms, oral ulcers, fatigue, paraesthesias in feet and arthralgias without any systemic or inflammatory joint symptoms.
General examination showed livedo reticularis rash on both elbows and legs up to the knees. There was no evidence of peripheral joint synovitis but she had nodal osteoarthritis in her hands. Systemic examination was unremarkable.

Method
Investigations revealed anaemia, pancytopenia, ESR of 77, low C4 and urine dipstick positive for leucocytes, nitrates, protein and blood. Schirmer's test, ANA and ENA screen was positive with positive RNP and SMdp antibody. She also had hypergammaglobulinaemia in a polyclonal pattern. Nerve Conduction and EMG studies revealed mild axonal sensory neuropathy.

Results
This lady appeared to have mixed connective tissue disease and commenced on Hydroxychloroquine but stopped it shortly after developing floaters in her eye. She had poor response to Depomedrone injection. She had ongoing symptoms of fatigue, weight-loss, loose stools and abdominal pain, investigated further and CT scan showed hyperdense liver lesions and mesenteric lymphadenopathy. Oesophagoduodenoscopy showed oesophageal candidiasis. She was admitted with progressive symptoms. Further investigations showed a positive HIV test and liver biopsy came back positive for anaplastic lymphoma, Later she was diagnosed with advanced HIV disease, rapidly deteriorated with neutropenic sepsis and multi-organ failure and unfortunately died.

Conclusions
This lady initially presented with symptoms of connective tissue disease and investigations in keeping with this diagnosis. Unfortunately by the time she was tested for HIV infection, it was already too late. There could be overlap of symptoms of connective

tissue disease and viral infections e.g. HIV infection. Autoantibodies may be falsely positive in infections e.g. HIV and in malignancy. Risk factors of HIV infection should be considered during assessment of multisystem diseases like connective tissue diseases particularly prior to immunosuppression. Viral screening including HIV test should be considered in all high risk patients and particularly if symptoms are atypical and do not quite fit as well as part of investigations for weight loss. Early diagnosis and treatment of HIV can prevent life-threatening complications.

(20S104) CLINICAL CASE 3

Is remission achievable?

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Introduction
A 19 years old lady was admitted with non-resolving sinusitis. Her symptoms included a 3-week history of purulent nasal discharge, haemoptysis, night-time fevers and malaise. Initial investigations showed a CRP of 105.8, normal WCC, elevated platelet count at 514.
CT of the sinuses showed diffuse mucosal wall thickening.
Bronchoscopy showed dramatic tracheal inflammation with purulent secretions, oedematous mucosa with contact bleeding.
Further investigations showed a positive c-ANCA with positive PR3 with a titre of 49 (nr<3), negative MPO.
A diagnosis of GPA was made.

Aims/Background
IV pulsed methylprednisolone was started followed by oral prednisolone 60mg. A CT Thorax showed numerous cavitating lung nodules. Histopathology from a nasal endoscopy confirmed GPA. PFTs showed moderate fixed airflow obstruction (FEV1/FVC of 52.75%). We decided to use rituximab to induce remission because of her child-bearing age.

Method
Treatment with rituximab was delayed due to latent Tb screening as 2 quantiferon results were indeterminate.
Eventually a 3rd quantiferon came back negative; she received the first dose of rituximab.

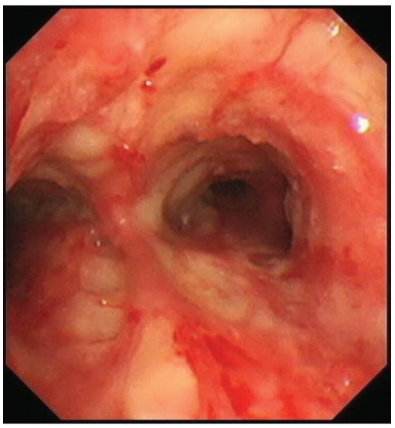
Results
On the day of receiving her 2nd rituximab, she was tachycardic with a low grade pyrexia. Rituximab was postponed; IV antibiotics and pulsed with IV methylprednisolone were started and oral steroids dose was then increased back to 60mg. Repeat CT thorax showed disease progression with enlarged cavitating lesions. After 5 days of antibiotics, she received the 2nd dose of rituximab, azathioprine was added 2 days later.

Conclusions
Further complications ensued:

1. Sputum samples persistently grew staphylococcus aureus, she was treated with IV flucloxacillin for 1 month.
2. A new leucopenia, likely due to azathioprine, so this medication was withheld.
3. A new onset stridor; repeat bronchoscopy showed improved mucosal inflammation but there was significant narrowing of the right main and left main bronchi due to scar tissue.
4. Serum Beta-D-glucan, a non-specific marker for invasive fungal infection was positive, but fungal cultures on the BAL were negative.

She has been discharged on oral prednisolone and awaits review in clinic to decide on future therapy. She will be considered for argon laser treatment to the bronchial stenotic lesions by the respiratory service, depending on repeat PFTs and repeat bronchoscopy in the near future.

Figure



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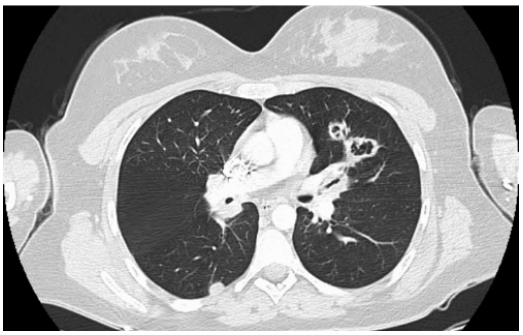

(20S105) CLINICAL CASE 4

'It's in the bones'

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Introduction
A 53 years old gentleman was referred to the rheumatology OPD. He complained of a 2-year history of constant lumbar spinal pain located over the T7/T8 area, which occasionally spreads inferiorly. He did not have any other significant arthralgia aside from intermittent left shoulder pain.
The back pain was worst at night, without morning stiffness, exercise made his pain worst. There was no fever, night sweats, and weight loss. Pain is relieved by acupuncture and NSAIDs.

Aims/Background
His background history included
• Sarcoidosis; pulmonary stage I with persistent adenopathy (now healed), diagnosed in 2009
• Malignant malenoma excised from his chest wall in 2005, local relapse in 2009

Method
Physical examination did not reveal any peripheral joint synovitis. There was no midline spinal tenderness, no sacroiliac joint tenderness, Schober's was 22cm. There was mildly reduced lumbo-thoracic lateral flexion.
Results
A series of investigations were carried out as follows:
MRI of whole spine noted multiple spinal bone lesions throughout the entire spine; suspicious for multiple spinal metastases.
CTTAP performed 1 week later did not show evidence of a primary or metastatic disease. The previously demonstrated bone lesions on MRI were not confidently seen on CT.
In addition; tumour markers (CEA, CA 19-9, CA 125, B-HCG, PSA) were normal, the rest of his bloods were unremarkable aside from serum ACE of 85 (nr 8-52), vitamin D of 42 (nr>50), ASOT 225 (recent streptococcal throat infection, nr<200)
PET scan confirmed multiple avid bone lesions; diffuse splenic involvement but no evidence of any primary site. The differential included lymphoproliferative/myeloproliferative disorder and recurrence of malenoma.
CT guided biopsy of lesions in posterior left ilium was performed; however there was no lesion visible on CT. Histology showed a small focus of non-necrotizing granuloma suggestive of sarcoid.
Repeat MRI whole spine showed stable multifocal spinal lesions.
Conclusions
Overall, imaging and biopsy suggest bone sarcoid. The differential is metastatic cancer given patient's past history. The fact that his back pain is ongoing for 2 years, without constitutional symptoms was reassuring; we felt that malignancy was less likely.
We decided to organize one further biopsy to more concretely exclude cancer and to perform imaging surveillance.

(20S106) CLINICAL CASE 5

Novel use of tofacitinib for checkpoint inhibitor induced inflammatory arthritis

Author(s)
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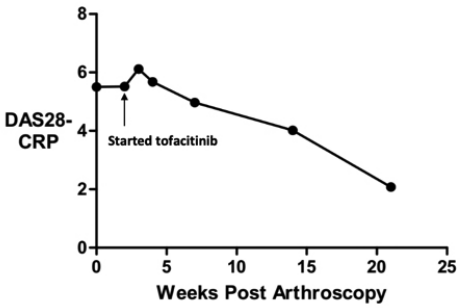
Introduction
Antibody mediated blocking of the T cell dominant immune check point inhibitor, Programmed cell Death protein-1 (PD-1), is a novel efficacious treatment for several malignancies including melanoma and lung cancer. However, compromising the PD-1 inhibitory pathway can cause immune-related adverse effects (irAEs) including inflammatory arthritis (IA). Our group has previously shown increased serum and synovium PD-1 levels in RA with less PD-Ligand 1 available for interaction. Thus, the PD-1 agonist pathway is down-regulated, suggesting agonistic PD-1 antibody-based therapies may be a novel RA treatment.
Aims/Background
A 56-year-old male was diagnosed with pulmonary adenocarcinoma and treated with the PD-1 inhibitor, Pembrolizumab. Following this, he developed a sero-negative inflammatory polyarthritis unresponsive to methotrexate and sulfasalazine.
Method
At arthroscopy, the patient had 4/28 tender and 6/28 swollen joints and a DAS28CRP of 5.5. Arthroscopy showed 70% synovitis, 60% vascularity and tortuous vessels. Histology revealed a hyperplastic synovium with CD3+ T cell infiltration. Flow cytometry with Simplified Presentations of Incredibly Complex Evaluations (SPICE) algorithm analysis of single cell synovial tissue suspension, demonstrated marked CD4+ T cell polyfunctionality with high expression of many pro-inflammatory cytokines (Figure 1).
Results
Based on the clinical, histological and flow cytometric data of T cell involvement, prednisolone and the Janus Kinase Inhibitor, tofacitinib were commenced at 2 weeks post arthroscopy. The patient had an excellent clinical response with disease remission (Figure 2). Repeat biopsy showed decreased synovitis (50%), vascularity (50%) and T Cell synovial infiltration. Preliminary flow cytometric analysis of synovial T cells at the time of the second arthroscopy, suggests downregulation of synovial CD4+ T cell expression of the pro-inflammatory cytokines TNF, IFN-gamma and GM-CSF.
Conclusions
Checkpoint inhibitors (CI) have dramatically improved cancer prognosis via T-cell pathway manipulation. But >80% of patients develop irAEs including life threatening hypophysitis and colitis. 1-7% of CI patients develop an IA. There are no randomised control trials to determine the best treatment for these patients. To our knowledge, this is the first CI-induced arthritis patient treated with tofacitinib. Given the good clinical response, we propose JAK inhibition as a novel therapeutic option in CI-induced IA.

Figure

Figure



(20S107) CLINICAL CASE 6

A Rare Presentation of Seronegative Spondyloarthropathy

Author(s)
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Introduction
Spondyloarthritis is a group of diseases with clinical, laboratory and genetic features in common. The most important of these is the association with human leukocyte antigen HLA-B27 in 90% of patients. The worldwide prevalence of SpA is estimated to be between 0.5% to 1.9%.
Spondyloarthritis affects both men and women but is most frequently seen in men. The disease usually starts between the second and the fourth decades of life and is rarely found after the age of 40 years
Enthesopathy is a prominent clinical feature in patients with spondyloarthritis, usually affecting the axial skeleton and peripheral and sacroiliac joints

Aims/Background
A 50 year old Irish lady was seen in the rheumatology clinic with 8 months history of shoulders, right elbow, hips and lower back pain. She has neck and lower back stiffness in the morning more than 1 hour. Her lower back pain sounds mechanical. She denies any waking up at night or buttock pain.
Her past medical history includes hypertension.
On examination revealed reduced range of movement in both shoulders with abduction less than 90 degrees bilaterally as well as some hip girdle stiffness. There was some possible of mild swelling in the knees. Her Modified Schober Test was 3cm and Faber's Test was negative.

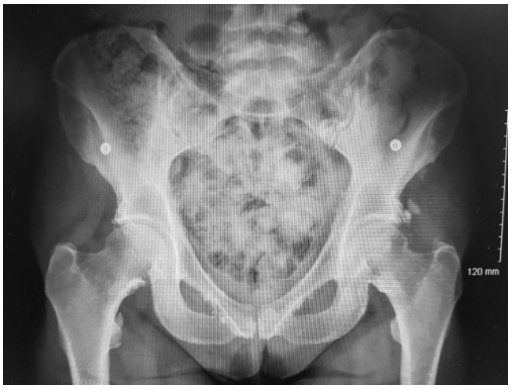
Method
Initial investigations
Hb 13.9 Urea 6.0
WCC 21.9 Creatinine 89
Platelet 485 eGFR 62
ESR 48 LFTs Normal
Ferritin 204 CRP 42
C.Calcium 2.32 RF Negative
Immunoglobulins Normal Anti-CCP Negative HLA-B27 Negative

Results
Radiology
Pelvic X-Ray shows multiple enthesopathic changes suggestive of chronic arthropathy.
MRI Whole Body revealed features of inflammatory arthritis in the shoulders, knees and ankles as well as active enthesitis at the hamstrings, with possible fatty Romanus lesions and syndesmophytes. The sacroiliac joints were normal.
This patient is subsequently diagnosed with seronegative peripheral spondyloarthritis with possible axial involvement.
She was then commenced on Etanercept 50mg subcutaneously weekly.

Clinical Progress
This lady was seen in the clinic again after 4 months. She reported 90% improvement and she was able to drive her car. Her inflammatory markers have nearly normalised.

Conclusions
This is a very unusual presentation of peripheral spondyloarthropathy with unusual findings on the x-rays with multiple calcification and enthesopathic changes in the joints involved.

Figure



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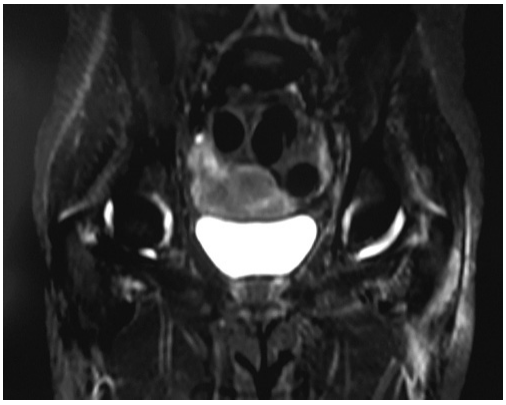

(20S108) CLINICAL CASE 7

Interstitial Lung Disease with autoimmune features: Hypersensitivity Pneumonitis – a rare complication of treatment with cyclophosphamide.

Author(s)
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Introduction
Interstitial lung disease (ILD) is a condition linked with a number of pathologies. Treatment for ILD with autoimmune features includes cyclophosphamide. We present a patient who received treatment with pulsed cyclophosphamide therapy and subsequently deteriorated.

Method
A 60 year old man was given the diagnosis of ILD with autoimmune features. He was noted to have a positive anti-chromatin > 8, positive homogenous ANA and anti-CCP of 28.
Throughout the course of the year, he had a rapidly progressive disease course despite maintenance steroid therapy and was established on LTOT, his HRCT confirmed progressive bilateral extensive fibrosis.
He was subsequently discussed at the ILD MDT. Six pulsed sessions of cyclophosphamide therapy was the recommended outcome.
He received his first session of therapy with standard protocol followed for the procedure. No signs of infections were noted. The following day, he presented to hospital hypoxic and pyrexic despite

use of his ambulatory LTOT.
He was treated with a higher dose of steroids, high flow nasal specs (HFNS), broad spectrum IV antibiotics and diuretics. Despite this, he necessitated admission to HDU for CPAP. In HDU he received IV methylprednisolone after an appropriate course of antibiotics. He improved and was weaned to HFNS with high dose oral steroids.
At ward level care, he was never weaned successfully from HFNS. Approximately one month after his initial presentation, he developed an acute- on-chronic hypoxia. His CXR showed rapidly progressive bilateral extensive nodular infiltrates with PJP and cultures negative to date. He unfortunately deteriorated and did not survive.

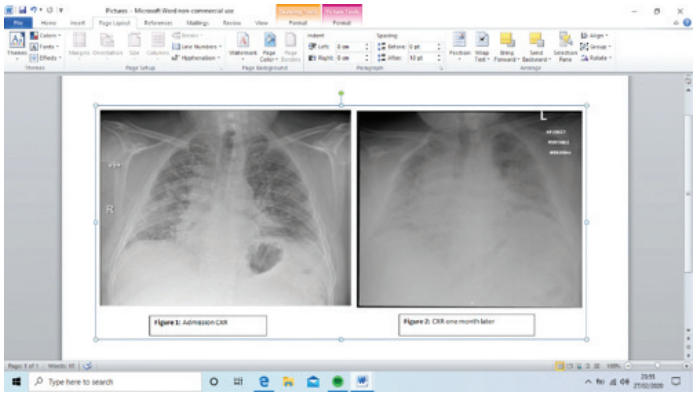
Results

Pneumotox lists less than 10 cases of pulmonary toxicity as a result of cyclophosphamide therapy, one manifestation is an acute hypersensitivity pneumonitis. Literature is sparse, but steroids seem to be the predominant management in this setting. Case reports suggest that the inflammatory stimulus of cyclophosphamide could last for months. The chronology of events in this case suggests a temporal relationship between the use of cyclophosphamide therapy and the hypersensitivity pneumonitis that led to the patient's deterioration.

Conclusions

This case highlights that pulmonary toxicity is a rare but important complication of cyclophosphamide that could be prolonged for months after discontinuation of the drug.

Figure



(20S109) CLINICAL CASE 8

The diagnostic challenge of GCA: A case of isolated vertebral arteritis in a healthy woman

Author(s)

Alwin Sebastian, Diana Prieto-Pena, Abdul Kayani, Alessandro Tomelleri, Bhaskar Dasgupta

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Introduction

Giant cell arteritis (GCA) is a vasculitis affects the cranial and extra-cranial arteries. Extracranial involvement in GCA is not uncommon, however, diagnosis remains challenging. Vertebral arteritis (VA), a rare finding in GCA, has been reported in few case reports and case series associated with stroke. In a retrospective study, 18F-FDG PET-CT showed 80% sensitivity and 100% specificity in identifying VA (Nielson BD et al. 2018). Here we report, an isolated case of VA presented without any neurological deficit.

Aims/Background

A 76 years old previously well woman was referred by GP to GCA fast track clinic for assessment. She presented with few days history of pressure like headache started on the left side and moved to the right, associated with scalp tenderness, mild neck pain and polymyalgic symptoms. She had constitutional symptoms including weight loss, night sweats and felt hot and cold. she denied any jaw, tongue or visual symptoms. Clinical examination was unremarkable including a neurological examination. She had raised inflammatory markers (CRP-100 mg/L, ESR- 38 mm/hour). Full blood count, renal, liver and bone profiles were within normal limit. Her pre-test probability score was 17. She had a normal temporal and axillary artery ultrasound (US). A single dose of Prednisolone 40 mg from her GP prior to referral gave her an excellent symptomatic relief. 18F-FDG PET-CT scan (Figure 1) showed bilateral vertebral arteritis. CT angiogram of carotids and aortic arch was normal. She has commenced on Prednisolone 40 mg daily with a tapering regimen and Leflunomide 10 mg daily was added.

Method

A case report

Results

She had dramatic clinical improvement and her inflammatory markers returned to normal.

Conclusions

VA should be considered with the presence of classical symptoms of GCA, good response to glucocorticoids, a high clinical probability and more importantly a negative cranial and axillary artery US. In our patient, our novel probability-based diagnostic algorithm allowed us to stratify the patient into a high category and paved the way to have an additional test, leading to a prompt diagnosis. PET-CT is a useful and reliable tool in diagnosing extra-cranial vasculitis such as VA.

Figure

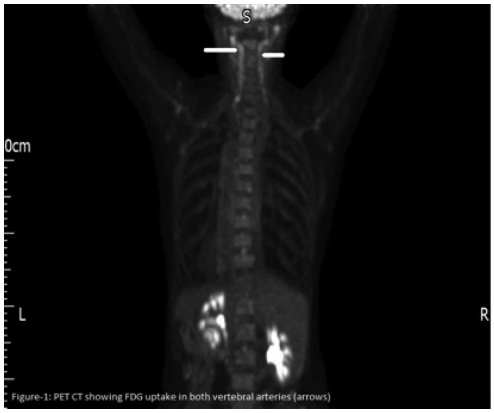


Figure-1: PET CT showing FDG uptake in both vertebral arteries (arrows)

Speakers - Autumn Meeting 2020

Dr John Stack, Consultant Rheumatologist and General Physician at the Mater Misericordiae University Hospital, Dublin.

Norah Campbell, associate professor of Marketing, Director of Undergraduate Teaching and Learning in Trinity Business School.

Dr Nicola Ambrose, Consultant Rheumatologist, Blackrock Clinic, Co Dublin

Dr Jean Fletcher, Associate Professor in Translational Immunology, TCD

(20S110) CLINICAL CASE 9

An unusual presentation of EGPA.

Author(s)
Dr. Shawn Chavrimootoo, Dr. Patrick Mulkerrin.
Department(s)/Institutions
Our Lady's Hospital Navan.

Introduction
1st presentation- JC 53yo male presents with 2 week history of epigastric pain. He has a background of worsening asthma since he was 40. He was on a course of steroids and antibiotics for infective exacerbation of asthma. Bloods showed high platelets, CRP. Urine dipstick was clear. He was seen by surgeons who arranged a CT abdo/pelvis and ultrasound. He was discharged on a tentative diagnosis of gastroenteritis.
2nd presentation- JC represents with periumbilical abdominal pain. He has right iliac fossa tenderness. Bloods show raised white cells and eosinophils also. A CT abdo pelvis was suspicious for appendicitis. He was transferred to theatre where he had an emergency appendicectomy and was discharged shortly after.
3rd presentation- Ten days post-operatively JC returns to ED with central abdominal pain. White cells, eosinophils, CRP remain raised. A repeat CT abdo/pelvis is sought querying post operative collection and sepsis- NAD. Histology of appendix returns with EGPA a consideration. Rheumatology opinion is sought.

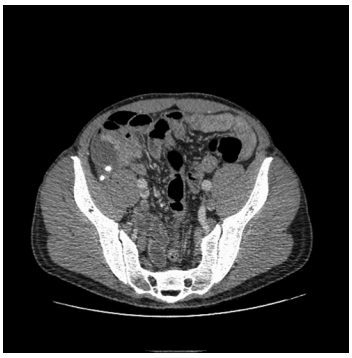
Aims/Background
The aims of this case are to highlight potential rheumatological causes of abdominal pain. GI vasculitis can result in perforation/mortality if not diagnosed/managed correctly.

Method
Diagnosis of EGPA is made.There is an absence of ENT, dermatological or joint involvement. ANCA result is negative, CXR shows chronic obstructive changes, repeat urine dipstick is clear. He is started on high dose steroids (1mg/kg) with bone protection and PPI cover. His five factor score is 2 therefore cyclophosphamide is added for 6 months. There is a massive improvement in symptoms, eosinophilia and inflammatory markers. He is switched to azathioprine as a steroid sparing agent after 6 months.

Results
1st presentation (on steroids)- FBC normal apart from platelets 459, CRP is 46.
2nd presentation(off steroids)- WCC 23.1, Eosinophils 13.2, CRP 32.3. CT abdo pelvis(pic2)- two calcifications which are apparent appendicoliths suspicious for appendicitis.
3rd presentation (post-operative)- WCC 23.7, eosinophils- 15.1, CRP-32.1. Histology(pic2)- necrotizing vasculitis of mural arteries and focal mesoappendiceal arteries, eosinophils are prominent. No background acute appendicits.

Conclusions
In this case we have a man with Eosinophilic Granulomatosis with Polyangitis confines to GI system. There was initial masking of the eosinophilia with steroids. Histological diagnosis was confirmed after emergency surgery.

Figure



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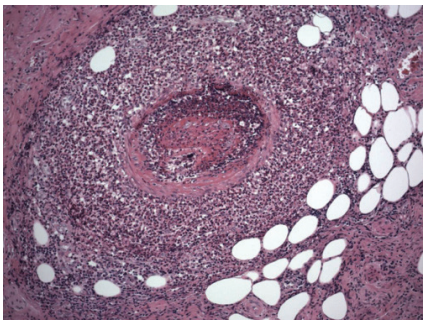

(20S111) CLINICAL CASE 10

No easy answer…Get your thinking CAPS on !

Author(s)
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Introduction
Systemic lupus erythematous (SLE) is a multisystem autoimmune disease with variable clinical course. Catastrophic antiphospholipid syndrome (CAPS) is a rare disease with high mortality characterized by widespread intravascular thrombosis resulting in multiorgan failure.

Aims/Background
A 60 year old male with a background history of SLE since 1996 manifested by lupus nephritis (biopsy proven), autoimmune haemolytic anaemia, interstitial lung disease, pyoderma gangrenosum, and antiphosholipid syndrome, was admitted electively for lumbar puncture for possible diagnosis of normal pressure hydrocephalus. Medications on admission were prednisolone 5mg, hydroxychloroquine, losartan, lercanidipine, bisoprolol, spironolactone, warfarin, omeprazole and calcichew. His immunosuppression (mycophenolate mofetil) had been held three weeks prior to this admission due to pneumonia.

Method
During his hospital stay, lumbar puncture was performed with normal CSF. He developed lupus flare with major organ involvement (Lung: acute on top of chronic pneumonitis and diffuse alveolar hemorrhage, kidney: worsening renal function and proteinuria), and he was treated with pulse IV methylprednisolone (500mg IV 3

days) and recommencement of mycophenolate. He had high titres of antibodies to beta-2-glycoprotein, anticardiolipin antibodies and thrombocytopenia (platelets 34 109/L), low complements C3 0.77 g/l, C4 0.08 g/l and raised inflammatory markers (ESR124mm/hr, CRP 150mg/l)

He later developed left distal upper limb weakness with multiple lacunar infarcts proven by MRI. Transoesophageal echocardiography showed changes suggestive of Libman–Sacks endocarditis with vegetations on mitral valve, and multiple blood cultures were negative. He also developed right foot purple discoloration with no ulcer.

Results

He was diagnosed with a catastrophic secondary antiphospholipid syndrome with multiple organ involvement (cardiac, renal insufficiency, embolic to CNS and skin).

He was immediately treated medically with anticoagulation, increased prednisolone 40mg and IVIG. Subsequent ECHO after 4 weeks showed no evidence of the sterile vegetation.

Subsequently his blood parameters showed improvement in hemoglobin, platelet, and INR with stabilization of renal function along with clinical improvement.

Conclusions

Improvement in survival depends on early recognition and management of CAPS. In the present case lupus flareup, immunosuppression and anticoagulation interruption were the main precipitating factors leading to catastrophic presentation.

(20S113) CLINICAL CASE 11

A microangiopahy - Rarely seen, Rarely heard

Author(s)

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Department(s)/Institutions

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Introduction

A 40-year-old Caucasian man, with no medical history, attended the emergency department with acute visual loss with associated retrobulbar pain and vertigo. Twenty-four hours from onset, his pain and vertigo subsided but his visual loss persisted. On further questioning, he described tinnitus and hearing loss involving both ears, which started 9 months earlier and had persisted.

The MRI of his brain showed lesions consistent with infractions. He had no traditional cardiovascular risk factors – no hypertension, dyslipidaemia or diabetes mellitus. He had an ECHO and carotid artery Doppler ultrasound, which were normal. He had a normal 24 hour cardiac holter monitor. He was treated for stroke, commenced on aspirin and clopidogrel and was discharged home.

Three months later, he had recurrence of his symptoms. He had visual colour disturbance, vertigo and, this time, associated ataxia and vomiting. Again, he was admitted to hospital and a repeat MRI of his brain showed progressive vascular changes, despite 3 months of dual antiplatelet therapy.

MRI 1, 2

His peripheral neurological examination was unremarkable, cardiovascular exam normal with all peripheral pulses were present with no vascular bruits. Fundoscopy, performed by an ophthalmologist, showed bilateral branch retinal artery occlusions. He and had audiology which identified bilateral sensorineural hearing loss.

His routine blood tests were unremarkable and other labs including C3/C4, ANA, ENA, dsDNA, antiphospholipid antibodies and ANCA were all negative. He had a normal trans-oesophageal ECHO. His CSF showed a high protein.

A diagnosis of Susac syndrome was made. His clinical presentation was consistent with the typical Susac triad and his MRI findings supported this diagnosis. He commenced prednisolone – initially 40mg dose and started mycophenolate. He successfully tapered and discontinued prednisolone over a 10-month period and remains on mycopheonolate with no disease flares.

Conclusions

Susac syndrome is a rare disease, first described by John O. Susac in 1979. It is an autoimmune endotheliopathy involving the arterioles of the brain, retina and cochlea. Partial or complete occlusion of the vessels can occur, resulting in temporary dysfunction or permanent damage to these organs.

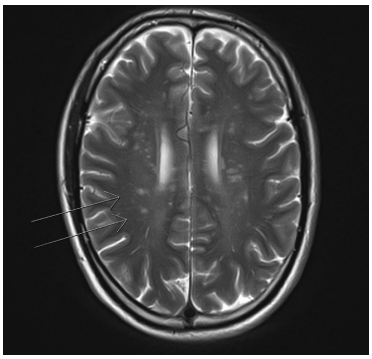
The 3 main clinical features of this syndrome are encephalopathy, vision loss due to branch retinal artery occlusions and sensorineural hearing loss.

MRI imaging of the Brain typically shows 'snowball' lesions in the corpus callosum.

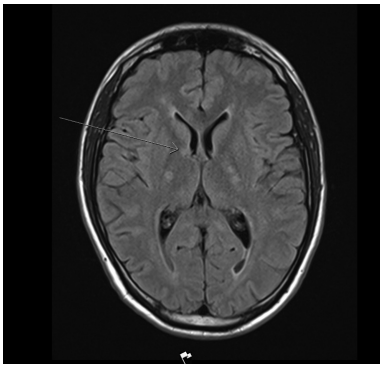
A diagnosis of Susac syndrome is made based on the presence of at least 2 components of the clinical triad and classic MRI findings.

Due to rarity of the disease, there have been no randomised controlled trials or prospective treatment studies. Based on cumulative clinical experience and cohort studies, all patients are commenced on prednisolone 40-60mg at diagnosis with addition of mycophenolate, azathioprine or tacrolimus. Cyclophosphamide, rituximab and IVIG can be considered in severe cases.

Figure



Figure



(20S114) CLINICAL CASE 12

Parechovirus in a Pathologist

Author(s)
Dr Claire Masih, Dr Andrew Cairns, Dr Michael Dolaghan (Department of rheumatology) Dr Robin Thompson (Department of radiology).
Department(s)/Institutions
Musgrave Park Hospital
Introduction
A 42 year old pathologist presented with a 1 week history of muscle pain and subjective weakness. CK level on 2 occasions was >3000.
The patient was systemically well with no past medical history, medication or foreign travel. He had 1 day history of shivering with no recorded pyrexia. He reported pain in his proximal muscles and neck and subjective muscle weakness and lack of finger dexterity with no objective findings.
Autoantibody panel and inflammatory markers were performed which were normal. Full blood count with differential white cell count including eosinophils was normal. There was a modest rise in transaminases. Myositis panel results are awaited. Full viral screen was positive for parechovirus with titre of 30 on several samples. MRI proximal musculature showed increased fluid signal in the perifascial region of both thighs primarily involving the hamstrings, not definitive for myositis but suggestive of fasciitis.
Conclusions
Parechovirus is a picornavirus, often causing mild gastrointestinal or respiratory illness but has been associated with epidemic myalgia and myositis during outbreaks of parechovirus in a Japanese population. The patient improved spontaneously with CK reduced to 187 and improved symptoms after 1 week. We expect a good outcome and will follow up with repeat MRI and clinical assessment/blood tests in a number of weeks.

(20A119) CLINICAL CASE 13

A case of eosinophilia granulomatosis with polyangitis causing appendicitis

Author(s)
Dr Patrick Mulkerrin, Dr Shawn Chavrimootoo.
Department(s)/Institutions
Our Ladys Hospital Navan
Introduction
JC presents to navan Ed with 2 week hx of abdominal pain. He presents to ED with periumbilical abdominal pain. This came on suddenly 2 weeks previously and is sharp and non radiating. He is a farmer who doesn't smoke. Vitals are within normal parameters. GI exam reveals RIF tenderness. Investigations including bloods,CXR, CT abdo/pelvis are arranged.
Aims/Background
Poorly controlled asthma(15 year hx). He is on Symbicort for this. He has no known allergies.
Method
CT abdo pelvis are discussed in results section. He is transferred to Drogheda where he has an emergency appendicectomy and is later discharged. He then represents to ED and is transferred back as there is concerns around a postoperative collection. The histology from the appendix result(discussed below) seems to suggest a GI vasculitis. He is started on high dose steroids and obtains symptomatic relief instantaneously. As his Five factor score is 2 he is started on cyclophosphamide for 6 months. He is currently maintained on methotrexate after an intolerance to azathioprine after a prolonged steroid course.
Results
WCC- 23.1, Eosinophils 13.33, CRP 32, U&E, LFT, amylase, lactate-normal. (first presentation).
Wcc 23.1,eosinophils 15.1, CRP 32.6. (representation)
CT Abdo pelvis initially showed appenicolith possibly indicative of appendicitis.
Repeat CT abdo pelvis- no evidence of collection.
Histology of appendix- necrotising vasculitis of mural arteries and focal mesoappendieceal arteries. Several vessels show a prominent of eosinophils within infiltrate. No background acute appendicitis.. Given prominent eosinophil count in blood film Churg-Strauss is a consideration.
WCC 9.0 Eosinophils 9.0 CRP1.00- Most recent bloods
Conclusions
Here we have a patient who was initially treated with emergency surgery. The histology from his removed appendix revealed a new diagnosis of EGPA. He has had an excellent response to high dose steroids, cyclophosphamide and methotrexate.

(20A121) CLINICAL CASE 14

Case report: The anti-PM/Scl 75/100 phenotype

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Introduction
Autoantibodies to PM/Scl 75/100 identify patients with features of myositis and systemic sclerosis. These antibodies are not commonly found, but are notable as they may predict likely clinical course and guide therapeutic decisions.
Aims/Background
A 19-year-old caucasian male presented with a 6-month history of inflammatory back pain and peripheral arthritis. He had had mild Raynaud's phenomenon for several years. Examination revealed synovitis of right knee and several PIP joints. Investigations showed a strongly positive ANA, negative dsDNA and ENA profile, CRP 22, CK 333, MRI spine/sacroiliac joints normal. Treatment for an undifferentiated connective tissue disease with prednisolone and methotrexate was initiated.
Method
He was lost to follow-up for 11 months and stopped all medication. His clinical phenotype had progressed significantly with weight loss, dysphagia, proximal weakness and worsening Raynaud's. There was digital ulceration, sclerodactyly, tight oral aperture, faint erythematous patchy rash, calcinosis cutis and normal lung auscultation. Investigations revealed persistent strong ANA and new positivity for dsDNA, SSA, PM/Scl 75, PM/Scl 100 antibodies, total IgG 28g/L, hypocomplementaemia, and serum CK 3008 U/L. He was treated with prednisolone, mycophenolate mofetil and Tadalafil. Arthralgia, dysphagia, muscle weakness and peripheral circulation resolved with healing of digital ulcers and CK fell to 96 over 6 months. Plaques of calcinosis cutis progressed, predominantly overlying bony prominences including elbows, anterior and posterior pelvis and knees. Due to the multiple autoantibodies and hypergammaglobulinaemia he received Rituximab 2g with a four-

month interval retreatment planned. Colchicine and Diazepam were added for calcinosis cutis.

Results

De Lorenzo reported features in 41 patients with PM/Scl 75/100 antibodies. She found a very high prevalence (>90%) of muscle involvement, early sclerodactyly (66%) and late development of ILD (>60%). Although ILD was not present in our case, susceptibility to this manifestation and evidence of a strong B cell drive prompted early Rituximab treatment. Extensive calcinosis cutis is a particular feature of our case, found more commonly in the cases reported by de Lorenzo than in anti-synthetase and dermatomyositis cases.

Conclusions

This case is one of rapidly progressive evolution into an overlap scleroderma/ myositis disorder with PM/Scl antibodies. It highlights the importance of regular clinical review and diagnostic suspicion.

(20A131) CLINICAL CASE 15

Weight Loss and Oesophageal Dysmotility as an Unusual Presenting Feature of Systemic Lupus Erythematosus in an Elderly Male

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Introduction

73 years old gentleman was admitted with collapse, hypotension, pyrexia and erythematous scaly rash on chest, abdomen, back and limbs. He lost 10kg in the past few months. There was no evidence of peripheral synovitis, Raynaud's or mouth ulcers.

Aims/Background

He is an ex-smoker and has a background of hypertension and Barrett's oesophagus.

Method

Investigations showed Haemoglobin of 78 with low MCV, lymphopenia and low iron levels. CRP and ESR were mildly raised at 35 and 29 respectively. CT thorax, abdomen and pelvis showed prominent heart with mild bilateral pleural effusions, minimal pericardial effusion and osteopenia. Patient continued to spike temperatures despite antibiotics, oral intake became poor and swallowing problems started to become more obvious from day 14 after admission. Video fluoroscopy suggested oesophageal dysmotility and silent aspirations with all consistencies.

Results

Further work-up showed positive autoantibody screen with positive Anti double-stranded DNA at a titre of 186, positive Anti Ro, Anti La and Anticardiolipin IgG. C3 was low and urine PCR was increased. Skin biopsy results showed lichenoid dermatosis consistent with SLE. Patient was diagnosed as Systemic Lupus Erythematosus and was commenced on steroids initially and Hydroxychloroquine and Mycophenolate were started later on. PEG was inserted due to swallowing problems. Patient showed significant improvement and started to eat and drink in 3 months' time. ESR and CRP normalized and anti ds-dna level fell down to 19. PEG tube was removed, patient started to gain weight and active features of SLE were resolved.

Conclusions

This is an unusual presentation of Systemic Lupus Erythematosus due to patient's age, gender and presenting symptoms of weight-loss and oesophageal dysmotility. Patients presenting with oesophageal dysmotility with no obvious cause should be screened for autoimmune conditions especially if they have features suggesting multisystem involvement as in this case there was evidence of serositis, skin rash and anaemia.

(20A136) CLINICAL CASE 16

Involuntary 'dance' in an elderly lady with a history of recurrent miscarriage -– a case report

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Introduction

A 72 year-old lady with underlying breast cancer diagnosed 2009 (lumpectomy and chemotherapy completed) presented with sudden onset of hyperkinetic choreoform movements of both hands, torso and orofacial muscle (left side worse than the right) for 3 months prior to admission in 2019. This was associated with emotional lability, mouth ulcers and significant weight loss. She denied arthralgia, myalgia, photosensitivity, alopecia and sicca symptoms. On further history, she had history of two miscarriage at 11 weeks with two live births. Otherwise, she had intact memory with no muscle weakness or numbness. There was no history of stroke or cardiovascular disease. No family history of autoimmune disease or movement disorder. She is an ex- smoker, stopped for 10 years and there is no history of illicit drug usage.

Clinical examination revealed stable vital signs. She was noted to have chorea movement involving all extremities and orofacial muscle. Her muscle power were full with normal reflexes. Cardiovascular,respiratory and abdominal system examination were unremarkable.

Her serum ANA was positive, homogenous with serum B2Glycoprotein Ig G was 199.0 U/ml (pre-treatment) and repeated at 12 weeks also positive. Lupus anticoagulant was positive with normal CRP and full blood count.

Her DsDNA, ENA, complements level, ANCA, immunoglobulin, fibrinogen, APTT level, renal and liver functions test and were all within normal range. Her serum anti-NMDA antibody was negative. CSF was normal with normal protein and sugar level, no cells and negative for staining and culture. Urinalysis was negative. Her MRI brain was normal and CT TAP showed no malignancy or metastasis. EEG showed no definitive epileptiform abnormalities or seizures activity. Her mammogram showed normal results.

She was co-managed by rheumatology and neurology team. Intravenous methylprednisolone 500mg daily was given for 3 days followed by prednisolone as maintenance therapy. She had significant clinical improvement overtime with no abnormal movement during review 4 months later. Repeated Ig G B2Glycoprotein was 32U/ml (post therapy). Current medication was hydroxychloroquine, tapering prednisolone dose, aspirin, calcium and vitamin D supplement.

Conclusions

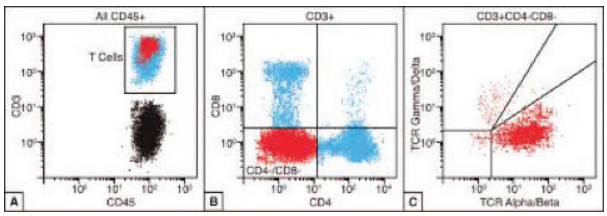
Chorea is an unusual but recognised clinical manifestation of antiphospholipid syndrome and obstetric history was important in making the diagnosis.

Figure

Diagnosis	Required criteria	Primary criteria	Secondary criteria
	1. Chronic (>6 mo) **lymphadenopathy and/or splenomegaly** (nonmalignant, noninfectious) 2. Elevated DNT cells ($CD3^+TCR\alpha\beta^+CD4^-CD8^-$) ≥1.5% of total lymphocytes or >2.5% $CD3^+$ cells in setting of normal or elevated lymphocyte counts	1. Defective lymphocyte apoptosis (in 2 assays) 2. Mutation in *FAS*, *FASLG*, or *CASP10* (somatic or germ line)	1. Elevated plasma marker (sFASL, IL-10, IL-18, or vitamin B12) 2. Typical immunohistological findings 3. **Autoimmune cytopenias** (hemolytic anemia, thrombocytopenia, or neutropenia) AND elevated IgG (polyclonal hypergammaglobulinemia) 4. Family hx of nonmalignant lymphoproliferation +/− autoimmunity
Definitive	Required criteria (2 of 2)	Primary criteria (1 of 2)	—
Probable	Required criteria (2 of 2)	—	Secondary criteria (1 of 4)

*Abbreviations: DNT, double negative cells; hx, history; sFASL, soluble FAS ligand. Boldface indicates two predominant features of the syndrome.

Figure



(20A137) CLINICAL CASE 17

Pulmonary limited MPO –ANCA Microscopic Polyangiitis in a gentleman and his response to Rituximab – a case report

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Introduction
A 53 year-old gentleman, ex-smoker (stopped 10 years ago) was initially treated as chronic pulmonary fibrosis with emphysema when he presented to respiratory team 2017 with complaint of productive cough with whitish sputum, breathlessness and reduced effort tolerance. He also complained of polyarthralgia and significant weight loss. He denied any ENT symptom, abnormal sensation over the limbs or prolonged fever. No sicca symptoms, oral ulcer, abdominal discomfort or parotid gland enlargement.

Aims/Background
Clinical examination revealed normal vital signs and oxygen saturation. Peripheral examination noted presence of clubbing with no cyanosis noted. The lung was clear on auscultation with no loud P2. There was no active synovitis or skin rash noted. Cardiovascular, abdomen and neurological examinations were unremarkable.

Method
Serum ANA, DsDNA, ENA and RF were all negative. His serum PR3 and complement level were within normal range. Urinalysis was negative.
ESR and C-reactive protein were rated at 16 /hr and 11 mg/L respectively. His MPO level was initially 134 IU/ml at diagnosis and reduced following treatment with Rituximab and have remained low/normal (latest result 8.3 IU/ml June 2018). His eosinophil count is normal.
CXR showed extensive bibasal ground glass opacity and lung volumes appeared decreased.
Initial HRCT of the lung (2017) showed radiographic appearance of interstitial lung disease consistent with a possible UIP type pattern. Repeat scan (2018) showed stable and similar appearance of established bilateral changes of pulmonary fibrosis with no significant changes.

Results
His initial DLCO was 32 % predicted at diagnosis (pre-Rituximab) and 44% predicted following the first 2 infusions of Rituximab measured early 2019. He was given a total of 4 full treatment course (day 1 and day 15, 1g on each infusion) started November 2017. During this time his prednisolone has been between 20mg and 40mg per day, currently maintained on 20mg daily. His condition is stable with Rituximab as induction and maintenance therapy.

Conclusions
High titer of MPO is associated with poor outcome and pulmonary limited MPO vasculitis can be life threatening if not treated with immunosuppression therapy early and it is suggest that every patient with idiopathic pulmonary fibrosis should be screened for MPO vasculitis.

(20A151) CLINICAL CASE 18

Straight from the cradle: Autoimmune lymphoproliferative syndrome (ALPS)

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Introduction
Autoimmune lymphoproliferative syndrome (ALPS) is a rare inherited disorder of immune function due to defective lymphocyte apoptosis, characterised by lymphoproliferation, autoimmune disease, and sometimes lymphoid malignancies. Most commonly, pathogenesis is linked to mutations affecting the FAS signalling pathway1. Here, we describe the case of a 29-year-old man with a history of recurrent infections since birth, autoimmune hepatitis, positive direct Coombs, hepatosplenomegaly, interstitial lung disease, diffuse mediastinal lymphadenopathy, and cutaneous vasculitis, who presented with confirmed influenza B infection.

Aims/Background
Case Report

Method
After developing jaundice as a two-year-old boy, our patient had a liver biopsy and was diagnosed with autoimmune hepatitis. His childhood was marked by immunosuppressive therapy, recurrent infections, and a nasal basal cell carcinoma. More recently, an incidental right lung nodule was noted on liver ultrasound. CT thorax showed bilateral consolidative nodules and extensive lymphadenopathy, but subsequent work-up ruled out malignancy or infection. Interval changes on follow-up CT thorax led to video-assisted thoracoscopic surgery (VATS) for nodule resection and lymph node biopsy. In combination with a reduced diffusion capacity and restrictive pattern on pulmonary function testing, he was diagnosed with non-specific interstitial pneumonia. At that time, the patient was also found to be direct Coombs positive. When the patient's steroid dose was reduced in preparation for MMR vaccination, he experienced worsening of a 6-month-old rash covering mostly his back. A skin biopsy showed a mixed dermal infiltrate in a pattern consistent with leukocytoclastic vasculitis. On this presentation, the man was wheezy, breathless, and his green sputum had recently become blood-streaked in keeping with influenza B infection. In light of his complex and extensive past medical history, flow cytometry of peripheral blood and ALPS-specific mutational analysis were carried out prior to his diagnosis with ALPS.

Results
Images will be referenced:
Diagnostic criteria for ALPS [Cheng & Andersen (2012)]
Characteristic flow cytometric findings [Matson & Yang (2020)]
Conclusions
ALPS, whose true incidence unknown, is a rare genetic dysregulation of immune function that develops early in life with great variability in phenotype. It is characterised by heterozygous mutations within the FAS signalling pathway, leading to lymphoproliferation, different forms of autoimmune disease, and sometimes lymphoid malignancy2.

(20A153) CLINICAL CASE 19

A Case report on Reactive Arthritis secondary to Intravesical BCG treatment of Bladder Ca

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Introduction
Intravesical instillation of BCG has been used in the treatment of intermediate and high-grade bladder carcinoma in situ. Osteoarticular side effects including reactive arthritis, following BCG treatment have been rarely reported.
Aims/Background
Case report on a patient with reactive arthritis post intravesical BCG therapy.
Method
58 year male presented with a three week history of bilateral eye pain and blurred vision, swollen painful large joints including left knee, ankle, wrist and elbow associated with morning stiffness for about 45 minutes and significant restriction in mobility. His past medical history was significant for Bladder cancer for which he was on Intravasical BCG treatment. Last BCG was four weeks ago. Eye symptoms started 1 week after the BCG treatment followed by arthralgia initially involving the left knee. Arthralgia progressed to involve the ankles, left wrist and left elbow. He was admitted to Wexford General Hospital. The case was discussed with our rheumatology department in University hospital Waterford after his arthralgia got pregressively worse. Prednisolone was started considering the diagnosis of reactive arthritis secondary to intravasical BCG treatment. The symptoms improved but recurred and rapidly deteriorated after Prednisolone was tapered too quickly. This time he presented to Emergency department in University hospital Waterford and was admitted under medical team.
On examination: He had conjunctivitis in both eyes. There was left knee effusion. Movements were significantly restricted at left knee. There was sinovitis at left wrist.
Lab investigations showed high inflammatory markers with CRP of 82, Normal Kidney and Liver functions, Auto antibodies were negative and Rheumatoid Factor was normal. Joint fluid aspirate ruled out septic arthritis and crystal arthropathy. X rays of left knee was normal.
Left knee was injected with Depomedrone and Lignocaine. Patient was discharged on Prednisolone 30 mg OD in reducing dose with follow up in Rheumatology clinic.
Results
He improved on Prednisolone but was difficult to taper off Prednisolone completely.
Methotrexate was started and maintained with complete resolution of symptoms.
Conclusions
Although rare, reactive arthritis secondary to treatment with intravesical BCG should be considered in patients presenting with arthralgia and arthritis. Most patients respond favorably to NSAIDs and corticosteroids used alone or in combination. Rarely, as in our patient, immunosuppressants like methotrexate are required.

(21SP101) CLINICAL CASE 20

Relapsing polychondritis associated encephalitis: a case report

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Introduction
Relapsing polychondritis (RP) is an autoimmune disease characterised by progressive inflammation of cartilaginous structures in multiple organs. Central nervous system (CNS) manifestation in RP is rare, but cases of limbic encephalitis have been reported.
Aims/Background
We present a case of a man who was diagnosed with RP associated encephalitis after consulting a number of different specialties for a variety of symptoms over many years.
Method
A 46 year-old Caucasian man presented with status epilepticus. He was intubated and ventilated on arrival, and treated initially as a suspected CNS infection with antibiotics and acyclovir. Upon reviewing his medical records, he had complained of bilateral pinna erythema and swelling, memory problems, emotional lability, bilateral ophthalmalgia, and joint pains over the last 6 years, and had consulted a number of specialties, but no unifying diagnosis was made.
Blood tests showed mildly raised inflammatory markers. T2 weighted brain magnetic resonance imaging (MRI) revealed abnormal hyperintensity involving the white matter and deep grey nuclei consistent with non-herpetic viral and less likely autoimmune encephalitis. Cerebrospinal fluid analysis revealed mildly raised protein at 1.08 g/L but negative for bacteriology and virology. All other investigations including autoimmune screen and computed tomography imaging were negative. A brain biopsy revealed inflammation of the leptomeninges cortex and white matter with cortical micro-infarcts.
Despite acyclovir therapy, his repeat MRI 2 weeks later showed evidence of progression. Rheumatology team was consulted and a diagnosis of RP associated encephalitis was made.
Results
He received 3 days of pulsed IV methylprednisolone, and commenced on oral prednisolone thereafter. He also received 5 days of intravenous immunoglobulin (IVIg). On follow-up review his auricular swelling and joint pains had disappeared, and repeat MRI demonstrated resolution of left periventricular white matter enhancement. Unfortunately he still had ongoing memory problems albeit not as severe.
Conclusions
CNS involvement in RP is rare and can be associated with severe morbidity or mortality. To date, there are no prospective randomised controlled trials studying treatments for RP with CNS manifestations, and efficacy of therapies are based on case reports. Clinicians should have a low index of suspicion of possible neurological involvement in patients with an autoimmune disease.

(21SP102) CLINICAL CASE 21

See No Evil, Hear No Evil, Speak No Evil: Giant Cell Arteritis – An Assault on the Senses.

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Introduction
A case of Giant Cell Arteritis (GCA) manifesting as bilateral tongue ischaemia with focal necrosis, central retinal artery occlusion(CRAO), headache and hearing loss. The case emphasises the importance of early recognition and treatment of GCA.

Aims/Background
MK, an 87 year-old female presented with acute-onset, painless, right monocular visual loss on a background of hypertension and atrial fibrillation on anticoagulation. Fundoscopy revealed a cherry-red spot consistent with CRAO, and a TIA/Stroke workup ensued. A CRP of 65 was not further investigate. She was commenced on aspirin and discharged, but re-presented four days later with left temporal headache, scalp tenderness, worsening left-sided hearing loss and a swollen, painful tongue with associated odynophagia and anorexia.
Examination revealed right monocular blindness, absent temporal pulses bilaterally, absent right radial pulse and tenderness on palpation of the left fronto-parietal region. A dusky-blue discolouration involving the tongue bilaterally was noted, with well-defined anterior ulceration. The tongue was tender to palpation.

Method
Bloods revealed a leukocytosis (11.9x10^9/L) with neutrophilia (9.8x10^9/L), thrombocytosis (425x10^9 /L) and an AKI (Creatinine-130µmol/L, Urea-18.2mmol/L). ESR and CRP were elevated (72mm/hr and 43.8mg/L, respectively), concordant with an acute inflammatory process. ANCA and ANA were negative. CT-Brain was unremarkable. Ultrasound confirmed bilateral temporal artery occlusion. CT-angiogram demonstrated intimal irregularity of intra- and extra-cranial vessels with prominent intimal thickening throughout the left vertebral artery, with an area of focal dissection.

Results
MK fulfilled the ACR-endorsed criteria for GCA and was commenced on high-dose steroids. Investigations for arteritis were omitted on initial presentation. Delay in diagnosis facilitated progression to tongue ischaemia, a rare occurrence given its rich vascularity. Current literature suggests that GCA-associated tongue necrosis often presents as lingual pain without typical symptoms, delaying diagnosis(9). MK had permanent right monocular blindness, though headache, scalp pain and hearing loss improved with steroids. Slow-healing ulceration compromised nutrition, necessitating nasogastric tube insertion. Given the above symptoms, communication proved difficult. MK became withdrawn and disengaged from MDT services, overcome by the psychological and physical impact of her disease.

Conclusions
Expedient recognition of GCA could significantly reduce physical and psychological morbidity. MK's case serves as a cautionary tale of the detrimental sequelae of a missed vasculitis.

Figure

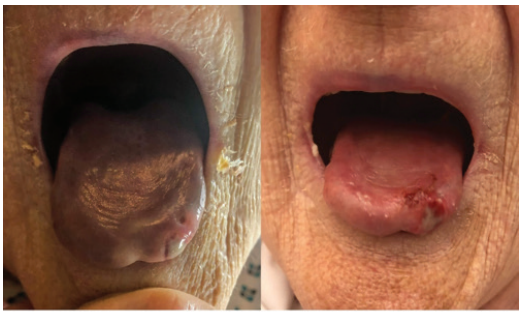

Image on left: Before commencing intravenous steroids. Image on right: One week after commencement of intravenous steroids, showing increased tongue perfusion, reduced swelling and healing ulceration.

(21SP103) CLINICAL CASE 22

The less seen side of IgG4 related disease

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Introduction
IgG4 related disease is a rare immune-mediated systemic fibroinflammatory disease with or without elevated plasma IgG4 levels. The prevalence continues to be unknown as the condition is often unrecognised or misdiagnosed. While there are a few classic presentations documented, other organ manifestations,like cardiac and CNS involvement, are significantly less often encountered in clinical practice. Ideally the diagnosis should rely on histopathological findings of dense infiltration of IgG4-positive plasma cells and a characteristic storiform fibrosis though not always possible in day-to-day practice.

Aims/Background
Case Report

Method
We report the case of a previously healthy 37 years old woman who presented to primary care with 8 weeks history of abdominal pain and constitutional symptoms. Following a CT abdomen she was referred into hospital with findings of a retroperitoneal mass suspicious of Lymphoma. During admission she underwent a PET-CT that showed moderately enhancing retroperitoneal mass and a bulky pancreas, no lymphadenopathy, inclining the diagnosis towards IgG4-RD. She went on to develop right sided headache and diplopia. What was initially suspected as Tolosa Hunt syndrome, proved to be on MRI brain a pachymeningitis in the context of IgG4-RD. Unfortunately none of the sites involved were amenable to biopsy
After an initial positive response to iv steroids,she represented with central chest pain due to an acute anterolateral myocardial infarction. Cardiac catheterization revealed 70 percent luminal stenosis in the first diagonal. Steroid induced coronary artery dissection was raised as a suspicion as well as atherosclerosis though the patients age and risk profile does not support the latter. Cardiac MRI showed significant burden of anterolateral myocardial edema and ischemia without thrombus.

Results
Immunosuppression with Rituximab was started given the multi organ involvement and showed excellent results with substantial reduction of para aortic fibrosis and the dural thickening of the temporal lobe on the repeat scans at six weeks.

Arthritis Ireland

“I’ve just been diagnosed and I feel completely lost

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Conclusions
IgG4 related disease is a rare condition, the diagnosis of which can at times pose some challenges.Manifestations like coronary and CNS involvement are exceedingly rare and can cause serious complications.Keeping an open mind and looking for a unifying diagnosis will help institute early treatment with immunosuppressive agents and a favourable outcome.

(21SP104) CLINICAL CASE 23

A curious case of Dermatomyositis/Localised Scleroderma Overlap

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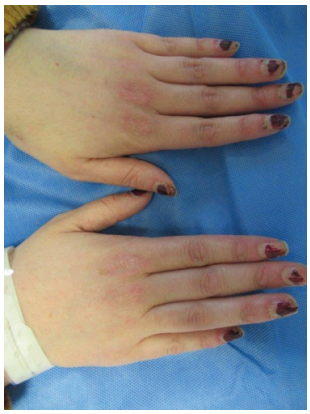
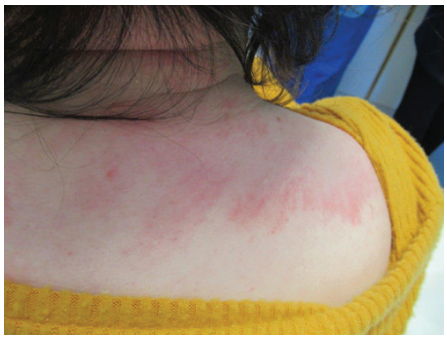
Introduction
Correlation between Dermatomyositis and Systemic Sclerosis is well described in the medical literature. However, association between Dermatomyositis and Localised Scleroderma is rare. We present a case report of Localised Scleroderma and Dermatomyositis presenting within 6 months of each other.
Aims/Background
A 28 year old patient presented with a 3 week history of progressively worsening proximal muscle weakness more prominent in the lower limbs. Rash with itch was reported over her metacarpophalangeal joints bilaterally. The patient denied any cardiac, respiratory or gastrointestinal symptoms. Medical history was significant for a diagnosis of Localised Scleroderma confirmed on biopsy 3 months previously.
Method
Examination revealed significant proximal muscle weakness. Nailfold dermoscopy showed ragged cuticles, and nailbed telangiectasia. Gottron's papules were present at the MCPs and proximal interphalangeal joints in a symmetrical pattern. An erythematous rash in a shawl distribution on her back was present. Healing Morphea lesions were present on both thighs. Laboratory work up noted Creatine Kinase of 8716 U/L, Troponin 616 ng/L and Lactate Dehydrogenase 889 U/L. Anti-nuclear antibody was positive. Myositis antibody panel revealed anti MI-2 antibodies consistent with a diagnosis of Dermatomyositis.
Results
MRI showed significant muscle oedema involving the quadriceps and obturator muscles. Deep muscle biopsy was performed which showed perifascicular and perivascular inflammation and atrophy in keeping with an inflammatory myopathy such as Dermatomyositis.
Conclusions
Scleromyositis – an overlap between systemic sclerosis and dermatomyositis, or more commonly polymyositis, is well described. However, on a review of published case reports, very few cases describing an overlap between morphoea and dermatomyositis exist. Rituximab has previously been used in treatment of scleroderma/myositis overlap to good effect, and was also effective in our patient. In cases of scleromyositis, muscle involvement has been described as mild, with minimal elevation in muscle enzymes, and patients generally respond well to low to moderate doses of steroid therapy. However, this patient had no features of systemic sclerosis, only presenting with localised plaque morphoea. Additionally, the patient's symptoms worsened when high dose steroids were weaned and she required treatment with an alternative immunomodulatory agent. This makes her case an unusual one, which has rarely been described in previous literature.

Figure



Figure



(21SP105) CLINICAL CASE 24

Visceral Leishmaniasis, mimicking SLE in a 79 year old Irish male with limited foreign travel

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Introduction
With increasing immigration, long distance travel and climate change, the range of tropical disease in Ireland is evolving. We wrote up this case because of the unfamiliarity of this diagnosis in an Irish population and to emphasise the importance of an accurate travel history.
We also felt it was important to highlight how autoimmune phenomena with false positive antibodies can occur with VL as has been described by numerous case reports.
Aims/Background
Our patient presented with a 6 month history of shortness of breath, fatigue and weight loss. He had a background of diabetes, hypertension, a single kidney post donation and a 50 pack year smoking history.
He was a retired mechanic who enjoyed playing golf and watching sports. Travel history included yearly holidays to Spain, the Canary Islands and Cyprus.
Exam revealed massive splenomegaly and initial blood work showed

pancytopaenia.

Method

We used a systemic approach investigating haematological, rheumatological and infectious diseases. Investigations included bloodwork, imaging and invasive procedures such as bone marrow biopsy.

With a normal initial bone marrow trephine and biopsy, lab results then pointed to a diagnosis of an autoimmune condition such as SLE or MCTD. There was no clinical improvement with hydroxychloroquine and Rituximab was started. With further deterioration in FBC indices and a rising ferritin, we revisited our differentials and a repeat bone marrow was performed.

Results

- DNA Elisa 49.00 IU/mL
- B2 microglobulin 14.10 mg/L
- C4 0.08 g/l
- ANCA: weak positive
- RF 48.1 IU/ml
- Anti PR3 3.60 IU/ml
- Anti cardiolipin antibody negative
- Myositis panel positive
- Serum: DAT positive (1 in 102499). Leischmania K39 antibody positive
- Bone marrow culture - abundant macrophages containing numerous spherical structures consistent with leischmanisis.

Interestingly these findings were not present on the initial bone marrow. We believe immunosuppression with rituximab is the most likely explanation for unmasking of the disease.

Conclusions

- VL can often mimic autoimmune diseases with false positive antibodies
- Diagnosis is confirmed by bone marrow biopsy
- Treatment is with amphotericin B and progress is monitored by improvement in spleen size, pancytopenia and weight.

(21SP106) CLINICAL CASE 25

A case of a retro-odontoid pseudotumor secondary to CPPD disease

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Introduction

Retro-odontoid pseudotumors are non-neoplastic, soft tissue masses adjacent to the odontoid process (dens) of C2. Aetiology can include inflammatory conditions such as rheumatoid and crystal deposition diseases. It can cause cervicomedullary compression, and the prevalence is thought to increase significantly with age1.

1. 1. Shi, J., Ermann, J., Weissman, B.N. et al. Thinking beyond pannus: a review of retro-odontoid pseudotumor due to rheumatoid and non-rheumatoid etiologies. Skeletal Radiol 48, 1511–1523 (2019). https://doi.org/10.1007/s00256-019-03187-z

Aims/Background

An 89 year old gentleman presented from home with acute onset dysarthria and right facial weakness. MRI brain confirmed an acute infarct, and incidentally noted a soft tissue mass at C1-C2. Dedicated C-spine imaging revealed a 3.9cm mass in C2, extending superiorly along the odontoid process (dens). There was mild effacement of the upper cervical cord; however normal cord signal was maintained. The patient had a longstanding history of osteoarthritis and radiological evidence of CPPD disease. On further questioning he had suffered with intermittent neck pain and stiffness over the previous few years. He had normal inflammatory markers and bloods were otherwise unremarkable. A combined clinical and radiological diagnosis of a retro-odontoid pseudotumor secondary to CPPD disease was made. He was commenced on colchicine and hydroxychloroquine for prevention of future flares, with outpatient rheumatology follow up.

Method

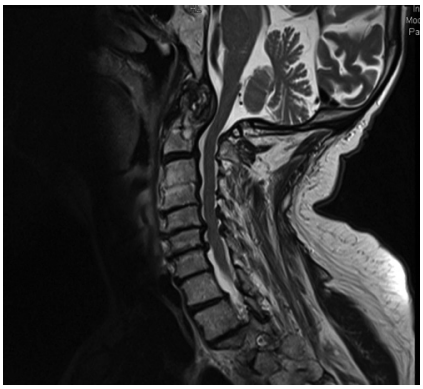
Clinical case

Conclusions

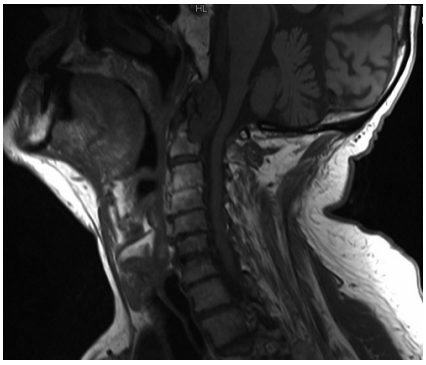
This case illustrates one of the rarer manifestations of crystal deposition diseases. Although patients are often asymptomatic, it is important to note acute inflammation of the tissue can result in acute presentations with severe neck pain, neck and shoulder girdle stiffness, pyrexia and elevated inflammatory markers (crowned dens syndrome). This is often misdiagnosed, and results in unnecessary investigations. It responds favourably to NSAIDS or colchicine2.

2. Godfrin-Valnet M, Godfrin G, Godard J, Prati C, Toussirot E, Michel F, Wendling D. Eighteen cases of crowned dens syndrome: Presentation and diagnosis. Neurochirurgie. 2013 Jun;59(3):115-20. doi: 10.1016/j.neuchi.2013.03.003. Epub 2013 Jun 24. PMID: 23806762.

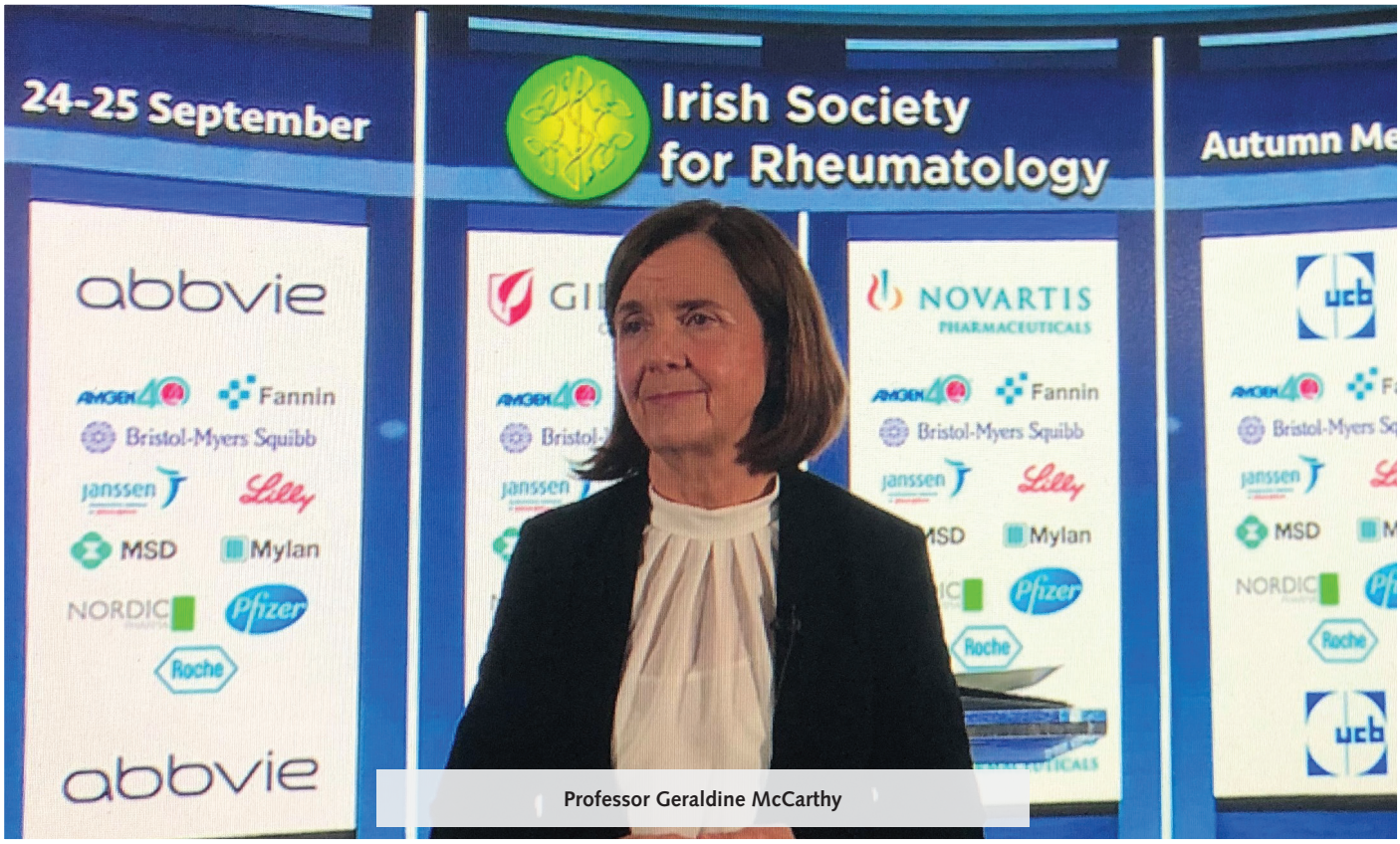
Figure



Figure



Speakers at the Autumn 2020

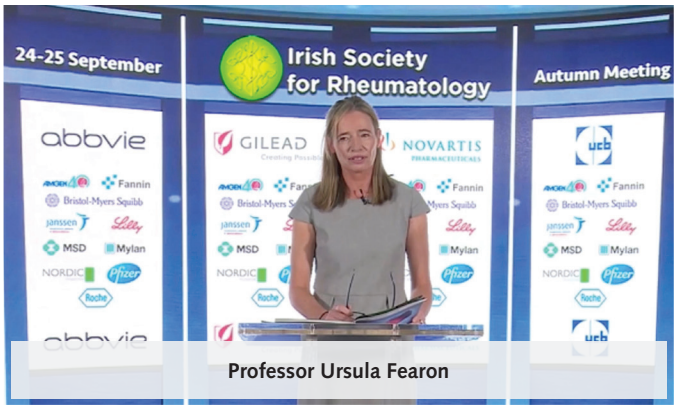


Professor Geraldine McCarthy

Dr Oran Kennedy, Bio Engineer, RCSI

Dr Richard Conway, UCD

Professor Rose Anne Kenny, Trinity College & SJH



Professor Ursula Fearon

ISR Meeting
Spring, 2021 Sponsors

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The Pharmas listed above have all supported this meeting.
They have had no organisational involvement in any other aspect of this meeting.

American College of Rheumatology IRNF Highlights Meeting

Thursday, 22 April 2021: Virtual – Commencing at 19.00

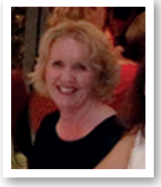
Agenda:

19.00 **Angela Camon,**
ANP Tullamore – Welcome and Introduction.

19.05 **Stephanie Narramore,** ANP, TUH –
"Janus kisane (JAK) inhibitors versus Biologic therapies in the management of R.A."

19.20 **Louise Murphy,**
ANP, CUH – "Update on Osteoporosis"

19.40 **Coffee**



19.50 **Una Martin,** ANP, UHW –
"Working with Wellness in Rheumatology"

20.10 **Angela Camon,**
ANP - Round up, Thanks and Conclusion

20.20 **AbbVie Satellite Meeting** (for HCP's only)
Deirdre Moran, 'AbbVie Medical Manager'
"Upadacitinib safety and efficacy/monitoring requirements"

This meeting was held virtually on Thursday 22 April 2021 to which an audience of 38 people logged in. The meeting was excellently chaired by Angela Camon from Tullamore. The presentations were developed and delivered by expert members of IRNF followed by short Q & A sessions. Anybody who was unable to login can view the whole session on www.isr.ie under past events. Well done to all concerned and in particular to AbbVie Ltd for their continued sponsorship.