TABLE 122-9 IDIOPATHIC INFLAMMATORY MYOPATHIES

MYOPATHY	SEX	TYPICAL AGE AT ONSET	PATTERN OF WEAKNESS	CREATINE KINASE	MUSCLE BIOPSY	RESPONSE TO IMMUNOSUPPRESSIVE THERAPY
Dermatomyositis	Women > men	Childhood and adult	Proximal > distal	Increased (up to 50× normal)	Perifascicular atrophy, inflammation, complement deposition on capillaries	Yes
Polymyositis	Women > men	Adult	Proximal > distal	Increased (up to 50× normal)	Endomyseal inflammation; invasion of non-necrotic fibers	Yes
Sporadic Inclusion Body Myositis	Men > women	Elderly (>50 yr)	Proximal and distal; predilection for finger and wrist flexors, knee extensors	Increased (<10× normal)	Endomyseal inflammation, rimmed vacuoles; electron microscopy: 15- to 18-nm tubulofilaments	No

involvement, difficulty swallowing, or cardiomyopathy. Both DM/PM can be associated with underlying malignancy (dermatomyositis more frequently than polymyositis), so screening for malignancy is recommended especially in patients over age 40.

Diagnosis and Differential Diagnosis

Diagnosis is based on clinical history and examination findings in conjunction with irritable changes on electromyography (e.g., fibrillation potentials and positive sharp waves) and characteristic muscle biopsy. Both can be associated with autoantibodies (e.g., ANA). The most useful is the anti-Jo-1 antibody, which can be seen more frequently in patients with pulmonary involvement.

Treatment

For both DM/PM the first line of treatment is prednisone. Steroid-sparing immunosuppressive therapies (e.g., methotrexate, azathioprine) are often added for those patients requiring long-term therapy in order to reduce the required dose of prednisone or to replace prednisone completely. In patients who do not respond to conventional therapy, intravenous immunoglobulin or rituximab may be effective.

Prognosis

Most patients respond to immunosuppressive therapies.

Sporadic Inclusion Body Myositis

Definition and Epidemiology

Sporadic inclusion body myositis (s-IBM) is an idiopathic, slowly progressive muscle condition in older adults (occurring in more men than women), associated with inflammation and characteristic pathological changes on muscle biopsy. It is the most common inflammatory muscle disease in patients over 50, affecting 3.5 per 100,000.

Pathology

Muscle biopsies resemble polymyositis with endomyseal inflammatory infiltrates and invasion of non-necrotic fibers. Distinctive for IBM are vacuoles rimmed by mitochondria and electron microscopy, which shows 15 to 18 nm tubulofilamentous inclusions.

Clinical Presentation

S-IBM is a slowly progressive, often asymmetric weakness occurring usually after 50 years of age initially in a distinctive pattern, including distal forearm muscles (distal finger flexors) and quadriceps wasting and weakness. This can progress to involve almost any muscle and can affect swallowing in up to 70% of patients.

Diagnosis and Differential Diagnosis

Diagnosis is based on clinical history and examination and characteristic muscle pathology. The main differential diagnosis is other idiopathic inflammatory myopathies or late-onset inherited myopathies, including a hereditary form of IBM.

Treatment

Unlike the other inflammatory myopathies, IBM does not respond to immunosuppression. Treatment is supportive.

Prognosis

Most patients with s-IBM progress to need a wheelchair over 10 to 15 years. Swallowing difficulty can be life threatening.

Infectious Myositis

An acute viral myositis can occur in the setting of an influenza viral upper respiratory tract infection. In addition to typical influenza-associated myalgias, affected patients develop muscle pain, proximal weakness, and elevated CK levels. The disorder is self-limited, but when severe it is often associated with myoglobinuria and occasionally with renal failure. A similar syndrome can complicate infections with other viruses.

An inflammatory myopathy can occur in the setting of human immunodeficiency virus infection, either in early or in later acquired immunodeficiency syndrome. The clinical presentation resembles polymyositis. The patient's condition may improve with corticosteroid therapy. The disorder must be distinguished from the toxic myopathy caused by zidovudine, which responds to dose reduction. Although rarely seen, tuberculosis can present with muscle abscess (pyomyositis) either in the setting of pulmonary or disseminated disease or in isolation.

Myopathies Caused by Endocrine and Systemic Disorders

Thyroid studies should be obtained in any adult coming in with a new complaint of muscle weakness. Patients with hyperthyroidism often have some degree of proximal weakness but this is rarely the presenting manifestation of thyrotoxicosis. Hypothyroid myopathy is associated with proximal weakness and