

Inflammatory Myopathy: Clinical Features, Pathophysiology and Management

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Description

Inflammatory myopathy is a group of rare autoimmune diseases characterized by chronic inflammation of the muscles, leading to weakness, pain and potential loss of muscle function. This condition can significantly impact an individual's quality of life, affecting their ability to perform daily activities and potentially leading to disability if not properly managed. The most common forms of inflammatory myopathy include polymyositis, dermatomyositis and inclusion body myositis, each with distinct clinical features but sharing common underlying mechanisms of muscle inflammation. Polymyositis is characterized by progressive muscle weakness, usually affecting the proximal muscles such as those of the hips, shoulders and neck. This form of myopathy primarily involves the muscle fibers themselves and is associated with an increase in inflammatory cells within the muscle tissue. Dermatomyositis, on the other hand is notable for its association with a distinctive skin rash in addition to muscle weakness. The rash which may appear as heliotrope purple rash on the eyelids or as a rash on the back or chest often precedes or coincides with muscle symptoms. This form of inflammatory myopathy is more common in children than adults and may be associated with underlying malignancies particularly in adults.

Pathophysiology of inflammatory myopathies

The pathophysiology of inflammatory myopathies involves an aberrant immune response where the body's immune system mistakenly targets its own muscle tissue. In polymyositis CD8+ T cells infiltrate the muscle fibers leading to inflammation and muscle damage. In dermatomyositis both CD4+ T cells and B cells contribute to the inflammatory process and the presence of autoantibodies such as anti-Jo-1 is a key feature. Inclusion Body myositis is distinguished by the presence of characteristic inclusions within muscle cells and involves a more complex exchange

of immune and degenerative processes. Diagnosis of inflammatory myopathy typically involves a combination of clinical evaluation, laboratory tests and imaging studies. Muscle biopsy is a critical tool for diagnosis, revealing characteristic histopathological features such as inflammatory infiltrates and muscle fiber damage. Blood tests may show elevated levels of muscle enzymes like Creatine Kinase (CK), while electromyography can identify abnormal electrical activity in the muscles. In some cases, autoantibody tests can help identify specific autoimmune markers associated with different forms of myopathy.

Treatment of inflammatory myopathy

Treatment of inflammatory myopathy often begins with corticosteroids such as prednisone, which help reduce inflammation and muscle damage. In cases resistant to steroids other immunosuppressive agents like methotrexate or azathioprine may be used to control the immune response. Physical therapy plays a crucial role in managing the symptoms and improving muscle strength and function. For dermatomyositis addressing any underlying malignancy is essential as it may lead to improvement in muscle symptoms. The course of inflammatory myopathy varies widely among individuals. Some may experience periods of remission and improvement while others may face progressive muscle weakness and disability. To control symptoms and enhance results, early diagnosis and treatment are essential. Ongoing research into the mechanisms underlying these diseases and the development of targeted therapies continues to enhance our understanding and treatment of inflammatory myopathies. While each type presents with unique clinical features and challenges, advances in diagnosis and treatment offer hope for better management and improved quality of life for affected individuals.