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A CASE OF IMMUNE MEDIATED NECROTIZING MYOPATHY

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ABSTRACT

Background: Immune-Mediated Necrotizing Myopathy (IMNM), one of the Inflammatory Myopathies, is a rare, thought to be complement-mediated muscle disease. IMNM can present much like polymyositis and causes muscle cell death (necrosis) that leads to weakness of the skeletal muscles on both sides of the body (symmetrical), commonly those closest to the body's core (proximal), such as the hips, thighs, upper arms, shoulders, and neck. Muscle weakness, which is often severe, can develop over a period of days, weeks, or months. Specific Antibodies - Anti SRP and Anti HMGCoA reductase antibodies-Presence of aniti SRP antibodies shows relatively refractory course, Aggressive, More difficult to treat, aggressive immunotherapy required.

Method: A Case of middle aged female presented with bilateral proximal myopathy taken detailed study and laboratory investigations done

Discussion: A 47 year old female presented with B/L Symmetrical muscle weakness insidious in onset, proximal more than distal, lower limb more than upper limb with Anti SRP ab positive suggestive of Immune mediated necrotizing myopathy.

Keywords: Immune mediated necrotizing myopathy, Antibodies, Inflammatory myopathy

INTRODUCTION

IMNM, or autoimmune necrotizing myopathy, is characterized by the acute or insidious onset of symmetric, proximal more than distal weakness. Dysphagia, dysarthria, or myalgia may occur. Patients may have an underlying CTD (usually scleroderma or MCTD) or cancer (paraneoplastic necrotizing myopathy), or the condition may be idiopathic. There are at least two distinct forms of IMNM associated with specific autoantibodies (anti-3-hydroxy-3-methyl-glutaryl-coenzyme reductase [HMGCR] and anti-signal recog-nition particle [SRP]). Anti-HMGCR myopathy can be seen in patients receiving statins, inhibitors of HMGCR, particularly in patients aged >50 years. However, anti-HMGCR myopathy can develop in children and young adults without a history of statin use and can mimic a limb girdle muscular dystrophy. Unlike the more common "toxic" myopathy associated with statin use, anti-HMGCR myopathy does not improve when statins are discontinued. Anti-SRP myopathies are notable for the presence of anti-SRP antibodies and a typically subacute, aggressive, and relatively refractory course.

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CASE PRESENTATION:

A 47 year old female presented with

- % - B/L wrist pain since 5 months
- Difficulty in getting up from sitting position since 2 month
- Facial and pedal edema since 2 month
- Difficulty in lifting hand above shoulder since 1 month

Patient's weakness was insidious in onset, gradually progressive in nature, Symmetrical, proximal more than distal muscles involved.

Negative History

No % Thinning of limbs, involuntary movements, Altered Sensorium, Skin Rash, Drug ingestion, Toxin, Sensory loss, oral ulcers, Hair loss, Dry mouth

General Examination:-

I have examined the patient in proper light and exposure. Patient is conscious and oriented to time, place and person. T: Normal
P: 86/min
BP: 110/66 mmHg
RR: 16/min
No any signs of Pallor, Icterus, Clubbing, Cyanosis, Lymphadenopathy.

CNS Examination:-

Patient is conscious, oriented to time, place and person

- Cranial nerves - Normal
- Tone- Normal in all 4 limbs

Nutrition - Normal in all 4 limbs, No wasting or hypertrophy
Sensory Supply:
Crude touch - Normal
Fine touch - Normal
Joint position - Normal
Vibration - Normal

Table 1: Discription of Power

	Shoulder (Rt/Lt)	Elbow (Rt/Lt)	Wrist (Rt/Lt)	Hip (Rt/Lt)	Knee (Rt/Lkt)	Ankle (Rt/Lt)
Flexion	4/4	5/5	5/5	4/4	5/5	5/5
Extension	4/4	5/5	5/5	4/4	5/5	5/5
Abduction	3/3	5/5	5/5	3/3	-	5/5
Adduction	3/3	5/5	5/5	3/3	-	5/5
E. Rotation	4/4	5/5	5/5	3/3	-	5/5
I. Rotation	4/4	5/5	5/5	3/3	-	5/5

Table 2: Differential Diagnosis

Infections	Inflammatory	Malignancy	Miscellaneous
• HIV • Guillan-Barre Syndrome • Botulism	• Dermatomyositis • SLE • Scleroderma • Muscular Dystrophy	• Paraneoplastic Syndrome	• Spinal Cord Injury • Spinal Cord Compression • Amyotrophic Lateral Sclerosis

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			• Multiple Sclerosis

Table 3: Investigation

	10/07/2023	**24/07/2023**
Hemoglobin [12.0-16.0g/dl]	11.9	12.3
Total RBC count [4.2-5.4mill/cmm]	4.13	4.36
Total WBC count [4000-11000/cmm]	4900	17580
Platelet count [150000-450000]	217000	371000
Absolute Neutrophil Count [1600-7000/ul]	-	15740
RDW [11.5-14.0%]	-	15

	13/07/2023	**16/07/2023**	**24/07/2023**
LDH	-	1516	-
ESR (3-12)	44	-	-
CRP (5)	29	-	-
ALT [14-63U/L]	-	-	374
AST [15-37U/L]	-	-	669
CPK [25-195U/L]	12140	6220	8428

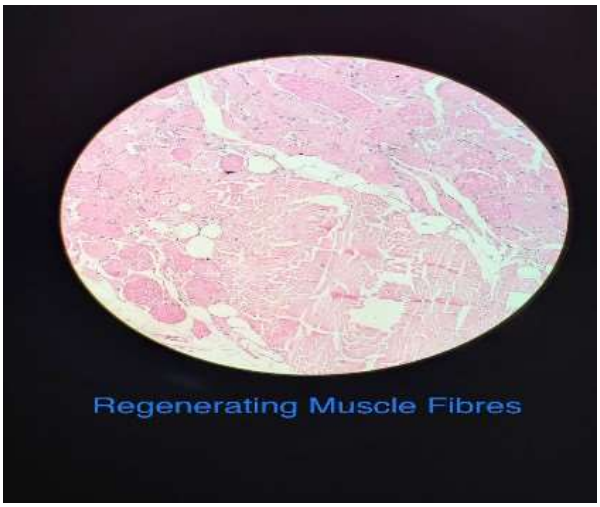
ADDITIONAL INVESTIGATION

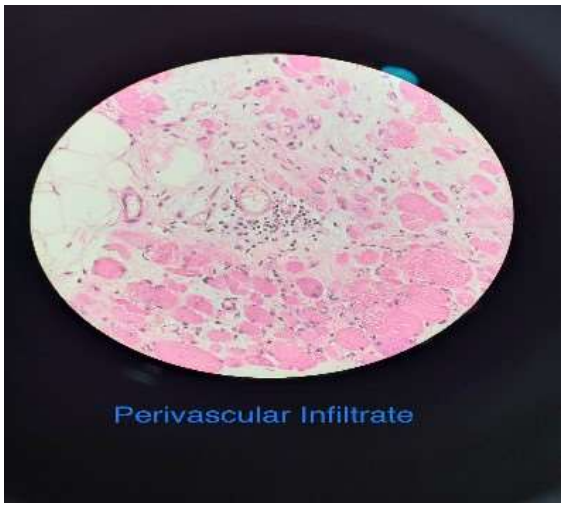
A myositis profile (IgG - against 16 antigens) was reported on 17/07/2023 which gave positive results for:

1. Anti SRP (ref - negative) - observed, 133
2. Anti PL-7 (ref - negative) - observed, 71
3. Anti RO-52 (ref - negative) - observed, 136

Biopsy:

- Muscle biopsy shows scattered necrotic fibers with inflammatory infiltrates with few regenerating fibers





Mri whole spine screening + pelvis & upper thigh screening

- Diffuse hyperintensitive signal with B/L posterior paraspinal muscles, B/L psoas muscle, B/L Iliacusmuscle, B/L pelvic girdle

Diagnosis - Immune mediated necrotizing myopathy

Treatment:

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- Steroids - Inj Methylprednisolone (500 mg) IV over 1 hr for 5 days
- Rituximab - Inj. Rituximab (500 mg) IV slowly for 6 hrs for 3days, IVIg - Inj. IVIg 1 gm/kg/day for 3 days

Other treatment that can be used-

- Azathioprine
- Cyclosporine
- Methotrexate
- Mycophenolate

Prognosis:

IMNM is the most severe idiopathic inflammatory myopathy in terms of muscle damage, and relapses are frequent. Early age of onset is a poor prognosis factor for muscle strength recovery.

DISCUSSION

In our case, A 47 year old female presented with B/L Symmetrical muscle weakness insidious in onset, proximal more than distal, lower limb more than upper limband

Muscle biopsy shows scattered necrotic fibres with inflammatory infiltrates with few regenerating fibres and Mri pelvis shows Diffuse hypertensive signal with B/L posterior Para spinal muscles ,B/L psoas muscle ,B/L Iliacus muscle, B/L pelvic girdle and with following reports- CPK Total -12140, LDH -1516, ESR-44, CRP-29, Anti SRP Ab observed suggestive of Immune mediated necrotizing myopathy.

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