DERMATOMYOSITIS

An Autoimmune Inflammatory Muscle Disorder

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Dermatomyositis is considered one of the most common of the autoimmune inflammatory muscle disorders.

What is Dermatomyositis

Dermatomyositis (DM) is an autoimmune disease formally classified as one of the inflammatory myopathies (inflammatory muscle diseases). The muscle inflammation of dermatomyositis, which affects multiple muscles causing a condition of polymyositis, is accompanied by a rash. The rash of DM often precedes the muscle disorder. In addition to the rash and myopathy, systemic symptoms such as heart and lung involvement or arthritis may also occur.

Who is Affected?

Dermatomyositis affects people of all ages, including children, and females are more likely to be affected than males. The peak ages affected are 50 years and 5-10 years. The incidence of dermatomyositis/polymyositis appears to be increasing.

Symptoms

The rash of dermatomyositis, which is frequently described as a heliotrope rash, resembles a patchy, bluish-purple to dusky discoloration that tends to appear on the face, particularly around both eyes, and also on the neck, shoulders, upper chest, elbows, knees, knuckles, and back. A splotchy rash may develop over the neck and upper chest and back in the pattern of a shawl (shawl sign). The rash in DM may also be accompanied by calcium deposits that occur as hard lumps beneath the skin known as Gottron papules.

Gottron papules, which occur in up to 40 percent of adolescents and are seen less often in adults, are frequently found on bony prominences such as those found on the knuckles, elbow, knees, toes, and feet. The rash in DM may itch (urticaria), appear scaly, appear as white plaques on the insides of the cheeks, or appear as thickening on the palms of the hands (mechanic's hands). At the time of disease onset, the skin manifestations may be the sole complaint.

Symptom severity varies. Most patients survive although death may occur if the muscle weakness is severe or affects the lungs or heart. Children with severe DM may develop muscle contractures, and calcinosis may cause vascular complications. Malignancy may occur in patients with DM, particularly those older than 60 years.
Disease Course

The onset of DM is often severe, and the muscle weakness targets the muscles closest to the trunk of the body. Eventually, patients with DM experience difficulty rising from a seated position, climbing stairs, lifting objects, or reaching overhead. Later in the course of the illness, the muscles distant from the trunk of the body may also be affected. Difficulty in swallowing (dysphagia) may occur, and the muscles may become tender to the touch. Other symptoms include fatigue, generalized discomfort, skin redness, hyperpigmentation, light sensitivity, Raynaud phenomenon, arthritis, arthralgia, arrhythmia, dyspnea (shortness of breath), dysphonia (vocal changes), weight loss, and low-grade fever.

Diagnosis

In 1975, criteria were established for diagnosing dermatomyositis and the related condition of polymyositis, which is not accompanied by rash. These criteria include: progressive symmetrical muscle weakness; elevated muscle enzymes; abnormal muscle biopsy results; and in dermatomyositis, the characteristic rash. Conditions of dermatomyositis and myositis may vary in severity and sometimes overlap. For instance, some patients have a well-controlled myopathy with a severe debilitating skin disease, resulting in a condition of postmyopathic dermatomyositis.

Patients with dermatomyositis have elevated levels of muscle enzymes, including creatine kinase (CK), aldolase, aspartate aminotransferase (AST), and lactic dehydrogenase (LDH). Patients with DM usually have positive tests for antinuclear antibodies (ANA). Anti-Mi-2 are highly specific for dermatomyositis but lack sensitivity. They're usually seen in patients with shawl rash and are a favorable prognostic sign. Anti-Jo-1 antibodies are often seen in dermatomyositis, especially in patients with pulmonary involvement. Anti-SRP antibodies are seen in severe disease involvement.

Treatment

Most patients with DM respond well to treatment although patients with co-existing heart or lung disorders may have a more severe disease course and not respond as well to therapy. Corticosteroids are usually prescribed as the first line of treatment. If corticosteroids prove ineffective, immunosuppressants such as azathioprine and methotrexate, are prescribed. In severe cases, intravenous immunoglobulins may be used. Physical therapy is recommended for preserving muscle function and preventing muscle wasting.

Resources:


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