A Rare Case of Sporadic Inclusion Body Myositis with Atypical Presentation

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Introduction

Sporadic inclusion body myositis (IBM) is the most common acquired inflammatory myopathy that occurs after the age of 50 years. IBM typically involves wrist and finger flexors and quadriceps, but all sporadic IBM may not have the classic presentation of distal arm and proximal leg involvement. Treating physicians must be aware of this atypical presentation to avoid the misdiagnosis of IBM, leading to treatment with immunosuppressive agents. The aim of this study is to increase the awareness among physicians about the atypical presentation of IBM and to emphasize the importance of muscle biopsy in such cases. Here we report a case of 52 years old male diagnosed with sporadic IBM by muscle biopsy presented with atypical presentation.

Case Report

A 55-year-old male presented with complaints of insidious onset of symmetrical flaccid weakness (proximal more than distal) involving bilateral upper and lower limb for the past 6 months. On further questioning, the patient described difficulty in standing and getting up from squatting or sitting posture. He also developed difficulty in climbing stairs, raising hands above the head, and difficulty in combing hairs. He also had erythematous skin rash over bilateral nasolabial fold, anterior aspect of chest, and back of the neck for the past 6 months.

On examination, he had the following pattern on muscular strength testing: neck flexors were 5/5, bilateral shoulder abduction (supraspinatus) was 3/5; bilateral shoulder adduction (pectoralis major and minor) was 3/5; bilateral shoulder flexion and extension (deltoid) were 3/5, bilateral elbow flexion (biceps) and extension (triceps) were 3/5; bilateral wrist flexion (flexor carpi ulnaris, flexor carpi radialis, flexor digitorum) and wrist extension (extensor carpi radialis, extensor carpi ulnaris, brachioradialis) were 3/5; bilateral ankle dorsiflexion and plantarflexion were 3/5; bilateral quadriceps and hamstrings were 3/5; bilateral intrinsic foot muscles were 3/5. There was no fasciculation, distal weakness, or atrophy. Muscle biopsy revealed rimmed vacuoles, endomysial lymphonuclear infiltrates, and endomysial fat infiltration.

Keywords
► sporadic inclusion body myositis
► endomysial lymphonuclear infiltrates
► rimmed vacuoles
► fat infiltration
► creatine kinase

Abstract

Sporadic inclusion body myositis (IBM) is the most common acquired inflammatory myopathy that occurs after the age of 50 years. IBM typically involves wrist and finger flexors and quadriceps, but all sporadic IBM may not have the classic presentation of distal arm and proximal leg involvement. Treating physicians must be aware of this atypical presentation to avoid the misdiagnosis of IBM, leading to treatment with immunosuppressive agents. The aim of this study is to increase the awareness among physicians about the atypical presentation of IBM and to emphasize the importance of muscle biopsy in such cases. Here we report a case of 52 years old male diagnosed with sporadic IBM by muscle biopsy presented with atypical presentation.

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Fig. 1  (A and B) Biopsy showing atrophy, multifocal rimmed vacuoles, endomysial lymphonuclear infiltrates involving the nonnecrotizing fibers.

Fig. 2  (A) T2-weighted short tau inversion recovery imaging shows diffuse hyperintensity noted in shoulder girdle muscles and edema noted in the intramuscular plane. (B) T2-weighted axial image shows high intensity in the muscle suggestive of chronic inflammation with fat infiltration. In addition, a nerve biopsy from the sural nerve showed axonal neuropathy.

Finally, the patient was diagnosed with sporadic IBM with an atypical presentation, and the patient was started on corticosteroids. He improved symptomatically and was on regular follow-up.

Discussion

Sporadic (IBM) is one of several chronic adult inflammatory myopathies. Its prevalence varies, but it may be as high as 35 per 1 million adults over age 50, with a slight male predominance. IBM typically manifests as slowly progressive weakness of quadriceps muscle more than hip flexors leading to frequent falls or difficulty in standing and next common problem would be finger flexor weakness leading to loss of dexterity.

However, all sporadic IBM may not have the classic presentation of distal arm and proximal leg involvement. In this case, it can present with symmetrical weakness where the proximal group of muscles involved more than the distal group of muscles without complications like dysphagia and most commonly misdiagnosed as polymyositis or dermatomyositis.

In such case, muscle biopsy and imaging of muscle play an important role in diagnosis. The recent identification of a serum autoantibody against anti-5′-nucleotidase, cytosolic IA in patients with sporadic IBM has offered a new clinical tool.

Anticytosolic 5′-nucleotidase is a highly specific diagnostic marker for IBM among patients with myopathy. Other blood biomarkers for IBM include an abnormal population of large granular lymphocytes on flow cytometry and a reduced CD4/CD8 ratio with an increased CD8 count.

High-dose corticosteroids are considered the first-line treatment. Unfortunately, IBM does not typically respond to any known immunotherapies. The mainstay of treatment is physical and occupational therapy to improve function and swallowing therapy. Patients with dysphagia may benefit from intravenous immunoglobulin therapy, along with esophageal balloon dilation or cricopharyngeal myotomy. Most patients require a wheelchair within 10 to 15 years of onset of symptoms. Life expectancy is not significantly altered in IBM.
Conclusion

Sporadic IBM with atypical presentation is still difficult to diagnose and unfortunately remains frequently misdiagnosed; in such cases, muscle biopsy and muscle imaging play an important role in diagnosis. Although sporadic IBM is rare and without effective therapy, accurate diagnosis is crucial to providing adequate patient counseling and information about the prognosis and course of the disease. Patients with IBM are highly motivated and should be encouraged to participate in clinical trials.

Conflict of Interest

None declared.

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