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Case Report

Internal Medicine Section

Rheumatoid Myositis: A Rare Case Report

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ABSTRACT

Rheumatoid Arthritis (RA) and inflammatory myositis are distinct clinical syndromes. Muscle involvement in RA is frequently secondary to therapy with steroids or immune-modulators medication. However, the association of myositis in a treatment- naive patient with RA is rare. Myositis represents a distinct entity with specific clinical, biological, imaging, and histological patterns. Here, authors describe a case of RA presented with myositis. A 30-year-old female presented with insidious onset gradually progressive to paraparesis lasting for six months. This was preceded by a history of inflammatory arthritis involving small and large joints of the upper and lower extremities. Clinically, she exhibited pure motor weakness of the upper and lower limbs (proximal>distal) with hand, elbow, and knee deformities. Her Anti-Cyclic Citrullinated Peptide antibody (Anti-CCP) and inflammatory markers were elevated. Muscle-specific enzymes were also elevated, along with positive myositis-specific antibodies. Electromyography (EMG) and Magnetic Resonance Imaging (MRI) of the thigh revealed findings consistent with inflammatory myositis. The patient was managed as a case of Rheumatoid Myositis (RM) with steroids, conventional, and biological immune-modulators Disease-Modifying Antirheumatic Drugs (DMARDs). She experienced remarkable improvement in her symptoms and quality of life.

Keywords: Anti-citrullinated, Arthritis, Electromyography, Imaging, Inflammatory

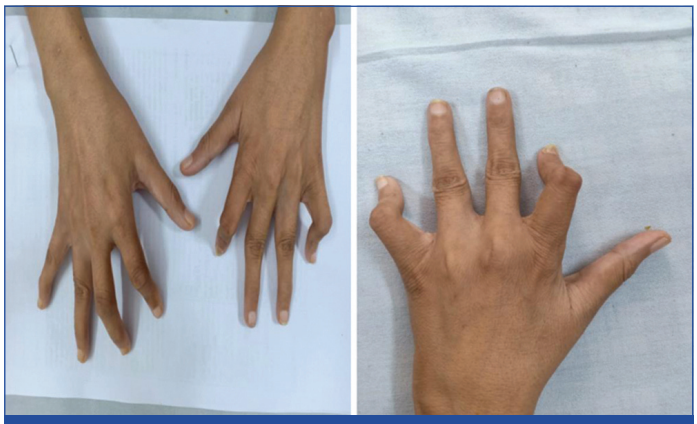
CASE REPORT

A 30-year-old female presented to the Accident and Emergency department with the following complaints: (a) Pain and swelling of joints for the past four years; (b) Weakness in both upper and lower limbs over a six-month period. Four years ago, she experienced the onset of symmetrical polyarthritis, starting in the hands and wrists and progressing to the elbows and knees. She partially found relief with analgesics but gradually lost function and developed deformities in her hands and elbows over the past year. In addition, she experienced insidious onset paraparesis that progressed to quadriparesis, with weakness in the trunk. This has resulted in difficulties with daily activities and dependence on others.

Her medical history was negative for autoimmune, inflammatory, chronic infection, or metabolic diseases. There was no family history of similar complaints. On examination, she exhibited pure motor weakness (more pronounced proximally than distally) without sensory symptoms, cranial nerve involvement, or fasciculations. On musculoskeletal examination patient had flexion deformity of both wrist, right elbow, tender and swollen joints, boutonniere deformity of hands [Table/Fig-1]. Vital signs were within normal limits, but she presented with anaemia and elevated muscle enzymes. Myositis-associated antibodies (RO-52 positive) were detected, along with elevated Erythrocyte Sedimentation Rate (ESR) and C-Reactive Protein (CRP).

Rheumatoid factor and Anti-CCP antibody (Anti-CCP) tests were performed [Table/Fig-2]. Anti Nuclear Antibody (ANA) by Indirect Immunofluorescence assay (IIF) showed a 1+ Nuclear, speckled pattern (1:100), and the Extractable Nuclear Antigen (ENA) panel was negative. EMG revealed increased insertional activity and early recruitment, indicating a myogenic pattern [Table/Fig-3]. The MRI findings of the thighs [Table/Fig-4] were consistent with inflammatory myositis.

The patient was diagnosed and managed as a case of RA. Treatment included corticosteroids (Tab Prednisolone 40 mg once daily, tapered over a three-month period), conventional Immune-modulators anti-rheumatic drugs (DMARDs) (Tab Methotrexate 10 mg/week, Tab Hydroxychloroquine 200 mg once daily), and subsequently, biological DMARDs (Injection Rituximab 1 gm



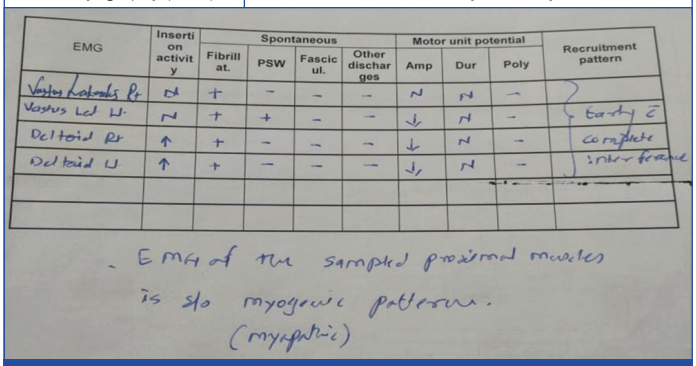
[Table/Fig-1]: Boutonniere deformity.

Parameters	Patient value	Normal range
Haemoglobin (Hb) (g/dL)	10.2	15.5±2,5
Mean Corpuscular Volume (MCV) (fL)	87	80-100
Total leucocyte count (TLC) (/cumm)	6100	4000-11000
Differential Leucocyte Count (DLC)	Neutrophils (N)-69 Lymphocytes (L)-18	N (40-75) L (20-45)
Platelets (lacs/cumm)	2.82	1.50-4.00
Peripheral Blood Smear (PBS)	Normocytic Normochromic (NCNC)	
Prothrombin time/International Normalised Ratio/PTTK (Partial Thromboplastin Time with Kaolin)	14.3/1.12/31.5	11-15/35-45
Urea/Creatinine (mg/dL)	26/0.8	10-50/0.8-1.2
Sodium/Potassium (Na/k) (meq/l)	136/4.1	135-145/3.5-5.0
Calcium/Phosphorus (Ca/P) (mg/dL)	8.7/3.5	8.5-10.2/2.5-4.5
Bilirubin (total) (mg/dL)	0.7	0.1-1.0
Aspartate aminotransferase/Alanine aminotransferase (AST/ALT) (IU/L)	140/15	15-37, 16-63
Uric acid (mg/dL)	4.1	3.5-7.2
Total Protein/Albumin/Globulin (g/dL)	6.7/3.2/3.4	6-8/3.5-5.5/1.8-3.6
Creatine phosphokinase (U/L)	360	(26-308)

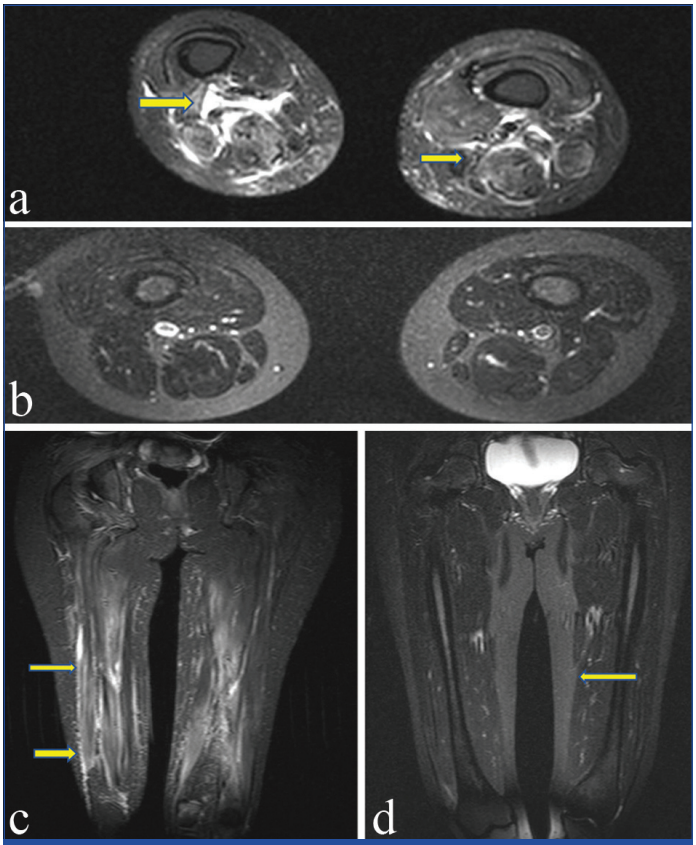
Erythrocyte Sedimentation Rate (ESR)	28 mm fall 1st hour	Male: (0-10)
C-Reactive Protein (mg/L)	13.3	0-5
Rheumatoid Factor (IU/mL)	26	<20
Anti-CCP (U/mL)	399.2	<17
Muscle specific antibody	RO- 52: Strong Positive	
ANA by IIF	1+Nuclear speckled (1:100)	

[Table/Fig-2]: Summary of the investigations.

Nerve conduction study	Normal
Electromyography (EMG)	Increased insertional activity and early recruitment



[Table/Fig-3]: Electromyography (EMG) of sampled proximal muscles.



[Table/Fig-4]: Magnetic Resonance Imaging (MRI) of thighs: a) Patient-STIR (Fluid sensitive) axial images; b) Normal-STIR axial images; c) Patient-STIR coronal; d) Normal-STIR coronal. Hyperintensity seen involving the visualised muscles.

intravenous infusion two weeks apart). Bilateral knee replacement surgery was also performed. The patient showed improvement in symptoms, became independent in daily activities, and is currently on regular follow-up.

This case represents a rare association of myositis in treatment-naive patients with RA.

DISCUSSION

This case represents an unusual clinical progression of RA leading to inflammatory myositis. The development of myositis in RA patients receiving steroids and DMARDs has been reported in a few cases [1,2]. However, the co-existence of RA and myositis in treatment-naive patients is a rare occurrence. Despite the availability of newer diagnostic techniques, myositis in the context of RA remains underdiagnosed, with limited published studies on the topic [3,4].

Myositis tends to occur early in the course of RA, especially in patients with active disease and elevated acute phase reactants such as ESR. Diagnosis relies on a combination of clinical presentation, serology, myositis-specific antibodies, EMG, imaging (MRI), and histopathology [5]. In the case of inflammatory myositis associated with RA, the muscle involvement is typically patchy and moderate, primarily affecting the proximal muscles in a symmetrical and bilateral pattern. Multiple sections of analysis may be required for accurate diagnosis due to the patchy nature of the condition [3,6]. While muscle biopsy is considered the gold standard, it is an invasive procedure and not practical to perform multiple biopsies. MRI, on the other hand, is a more feasible and less invasive modality that can screen a majority of the affected muscles and even detect subclinical inflammation [7,8]. Additional diagnostic aids include muscle enzymes, myositis-specific antibodies, and EMG [5,6]. In this case, the diagnosis of RA was confirmed by the presence of inflammatory arthritis in small and large joints, elevated Anti-CCP and RF levels, and increased ESR and CRP [Table/Fig-2]. The diagnosis of RM was confirmed by the patient's presentation of pure motor quadriparesis (proximal>distal), elevated muscle enzymes, positive myositis-specific antibodies [Table/Fig-2], myopathic pattern in EMG [Table/Fig-3], and evidence of inflammatory myositis in the thigh MRI [Table/Fig-4]. The criteria for American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) 2010 is shown in [Table/Fig-5].

Domain	Category	Score
A	**Joint involvement**	
	1 large joint	0
	2-10 large joints	1
	1-3 small joints	2
	4-10 small joints	3
	>10 joints (≥1 small joint)	5
B	**Serology (≥1 test result needed)**	
	Negative RF and Negative ACPA	0
	Low positive RF or low positive ACPA	2
	High positive RF or high positive ACPA	3
C	**Acute phase reactants**	
	Normal CRP and normal ESR	0
	Abnormal CRP or abnormal ESR	1
D	**Duration of symptoms**	
	<6 weeks	0
	≥6 weeks	1

[Table/Fig-5]: ACR EULAR criteria for RA 2010 [9,10].
ACPA: Anti-citrullinated protein autoantibodies; RF: Rheumatoid factor; CRP: C-reactive protein; ESR: Erythrocyte sedimentation rate

There is no definitive guideline for the management of RA myositis due to its rarity. However, reported cases have shown a positive response to high-dose oral steroids and both (Bilatrel Knee Arthroplasty) DMARDs [3,6]. In this particular case, the patient showed significant clinical improvement with the combination of conventional and biologic DMARDs, along with high-dose systemic corticosteroids. This case highlights the rarity of RA myositis and the lack of sufficient literature, which can lead to delay in diagnosis and treatment.

RM is a rare clinical manifestation in treatment-naive RA patients, characterised by varied clinical presentations and moderate, patchy muscle involvement primarily affecting the proximal muscles in a symmetrical and bilateral manner [4]. Limited studies have documented this rare clinical association (Iyagba A and Altraide D, Ancuta C et al., and Paul R et al.,) [3,4,6].

RM is distinct from other forms of inflammatory myositis due to its patchy muscle involvement, unique electrodiagnostic findings, and non specific myopathy. The patchy nature of muscle involvement in RM makes non invasive modalities such as MRI and EMG essential for diagnosis [5,7].

The co-existence of myositis and RA has shown favourable clinical outcomes with the use of steroids, conventional DMARDs, and biological DMARDs, as observed in this patient.

CONCLUSION(S)

RM is still poorly characterised, although the concept of muscle involvement in RA is well-known, ranging from medications to impaired joint flexibility and sedentarism. This study provides information about myositis in treatment-naive patients with RA. The co-existence of myositis and RA leads to different clinical manifestations compared to typical presentations. The presence of non specific myopathy and patchy muscle involvement makes non invasive modalities like electrodiagnostic studies and MRI the preferred diagnostic tools for RM. The treatment approach for RM also varies, and the use of steroids, conventional DMARDs, and biological DMARDs has been associated with better clinical outcomes. This study highlights the diverse clinical presentation of RM and emphasises the importance of non invasive diagnostic modalities for this rare condition, as well as the treatment approach to RM.

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