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Clinical features and final diagnosis in patients with myositis associated and myositis specific antibodies

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INTRODUCTION

The idiopathic inflammatory myopathies (IIMs) are a heterogeneous group of conditions characterized by proximal muscle weakness. Autoantibodies are recognized in more than 80% of patients with polymyositis (PM) or dermatomyositis (DM). Some are also found in other connective tissue diseases (CTD), while others are more specific to IIM. Thus, they are classified into two categories named myositis associated antibodies (MAA) and myositis specific antibodies (MSA). MSA have been reported as being 90% specific for IIM while MAA are found in up to 50% of myositis patients.

AIMS AND OBJECTIVES

The aim is to assess the associations of these antibodies with clinical manifestations and final diagnosis in our cohort.

METHODS

A retrospective chart review study was conducted at St. James's Hospital from 2015-2019. All positive myositis panels were obtained. The MAA evaluated were PMScl (100/75), U1snRNP, Ku and Ro52, while MSA were Mi2a, Mi2b, TIF1, MDA5, NXP2, SAE1, Jo-1, SRP, PL-7, PL-12, EJ and OJ. Associations with clinical features and final diagnosis were examined.

	MAA	MSA	ILD	Arthritis	Arthralgia	Myositis	Raynauds	Cutanous	Malignancy	Cardiac	Final diagnosis
Inflammatory myositis											
1	Ro52			+	+	+		+			Dermatomyositis Hidradenitis suppurativa
2		NXP2				+		+			Dermatomyositis Myasthenia gravis
3	Ro52							+	+		Paraneoplastic dermatomyositis, stage 4 high-grade serous ovarian carcinoma
4		MDA5			+			+			Amyopathic dermatomyositis
5		SAE1				+		+			Dermatomyositis
Interstitial lung disease											
6	Ro52	PL12	+								IPF
7		SAE1/OJ	+								IPF
8	Ro52		+								IPF
9	Ro52		+		+			+			IPF Pyoderma gangrenosum
10	Ro52	PL7	+	+	+		+	+			Anti-synthetase syndrome
11		PL7	+			+					Anti-synthetase syndrome
12		JO-1	+				+				Anti-synthetase syndrome
Connective tissue disease											
13	Ro52										SLE
14	Ro52			+	+						Sjogren
15	Ro52										Sjogren
16	Ro52				+				+		Sjogren Breast cancer
17	Ro52		+		+						Sjogren with lung manifestation
18	Ku/Ro52				+			+			Undifferentiated CTD
19	U1snRNP			+	+		+				Undifferentiated connective tissue disease
20	U1snRNP/ Ro52	OJ		+	+	+		+			MCTD Autoimmune hepatitis
21		SRP					+	+			Limited cutaneous scleroderma
22	PMscl 100/75						+	+			Scleroderma Scleroderma renal crisis
Others											
23		NXP2			+						Polymyalgia rheumatica
24	ku	Mi2b			+						large vessel vasculitis
25		PL12		+	+			+			PsA
26		Mi2b									PBC
27	Ro52										Liver cirrhosis
28	Ro52										Autoimmune limbic encephalitis
29	Ro52				+						Fibromyalgia
30	Ku/Ro52										Chronic spontaneous urticaria Hypothyroidism
	Total		8	6	14	5	5	12	2		

RESULTS

30 patients were positive for one or more MSA/MAAs. The mean age was 54.9 years, the majority were female (73.3%).The most prevalent MAA was anti-Ro52 (17/30) followed by anti-Ku (3/30) and anti-U1RNP (3/30), anti-PMScl 100/75 was seen in (1/30). The prevalence of MSA were (3/30) for anti-OJ followed by (2/30) for each of anti-Mi2b, anti-PL7, anti-PL12, anti-SAE1 and anti-NXP2 and (1/30) for each of anti-Jo, anti-SRP and anti-MDA5. The most common observed clinical phenotypes in our cohort were arthralgia (14/30) and cutaneous manifestations (12/30). Less than one third of the studied population had ILD (8/30), arthritis (6/30), myositis (5/30), raynauds (5/30) and malignancy (2/30). Various diagnosis were allocated for these patients, while only five cases were diagnosed having dermatomyositis. Figure1

CONCLUSION

IIM was the final diagnosis in only 16% of positive myositis panels in our study, signifying low specificity in our cohort.

REFERENCES

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