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Case Report

Case of Polymyositis Associated with Celiac Disease: A Case Report

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ABSTRACT

Polymyositis is an important subtype of idiopathic inflammatory myopathies characterized by muscle inflammation and weakness, often associated with other systemic diseases and malignancies. A male, 29 years of age, with a history of neck stiffness, generalized weakness and body aches, has been described. Physical examination, laboratory workup, electromyography and muscle biopsy suggest Polymyositis. Tumour markers were also checked to rule out any malignancy. This report highlights the importance of immunological workup in patients presenting with generalized symptoms.

Keywords: Idiopathic Inflammatory myopathy, Polymyositis, Muscle biopsy.

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INTRODUCTION

Polymyositis (PM) is one of several idiopathic inflammatory myopathies. It is a rare disorder with an estimated prevalence of 5-22 per 100,000 persons, but its incidence increases over time due to an increase in its early detection rate.[1] Its mean age of onset is 50-60 years, with rarely occurring in children.[2]

The most commonly used criteria for PM/DM were given by Peter and Bohan, which include: proximal muscle weakness, increase in serum muscle enzymes, myopathic abnormalities on electromyography (EMG), characteristic histopathological findings on muscle biopsy and typical rash for DM.[2,3] PM is an autoimmune disease secondary to defective cellular immunity, often associated with other systemic autoimmune diseases. In addition, environmental factors like viral infections, drugs, and genetic susceptibilities in certain populations may trigger it.[4]

Muscle biopsy of involved muscles serves as a differentiating tool for diagnosing polymyositis.[5] We report a case of an adult male with biopsyproven celiac disease with Polymyositis.

CASE REPORT

The male patient, 29 years of age, was in his usual state of health two years ago when he started to develop generalized weakness and body aches. Weakness was sudden onset, progressive, and associated with pain in the upper and lower limbs with frequent neck spasms. Due to this disabling weakness, the patient faced difficulty performing daily activities at work. His past medical and family history was insignificant for any chronic ailments. He also reported to have lost 7kg weight over eight months. The patient consulted several physicians and got treated for over a year with vitamin supplements for his weakness and painkillers for his body aches.

Physical examination revealed a male adult with a lean build. The patient had no rash and was vitally stable. Joint and musculoskeletal examination revealed reduced muscle mass in the upper limbs. Weakness of proximal muscles of upper and lower limbs with a reduced power of 4/5 was noted. Muscles of neck flexion and neck extension were normal. He faced difficulty in standing up (Gower sign positive). The rest of the systemic examination was unremarkable.

The patient underwent an extensive serological and immunological workup. His serum creatine kinase level was elevated (3246U/l), alanine aminotransferase (50U/l) with normal serum levels of aspartate aminotransferase, bilirubin, urea and creatinine. Urinalysis was unremarkable, and serum electrolytes were in the normal range.

Autoimmune workup revealed elevated levels of Anti Mi-2 alpha (22AU), Anti NXP2 (50AU), Anti Ku (30AU) and Anti EJ (52AU) antibodies. However, Anti Sm, Anti RNP, Anti SSA (Ro), Anti SSB (La), Anti Jo 1, Anti Scl 70, and Anti gliadin antibodies were negative. Tumour markers were within normal limits, including PSA, CEA, CA 19-9 and AFP.

NCS and EMG studies were done, which were consistent with the myopathic process exhibiting

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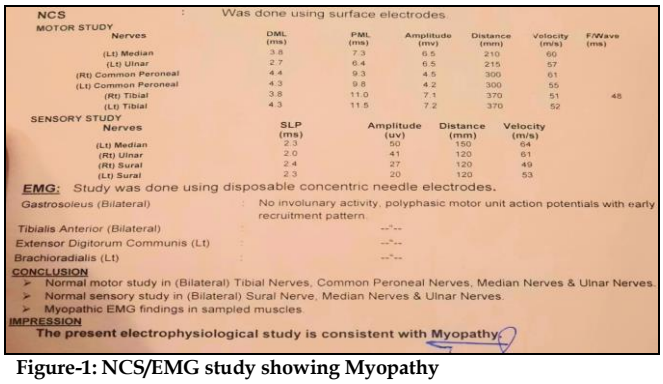
polyphasic motor unit action potentials with early recruitment patterns.

A muscle biopsy of gastroscopes muscle was taken, which showed the atrophy of individual muscle fibres with compensatory hypertrophy of surrounding fibres. Internalization of nuclei and necrosis of fibres with infiltration by histiocytes was also seen. There was minimal lymphocyte infiltration around vessels.

A duodenal biopsy was done, which showed moderate villous atrophy consistent with the findings of celiac disease. HRCT chest was normal and did not show any signs of interstitial lung disease.

The patient was started on prednisone 1mg/kg/day. This dose was gradually tapered once the patient regained remission and creatine kinase levels returned to normal. His muscle strength improved gradually. He was started on a gluten-free diet with dietary supplements.


NCS : Was done using surface electrodes.

MOTOR STUDY

Nerves	DML (ms)	PML (ms)	Amplitude (mv)	Distance (mm)	Velocity (m/s)	F/Wave (ms)
(Lt) Median	3.8	7.3	6.5	210	60	
(Lt) Ulnar	2.7	6.4	6.5	215	57	
(Rt) Common Peroneal	4.4	9.3	4.5	300	61	
(Lt) Common Peroneal	4.3	9.8	4.2	300	55	
(Rt) Tibial	3.8	11.0	7.1	370	51	48
(Lt) Tibial	4.3	11.5	7.2	370	52	

SENSORY STUDY

Nerves	SLP (ms)	Amplitude (uv)	Distance (mm)	Velocity (m/s)
(Lt) Median	2.3	50	150	64
(Rt) Ulnar	2.0	41	120	61
(Rt) Sural	2.4	27	120	49
(Lt) Sural	2.3	20	120	53

EMG: Study was done using disposable concentric needle electrodes.

Gastrosoleus (Bilateral) : No involuntary activity, polyphasic motor unit action potentials with early recruitment pattern.

Tibialis Anterior (Bilateral) : --"--

Extensor Digitorum Communis (Lt) : --"--

Brachioradialis (Lt) : --"--

CONCLUSION

- Normal motor study in (Bilateral) Tibial Nerves, Common Peroneal Nerves, Median Nerves & Ulnar Nerves.
- Normal sensory study in (Bilateral) Sural Nerve, Median Nerves & Ulnar Nerves.
- Myopathic EMG findings in sampled muscles.

IMPRESSION

The present electrophysiological study is consistent with Myopathy.

Figure-1: NCS/EMG study showing Myopathy

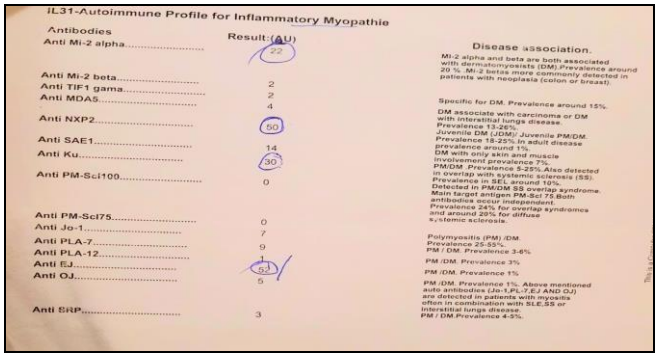


Figure-2: Autoimmune Profile for Inflammatory Myopathy

DISCUSSION

Inflammatory myopathies have occasionally been reported with other autoimmune diseases and malignancies.[6] Studies have reported a high incidence of celiac disease in inflammatory myopathies. The mechanism involved is believed to be due to autoimmunity involved in the pathogenesis of these conditions. Scientists have found an association of HLA B8-DR3 between celiac disease and inflammatory myopathies.[7]

Polymyositis has several systemic manifestations, including rheumatoid-like arthralgias and myalgias. It may involve the lungs leading to interstitial lung diseases.[8] Cardiac involvement is rare but severe and has a poor prognosis. In rare cases, patients may develop dysphagia which may be treated with IVIG. Early detection of this disease is vital to its management. Therapy aims to reduce disease severity, slow its progression and improve patients' quality of life.[9,10]

Poor prognostic factors are delayed recognition and treatment of disease, cardiac involvement and dysphagia. Therefore, patients are closely followed up for adverse reactions to steroids with laboratory workups after every 2-3 weeks to document creatinine kinase levels and check muscle strength.

Authors Contribution

Following authors have made substantial contributions to the manuscript as under:

ARA & MA: Conception, drafting the manuscript, approval of the final version to be published.

NA & RT & AA: Critical review, data acquasation, drafting the manuscript, approval of the final version to be published.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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