Inclusion body myositis

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1 Inclusion body myositis is the most common type of late-onset inflammatory myopathy

Inclusion body myositis is an idiopathic inflammatory myopathy. Whether its cause is primarily autoimmune or degenerative is debated. Its prevalence is 18 per 100 000 people, higher than dermatomyositis or amyotrophic lateral sclerosis (ALS). Inclusion body myositis predominantly affects males (3:1 to females), usually becoming symptomatic after age 50 years.[1]

2 Typical features are insidious, painless, and progressive weakness and muscle atrophy that is asymmetric and multifocal

Inclusion body myositis affects proximal and distal muscles, causing striking weakness of the quadriceps and finger flexor muscles.[2] This leads to falls and loss of grip strength. Over time, foot drop and dysphagia also become prominent.

3 Electromyography (EMG) and muscle biopsy are the leading diagnostic tools

Serum levels of creatinine kinase are only moderately elevated at 300–2000 U/L. Needle EMG may show equivocal findings that can suggest both myopathy and neuropathy. Current auto-antibody tests (anti-NT5C1A) have high specificity (90%) but only moderate sensitivity (40%); results must be interpreted in the clinical context.[3] A muscle biopsy should be performed, which may reveal inflammation, rimmed vacuoles, and congophilic inclusions; interpretation requires expertise in neuromuscular pathology.[4]

4 Differential diagnosis includes motor neuron disease, other inflammatory myopathies, and late-onset muscular dystrophies

Lack of fasciculations and myopathic findings on EMG distinguish inclusion body myositis from motor neuron disease such as ALS. Asymmetry, selective weakness of finger flexors and muscle biopsy findings distinguish inclusion body myositis from other inflammatory myopathies or muscular dystrophies. Most patients benefit from referral to a neuromuscular specialist.

5 Treatment is supportive

No immunosuppressive therapy has been proven to be helpful for inclusion body myositis, in contrast to other forms of myositis. Progressive motor disability causes 60% of patients to require a wheelchair at 10 years. Life expectancy is, however, only minimally reduced. Patients do not usually require tube feeding or assisted ventilation. Management focuses on adaptive measures, home exercises, and optimized nutrition.[5]

References

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