Amyloid myopathy as the presenting feature of lymphoplasmacytic lymphoma

Sir,

Amyloid myopathy is a rare manifestation of systemic amyloidosis.[1] We report a case of Amyloid myopathy with underlying lymphoplasmacytic lymphoma that presented with clinical features closely mimicking motor neuron disease. A 61-year-old male with a past medical history of hyperlipidemia, atrial fibrillation, and congestive heart failure presented with ascending weakness, dysphagia, dyspnea, and unintentional weight loss for 2 months preceding hospitalization.

The patient denied sensory symptoms or sphincteric dysfunction. He gave no family history of neuromuscular disorders. Medications on admission were warfarin, furosemide, and amiodarone.

On examination, mentation was clear; cranial nerves were intact. Tongue did not show atrophy or fasciculations, and bulbar function was clinically intact. Muscle strength: proximal upper and lower extremities were 4-/5, and distal upper and lower extremities were 4/5. Frequent fasciculations were noted in upper extremity muscles. Deep tendon reflexes were hypoactive with plantar responses flexor bilaterally. Sensory examination was intact other than decreased vibration distal to ankle. Cerebellar function was normal. The patient was unable to ambulate without assistance.

Based on the presentation of progressive weakness, frequent fasciculations in extremity muscles, dysphagia, and dyspnea, the patient was initially suspected to have motor neuron disease.

Basic laboratories showed normal routine blood count and comprehensive metabolic panel. Creatine kinase was 164, lactate dehydrogenase 212, Vitamin B12 350, thyroid-stimulating hormone 2.2. HIV screen, acetylcholine receptor antibodies were negative.

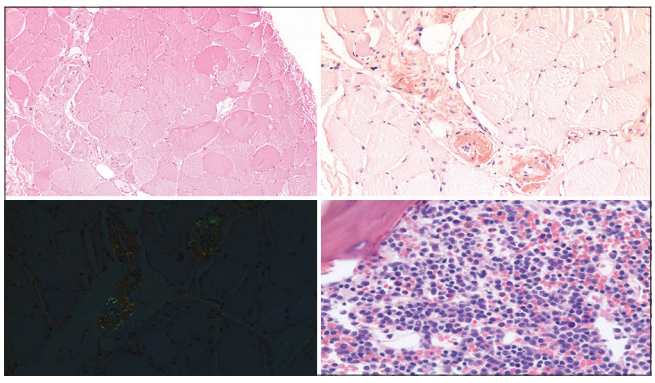

Figure 1: Top left: H and E, stain: Marked fiber size variation with frequent angulated atrophic fibers. There is no active degenerating/regenerating muscle fiber/inflammation. Top right: Congo red stain: prominent amyloid deposit in the connective tissue and vascular wall. Bottom left: Polarized amyloid deposits with Congo red stain: Muscle. Bottom right: Bone marrow biopsy: 25% of the biopsy is involved by a diffuse and interstitial lymphoplasmacytic infiltrate. Magnification ×175

Serum protein electrophoresis revealed hypoalbuminemia. Urine protein electrophoresis showed monoclonal gammopathy. M-spike was 659 mg/24 h. Immunofixation electrophoresis showed free lambda light chain monoclonal gammopathy (108 mg/dl) with free kappa free lambda ratio of 0.01. Anti-GM1, anti-GD1, Sulfate-3-Gluc.Parag. Ab IgM and anti-MAG antibodies were negative. Nerve conduction studies were normal. Electromyogram (EMG), all the muscles revealed diffuse increased insertional activity in the form of fibrillation potentials and positive sharp waves. Multiple muscles revealed predominance of myopathic potentials, i.e., small motor unit potentials with increased polyphasia and early recruitment. The EMG was not typical for motor neuron disease, and it prompted a muscle biopsy which showed marked fiber size variation with frequent angulated atrophic fibers and no inflammation. Amyloid deposits were noted in the connective tissues and vascular wall confirmed by Congo red stain and amyloid immunostaining, findings consistent with amyloid myopathy [Figure 1].

The patient underwent bone marrow biopsy which revealed evidence of lymphoplasmacytic lymphoma. He started chemotherapy but remained weak after multiple cycles.

Amyloid myopathy is a rare condition that can be found in patients with hematopoietic malignancies or patients with hereditary primary amyloidosis.[1,2] It usually afflicts middle-aged to elderly patients, manifesting as gradually progressive weakness.[3] Optimal use of fluorescent Congo red stain has improved the sensitivity on muscle biopsy.[4] There are reports of amyloid myopathy masquerading as polymyositis.[5] We report a case where patient's presentation closely mimicked motor neuron disease. EMG findings and subsequent muscle biopsy was instrumental in making the correct diagnosis. Since motor neuron disease is incurable and fatal disease, it is imperative to rule out other neuromuscular disorders, and amyloid myopathy should be considered in the differential diagnosis.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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Quick Response Code:

Website: www.annalsofian.org

DOI: 10.4103/0972-2327.194303

How to cite this article: Guduru Z, Purohit A, Pu C, Rana S. Amyloid myopathy as the presenting feature of lymphoplasmacytic lymphoma. Ann Indian Acad Neurol 2017;20:77-8.

Received: 06-06-16, **Revised:** 29-07-16, **Accepted:** 04-09-16