



# DERMATOMYOSITIS

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Dermatomyositis (DM) is a rare inflammatory muscle disease that affects both the muscles as well as the skin. DM can affect people of all races, sex and age. Although it affects both males and females equally in childhood, it is more common in females in adults. The exact cause of DM is unknown but it is believed to result from an immune-mediated process triggered by outside factors (e.g. malignancy, drugs, and infectious agents) in genetically predisposed individuals. Dermatomyositis can occur with other connective tissue disorders such as systemic lupus erythematosus, rheumatoid arthritis, **scleroderma**, Sjogren's syndrome, and mixed connective tissue disease.

Patients with dermatomyositis usually present with complaints of tiredness and loss of energy. The skin changes occur before the onset of muscle disease in most patients. The earliest signs of skin manifestations may begin with a red to bluish-purple rash most commonly in the sun exposed areas (face, neck, shoulders, upper chest and back). The eyelids may get the typical purple rash known as the heliotrope rash. Early clinicians thought that this violaceous rash around the eyes reminded them of the color of a heliotrope flower, and thus referred to this as the 'heliotrope sign'. The knuckles may have purple spots known as 'Gottron's papules'. The most important diagnostic feature of skin eruption of DM is poikiloderma, which is the pale, thin skin with blood vessels and dark spots located in the sun exposed areas. Hardened deposits of yellow to white lumps under the skin can develop especially in children and adolescents known as calcinosis. Progressive muscle weakness especially the hips, thighs, shoulders, upper arms and neck in a symmetric fashion may occur. In advanced disease, all muscle groups can be affected. Patients then complain of great fatigue and are often unable to complete simple tasks such as combing their hair or rising to their feet from a sitting position. The affected muscle groups are tender to touch in more advanced disease. Lung disease occurs in approximately 15-30% of patients.

In some cases the presence of dermatomyositis may be related to an internal cancer. The reported frequency of an internal cancer in adults with DM varies from less than 10% to 50%. Ovarian cancer, breast, lung, gastric, and other female genital cancers can sometimes be found in patients with DM.

The diagnosis of dermatomyositis is usually recognized by the typical heliotrope rash. Patients with a characteristic skin eruption should have a skin biopsy performed to confirm diagnosis. Additional tests can be ordered to help confirm the diagnosis or to determine the extent of the disease. Blood work measuring creatine kinase and aldolase can help measure the extent of muscle damage. The presence of circulating autoantibodies in the blood may indicate the autoimmune disease. MRI may detect inflammation in the muscles. Electromyography (EMG) testing done by a small electrode inserted into the muscle can measure electrical activity of the muscle. The muscle biopsy is a very important test for confirming the diagnosis and excluding other inflammatory muscular diseases.

There is no cure for dermatomyositis, but treatment can improve the symptoms. **Oral corticosteroids** such as prednisone may help by reducing the inflammation and thus improving the skin and muscular symptoms. Immunosuppressive drugs used alone or in combination with corticosteroids include **methotrexate**, azathioprine, cyclophosphamide, **cyclosporin**, and high dose intravenous immunoglobulin. Physical therapy can help patients with strength and flexibility improving daily function. Protection from the sun by using sunscreens and sun protective clothing can also improve the appearance of the skin.

**This information has been provided to you compliments of the American Osteopathic College of Dermatology and your physician.**

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