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Dermatomyositis

Dermatomyositis is a disease that involves muscle inflammation and a skin rash. Polymyositis is a similar inflammatory condition that also involves muscle weakness, swelling, tenderness, and tissue damage but no skin rash. Both are part of a larger group of diseases called myopathies, more specifically inflammatory myopathies.

Causes

The cause of dermatomyositis is unknown. Experts think it may be due to a viral infection of the muscles or a problem with the body's immune system. It may also occur in people who have cancer in the abdomen, lung, or other parts of the body.

Anyone can develop this condition. It most often occurs in children age 5 to 15 and adults age 40 to 60. It affects women more often than men.

Symptoms

Symptoms may include:

- Muscle weakness, stiffness, or soreness
- Problems swallowing
- Purple color to the upper eyelids
- Purple-red skin rash
- Shortness of breath
- Difficulty swallowing

The muscle weakness may come on suddenly or develop slowly over weeks or months. You may have trouble raising your arms over your head, getting up from a sitting position, and climbing stairs.

The rash may appear on your face, knuckles, neck, shoulders, upper chest, and back.

Exams and Tests

The health care provider will do a physical exam. Tests may include:

- Blood test to check levels of muscle enzymes called creatine phosphokinase and aldolase
- Blood tests for autoimmune diseases

- Electrocardiogram (ECG)
- Electromyography (EMG)
- Magnetic resonance imaging (MRI)
- Muscle biopsy
- Skin biopsy
- Screening tests for cancer
- Chest x-ray and CT scan of the chest
- Lung function tests
- Swallowing study
- Myositis specific and associated autoantibodies

Treatment

The main treatment is the use of corticosteroid medicines. The dose of medicine is slowly tapered off as muscle strength improves. This takes about 4 to 6 weeks. You may stay on a low dose of a corticosteroid medicine after that.

Medicines to suppress the immune system may be used to replace the corticosteroids. These drugs may include azathioprine, methotrexate or mycophenolate.

Treatments that may be tried when disease that remains active in spite of these medicines are:

- Intravenous gamma globulin
- Biologic drugs

When your muscles get stronger, your provider may tell you to slowly cut back on your doses. Many people with this condition must take a medicine called prednisone for the rest of their lives.

If a tumor is causing the condition, the muscle weakness and rash may get better when the tumor is removed.

Outlook (Prognosis)

It is important to be followed by a health care provider when you have dermatomyositis. Severe illness can lead to disability and can sometimes result in death.

Symptoms may go away completely in some people, such as children.

The condition may be fatal in adults due to:

- Severe muscle weakness
- Malnutrition
- Pneumonia
- Lung failure

The major causes of death with this condition are cancer and lung disease.

People with lung disease with the anti-MDA-5 antibody have a poor prognosis in spite of current treatment.

Possible Complications

Complications may include:

- Lung disease
- Acute renal failure
- Cancer (malignancy)
- Inflammation of the heart
- Joint pain

When to Contact a Medical Professional

Contact your provider if you have muscle weakness or other symptoms of this condition.

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