INTRODUCTION

Polymyositis (PM) and dermatomyositis (DM) belong to a heterogeneous group of disorders which affect the skeletal muscles, called the idiopathic inflammatory myositis (IIM). In a rheumatology clinic, these are seen infrequently, being rarer than systemic lupus erythematosus (SLE). Nonetheless, it is important to be familiar with the clinical presentations of these diseases, as early diagnosis and prompt treatment prevents chronic morbidity and even mortality. Like other systemic connective tissue diseases, the incidence and prevalence of this disease in our country is not known. In the Western countries, the incidence is 0.5 to 8.4 per 100,000 population per year.

Studies on a possible viral triggering agent have received considerable interest. Circumstantial evidence for such a theory emerges from observations of seasonal occurrence of a subset of cases. Viruses which have been implicated in patients are Coxsackie A9 and mumps. In experimental models, injection with Coxsackie B1 virus in mice induces a disease similar to myositis. An attractive theory is that antibodies against the virus crossreact with antigenic targets of the hosts (molecular mimicry).

GENETICS AND IMMUNOLOGICAL BASIS

That there is a predominant genetic and immunological basis for this group of diseases is undisputed. The presence of myositis specific autoantibodies, T-cells infiltrating the skeletal muscles, HLA associations and presence of co-existent autoimmune disease manifestations support this contention.

CLASSIFICATION

There are two classification systems that have been proposed for IIM. The Bohan and Peter’s is the more popular one and categorizes patients in to five subtypes:

1. Adult polymyositis
2. Adult dermatomyositis
3. PM/DM overlap with other connective tissue diseases
4. Juvenile polymyositis / dermatomyositis and
5. Myositis associated with malignancy

**ADULT POLYMYOSITIS/DERMATOMYOSITIS**

The disease is usually seen to affect individuals between the age of 45 to 60 years, with a female to male ratio of 2:1. The characteristic clinical picture is that of insidious onset pelvic and pectoral girdle muscle weakness evolving in weeks to months. Pain and tenderness is only seen in half the cases. Difficulty in getting up from a sitting position, climbing stairs and raising the arms to comb hair are the early symptoms. There is difficulty in walking and if left untreated, the patient becomes bedbound. Rarely, the patients may have an acute onset with rhabdomyolysis and myoglobinuria. In some dysphagia, nasal regurgitation of swallowed fluid and nasal intonation of voice are presenting features but, on direct questioning, the history of pelvic and pectoral girdle muscle weakness is easily elicited.

Being a systemic disease, the disease has a wide variety of extraskeletal muscle manifestations, like Raynaud’s, interstitial lung disease, myocarditis, symmetrical peripheral arthritis, sclerodactyly and localized skin thickening. The presence of these features may confuse the clinician as to whether there is associated connective tissue disease or not. There is often a prodromal phase of asthenia and a symmetrical polyarthritis. Many patients are misdiagnosed as rheumatoid arthritis early in the course of IIM and the limited movement on the part of the patient is attributed to joint pain and stiffness.

**ADULT DERMATOMYOSITIS**

The addition of the characteristic skin rash to the clinical picture of polymyositis describes dermatomyositis. The typical heliotrope rash is lilac coloured and involves the upper eyelid. It is accompanied by lid oedema, which may be mistaken for nephrotic syndrome. This classical rash is seen in only a minority of patients with this disease. The
other characteristic skin rash is the presence of Gottron’s papules over the metacarpophalangeal joints and elbows. At the onset, it is erythematous painless papule, and as it heals it leaves behind a thin scaly whitish papule. More common is the presence of a diffuse erythematous rash over the face and extremities. Patients with antibodies to synthetase (Jo-1) have a ‘mechanic’s hands’ (roughening of the inner aspect of index finger) and those with antibodies to Mi-2 have a typical V shaped rash over the necklace area called the “shawl sign”. The rash usually occurs concurrently with muscle weakness but sometime precedes it.

**JUVENILE POLYMYOSITIS/DERMATOMYOSITIS**

Onset of PM/DM in early childhood is characterized by vasculitis and myocarditis, along with muscle weakness. Small vessel vasculitis taking the form of palpable purpura, or bleeding, occur due to vasculitis. Another feature that occurs more frequently in the juvenile form is soft tissue calcinosis. The typical cutaneous manifestations of adult DM are also seen in juvenile DM. Some of the children present with dysphagia or dyspnoea, which carry a worse prognosis.

**MYOSITIS ASSOCIATED WITH CONNECTIVE TISSUE DISEASES**

Myositis can occur in SLE, systemic sclerosis, mixed connective tissue disease (MCTD) and Sjögren’s syndrome. It is generally milder symptomatically, with only elevation of muscle enzymes and electromyographic (EMG) abnormalities. The histological changes are similar to adult PM/DM. Myositis has been reported in RA, Wegener’s granulomatosis and adult onset Still’s disease.

**MYOSITIS AND MALIGNANCY**

There seems to be an association between malignancy and polymyositis/dermatomyositis, with the former appearing within two years of the onset of the latter. DM appears to have a stronger association than PM with carcinomas of ovary, breast and stomach. The onset of myositis after the age of 50 years, presence of erythroderma or vasculitis and a poor response to immuno-suppressive treatment usually provide the clue of an underlying malignancy and the need for a careful search. There is no consensus as to how rigorously
to investigate such a situation. The majority are of the opinion that a detailed clinical examination with non-invasive tests like a PAP smear, X-rays and ultrasound examinations are all that is required.

**INCLUSION BODY MYOSITIS**

Inclusion body myositis (IBM) resembles polymyositis with some difference. It presents at a later age with insidious onset and prolonged course. Several years pass by before the patient presents to the physician. Typically, there is distal muscle weakness, along with proximal muscle disease, unresponsive to prednisolone 40-60mg/d. The diagnosis is established by light and electron microscopy examination of the affected muscle tissue. The characteristic changes are the presence of intracellular vacuoles by light microscopy. Electron microscopy reveals either intracytoplasmic or intranuclear tubular inclusion bodies.

**DIFFERENTIAL DIAGNOSIS**

In the presence of proximal muscle weakness and a skin rash, the diagnosis of dermatomyositis is straightforward. The lilac coloured rash is sometimes difficult to recognize in dark coloured individuals, however, the lid oedema and the Gottrons’ rash are unmistakable signs. Because of the absence of rash, the diagnosis of PM clinically needs to be differentiated from other diseases that cause proximal muscle weakness. The table shows a list of clues to distinguish PM from other diseases or conditions.

**Table 2: Clinical clues that distinguish PM/DM from other diseases causing muscle weakness**

<table>
<thead>
<tr>
<th>Clue</th>
<th>Diagnosis</th>
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</thead>
<tbody>
<tr>
<td>1. Family history of similar illness</td>
<td>Myopathy</td>
</tr>
<tr>
<td>2. Facial muscles involvement</td>
<td>Myasthenia/myopathy</td>
</tr>
<tr>
<td>3. Episodic involvement</td>
<td>Hypokalemic periodic palsy</td>
</tr>
<tr>
<td>4. Hypocalcaemia</td>
<td>Osteomalacia</td>
</tr>
<tr>
<td>5. Asymmetrical involvement</td>
<td>Myopathy</td>
</tr>
<tr>
<td>6. Other neurological symptom</td>
<td>Guillain Barre syndrome</td>
</tr>
</tbody>
</table>
7. Other endocrine symptoms
   Hypo/hyperthyroidism; Cushing’s Syndrome
8. History of drug ingestion
   Phenytoin, D-penicillamine, chloroquine, corticosteroids, ethanol, zidovudine
9. Infections
   Toxoplasma, trichinella, cysticercosis, human immunodeficiency virus, hepatitis B

It is worth remembering that PM/DM is a symmetrical, progressive proximal muscle weakness

**DIAGNOSIS**

The diagnostic criteria of Bohan and Peter’s requires documentary evidence of muscle inflammation in presence of symmetrical muscle weakness. Any two out of the following three features are required to be present.

**Raised muscle enzymes in the serum**

The levels of creatine phosphokinase (CK), aldolase, ALT and AST are elevated several times the normal level in IIM. The level of CK is usually more than 1000 IU/ml (normal 250 IU/ml) in adult/juvenile PM/DM. There is no good correlation between the extent of muscle damage and the elevated enzyme levels, although this is a useful marker to monitor patients on treatment.

**Electromyogram (EMG)**

In the EMG, short, small polyphasic motor unit with insertional irritability, along with bizarre, high frequency repetitive discharges suggest muscle degeneration. A sluggish motor or sensory nerve conduction abnormality will suggest alternative diagnosis, such as peripheral or anterior horn cell damage.

**Histology**
This is the “gold standard” for establishing the diagnosis of IIM as well as IBM. An open biopsy of the quadriceps or biceps is desirable. The hallmark of the disease is inflammatory cell infiltrate with myofibrillar degeneration and regeneration. The distribution of the infiltrate is different in DM as compared to PM. There is perivasclar and perifascicular cell infiltrates with perifascicular atrophy in DM; whereas in PM, the inflammatory cell infiltrate is within the fascicles and the necrotic fibres are distributed throughout. Additionally, necrosis and phagocytosis of muscle fibres are seen.

**Myositis specific autoantibodies**

Antinuclear antibodies are present in only up to 20% of cases. Antibodies to histidyl-transfer RNA (tRNA) synthetase (Jo-1) enzyme is seen in up to one third of patients with myositis.

**TREATMENT**

Despite the fact that there have been no controlled trials proving their value, prednisolone is the treatment of choice in a daily dosage of 1-2mg/kg of body weight; higher dose is required in case of acute and severe disease. Usually, improvement is noticeable by 6-8 weeks and the higher dose of prednisolone should be continued for 12 weeks. If there is substantial recovery of the muscle power, prednisolone should be reduced at 5mg/d at weekly interval till the dose is 0.5mg/kg/d and thereafter 5mg/d every fortnight till the daily dose is down to 0.25mg/kg. A slower reduction by 2.5mg-5mg/d every month is recommended. There is some wisdom in giving a maintenance dose of prednisolone 0.15mg/kg/d for 6-9 months before reducing by 1mg every month till it is discontinued.

If there is no improvement at the end of 12 weeks, the diagnosis needs to be reviewed, preferably with the pathologist. One should look for other metabolic or neurological diseases or the possibility of inclusion body myositis. If the diagnosis of PM/DM is confirmed, then addition of either daily azathioprine 2-3mg/kg/d or weekly methotrexate 7.5-15mg is helpful. In severe cases of PM/DM, there may be rapid deterioration at the initiation of therapy with acute respiratory failure or myocarditis. In some cases, IV
methylprednisolone 20mg/kg for 3-5 days can be life-saving. In case of respiratory muscle involvement, one needs to be very careful and prepared to do intubation and ventilatory therapy at short notices. IV immunoglobulins and cyclosporine have been useful in juvenile DM/PM. Prompt institution of treatment is life saving. A recent study suggests that a combination of oral methotrexate and azathioprine may benefit patients with treatment resistant myositis, including those who previously had inadequate response.

**How long to treat?**
This is difficult to answer, perhaps a safe limit is two years after complete remission. It has been definitely established that overzealous tapering is associated with a flare of the disease that is more difficult to control than when treating the disease in the first instance.