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Case Report

OVERLAP OR MIXED?

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ABSTRACT
Rheumatoid arthritis is common in India with prevalence of 15 % i.e. over 180 million people in India. Rheumatoid arthritis also coexists with other connective disorders which occur as overlap syndrome. In India, no definite literature defines the prevalence of overlap syndrome of rheumatoid arthritis with myositis. Often overlap syndrome goes unnoticed because of severe arthritic manifestation. Until unless one has a thorough vigilance and knowledge to look for other system involvement about overlap Syndrome, the disease goes undiagnosed. Hereby we present a case of MCTD, who presented initially with severe myositis and features of RA, a high vigilance formyositis and search for overlap syndrome guided in diagnosing this case of MCTD. MCTD is distinguishable from RA-myositis overlap in the form that it has elevated Anti U1-RNP, the muscle biopsy revealed features of myositis which is not found with RA myositis overlap syndrome. Patient was treated with pulse methyl prednisolone therapy for three days, continued with oral steroids and methotrexate. Patient showed significant improvement in muscle pain and weakness.

Keywords: *RA, MCTD, Methotrexate, Muscle, Myopathy (RA-Rheumatoid Arthritis, MCTD – Mixed Connective Tissue Disorder)*

INTRODUCTION
Connective tissue diseases are a group of disorders of unknown etiology. Their classification depends upon identifying mixture of clinical and lab features. Mainly major diffuse connective tissue disorder includes systemic lupus erythematosus; scleroderma; polymyositis; dermatomyositis; and rheumatoid arthritis.
Mixed Connective Tissue Disorder is a distinct syndrome in which the combination of features similar to those of systemic lupus erythematosis, systemic sclerosis, dermatomyositis/polymyositis and rheumatoid arthritis are seen. MCTD is associated with autoantibodies to a Ribonuclease – sensitive component of extractable nuclear antigen (ENA) now known as U1RNP and hence it is unique.

CASES
A 51 -year -old lady presented with complaints of multiple joint pain since three years and pain in both her lower limbs for eleven months. She had symmetrical joint involvement with the small joints of her hand being involved initially with early morning joint stiffness lasting for at least one hour. She had been prescribed analgesics and steroids on multiple occasions for the same. Her symptoms worsened over a period of one year when she started noticing pain at her calf muscles. She also complained of small swellings which appeared over ankle joint, metacarpal joints.
For last six months she had difficulty in getting up from squatting position and had pain in her thigh muscles too. She managed to walk without any support. There was no history of skin rashes, recurrent genital ulcer, photosensitivity. She has no past history of Diabetes, Hypertension, Bronchial asthma, Seizures, TB. There was no family history of similar complaints, diabetes mellitus, hypertension, epilepsy or asthma.

On Examination
She was conscious, oriented, a febrile and alert. Examination revealed that she had mild puffiness of face. The fingers showed spindle shaped deformity affecting proximal interphalangeal joints and there was swelling of the metacarpophalangeal joints also (Figure 1). Her pulse was 90/min, regular, normal volume, normal character with no vessel wall thickening and her B.P was 140/ 90 mmHg. During

systemic examination, diffuse tenderness was present over both thighs (R>L), with power of 4+/5 in both lower limb (proximal group).

Power in all other group of muscles was 5/5. Reflexes were normal. Rest of the systems was within normal limits. A clinical diagnosis of probable MCTD with varied severity of polymyositis, rheumatoid arthritis and Systemic Lupus Erythematosis was made based on Kasukawa criteria (Table 2) and further investigated.

Investigations

Imaging of hands showed juxta articular osteopenia suggestive of RA. A muscle biopsy was taken from the right thigh revealed myositis along with inflammatory infiltratein the regions surrounding the normal muscle fibres (Figure 3). CBC, RFT, Electrolytes, LFT was normal. HRCT revealed fibrotic strands in the lower lobe of left lung.

Table 1: Serological Reports

HIV	**NEGATIVE**
HbsAG	NEGATIVE
Hepatitis B & C	NEGATIVE
Toxoplasmosis	NEGATIVE
Cytomegalovirus	NEGATIVE
ECHO	NORMAL
PFT	RESTRICTIVE
RA FACTOR	POSITIVE
U1-SNRNP	66 (POSITIVE)
CPK TOTAL	1856

Figure 1: Title Missing

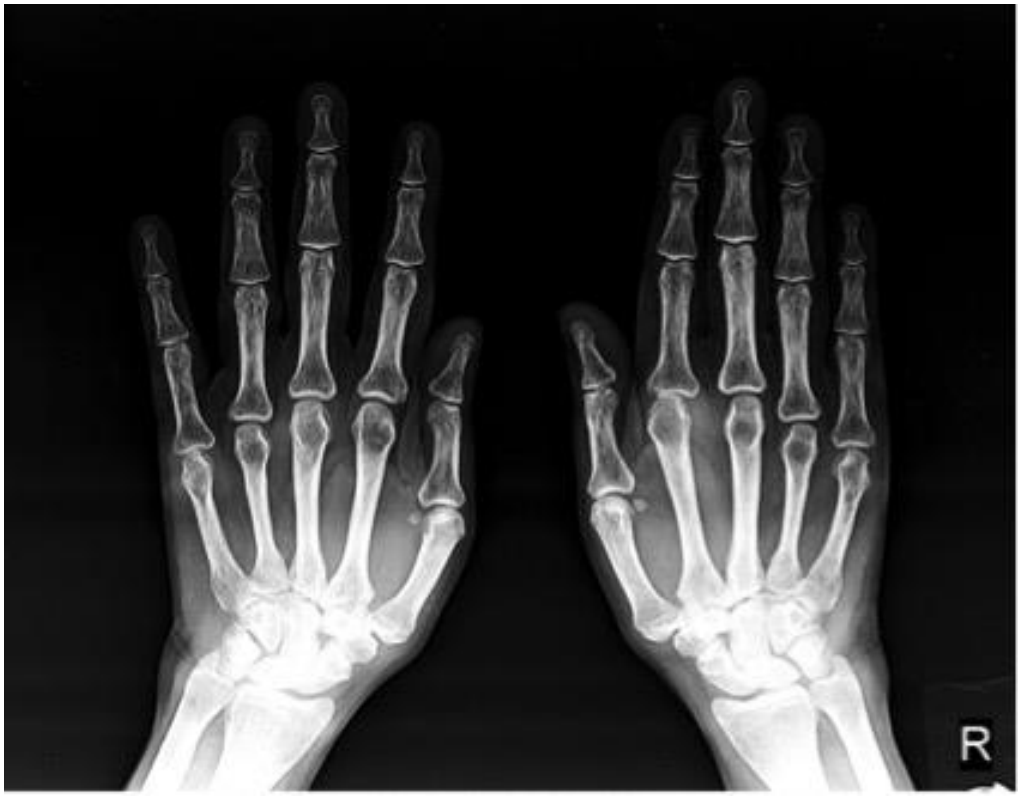

Figure 2: Xray of hand showing juxta articular osteopenia

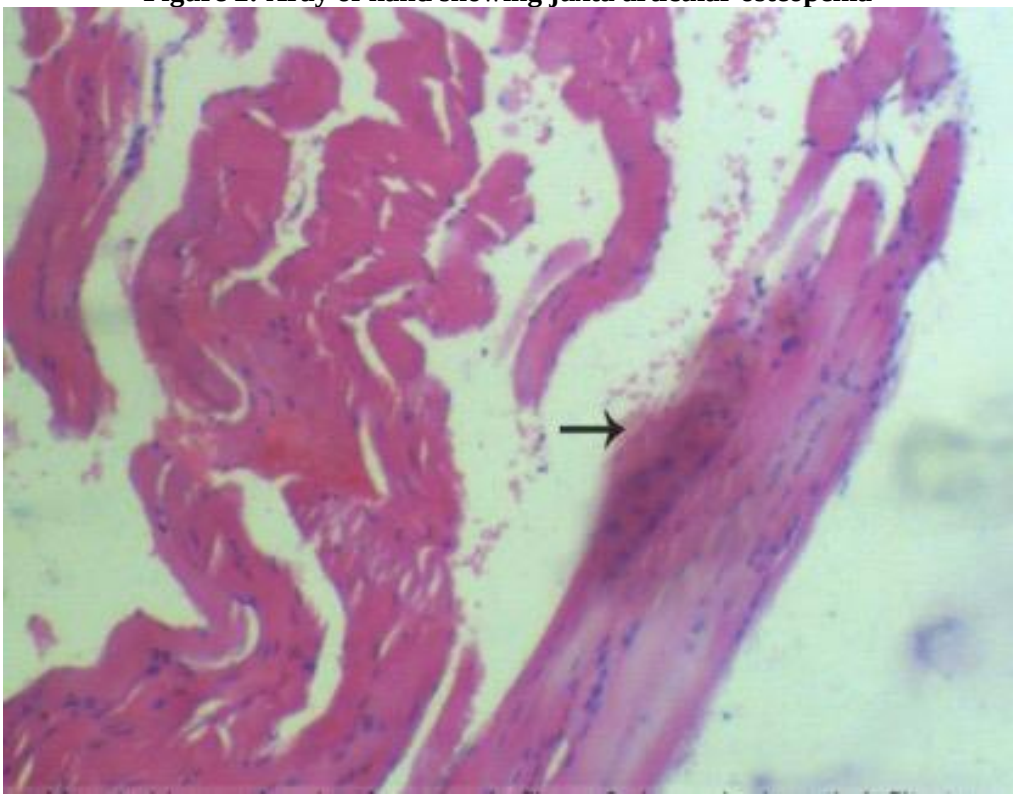

Figure 3: Muscle biopsy showing few atropic fibres & dense leukocytic infiltrates

Table 2: Diagnostic Criteria for MCT

Kasukawa	Common symptoms	Mixed symptoms	Requirement for diagnosis
	1. Raynaud's Phenomenon	**1. SLE-like symptoms:** i. Polyarthritis ii. Pericarditis or Pleuritis iii. Facial erythema iv. Leukothrombocytopenia v. Lymphadenopathy	At least 1 or 2 common symptoms plus positive for anti RNP plus one more of the mixed symptoms in at least 2 of the 3 disease categories
	Swollen fingers or hands Anti-RNP antibody positive	**2. Scleroderma-like findings** i. Sclerodactyly ii. Hypomotility or dilatation of esophagus iii. Pulmonary fibrosis, restrictive changes of lung or reduced diffusion capacity	
		3. Polymyositis-like findings: i. Muscle weakness ii. Myogenic pattern on EMG iii. Eleveated serum levels of muscle enzymes (CPK)	

DISCUSSION

In the present study, we have described a rare case of one female patient with MCTD who presented initially with severe myositis and features of interstitial lung disease as diagnosed by restrictive pattern in PET & Fibrotic strands in HRCT chest. The disease would have progressed to CREST syndrome. Nearly 60% of patients who have inflammatory myositis present with subclinical myopathy (Kasukawa, 1999). However, one can achieve quick clinical and laboratory response with low dose corticosteroids (Lundberg and Hedfors, 1991).

Our patient presented with severe myositis on biopsy and elevated creatine kinase levels. It was this observation that made us initiate treatment with high dose corticosteroids and methotrexate. Our patient had fulfilled the international diagnostic requirements proposed by Kasukawa for MCTD, after excluding other infectious diseases via serologic tests and after ensuring there was no history of any myotoxic drug intake. In the literature only one previous report was found that described a severe and refractory case of myositis in an adult with MCTD (Bonin *et al.*, 2010).

This work has reported an extremely rare case of an adult patient with MCTD characterized by a severe myositis and high titres of Anti U1-RNP levels at presentation that was effectively treated after the administration of pulse methyl prednisolone, and methotrexate. A high vigilance for myositis and search for overlap syndrome, guided in diagnosing this case of MCTD

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