CASE REPORTS

Polymyositis, not polymyalgia rheumatica

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Abstract

The distinction between polymyalgia rheumatica and polymyositis is important for treatment and prognosis. Four elderly patients were diagnosed and treated for polymyalgia: raised creatine kinase led to muscle biopsy and a diagnosis of polymyositis.

Polymyositis and polymyalgia rheumatica may both present with a similar distribution of proximal muscle pain but are not normally confused. In polymyositis tenderness or weakness of muscles may be marked and there may be other helpful diagnostic features—for example, dysphagia, whereas muscle pain or stiffness without weakness are the more prominent symptoms in polymyalgia rheumatica. In older patients the differentiation may not be easy, especially when muscle weakness is not marked. We report four such cases, seen in one year in one rheumatologist's practice, who were originally diagnosed and treated as polymyalgia rheumatica when the correct diagnosis was polymyositis.

Case reports**CASE NO 1**

An 83 year old woman presented to her general practitioner with proximal muscle pain and stiffness. The erythrocyte sedimentation rate (ESR) was 65 mm/h, but no muscle enzyme determinations were performed. A diagnosis of polymyalgia rheumatica was made and treatment was started with prednisolone 30 mg/day, which led to improvement and the ESR fell to 10 mm/h. Three months later treatment continued with prednisolone 10 mg/day. At this stage she was referred because of further pain and stiffness and a recent onset of proximal muscle weakness. On admission to the rheumatology ward she was found to have wasting and weakness of shoulder and pelvic girdle muscles. Investigations showed the haemoglobin concentration to be 129 g/l, the ESR 56 mm/h, aspartate transaminase was normal, but creatine kinase was grossly raised at 517 IU/l (normal range 20-205 IU/l). Needle muscle biopsy confirmed a moderate myositis.

CASE NO 2

A 74 year old woman presenting with proximal muscle pains and stiffness had an ESR of 96 mm/h, but no muscle enzyme results were obtained. A diagnosis of polymyalgia rheumatica was made and treatment was started with

prednisolone 30 mg/day; two weeks later there was improvement and the ESR had fallen to 14 mm/h. Six months later she again had proximal muscle pain but also weakness and she reported numerous chest infections, all responding poorly to antibiotics. Hospital referral and subsequent admission confirmed a broncho-pneumonia and marked proximal muscle wasting and weakness. Investigations showed a haemoglobin concentration of 119 g/l and the white blood cell count was raised at $14.9 \times 10^9/l$ with a neutrophil leucocytosis. The ESR was raised at 100 mm/h and creatine kinase was raised at 600 IU/l. Needle biopsy confirmed a moderate myositis.

CASE NO 3

A 77 year old woman presented to her general practitioner with pain and weakness of the proximal muscles of her arms and legs. She had difficulty climbing stairs and in rising from a chair. Investigations showed a normal full blood count and an ESR of 6 mm/h; no muscle enzyme determinations were requested. Polymyalgia rheumatica was diagnosed and prednisolone 15 mg/day started; there was no improvement after four weeks so she was referred to us. Examination confirmed marked proximal tenderness and weakness. Investigations showed a normal ESR, but the creatine kinase was 2493 IU/l and aspartate transaminase 251 IU/l (normal range 10-35 IU/l). Needle muscle biopsy confirmed a severe myositis.

CASE NO 4

A 69 year old man presented to his general practitioner with a one month history of pain and stiffness in both thighs and upper arms. The stiffness was most marked in the morning and he had difficulty dressing. He was referred to us when investigations showed a haemoglobin concentration of 147 g/l, an ESR of 18 mm/h, normal aspartate transaminase, and creatine kinase raised at 230 IU/l. A diagnosis of polymyalgia rheumatica was made and treatment was started with prednisolone 15 mg/day with subsequent improvement. Over the next few months his creatine kinase rose to 747 IU/l and aspartate transaminase to 42 IU/l. Although he felt well and muscle weakness was not present a muscle biopsy was performed, which confirmed a mild myositis.

Discussion

Our four cases highlight the occasional difficulty in distinguishing polymyositis from poly-

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myalgia rheumatica; the distinction is important because a diagnosis of polymyositis has different implications, such as a higher risk of underlying malignancy and the need for a higher initial dose of steroids, and the possibility of immunosuppressive treatment. Both patients 1 and 3 responded to an increase in their steroid dose and the initiation of azathioprine. Patients 2 and 4 responded to an increased steroid dose only.

Polymyalgia rheumatica and its close relative, giant cell arteritis, are not rare conditions and their incidence increases with age.¹ The diagnosis of polymyalgia rheumatica is one of exclusion as firm diagnostic criteria do not exist. Patients describe muscle pain and stiffness often around the shoulder girdle initially but later spreading to affect the pelvic girdle too. Stiffness is usually the main feature and is more marked after resting; it may prevent the patient rolling over in bed or from getting up in the morning. Criteria for the diagnosis of polymyalgia rheumatica were tested by Bird *et al.*² The seven most useful criteria were: bilateral shoulder pain and stiffness; onset of illness of less than two weeks' duration; initial ESR greater than 40 mm/h; duration of morning stiffness exceeding one hour; aged 65 or over; depression or weight loss, or both; and tenderness of both upper arms. They suggested that three of these criteria made a diagnosis of polymyalgia rheumatica probable. These criteria were not tested in patients with polymyositis, however, and at least three of these criteria were satisfied in all four of our patients.

In the vast majority of patients with polymyalgia rheumatica the ESR is raised.^{3,4} A completely normal ESR therefore makes the diagnosis unlikely but not untenable. Anaemia may occur, which may be either hypochromic or normochromic. Abnormal liver function tests, especially increased transaminases may occur,³ but muscle specific enzymes such as creatine kinase are normal.

Non-steroidal anti-inflammatory agents may be partially effective in the treatment of polymyalgia, but corticosteroids have a more dramatic and sustained effect.³ Polymyalgia can be controlled in most patients with 15–20 mg prednisolone/day⁴ or less,⁵ though the rate at which this dose can be reduced varies greatly. The average length of treatment needed in one series was 31 months.⁵ Polymyositis features in the differential diagnoses of polymyalgia, but is rare compared with the musculoskeletal syndromes associated with various malignancies or with hypothyroidism, cervical spondylosis, or rheumatoid arthritis.

Polymyositis is a rarer condition with an estimated incidence of 0.5 per 100 000. In adults the peak incidence is between the ages of 45 and 54 and decreases thereafter.⁶ The most common presenting symptom is proximal muscle weakness, but muscle pain and tenderness may also be prominent. Other clinical features may be present, including dysphagia, Raynaud's phenomenon, and arthritis. In one series the ESR was normal in 45% of cases of polymyositis and only 19% had an ESR of over 50 mm/h.⁷ Muscle enzyme activity, especially of creatine kinase, is usually raised in active polymyositis and falls with successful treatment. Electromyography is abnormal in about 90% of cases and muscle biopsy is usually diagnostic.^{7,8} In about 20% of cases an underlying connective tissue disease will be present and there is also an association with malignancy.⁹

Polymyositis is managed with high dose steroids and initial daily dosages of prednisolone of 60 mg/day or even higher are often needed depending on disease severity; these high doses may need to be maintained for long periods. Immunosuppressive drugs, such as azathioprine or cyclophosphamide, are often used in association with steroids in a steroid sparing regimen: they are not helpful in polymyalgia.

In summary, the distinction between polymyositis and polymyalgia rheumatica is not always easy at presentation; patients presenting with proximal muscle pain, stiffness, or weakness should therefore have a careful history taken, a full clinical examination to distinguish stiffness from weakness, and initial blood tests should include an ESR and determination of muscle specific enzymes such as creatine kinase.

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