INFLAMMATORY MYOSITIS
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Definition

Inflammatory myositis is a systemic autoimmune disease that has four forms: polymyositis, dermatomyositis, necrotizing autoimmune myositis and inclusion body myositis. Inflammatory myositis is more common in African-Americans compared to whites and in woman compared to men. Polymyositis affects adults in their 30’s to 50’s. Dermatomyositis commonly affects children ages 5-15 and adults ages 45-65. Myositis can occur in combination with other autoimmune diseases (e.g. scleroderma and mixed connective tissue disease). The exact cause is unknown, but we do know that it is an autoimmune disease (the body’s own immune system attacks the muscles).

Symptoms

Myositis is characterized by the onset over several weeks of weakness in muscles close to the trunk - the upper arms/shoulders, thighs and sometimes the neck. Weakness is due to muscle inflammation and subsequent muscle damage. While weakness is the most prominent symptom, muscle soreness can also occur. Patients may notice difficulty getting out of a chair, putting things away in overhead cabinets, or trouble climbing stairs due to weakness rather than pain. Other symptoms can include shortness of breath from lung inflammation, joint pain, Raynaud’s syndrome (finger color changes and numbness in cold weather), and trouble swallowing. Patients with dermatomyositis often develop several different kinds of rashes including one on the hands (Gottron’s papules), a violet discoloration or swelling around the eyes (heliotrope), and/or across the back and chest (shawl rash; see images below).

Figure 1. Dermatomyositis Rashes (Gottron’s, Heliotrope, Shawl)
**Diagnosis**

The diagnosis of inflammatory myositis relies on the history (e.g. how fast are the symptoms progressing?), physical exam, lab tests (e.g. how high are the muscle enzymes or tests that measure levels of muscle inflammation in the blood?), imaging such as MRI of the thigh muscles that can reveal muscle inflammation, nerve conduction studies and a muscle biopsy. The above results will help your rheumatologist (sometimes working with a neurologist) to exclude other diseases that can cause muscle weakness and thereby mimic inflammatory myopathies. Such mimickers include thyroid disease, toxin-induced myopathies (e.g. associated with the use of cholesterol lowering medications called statins), neuromuscular disorders (e.g. muscular dystrophy), infections, and metabolic myopathies (e.g. McArdle’s, mitochondrial myopathies). It is important to establish the correct cause of the muscle weakness because each type of myositis is treated differently and each has a different prognosis. Importantly, polymyositis and dermatomyositis can be associated with cancer. Your doctor will take a thorough history to make sure you are up-to-date with age-appropriate cancer screening (e.g. colonoscopy if you are over age 50).

**Treatment**

The initial treatment for inflammatory myopathies is steroids taken by mouth and sometimes intravenously, depending on how severe symptoms are at diagnosis. No other medication works as quickly. The eventual goal is to taper the steroids slowly and substitute a medication that is safer to take in the long term such as mycophenolate mofetil or azathioprine. In severe cases of myositis, especially when the muscles used for swallowing are impacted, we sometimes use intravenous immunoglobulin, or IVIg, a medication given monthly by infusion. Physical therapy is an important part of myositis treatment to help patients regain strength.