General practitioner information on sporadic inclusion body myositis (sIBM)

Sporadic inclusion body myositis (sIBM) is a rare, relentlessly progressive multifactorial disorder of skeletal muscle cells, leading to severe atrophy and weakness.

Incidence: sIBM is an age-related disease, usually appearing after age 50. It is the most common acquired muscle disorder seen in older patients although about 20% of cases display symptoms before the age of 50. It is more common in men, (2 to 3 males to 1 female). Its prevalence is estimated at between 4.3 and 9.3 per 1,000,000, (from ~1500 to 3100 cases diagnosed per year overall) rising to 35.3 per 1,000,000 for people over 50 (~2900 cases diagnosed per year in over 50s). Total IBM patients alive today in Canada and USA estimated at from about 30,000 to 62,000).

Differential diagnoses: IBM is commonly initially diagnosed as polymyositis. A course of prednisone is typically completed with no results and eventually sIBM is confirmed. sIBM weakness comes on over months or years and progresses steadily, whereas polymyositis has onset of weeks or months.

Diagnosis: Elevated CK levels (10 times normal) are typical in sIBM but patients can also present with normal CK levels. Electromyography (EMG) studies will display abnormalities. Muscle biopsy is necessary to confirm diagnosis with several common findings including; finding inflammatory cells invading the muscle cells, vacuolar degeneration, inclusions or plaques of abnormal proteins (primarily beta amyloid), abnormal strands of phosphorylated tau protein ("paired-helical filaments" PHFs) that react to an immunocytochemical stain called SMI-31 monoclonal antibody (gold standard diagnosis).

Pathology: sIBM muscle has two major aspects, 1) autoimmune (with invasion by CD8+ lymphocytes of muscle fibres expressing MHC-I antigens) and 2) degenerative (cytoplasmic and intranuclear inclusions containing amyloid beta). These processes occur in parallel.

Presentation: Age of onset may vary from the early forties to the nineties and the rate of progression of sIBM may also vary widely. The quadriceps muscles are commonly affected first. Weakening of the deep finger flexor and wrist flexor muscles is a common early symptom. sIBM commonly leads to major or "total disability" within 10 to 15 years of symptom onset, in up to 60% of cases, patients also develop dysphagia. Examination should rule out diaphragmatic involvement.

Genetics: sIBM is apparently not caused by one genetic mutation but it has a clear genetic association with genes in the major histocompatibility complex (MHC) and is one of the strongest HLA–disease connections recorded: present in ~75% of sIBM cases.

Treatment/management: sIBM patients do not respond to the anti-inflammatory, immunosuppressant, or immunomodulatory drugs available today. There is no established therapy to slow or stop the progression of the disease. Management is symptomatic – a preventive strategy should be used as sIBM patients are very vulnerable to injury from falling.


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