Johns Hopkins Myositis Center Experience with Inclusion Body Myositis

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Inclusion Body Myositis (IBM) is the most common muscle disease in adults over the age of 50. IBM is a slowly progressive muscle disease of unclear etiology and limited available treatments. Diagnosis involves both clinical and pathological findings, and misdiagnoses are common when pathological findings are overemphasized (eg diagnosing polymyositis when rimmed vacuoles (RVs) are not observed and diagnosing IBM in patients with muscular dystrophy due to appearance of RVs and inflammation). New immunohistochemistry markers such as TDP-43 or p62 may improve diagnostic accuracy. Overlap of IBM with autoimmune and infectious diseases including Sjogren's syndrome, HIV, and Sarcoidosis suggests dysregulation of the immune system in IBM; however, IBM patients rarely have lasting response to immunotherapy. Dysphagia is a common symptom in IBM, and patients often respond well to esophageal dilatation. Finger flexor weakness is a major cause of disability in IBM, and patients may benefit from orthopedic procedures (eg tendon transfer) to improve grip strength. Therapies that stimulate autophagy to remove toxic misfolded proteins are being considered for IBM clinical trials.