Inclusion Body Myositis: A Diagnostic Dilemma

Brianna Cunningham DO, Shawn Mathew DO, Mahmood Danishwar MD, Robert Libera DO, Christopher Reggio DO

Introduction

Inclusion body myositis (IBM) is a rare disease, with a prevalence of 15-70 cases per million adults. Presentations of IBM are insidious often making it difficult to diagnosis in the landscape of other inflammatory myopathies. Current proposed diagnostic criteria, based on expert opinion, includes a muscle biopsy. We present a case of suspected IBM with classic findings where the harm of biopsy was deemed to outweigh the benefit.

Case Description

A 68-year-old male presented to the hospital for acute hypoxic and hypercapnic respiratory failure and aspiration pneumonia. His past medical history included progressive weakness in bilateral lower extremities. Fifteen years ago, the patient first noticed subtle bilateral calve weakness. This manifested as difficulty with prolonged walking or standing. He had no alterations in sensation and reflexes were 2+. After ten years, weakness progressed to the upper extremities. Spine imaging did not demonstrate any pathology. His progressive, symmetric, painless weakness which spared sensation, raised suspicion for IBM. Additionally, he had finger flexor weakness and bilateral foot drop. He had mildly elevated CK level of 339. Given characteristic findings for IBM, antibody testing was performed and came back positive for NT5CA1. The disease is progressive and does not have any definitive treatment. The patient was trialed on a steroid course without any improvement, unlike other inflammatory conditions such as polymyositis, dermatomyositis, or polymyalgia rheumatica. He was treated for pneumonia but continued to have significant coughing, and was then determined to have significant dysphagia. This is a

late finding of IBM. A multidisciplinary team (MDT) including rheumatology, neurology, and pulmonology determined that with his progressive course, a biopsy may only introduce harm without altering management, since he previously failed to respond to steroids. The patient was recommended to remain NPO and chose to pursue hospice care upon discharge.

Discussion

IBM classically presents with progressive proximal and distal muscle weakness without sensory abnormalities and can eventually progress to dysphagia. This patient's clinical findings, positive antibody, and lack of response to steroids were highly indicative of IBM. While current, available guidelines recommend biopsy, a MDT determined that the harm would outweigh the benefit.