Introduction: Inclusion body myositis (IBM) is the most frequently acquired muscle disease seen after age 50. It is a type of muscular dystrophy: 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. It appears that IBM is a type of autoimmune disorder with large numbers of clonal, highly differentiated cytotoxic CD8+ T cells found in muscle cells and blood but muscle also shows 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 cell-killing 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. For some, IBM begins with the finger flexors causing weakness in wrists and fingers, causing 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. Commonly 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 tends to have a much faster onset). Other forms of muscular dystrophy (e.g. limb girdle, Becker) as well as ALS must be ruled out. IBM has a unique presentation seen on examination (above) but many doctors do not recognize these unique physical features.

Treatment: No standard course of treatment has been shown to be effective or even helpful. IBM is resistant to immunosuppressive treatment. Prednisone, corticosteroids or IVIG do not impact the immune attack. ATG and alemtuzumab are contra-indicated.

Management: The best care for most patients with IBM involves strictly nonpharmacological management, including emotional support, physical therapy, and education on exercise. Evaluation and monitoring of potential complications is vital (depression, fatigue, dysphagia and respiratory involvement). Prevention of falls is another early, vital consideration. Integration of palliative care is helpful at end stages.

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: reduced lung volumes may raise carbon dioxide levels in arterial blood (hypoventilation causes hypercapnia).

Pathology: T cells, myeloid dendritic cells, macrophages and plasma cells all invade IBM muscle, but it is the infiltration of muscle by 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.

References: Please see http://www.ibmyositis.com for further information.

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