Information for General Physicians about inclusion body myositis (July/2019).

Introduction: Inclusion body myositis (IBM) is the most frequently acquired muscle disease seen after age 50. It is a poorly understood, relentlessly progressive disorder. Characteristic weakness and atrophy of finger flexor and quadriceps muscles develop over months or years, leading to profound disability (wheelchair use). No effective treatment exists. IBM may be an autoimmune disorder with large numbers of clonal, highly differentiated cytotoxic CD8+ T cells found in muscle cells. Muscle cells also show degenerative aspects.

Incidence: IBM is age-related. Typical onset is 61-68 with wide variation: 20% show symptoms before 50, some in the 30's. Prevalence is 46 per million in the overall population.

Presentation: IBM has unique clinical and pathological features (finger flexor and quadriceps weakness and the presence of CD8+ T cells invading muscle cells). Presentation varies widely. People are affected in different ways, to different degrees, and at various rates. Progression tends to be more rapid in men and in those with late onset. The quadriceps are often affected first; toe drop, falling, and tripping are common first symptoms. Walking difficulties typically result from knee buckling, owing to knee extensor weakness, or tripping owing to ankle dorsiflexion weakness. For some, IBM begins with the finger flexors causing weakness in wrists and fingers; difficulty pinching, buttoning, making a fist, and gripping objects. Relative sparing of shoulder and hip abductors, and neck muscles. May weaken the diaphragm but does not directly affect heart muscle. Muscle pain is not noted in the literature but commonly reported by patients. Seen more often in men (1.6 m to f ratio).

Diagnosis: Usually diagnosed by a neurologist by clinical exam, EMG, biopsy, MRI, and serology. IBM is often initially misdiagnosed as polymyositis or arthritis: average from symptom onset to correct diagnosis is 4.6 to 5.8 years. IBM weakness comes on over months or years and progresses steadily (polymyositis has a much faster onset). Other forms of muscular dystrophy (e.g. limb girdle, Becker) and ALS must be ruled out. IBM has a unique clinical presentation but many doctors do not recognize this unique presentation.

Treatment: Standard of care for most patients with IBM involves strictly nonpharmacological management, including emotional support, physical therapy, education on fall precautions and exercise, and dysphagia evaluation. Dysphagia can be treated transiently with pharyngoesophageal dilatation or cricopharyngeal myotomy. Few physicians routinely prescribe immunosuppressant therapies, with the belief that such therapies result in transient responses at best.

Management: Evaluation and monitoring of potential complications is vital (depression, fatigue, dysphagia and respiratory involvement, etc.). Integration of palliative care is helpful at end stages.

Progression: IBM is a progressive disease, on average, from symptom onset, in 7.5-10 years patients require assistive devices (cane, walker, rollator) and within 13-15 years, patients usually require a power wheelchair.

Complications: Cause of death commonly relates to respiratory dysfunction, aspiration, or dysphagia. Many patients develop progressive, often severe dysphagia. Respiratory dysfunction caused by diaphragmatic weakness may present as sleep apnea—hypoventilation causes hypercapnia. Comorbidities in IBM patients include higher rates of hypertension (66% vs 22%), hyperlipidaemia (47% vs 18%), diabetes mellitus (34% vs 10%), myocardial infarction (13% vs 2%), congestive heart failure (17% vs 5%), pneumonia (11% vs 2%; aspiration pneumonia, 6% versus 0.3%) and anaemia (32% versus 7%).

Pathology: T cells, myeloid dendritic cells, macrophages and plasma cells all invade IBM muscle, but it is the infiltration of muscle by CD8+ T cells that is the most obvious microscopic feature of IBM muscle.

Causes: No cause or initial trigger event is known. Major theories: inflammation-immune reaction or degenerative protein disorder. Complex interplay of environment, genetic predisposition and aging is implied. IBM is not a genetic disorder but may be predisposed by genetic factors.

More information: Please see http://www.ibmmyositis.com .

Written by Bill Tillier. Reference: https://www.nature.com/articles/s41584-019-0186-x