Figure 1. Correlations between PASTUL and SSPRO (A) and mRSS (B)

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AB0403

ORBITAL MYOSITIS: AN UNCOMMON OPHTHALMIC PRESENTATION IN RHEUMATIC DISEASES - CASE SERIES AND SYSTEMIC REVIEW OF CASE REPORTS

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Background: Ophthalmic conditions are common manifestations in patients with rheumatic diseases (RDs), orbital myositis (OM) remain rare. Only 1 case each for scleroderma and undifferentiated connective tissue disease (UCTD) reported. **Objectives:** Report frequency of Rheumatic Diseases with OM (RDs-OM), diagnosis and treatments.

Methods: Patients database were obtained from trust electronic record and literature review of case reports were performed.

Results: 4 out of 7 patients in our clinic with RDs-OM. Both Scleroderma-myositis (SCM) patients were positive for anti-PM/SCL antibody. All received glucocorticosteroid (GCS) and Mycophenolate as steroid-sparing or rescue therapy with good tolerance and outcome. One idiopathic OM has residual muscle paresis.

Table 1. Case series of patients with Orbital Myositis

	Idiopathic OM (n=3)	Rheumatology Diseases (n=4)	p-value
Age (years)	48 (27-72)	42.75 (21-60)	P= 0.8571
Gender – Female (Male)	2 (1)	4(0)	
Median Period Of Follow Up (months)	24.60 (7-37)	47.25(1-120)	
Underlying RD			
I.UCTD	-	1	
II.Overlap Scleromyositis	-	2	
III.Behcet's Disease	-	1	
Biopsy			
I.Pathological significant	1	1	
II.Normal	1	1	
III.Not done	1	2	

Total 93 cases with RDs-OM reviewed and the characteristic of patients represented in Figure 1.

Figure 1. Characteristics of orbital myositis in patients with rheumatic diseases. F: female, M: male, NS: not-specified, PrePD: onset before RD, AtD: onset with RD, PostPD: onset after RD, GCS 1st: Glucocorticoid as first line treatment, GCS res: Glucocorticoid responder, BxOM: biopsy of extra-ocular muscle, OtherTs: Other Tissue A: Median age at diagnosis, Sarc: sarcoidosis, SLE: Lupus, EGPA: Eosinophilic Granulomatous Polyangiitis, GPA: Granulomatous Polyangiitis, RA: rheumatoid Arthritis, BD: Behcet's Disease, RP: Relapsing Polychondritis, DM: Dermatomyositis, LoSSC: Localized Scleroderma, SPA: Spondyloarthropathy, ASOD: Adult Onset Still's Disease, RPF: Retroperitoneal Fibrosis, GCOM: Giant Cell OM, SJS: Sjogren Syndrome, DL: Discoid Lupus, KD: Kawasaki Disease, SAPHO: Synovitis, acne, pustulosis, hyperostosis, and osteitis, APLS: Antiphospholipid Syndrome.

OM although is rare but can present in most RDs. Our review showed OM commonest in patients with RA, GPA, EGPA and sarcoidosis. Only 1 each for SCM, APLS, UCTD, SAPHO and Kawasaki Disease.

Most RDs-OM patients are female. 89.2% OM diagnosed concurrently with primary RD diagnosis. Majority RA, BD and GPA developed OM along the course of RDs (83%-90%).

Except for routine RDs screening, thyroid eye disease, infections, malignancy (eg. lymphoma) and myasthenia gravis, atypical infective screenings include mycobacterium, fungal, *Borrelia* and spirochetes were excluded. Histopathological diagnosis is important diagnostic tool for IgG4 disease, neoplasm, sarcoidosis and vasculitis. 59% patients underwent tissue biopsy, 71% of these were sampled from EOM, some from involved organs. Orbital MRI or CT were the commonest diagnostic imaging, 5 reported had Ocular ultrasound as single or complementary imaging and 1 had PET-CT for exclusion of other diagnosis.

Only 40% GPA and 20% of EGPA patients were positive for ANCA either c-ANCA, p-ANCA and MPO which is similar in other literatures.

97.8% (n=91) patients received GCS with 79% of these patients reported improvement or resolution of ocular symptom with or without steroid-sparing agents. Immunosuppressants used are those not foreign among rheumatologists while biologics prescribed for refractory cases were Rituximab(2), Infliximab(4), Adalimumab(2) and Tocilizumab(1). Orbital decompression and orbital radiation were other measures. 13 patients had residual EOM paresis.

Conclusion: 1.RDs-OM is increasing recognized.

2.Most OM in RA, BD and GPA occurs along its primary RDs.

3.40% GPA and 20% of EGPA patients were ANCA positive.

4.GCS remain the primary treatment, three quarter patients responded.

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AB0404

EXTENDED MYOSITIS PANEL AND THE CLINICAL ASSOCIATION IN PATIENT WITH SUSPECTED INFLAMMATORY DISEASE, A RETROSPECTIVE STUDY

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Background: The idiopathic inflammatory myopathies (IIMs) are a heterogeneous group of conditions characterized by proximal muscle weakness. Autoantibodies are identified 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. ¹

Objectives: The aim of the study was to evaluate the myositis antibody prevalence and to assess the associations of these antibodies with clinical manifestations, final diagnosis, medication received and outcomes.

Methods: A retrospective chart review study was conducted at St. James's Hospital from 2015-2020. 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.

Results: We identified 52 patients who were positive for one or more MSA/MAAs. The mean age was 58.9 years, the majority were female (65.3%). The most prevalent MAA was anti-Ro52 (29/52) followed by anti-PMScl 100/75 (7/52), anti-Ku and anti-U1RNP were seen in (3/52) each. The prevalence of MSA were (4/52) for anti-PL12 and (3/52) for anti-SAE1 followed by (2/52) for each of anti-Mi2b, anti-NXP2, anti-Jo, anti-SRP, anti-PL7, anti-EJ and anti-OJ while only (1/52) for anti-MDA5. The most common observed clinical phenotypes in our cohort were arthralgia (20/52), ILD (18/52) and cutaneous manifestations (15/52). Less than a quarter of the studied population had arthritis (8/52), myositis (7/52), raynauds (7/52) and malignancy (4/52). Various diagnosis were allocated for these patients, while only eight cases were diagnosed with dermatomyositis. Medication received and the final outcome for those with strong positive MSA were summarized in table1.

Conclusion: IIM was the final diagnosis in only 15% of positive myositis panels and the presence of antisynthetase antibodies didn't necessarily indicate the presence of antisynthetase syndrome thus signifying low specificity in our cohort.

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