Myositis - Polymyositis and Dermatomyositis

Polymyositis and dermatomyositis are connective tissue diseases characterised by inflammation of muscles. Although dermatomyositis affects the skin and muscles, it may also affect other parts of the body such as joints, oesophagus, lungs and heart. Both polymyositis and dermatomyositis have an autoimmune basis.

Viral infection has been implicated in the form of the human retroviruses HIV and human T-cell lymphotropic virus type I (HTLV-I), the simian retroviruses, and Coxsackievirus B.

It is possible to divide the conditions into seven basic subgroups according to aetiology:[1]

- Primary idiopathic polymyositis in adults.
- Idiopathic dermatomyositis in adults.
- Childhood dermatomyositis or myositis with necrotising vasculitis.
- Polymyositis associated with connective tissue diseases.
- Polymyositis or dermatomyositis associated with malignancy.
- Inclusion body myositis
- Miscellaneous (eg, eosinophilic myositis, myositis ossificans, focal myositis, giant cell myositis)

Epidemiology

- Polymyositis tends to present between 30 and 60 years of age with a smaller peak at about 15 years of age.[1]
- Dermatomyositis can occur in people of any age. The peak age of onset in adults is approximately 50 years; the peak age of onset in children is approximately 5-10 years.[2]
- Dermatomyositis and polymyositis are twice as common in women as in men.[2]

Polymyositis

History

- There is an inflammatory myopathy with onset over weeks or months and steady progression.
- Diffuse weakness in the proximal muscles develops.
- Proximal myopathy causes difficulty rising from a low chair, climbing steps, lifting objects and combing hair. Fatigue, myalgia, and muscle cramps may also be present.
- Distal muscles are spared, and so fine motor movements of the hand, such as buttoning a shirt, writing, operating a keyboard or playing the piano are affected only late in the disease.
- Pharyngeal weakness causes dysphagia.
- Weakness may vary from week to week or month to month.
- Only a third have pain. There is no rash.
- There is no family history of neuromuscular disease, evidence of endocrine disorder or history of exposure to possible toxins.

Examination

Polymyositis produces muscle weakness. It is not painful, although a few patients complain of aches or cramps.

- Proximal muscle weakness occurs with comparative sparing of distal muscles until the disease is well advanced.
- External ocular muscles are unaffected. Facial muscles are affected only in severe disease.
- Forced flexion of the neck is weak and there may be difficulty just holding the head up.
- Muscular atrophy occurs with preservation of tendon reflexes, flexor plantar response and normal sensation.
Muscles may be tender on palpation and may have a nodular grainy feel.

**Investigations**

- Creatine kinase can be up to 50 times normal. It is rarely normal in active disease and the level is usually a good indicator of disease activity.
- About 20% have anti-Jo-1 antibodies. They indicate a poor prognosis with interstitial lung disease. This lung disease occurs in about a third.[4]
- Other enzymes to be elevated include aldolase, serum glutamic-oxaloacetic transaminase (SGOT), serum glutamic-pyruvic transaminase (SGPT), and lactate dehydrogenase (LDH). If SGOT is higher than SGPT, a myogenic cause should be suspected.
- The diagnosis is established by electromyography (EMG) and is confirmed by muscle biopsy. In polymyositis it is the definitive test.
- Cancer antigen 125 (CA-125) and cancer antigen 19-9 (CA19-9) may be useful markers of the risk of malignancy.[1]
- Autoantibody testing for myositis-specific antibody (MSA) and myositis-associated autoantibodies (MAA) may be useful to differentiate the patients with underlying malignancy.[5]

**Differential diagnosis**

Polymyositis is a diagnosis of exclusion and other considerations include:

- Hereditary neuromuscular diseases.
- Endocrine disease including thyrotoxicosis and Cushing's disease.
- Malabsorption syndromes, alcoholism, cancer, vasculitis, granulomatous disease, sarcoidosis or exposure to drugs or toxins that affect the muscles.

**Dermatomyositis**

**History**

Dermatomyositis affects children as well as adults. The muscle weakness has the same pattern as in polymyositis but there are other features too:

- Rash: see under ‘Examination’, below.
- There is systemic upset with fever, arthralgia, malaise and weight loss. It can resemble scleroderma with Raynaud's phenomenon and dysphagia.
- Possible cardiac disease including atrioventricular conduction defects, tachyarrhythmias and dilated cardiomyopathy.
- Gastrointestinal ulcers and infections.
- Thoracic muscles may be weak but there may also be interstitial lung disease in 30-50%.[6]
- Children tend to have more non-muscular features, especially gastrointestinal ulcers and infections.

**Examination**

- The rash includes blue-purple discoloration on the upper eyelids with periorbital oedema, a flat red rash involving the face and upper trunk and raised purple-red scaly patches over the extensor surfaces of joints and fingers. Ulcerative vasculitis and calcinosis of subcutaneous tissue may occur.
- The rash may affect knees, shoulders, back and upper chest and may be exacerbated by sunlight.
- Skin lesions may produce scaling, pigmentation or depigmentation of the skin and a shiny appearance.
- Dilated capillary loops at the fingernail base are typical of dermatomyositis. The cuticles may be irregular and thickened, and the palmar and lateral surfaces of the fingers may become rough and cracked.
- Muscle weakness is proximal and can vary from mild to extreme. Sensation is preserved and tendon reflexes are normal unless atrophy is severe.
- There is muscle pain and tenderness early in the disease.

**Investigations**

- Elevation of creatine kinase is not so reliable although it can be extremely high.
- SGOT, SGPT, LDH, and aldolase levels may also be raised.
Autoantibodies:
- A positive antinuclear antibody (ANA) finding is common in patients with dermatomyositis.
- Anti-Mi-2 antibodies are specific for dermatomyositis, but found in only 25% of patients with dermatomyositis.
- Anti-Jo-1 antibodies are more common in patients with polymyositis than in patients with dermatomyositis. They are associated with interstitial lung disease, Raynaud’s phenomenon and arthritis.

- MRI is not very helpful for making the diagnosis but it can help monitor activity and guide the best place for muscle biopsy.
- EMG may be helpful but can be normal in 15%. It can also guide a suitable place for biopsy.
- Muscle biopsy can be diagnostic.

Differential diagnosis
Dermatomyositis is much easier to diagnose, as the rash and subcutaneous calcification is typical.

- In systemic lupus erythematosus (SLE), the skin over the phalanges is involved and the skin over the metacarpophalangeal joints is spared. In dermatomyositis, the reverse is true.
- There can be some overlap with systemic sclerosis, rheumatoid arthritis, and Sjögren’s syndrome but the weakness is more than can be accounted for by arthritis alone.

Associated diseases
- Polymyositis and dermatomyositis may be associated with other autoimmune diseases such as myasthenia gravis, Hashimoto’s thyroiditis, systemic sclerosis and Waldenström’s macroglobulinaemia.
- In both polymyositis and dermatomyositis it may be necessary to hunt for an underlying malignancy.
  - Polymyositis and especially dermatomyositis may be part of a paraneoplastic syndrome.
  - Between 10 and 20% of patients with dermatomyositis have neoplasms.
  - In elderly patients with dermatomyositis there is often malignancy.
  - Breast cancer, lung cancer, ovarian carcinoma and gastric carcinoma are usually implicated.

Treatment

Nondrug
- Sun-blocking agents should be used.[7]
- Encourage physical activity within reason to maintain muscular strength. This may involve consultation with a physiotherapist and occupational therapist.
- Evaluation of swallowing may be required and a speech and language therapist may help with difficulties of swallowing.
- Monitor creatine kinase and clinical response but treatment can improve the former without benefiting the latter.

Drugs
- Early initiation of therapy is essential.[8]
- Steroids are the most important drugs. In mild disease, topical steroids may suffice. In more severe disease, high doses of systemic steroids are used and tapered off. Improvement is usually apparent by the second or third month. The usual precautions must be exercised when giving high doses of steroids for long periods.
- If steroids fail then immunosuppressive drugs such as azathioprine can be used. As an alternative, cyclophosphamide is usually better than methotrexate.
- Intravenous immunoglobulin is appropriate in patients with resistant dermatomyositis or aggressive disease.[9]
- Other treatments include antimalarial agents and immunomodulatory therapies.[7]
- Patients with anti-Jo-1 antibodies need long-term immunosuppression.[10]
- For lung disease, an aggressive combination regimen including ciclosporin A or tacrolimus with cyclophosphamide is recommended to be added to corticosteroids.[6]
- There have been reviews of treatment options but with few good controlled trials.[11][12]
Complications

These tend to be unrelated to muscles.

- Gastrointestinal ulceration can cause melaena or haematemesis. Infarction of the bowel may occur, especially in dermatomyositis.
- Dermatomyositis can cause subcutaneous calcification that punctures the skin with ulcerations, infection and ugly scars.
- Dermatomyositis is also associated with increased risk of malignancy, especially in patients older than 60 years, atrioventricular defects, tachyarrhythmias, dilated cardiomyopathies, joint contractures and lung involvement (due to weakness of thoracic muscles, interstitial lung disease).
- Complications of polymyositis may include:
  - Interstitial lung disease, aspiration pneumonia.
  - Heart block, arrhythmias, congestive heart failure, pericarditis, myocardial infarction.
  - Dysphagia, malabsorption.
  - Infection.
  - Carcinoma, especially breast and lung (the incidence of bladder cancer and non-Hodgkin’s lymphoma may also be increased, especially in the first year after diagnosis).
- Complications of steroid therapy - eg, osteoporosis, myopathy.

Prognosis

- Polymyositis:[1]
  - 5-year survival rates have been estimated at more than 80%. Mortality is most often related to associated malignancy or pulmonary complications.
  - Polymyositis usually responds well to treatment but residual weakness occurs in approximately 30% of patients.
  - Poor prognosis is associated with:
    - Older age, female sex.
    - Interstitial lung disease, cardiac involvement, associated malignancy.
    - Presence of anti-Jo-1 (lung disease) and anti-SRP antibodies (severe muscle disease, cardiac involvement).
    - Delayed or inadequate treatment.
    - Dysphagia, dysphonia.
- Dermatomyositis:[2]
  - Most patients survive but may develop residual weakness and disability.
  - It may spontaneously remit in as many as 20% of affected patients but about 5% of patients have a fulminant progressive course with eventual death.
  - Poorer prognosis is associated with malignancy, cardiac or pulmonary involvement and those aged >60 years.
  - Particular causes of death include muscle weakness, cardiopulmonary involvement or associated malignancy.

Further reading & references

- Myositis Support Group
- Pappu R et al; Polymyositis, Medscape, Sep 2011
- Callen JP; Dermatomyositis, Medscape, Oct 2012

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