Case Report

Elderly Lupus Patient Presenting with Myositis — An Uncommon Presentation of a Common Disease

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A 52-year-old married female presented with a history of generalized weakness for last 4 months aggravating over last two weeks. On clinical examination, we found Hyperpigmentation on her face and proximal muscle weakness in all four limbs, Alopecia; in routine investigations we found Anaemia, Thrombocytopenia, Raised Creatine Kinase. In imaging we found consolidation in left lower lobe, Nerve Conduction Velocity test showed brachial plexopathy and Sural Sensory Neuropathy, Electromyography showed fibrillation potentials and increased insertional activity in Gastrocnemius medial head, Tibialis anterior, Biceps brachii. We also performed muscle biopsy which showed findings suggestive of Dermatomyositis. Summarizing all the findings, we thought the cause to be an underlying Connective Tissue Disorder, hence we send samples for ANA, ENA profile which showed ANA 4+ homogenous, Ro-52+++, RNP++, SS-A++; which led us to the final diagnosis of Systemic Lupus Erythematosus.

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Key words **: Lupus, Myositis and Autoimmunity, Systemic Manifestations of Lupus, Proximal Myopathy in Elderly.**

Lupus is commonly seen in females in the child bearing age group; most of the patients present at a young age; but this patient presented with Lupus in her fifth decade that too with proximal muscle weakness as her presenting feature. As the presentation and the age; both are rare so we decided to write this case as a "food for thought" for all the clinicians that an elderly female with chronic muscle weakness can be a manifestation of Lupus.

Case Report

Our patient presented with a history of proximal muscle weakness for last four months, aggravated over last two weeks. On examination we found that she had Alopecia, Hyperpigmentation over her face and was unable to stand without aid and unable to comb her hair; muscle wasting in both lower limbs; deep tendon jerks were absent in all four limbs; she could not move her lower limbs against gravity, while she could move her upper limbs against gravity, she could not do so against resistance. In routine investigations, we detected Anaemia, Leukopenia, 650 mg proteins in 24 hours urine sample, raised Creatine Kinase. In High Resolution CT scan of thorax, we found consolidation right upper, middle segment of lower lobe. As this was a case of proximal muscle weakness in an elderly female and we had already found raised Creatine Kinase so we were thinking it to be either a case of Myositis caused by an underlying malignancy or a Connective Tissue Disorder. However, she lacked the skin manifestations of Dermatomyositis such as heliotrope rash, gottron papule making Dermatomyositis an unlikely diagnosis. As her condition was deteriorating so we started her on pulse Methylprednisolone Therapy for 3 days followed by oral steroids and was also started on Immunomodulator therapy. To look for malignancy we needed to perform imaging investigations; Contrast Enhanced CT scan of thorax showed consolidation in left lower lobe, Contrast Enhanced CT scan of abdomen showed no abnormality. Nerve conduction velocity test showed brachial plexopathy and sural sensory neuropathy; while Needle Electromyography showed increased insertional activities and fibrillation potentials in Gastrocnemius medial head, Tibialis anterior, Biceps brachii. At this point we sent samples for ANA, ENA profile; did muscle biopsy from the Gastrocnemius medial head; the results showed ANA 4+, nuclear homogenous;(Ro-52 +++, SS-A ++, RNP ++); muscle biopsy showed findings suggestive of Dermatomyositis. Hence, we concluded this to be a case of Lupus presenting as Myositis.

Editor's Comment :

- Myositis can be a presenting feature of Lupus.
- Lupus patients having myositis usually present at a young age, but in case they present at an older age they have worse prognosis than primary myositis.
- Vasculitis in Lupus may mimic myositis like changes in muscle biopsy.

Investigations

- *Haemoglobin: 8.9, TLC: 3100, Platelet:1,80,000/ cubic mm;*
- *Urine: trace proteins; 24 hours urine protein: 650 mg*
- *HBsAg, Anti HCV, ICTC: NON-REACTIVE*
- *Direct Coombs Test: Positive*
- *Chest x-ray: consolidation in left lower lobe; HRCT Thorax: Consolidation in right upper, middle segment of left lower lobe; CORADS-3; CECT Thorax:*

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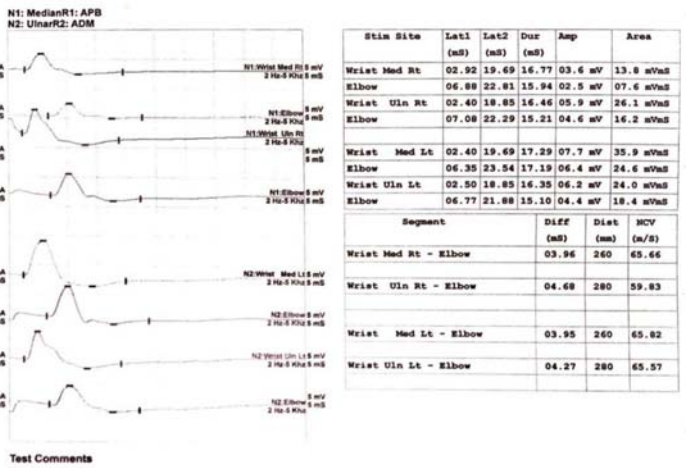
consolidation in left lower lobe

- *COVID RTPCR: NEGATIVE, Sputum for AFB, CBNAAT: NEGATIVE*
- *Serum Creatine Kinase: 8469 U/L (normal: 26-192 U/L)*
- *Nerve Conduction Velocity: Brachial plexopathy (C5, C6) and sural sensory neuropathy*
- *Electromyography: Increased insertional activities and fibrillation potentials in Gastrocnemius medial head, Tibialis anterior, Biceps brachii (Fig 1).*
- *ANA 4+, nuclear homogenous, Ro-52+++, SS-A++, RNP++.*
- *Muscle biopsy showed severe myofiber degeneration with moderate endomysial fibrosis perifascicular atrophy seen, hypertrophy seen in 20% fibers, moderate perivascular inflammation seen with Lymphocytes and Plasma cells; suggestive of Dermatomyositis (Fig 2).*



Stim Site	Lat1 (mS)	Lat2 (mS)	Dur (mS)	Amp	Area
Wrist Med Rt	02.92	19.69	16.77	03.6 mV	13.8 mVmS
Elbow	06.88	22.81	15.94	02.5 mV	07.6 mVmS
Wrist Uln Rt	02.40	18.85	16.46	05.9 mV	26.1 mVmS
Elbow	07.08	22.29	15.21	04.6 mV	16.2 mVmS
Wrist Med Lt	02.40	19.69	17.29	07.7 mV	35.9 mVmS
Elbow	06.35	23.54	17.19	06.4 mV	24.6 mVmS
Wrist Uln Lt	02.50	18.85	16.35	06.2 mV	24.0 mVmS
Elbow	06.77	21.88	15.10	04.4 mV	18.4 mVmS

Segment	Diff (mS)	Dist (mm)	NCV (m/S)
Wrist Med Rt - Elbow	03.96	260	65.66
Wrist Uln Rt - Elbow	04.69	280	59.83
Wrist Med Lt - Elbow	03.95	260	65.82
Wrist Uln Lt - Elbow	04.27	280	65.57

Fig 1 — NCV AND EMG showing brachial plexopathy, and increased insertional activities in Gastrocn

MICROSCOPY : Fatty infiltration - Absent.
Endomysial fibrosis - Moderate.
Myofiber degeneration - Severe.
Necrosis - Moderate.
Myophagocytosis - Absent.
Basophillic fibers, large nuclei - Present in 70% fibers.
Hypertrophic fibers - Present in 20% fibers.
Atrophy/Hypotrophy - Present.
All fibers within the specimen.
Perifascicular distribution - Found.
Myopathy-associated pathological structures - not present.
Inflammation - Moderate.
Perivascular.
Associated with myofiber damage.
Granulomas - Absent.
Inflammatory cells identified - Lymphocytes and plasma cells.
No abnormal storage substance identified.

IMPRESSION : **Above histological findings suggestive of active myopathy with inflammation, suggestive of Dermatomyositis.**

Fig 2 — Muscle biopsy showing perivascular inflammation with lymphocytes and plasma cells

Differential Diagnosis

Our patient presented with proximal muscle weakness; we started to progress with the case in the lines of Myositis. As we progressed the investigations revealed evidence of Myositis such as raised Creatine Kinase, Electromyography showing fibrillation potentials, as well as peripheral neuropathy suggested by Brachial plexopathy and sural sensory neuropathy on Nerve conduction velocity test. Now in this case we are dealing with an elderly patient so at this stage we were thinking that this could be a case of myositis itself or any underlying malignancy with Myositis as a paraneoplastic feature; or a Connective Tissue Disorder presenting as Myositis. Finally, we diagnosed this as a case of Systemic Lupus Erythematosus; with a rare presentation such as Myositis.

Treatment

- *Injection Methylprednisolone 1000 mg IV once daily for 3 days; followed by Tab Prednisolone 40 mg once daily*
- *Injection Cyclophosphamide 500 mg in 1000 ml normal saline over 3 hours*
- *Tab Hydroxychloroquine sulphate 200 mg at bedtime*
- *Tab Ramipril 5 mg once daily after breakfast*
- *Sunscreen lotion to be applied 30 minutes before going outside*
- *Physiotherapy as advised for the proximal muscle weakness*
- *Tab Azithromycin 500 mg once daily for 5 days·*
  Tab Rabeprazole 20 once daily in empty stomach

Outcome and Follow-up

We discharged the patient with the aforementioned medications and was asked to come at our Out-patient Department (OPD) after 4 weeks; however, we got the news of her demise 2 weeks after being discharged from our hospital.This showed us that in case of Lupus, age at the time of presentation determines the prognosis to a great extent; and it is poor prognosis in case of elderly Lupus patient.

Discussion

Myositis is a rare but significant complication of Lupus, although it is thought to be milder than primary Myositis but it has been found that Lupus myositis is often as severe as primary Myositis hence should be treated with equal vigour[1]. SLE patients of black race with childhood onset SLE, who possess myositis specific antibodies or Myositis associated antibodies should be regularly screened for Myositis[2]. The point prevalence of myositis was 2.6% in SLE patients. The significant association of Alopecia, Leukopenia and active disease with myositis suggests that organ damage, haematological abnormality, and high disease activity promote the progression of Myositis in Lupus patients[3]. We looked for Lupus cases with Myositis and in all such cases the age of the patient was below 30; hence we decided to write this case report to emphasis on the fact that an elderly patient can also present with Myositis as a feature of Lupus in her fifth decade!

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