

REVIEW

Polymyositis/dermatomyositis: the current position

A Urbano-Márquez, Jordi Casademont, Josep M Grau

Polymyositis/dermatomyositis are a heterogeneous group of diseases characterised by skeletal muscle inflammation and necrosis.^{1,2} Since an excellent clinical description in 1903 by Steiner of dermatomyositis, which is essentially still valid,³ much progress has been made towards our understanding of this group of diseases. The most widely used clinical classification of idiopathic inflammatory myopathies is the one proposed by Bohan and Peter in 1975.⁴ That was a landmark, providing guidelines in clinical practice to accurate diagnosis of inflammatory myopathies and standardisation of studies. Nonetheless, the classification was based on clinical data. In view of recent histological and immunological studies the classification proposed by Karpatis *et al* in 1987 seems better to fit our current view of such diseases⁵ (table 1). In this review we focus on recent developments in polymyositis/dermatomyositis, analysing separately the currently considered third major form of inflammatory myopathy—inclusion body myositis.

Epidemiology

Polymyositis/dermatomyositis are the most commonly acquired myopathies in developed countries⁶ if we do not consider subclinical forms of toxic myopathies,⁷ but still fairly uncommon. No more than 5–10 new cases per million persons per year are diagnosed in the United States.⁸ We have found a similar incidence in a large teaching hospital with dermatological clinic and muscle research unit. Nevertheless, the true incidence of polymyositis/dermatomyositis remains unknown for several reasons. Firstly, some patients with dermatomyositis are currently treated by their general practitioner because diagnosis is 'easy'. Secondly, some cases of polymyositis may have such an indolent clinical course that diagnosis is only made when a muscle biopsy is performed; some patients with polymyositis undergo diagnostic muscle biopsy after persistently raised serum creatine kinase activity with minimal or no clinical symptoms. Thirdly, there are few or no prospective studies of connective tissue disease associated myositis; fourthly, not all laboratories

use the different microscope techniques required to diagnose entities such as inclusion body myositis. There are striking differences between the prevalences of the three major forms of idiopathic inflammatory myopathies reported by different authors.^{6,9-12} Probably, dermatomyositis is the most prevalent form, but in reference centres, from where epidemiological data are usually obtained, other less well known entities, such as inclusion body myositis, are relatively more prevalent. Experience with idiopathic inflammatory myopathies in a single institution rarely exceeds 100 cases and thus multicentre studies are mandatory to achieve better knowledge about epidemiology, clinical manifestations, and therapeutic approaches. Considered as a whole group, polymyositis/dermatomyositis are more common in women than in men (3:1). A higher incidence has been reported in black women. Polymyositis/dermatomyositis are seen in all ages with two peaks of incidence—one in the first decade of life and the other in the fifth to sixth decade. Seasonal variation in the onset of polymyositis/dermatomyositis has been noted, suggesting that environmental factors may affect the development of the disease.¹³

The association of polymyositis/dermatomyositis with cancer remains controversial.^{14,15} The incidence of cancer ranges from 10 to 70% in different series, but in most studies the patients selected were not a sufficiently homogeneous group for definite conclusions to be reached. Attempts to identify patients with myositis at risk for developing cancer have been unsuccessful.^{16,17} Our current position is to consider at risk patients over 50 years, particularly men, with typical or no cutaneous lesions but who exhibit capillary damage at muscle biopsy. In such patients a careful clinical history, physical examination, search for occult blood in stools, routine chemistry and haematology tests, and abdominal ultrasound examination are the first step in clinical evaluation. The submission of patients to an exhaustive search for an occult neoplasia seems disproportionate.

Pathogenic mechanisms

Two major pathogenic mechanisms probably lead to polymyositis/dermatomyositis—namely, capillary muscle damage and lymphocyte and macrophage mediated cytotoxicity, each, in turn, affecting patients with dermatomyositis and polymyositis to a different degree. Several authors have shown the unequivocal and constant capillary damage in muscle tissue from patients with dermatomyositis,^{1,18,19} even as an early pathological feature.²⁰ Deposition of C₃b₉

Table 1 Classification of idiopathic inflammatory myopathies

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| <ul style="list-style-type: none"> Dermatomyositis <ul style="list-style-type: none"> Adult form Childhood form Polymyositis Inclusion body myositis Myositis associated with connective tissue disorders Miscellaneous (sarcoid, eosinophilic, graft versus host disease . . .) |
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(membrane attack complex) has also been reported in the capillary bed of patients with dermatomyositis.²¹ The presence of undulating tubules in the endothelial cells is of great value in the diagnosis of dermatomyositis. Ischaemia may be the cause of muscle changes, such as microinfarcts, focal loss of myofibrils, and perifascicular atrophy, all characteristic of biopsy specimens from patients with dermatomyositis.

In polymyositis there is no evidence for muscle ischaemia. Lymphocyte and macrophage mediated cytotoxic damage seem to cause muscle cell injury, as suggested by immunohistochemical^{22 23} and ultrastructural²⁴ studies. Recent papers have shown a strong expression of class I products of the major histocompatibility complex (MHC) in the sarcolemma of most polymyositis muscle cells, especially in the non-necrotic partially invaded cells, in contrast with dermatomyositis biopsy specimens, in which MHC I proteins are only expressed in atrophic cells of the perifascicular areas and in regenerating fibres.²⁵ This suggests that in polymyositis there is a cell mediated immune response against the muscle cell due to sensitisation to a surface associated antigen or to an antigen cross reactive with a component of the fibre surface.

Although the factors that lead to capillary damage and class I MHC expression in muscle cells are not known, viral triggering is possible as occurs in other virus related myopathies such as HIV.²⁶ Thus Bowles *et al* used reverse transcription to show the presence of coxsackievirus B RNA in four of seven dermatomyositis and one of two polymyositis muscle tissues but found none in controls.²⁷ Serological evidence for coxsackievirus B infection has also been published by Christensen *et al*.²⁸ Other factors have been suggested, but the causative agents of polymyositis/dermatomyositis remain unknown. A better knowledge of the different pathogenic mechanisms would enable more rational treatment with the diverse immunomodulators available.²⁹

Clinical picture

The major clinical feature of polymyositis/dermatomyositis is weakness. Far less common are tenderness and spontaneous muscle pain. In dermatomyositis muscle pain is commonly induced by exercise.¹ Atrophy and contractures appear late in the course of the disease. Sometimes, muscular calcifications may develop later, especially in childhood forms. The proximal muscles of the legs are usually the first affected, followed by the proximal arm muscles and the trunk. The presence or absence of capillary damage establishes two subgroups of inflammatory myopathies with different disease chronicity. The presence of capillary damage (dermatomyositis) is associated with a more acute form of the disease, with a mean time from the onset of symptoms to diagnosis of about 10 weeks, whereas in the absence of capillary damage (polymyositis) the mean time from onset to diagnosis is about 25 weeks.³⁰ Skin manifestations are the foremost criteria to differ-

entiate dermatomyositis from polymyositis. The only pathognomonic lesions are the heliotrope discoloration over the upper eyelid and the Gottron erythema over the knuckles.³¹ About 15% of patients with dermatomyositis do not present with these typical lesions, however.³² There are other dermatological changes—subtle rashes occasionally in a photosensitive distribution, vasculitic changes at the bases of the nails, and cuticular overgrowth—that make it difficult to differentiate between polymyositis and dermatomyositis on clinical grounds.

The most common gastrointestinal disorder is dysphagia. It is found clinically in about one third of patients with dermatomyositis, being more rare in polymyositis. It may complicate severe cases, possibly with nasal regurgitation and aspiration.^{33 34} It is usually due to cricopharyngeal striated muscle weakness, though oesophageal dysfunction may also occur. Delayed gastric emptying and decreased intestinal motility are more rarely found, suggesting a malfunction of the smooth muscle in these diseases.³⁵

Cardiac disease is occasionally seen in dermatomyositis but is rare in polymyositis. It presents as unspecific electrocardiographic changes, though arrhythmias, bundle branch blocks, pericarditis, and even congestive heart failure have been described.^{36 37} Some studies have found cardiac disease to be an important prognostic factor.³⁶

Arthralgia is common in dermatomyositis, whereas overt arthritis is rare. The affected joints are usually the small ones of the hands and wrists, but shoulders and knees may also be affected. Arthralgia is not a feature of polymyositis, being found only when associated with interstitial lung disease.^{38 39} Raynaud's phenomenon is often present when polymyositis is associated with other collagen vascular diseases.¹¹

An important systemic manifestation is pulmonary disease. Aspiration pneumonia, hypoventilation due to muscle weakness, opportunistic infections, and drug related pulmonary infiltrates may be found, but the most characteristic clinical picture is interstitial lung disease.^{40 41} It affects 5 to 10% of patients with myositis and presents as non-productive cough, dyspnoea, and hypoxaemia. Radiologically, interstitial lung disease is indistinguishable from idiopathic pulmonary fibrosis. The histological findings in polymyositis/dermatomyositis associated with interstitial lung disease cover a wider field than previously suspected and include some forms of bronchiolitis obliterans organising pneumonia and diffuse alveolar damage.⁴² We have found it to be associated equally with the presence or absence of skin manifestations, but muscle capillary lesions are always present. Fortunately, most cases of interstitial lung disease are responsive to immunosuppressive treatment.⁴³

Diagnosis

To diagnose polymyositis/dermatomyositis it is mandatory to rule out other inflammatory myopathies of known cause and other clinical

Table 2 Differential diagnoses in idiopathic inflammatory myopathies

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| Polymyalgia rheumatica |
| Endocrine myopathies (particularly hypothyroid) |
| Necrotising myopathy associated with cancer |
| Mitochondrial myopathy (adult onset) |
| Trichinosis |
| HIV myopathy (with or without concurrent zidovudine treatment) |
| Eosinophilia myalgia syndrome |
| McArdle's disease and other storage diseases |
| Chronic fatigue syndrome |

conditions that can mimic idiopathic inflammatory myopathy (table 2). Polymyositis/dermatomyositis and even the particular form of it may be suspected on clinical grounds, but further investigations are always required to confirm the diagnosis. The most widely accepted diagnostic criteria are those originally proposed by Bohan and Peter⁴ and modified by Hudgson and Peter⁴⁴ based on a compatible clinical picture, raised creatine kinase activity, multifocal electromyographic changes of 'myositis', and biopsy evidence of necrosis and inflammation. When all four criteria are satisfied a diagnosis of *definite* idiopathic inflammatory myopathy can be made. The presence of three or two criteria allows the diagnosis of *probable* and *possible* idiopathic inflammatory myopathy respectively. Probably not all the criteria should be given the same weight as the definition of polymyositis/dermatomyositis is based on histopathological criteria.^{1,2} We believe that the biopsy findings are of great importance in establishing the diagnosis. Without them the diagnosis will always be questionable, whereas with a biopsy definite forms can be diagnosed even in the absence of more than one of the other criteria. Among the 'muscle enzymes' creatine kinase activity is the most sensitive and useful biochemical determination.⁴⁵ It is usually, but not invariably, raised during the active phase of disease. Immunological studies such as rheumatoid factor, immunoglobulins, complement, and antinuclear antibodies are of little value in the diagnosis and management of any form of polymyositis/dermatomyositis. The exception is the Jo-1 antibody, an antinuclear antibody clearly related to the presence of interstitial lung disease.^{46,47} The significance of the antiendothelial antibodies, taking into account the importance of capillary damage as a causative factor of dermatomyositis, is not yet completely understood.⁴⁸

Electromyographic findings in dermatomyositis may be normal in about 10% of patients, or even more patients if the sample is not wide, owing to the multifocal nature of this

disease. Electromyography is not a good test for assessing disease activity or for establishing differences between the different forms of polymyositis/dermatomyositis.⁴⁹ Muscle scintigraphy after administration of ^{99m}Tc labelled phosphate complexes, ultrasonography, computed tomography, and magnetic resonance imaging are of limited value for diagnostic purposes but may be useful in identifying the site of biopsy.⁵⁰

Muscle biopsy is indispensable for the diagnosis of polymyositis/dermatomyositis. If proper techniques are used a distinction between the different forms is easily accomplished. We believe that biopsy is always indicated for adults. The biopsy must be done on affected but not wasted muscle, and by sensitised personnel. The convenience of an open biopsy is debatable. We performed simultaneous open and needle biopsies on 13 patients, and the diagnostic yield was the same. The advantages of an open biopsy are a better sample, better orientation, and the possibility of studying different areas of the same specimen—particularly useful in focal diseases such as dermatomyositis. We only use needle biopsies for follow up of patients.

Table 3 summarises the different pathological features of polymyositis and dermatomyositis, including also inclusion body myositis characteristics (see below). A not uncommon clinical dilemma is that of a patient receiving steroids who complains of weakness. Although a further biopsy—a needle biopsy would be indicated in this case—might help in differentiating steroid from inflammatory myopathies, this is not always the case. The most salient histopathological finding in steroid myopathy—type II fibre atrophy—can be found as an unspecific finding in polymyositis/dermatomyositis, and inflammation may be absent from a biopsy specimen of a patient with myositis receiving steroid treatment. Clinical judgment is probably most important in distinguishing between such entities.

Treatment and prognosis

Prognosis depends on the form of idiopathic inflammatory myopathy. Dermatomyositis seems to be a more acute and aggressive disease, but in turn responds more rapidly to immunosuppressive treatment. Its course tends to be remitting with periods of improvement and remission after treatment alternating with occasional relapses. The frequency and severity of relapses diminish over the years. Interestingly, there are patients with dermatomyositis who have relapses exclusively affecting the skin. Muscle strength, creatine kinase activity, and even successive muscle biopsy specimens are normal. In such cases a 'silent' neoplasia, such as prostatic gland carcinoma or low grade non-Hodgkin's lymphoma, has to be completely ruled out. Polymyositis, on the other hand, is a more insidious disease and the response to treatment slower. Treatment strategies have to be based on these points and on pathogenetic mechanisms. Adverse side effects must be avoided as far as possible, while remembering that the degree of residual weakness depends on

Table 3 Salient distinctive pathological features of polymyositis/dermatomyositis (PM/DM) and inclusion body myositis (IBM)

| | DM | PM | IBM |
|---|------------------|--------------|--------------|
| Perifascicular atrophy | + | - | - |
| Capillary damage | + | - | - |
| Lined vacuoles | - | - | + |
| Ragged-red fibres | - | - | + |
| Partial invasion | - | + | + |
| Cellular infiltration (predominant) | B Perivascular | T Endomysial | T Endomysial |
| MHC* class I expression | + Perifascicular | + | + |
| C ₃ b ₁₉ in vessels | + | - | - |

*MHC=major histocompatibility complex.

the accuracy of the initial treatment. In dermatomyositis we use high dose steroids (1 mg/kg daily). In polymyositis we use steroids in lower doses (0.3 mg/kg daily) together with azathioprine (2 mg/kg daily). With patients over 70 we follow the polymyositis strategy. After two to four months' treatment in dermatomyositis and four to six months in polymyositis the doses are gradually tapered over one to one and a half years. Similar schedules are followed by other authors.^{9 51}

There are single case reports of patients with polymyositis/dermatomyositis treated with cyclosporin, but results are contradictory.^{52 53} Only recently has the usefulness of this drug been clearly shown in refractory cases of juvenile dermatomyositis.⁵⁴ Our own experience with this drug is also encouraging not only in refractory cases, but as a first choice of treatment in some particular forms (aggressive juvenile forms of dermatomyositis). Important adjuncts to the drug treatment include a high protein diet and isometric muscle exercise after a brief period of bed rest. Overall mortality in polymyositis/dermatomyositis is about 10% if paraneoplastic forms are excluded.

Inclusion body myositis

Inclusion body myositis is currently considered one major form of inflammatory myopathy owing to its inflammatory exudates and the expression of class I MHC products in some muscle cells.^{9 55} It was first described in 1971⁵⁶ and characterised as a distinct clinical entity in 1978.⁵⁷ As the diagnostic criteria are mainly histopathological,⁵⁸ there are a wide range of clinical forms.^{10 57-60} The most typical is that of an old man whose only complaint is longstanding predominantly distal muscle weakness, or that of a patient with polymyositis resistant to treatment.⁵⁸ Anecdotal associations with other diseases have been reported.⁶¹⁻⁶³ The long clinical course, the particular distal distribution of weakness, the reported familial cases, and the lack of response to any immunosuppressive or antiviral treatment suggest that it is not an inflammatory condition. Some authors suggest that it should be considered a primary non-inflammatory muscle disease.^{55 64} It is currently managed by physiotherapy and general support. No epidemiological data on its mortality are available.

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Ann Rheum Dis 1991 50: 191-195

doi: 10.1136/ard.50.3.191

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