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Case Report
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Inclusion Body Myositis: A Demonstrative Case and Diagnostic Challenges

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Abstract

In this case, we present a case of a 65 year old patient with progressive muscle weakness, with asymmetric onset and progressive muscle weakness of limbs which an often be mistaken for amyotrophic lateral sclerosis. But Biopsy of muscle is often helpful in reaching a diagnosis.

Keywords: Inclusion Body Myositis (IBM); Proximal girdle weakness; Forearm muscle weakness; Asymmetric weakness of limbs; Inflammatory myopathy; Rimmed Vacuoles; Muscle weakness; Elevated CPK

Introduction

Inclusion Body Myositis (IBM) should be suspected in any patient with asymmetric proximal and distal muscle weakness and atrophy, especially those with prominent involvement of finger and wrist flexors in the upper extremities and/or quadriceps muscles in the lower extremities [1]. Definitive diagnosis is desired as IBM is refractory to existing treatments effective against other inflammatory myopathies, but requires muscle biopsy confirmation given nonspecific EMG findings, lack of diagnostic laboratory testing, and variable symptomatology which may overlap with other myopathic disorders.

Case Report

A 65-year-old male was referred to neurology clinic by his rheumatologist for a 4 year history of progressive muscle weakness and atrophy. The patient initially noted weakness of the right hand, but symptoms were otherwise symmetric, with proximal and distal involvement of all extremities. No temporal pattern or precipitating factors were identified. Review of systems was positive for Raynaud phenomenon, myalgia, arthralgia, numbness, paresthesia, and mild dysphagia for solids and liquids.

Home medications during the disease course included amlodipine, hydrochlorothiazide-olmesartan, clonazepam, gabapentin, ibuprofen, omeprazole, sildenafil, and as needed acyclovir. He had also completed several courses of antibiotics for a longstanding history of bilateral lower extremity lymphedema with intermittent episodes of cellulitis. There was no history of respiratory or cardiac disease. Family history was significant only for a brother with multiple sclerosis limited to an isolated clinical event in the remote past.

On exam, the patient's hands and feet were erythematous, though not distinctly over the joint extensor surfaces. There was no facial or truncal rash. Severe atrophy was noted throughout the upper extremities bilaterally. The lower extremities were also atrophic despite massive pitting edema being present. No fasciculations were observed. Muscle tone was normal. Weakness was most prominent in the distal upper extremities, with considerable difficulty in finger flexion. All major muscle groups had 4- to 4/5 strength bilaterally with the exception of knee extension and plantarflexion, which were near normal. There was no proximal to distal gradient or lateralizing weakness. Truncal musculature was extremely weak such that the patient lacked power to rotate his neck or thorax. Deep tendon reflexes were normal in the upper extremities and diminished in the lower extremities, with inconsistently obtained patellar reflexes and 1+ Achilles bilaterally. Plantar reflex was flexor bilaterally. Sensation was decreased to vibration in the distal lower extremities. He was unable to rise from a chair unassisted and had a waddling gait with bilateral foot drop, requiring a walker to ambulate.

Results

Per records, creatinine kinase (CPK) was only minimally elevated (290 U/L, normal range 24-204 U/L) at presentation. Aldolase was within normal limits. A limited myositis panel including fibril U3 RNP, MDA-5, NSP-2, and TIF1-GAMMA enzyme-linked immunosorbent assays was negative. Atypical perinuclear

anti-neutrophil cytoplasmic antibody (ANCA) was mildly positive at a titer of 1:160; otherwise, ANCA panel was negative. Antinuclear antibodies were negative.

Additional workup was limited by the patient's reluctance for any invasive procedures or even further laboratory testing. He could not tolerate electromyography (EMG) and refused muscle biopsy. MRI brain revealed numerous nonspecific, non-enhancing white matter lesions in pericallosal, periventricular, and juxtacortical distributions with infratentorial involvement, suggestive of a demyelinating process. MRI c-spine was negative for similar lesions or central stenosis. He declined lumbar puncture.

Based on clinical history, exam, and limited data, differential diagnosis included an inflammatory myopathy such as IBM, dermatomyositis or polymyositis versus amyotrophic lateral sclerosis (ALS) or muscular dystrophy. Though less likely, the patient did meet radiographic criteria for clinically isolated syndrome as well. No treatment was started as IBM was the presumptive diagnosis. He subsequently agreed to muscle biopsy, which showed evidence of an acquired inflammatory myopathy with rimmed vacuoles (Figure 1), confirming the diagnosis of IBM.

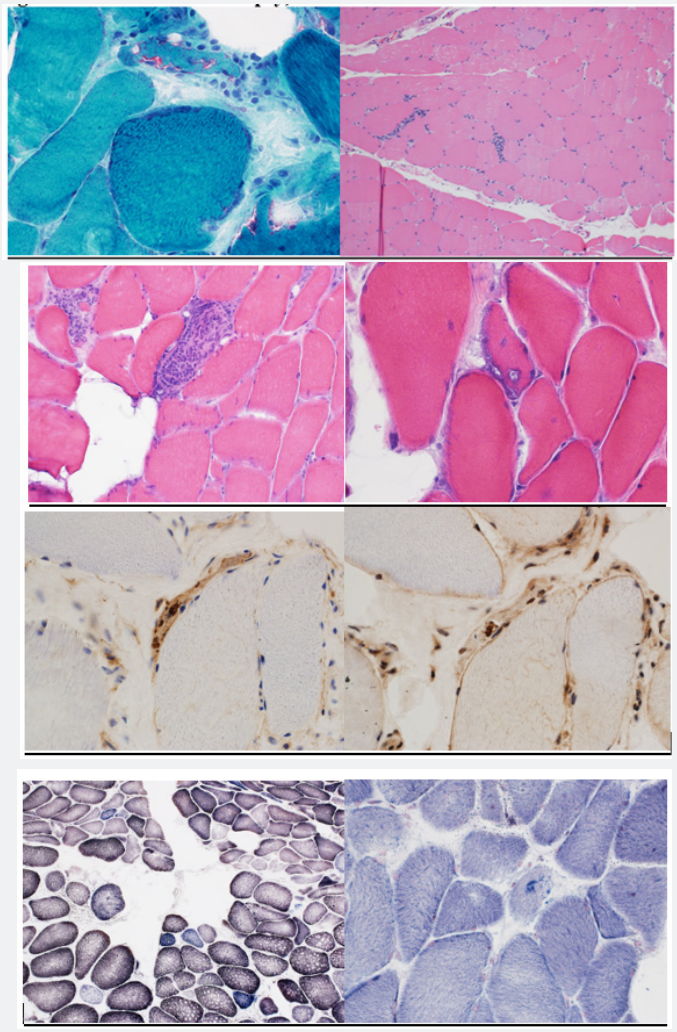

Figure 1: Skeletal muscle biopsy, left deltoid.
A. Paraffin section H&E stained: Mild chronic inflammation within the endomysium (blue arrows) and few regenerating fibers (black arrows).
B. Frozen section H&E: Mild patchy chronic inflammation within the endomysium (black arrows).
C. Frozen section H&E: Rare muscle fibers with rimmed vacuoles (black arrow).
D. Frozen section modified Gomori Trichrome: Rare muscle fibers with rimmed vacuoles (black arrow). E. Frozen section myoadenylate deaminase (MAD): Highlights rimmed vacuoles (black arrow).
E. Frozen section TDP43 immunohistochemistry: Rare muscle fibers with TDP43 positive inclusions (black arrow).
F. Frozen section p62 immunohistochemistry: Rare muscle fibers with p62 positive inclusions (black arrow).
G. Frozen section combined COX SDH stain: Increased number of blue colored COX-negative muscle fibers (black arrows).

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Discussion

IBM symptomatology, laboratory data and EMG findings are overall nonspecific and may be confounded by medical comorbidities in the individual patient. Definitive diagnosis is essential to exclude a treatable myopathy or alternate cause of muscle weakness that may have a less insidious course such as ALS, but relies solely on muscle biopsy. Serum cytosolic 5-nucleotidase 1A IgG antibody testing offers a specificity of 80-90% but has low sensitivity and is also associated with other forms of myopathy [2]. Noninvasive methods of diagnosis such as forearm ultrasound are being explored [3] and would be useful.

References

1. Needham M, Mastaglia FL (2016) Inclusion body myositis. In International Neurology RP Lisak, DD Truong, et al. (eds).
2. Herbert MK, Stammen-Vogelzangs J, Verbeek MM, Anke Rietveld, Ingrid E Lundberg, et al. (2016) Disease specificity of autoantibodies to cytosolic 5′-nucleotidase 1A in sporadic inclusion body myositis versus known autoimmune diseases. Ann Rheum Dis 75(4): 696-701.
3. Karvelas KR, Xiao T, Langefeld CD, Francis OW, Sapna Pathak, et al. (2019) Assessing the accuracy of neuromuscular ultrasound for inclusion body myositis. Muscle Nerve 59(4): 478-481.

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