Inclusion Body Myositis

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KEYWORDS

• Inclusion body myositis • Idiopathic inflammatory myopathies • Polymyositis
• Diagnosis • Pathology • Pathophysiology • Treatment • Prognosis

KEY POINTS

- Inclusion body myositis (IBM) is the most common inflammatory myopathy after age 50 years.
- Despite similarities with polymyositis (PM) inflammatory disorders, IBM histopathology shows marked degeneration and protein aggregation.
- The clinical phenotype of typical IBM is distinctive, manifesting as proximal leg or distal arm weakness, although in our experience there are several phenotypic variants.
- IBM is refractory to all known immunosuppressive therapies.
- Low-intensity exercise may slow the rate of functional decline.
- Patients with IBM are highly motivated and should be encouraged to participate in clinical trials.

EPIDEMIOLOGY

Inclusion body myositis (IBM) is a rare sporadic disorder with a male/female ratio of 2:1 to 3:1. Data on prevalence and incidence of IBM vary depending on methodology, countries, and regions. In Western Australia, the overall prevalence was 9.3 per million, whereas the age-adjusted prevalence of IBM in people more than the age of 50 years is 3.5 per 100,000, making it the most common idiopathic inflammatory myopathy (IIM) in this age group.[1] A recent study from South Australia yielded a higher overall prevalence rate of 51 per million, with an incidence of 2.9 per million.[2] The latter is comparable with the incidence observed in Sweden (2.2 per million)[3] but lower than the 7.9 per million recorded in Olmstead County, Minnesota, adjusted for sex and age to the

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2000 US Census population.[4] The highest reported prevalence is 71 per million inhabitants of Olmsted County, Minnesota, whereas it is lowest in the Netherlands at 4.9 per million and, when age-adjusted to those older than 50 years, it is 16 per million Dutch inhabitants.[3] IBM is rare in African Americans and in nonwhite people. IBM should be considered in patients with appropriate symptoms who are older than 30 years. Symptom onset before age 60 years occurs in 18% to 20% of patients.[5,6]

Clinical Presentation

In most cases, IBM classically presents with insidious weakness in the proximal leg and/or distal arm.[7,8] There is typically a 5-year to 8-year delay in presentation and diagnosis.[3,5,7,9–11] In the University of Kansas Medical Center (KUMC) IBM series fulfilling pathologic criteria for probable IBM (**Table 1**), delay to diagnosis in 51 cases ranged from 1 to 15 years (mean, 5.1 years).[12] IBM typically manifests as slowly

Table 1
Retrospective chart review of IBM from 2000 to 2010 at KUMC

Male/female ratio	1.7:1
Ethnicity (n = 51)	49 white; 2 Hispanic
Mean age at onset (y)	61 (45–80)
Symptom onset before age 50 y (%)	12
Mean time to diagnosis (y)	5.1 (1–15)
Mean follow-up period (y)	2.5 (0.5–8)
CK (IU/L)	609 (59–3000)
Nerve conductions with axon loss neuropathy (%)	32
Electromyography (%)	60 irritative myopathy 12 nonirritative myopathy 28 mixed neuropathic/myopathic pattern
Asymmetry (%)	90
Nondominant side weaker (%)	85
Typical phenotype: FF and (quads)	39/51 (76%): 13: classic phenotype (FF and quads weakest) 11: classic FF, no preferential quads weakness 6: classic quads, no preferential FF weakness 9: FF and quads weak but not weakest
Atypical phenotype	12/51 (24%): 5/12: classic FF with leg weakness sparing quads 4/12: limb-girdle weakness 3/12: other atypical phenotypes (FF arm only, hip flexion/ankle dorsiflexion, facioscapulohumeral)
Muscle pathology	43: inflammation and rimmed vacuoles 8: phenotypic IBM with inflammation
Mobility outcome	75%: recurrent falls 56%: assistive device use at mean 7.5 y 20%: wheelchair or scooter
Bulbar dysfunction (%)	51 dysphagia 55 facial weakness

Abbreviations: CK, creatine kinase; FF, finger flexor; quads, quadriceps.
Adapted from Estephan B, Barohn RJ, Dimachkie MM, et al. Sporadic IBM: a case cohort. J Clin Neuromuscul Dis 2011;12(3):18–9; with permission.

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