Contracturing granulomatous myositis: a separate entity

N J Simmonds, B I Hoffbrand

Abstract

Granulomatous muscle disease is most commonly seen in sarcoidosis, but may be seen in association with a wide variety of other disorders or in isolation. Patients with granulomatous myositis usually present with slowly progressive muscle pain and weakness affecting mainly proximal muscles. There are, however, a few reports of granulomatous muscle disease presenting with flexion contractures of the limbs. Two further patients with granulomatous muscle disease and flexion contractures of the limbs, but with no evidence of systemic granulomatous disease, is presented. It is suggested that such patients represent a separate clinical entity that is distinct from idiopathic granulomatous myositis presenting with muscle pain and weakness. The association of contracturing granulomatous myositis with a long-standing vasculitis in one patient suggests that the two conditions may be related.

Granulomatous muscle disease is most often seen in sarcoidosis, but occurs in a wide variety of disorders including Wegener's granulomatosis, polymyositis, dermatomyositis, mixed connective tissue disease, tuberculosis, Crohn's disease, myasthenia gravis and dysgerminoma.1-7 Granulomatous myositis has also been described in isolation, and in the absence of systemic granulomatous changes.8-11 These patients tend to present with slowly progressive muscle pain and weakness affecting mainly proximal muscles. There are, however, in addition, a few reports of granulomatous muscle disease presenting with flexion contractures of the limbs.12-15 In each case a diagnosis of sarcoidosis was made, but solely on the basis of the muscle biopsy. In none were there clinical, radiological or biopsy findings of an underlying granulomatous disorder to justify the diagnosis.

We present two further patients with granulomatous myositis and flexion contractures of the upper limbs with no evidence of sarcoidosis. We believe that these and the previously described cases of contracting granulomatous myositis12-15 may represent a separate clinical entity distinct from idiopathic granulomatous myositis. One of our patients had a longstanding associated systemic vasculitis which may be a pointer to the underlying pathology.

Case reports

Case 1
A 58 year old woman presented in 1976 with a six month history of progressive flexion contractures of the long finger flexors of both hands and weakness around the shoulder girdle. She had Scaglietti muscle slides, involving the release of the origins of the flexor muscles of the forearm at the elbow,16 and biopsies showed numerous sarcoïd-like granulomata with surrounding lymphocytes (figure). Investigation showed a strongly positive rheumatoid factor, but no evidence of generalised sarcoidosis. The chest radiograph and liver biopsy were normal, a negative Kveim test and a serum angiotensin converting enzyme (SACE) of 63 nmol/min/ml (normal range 16–53 nmol/min/ml).

Investigation in 1979 for ankle swelling, shortness of breath and aching in her hips revealed a normochromic, normocytic anaemia, with an erythrocyte sedimentation rate (ESR) of 80 mm/hr and increased plasma globulins. A bone marrow showed erythroid hypoplasia and spine radiographs showed osteoporotic collapse of L3. These symptoms resolved spontaneously, but later that year she developed a mixed motor and sensory neuropathy affecting the lower limbs.

Figure  Muscle biopsy showing a granuloma with surrounding lymphocytes (H & E × 270).
She then remained well until 1987, when she presented with a vasculitis; with lesions on toes, fingers, knees and elbows and ulcers on both ankles. She had marked livedo reticularis and flexion deformities of her hands, more marked than eight years previously, wasting of the small muscles of the hands and inability to dorsiflex her feet fully. Investigations showed: haemoglobin 8.8 g/dl, ESR 135 mm/hr, urea 12.6 mmol/l, creatinine 187 mmol/l, albumin 31 g/l with increased α and γ globulins, rheumatoid factor positive at 1/1520, antinuclear antibody positive at 1/80 with a diffuse pattern, normal C3, CH50 23% of normal, and binding complexes of C1q with IgM and IgA, SACE 42 mmol/min/ml, antineutrophil cytoplasmic antibodies (ANCA) strongly positive with staining of the nuclear membrane, using an indirect immunofluorescence technique. A skin biopsy showed a severe acute vasculitis involving medium sized arteries with foreign body giant cells near damaged vessel walls. A renal biopsy, performed a year later, showed evidence of previous inflammatory injury in the form of organising crescents.

She was treated with prednisolone and dapsone and made a good recovery. The prednisolone was stopped because of further vertebral collapse and she has since remained well taking only dapsone 25 mg once daily, but with continuing slow progression of her contractures.

Case 2
A 65 year old man presented in 1965 with pain and swelling in his right arm resulting in difficulty in straightening his elbow. This resolved spontaneously, but five years later he developed a painful swelling of the forearm muscles on the right associated with a contraction of his right ring finger. Biopsy of the affected area revealed a granulomatous myositis, with muscle fibres undergoing active necrosis and phagocytosis, numerous granuloma and perivascular infiltration of inflammatory cells, predominantly lymphocytes. Postoperatively there was further inflammation and progressive contracture formation.

In July 1971 he developed swelling of the left triceps which cleared spontaneously over the next month and then recurred in September 1971. His ESR at that time was 40 mm/hr. His full blood count, serum calcium, creatine kinase and proteins were normal. His tuberculin reaction was positive at 1:1000 and a Kveim test was negative. Once more, the swelling resolved spontaneously.

The following year the left triceps mass recurred and he was started on steroids with an initial marked improvement. From 1972–8 his clinical course was characterised by remissions and exacerbations of his myositis, mainly involving the left triceps, but also the right forearm and right sternomastoid. Azathioprine was added as maintenance therapy in 1973 and in 1978, after a prolonged remission, all medications were stopped. He then remained free of myositis until his death in 1984 from carcinomatosis.

Discussion
Pathological studies have failed to reveal any specific features that enable a distinction to be made between idiopathic granulomatous myositis and sarcoidosis and other granulomatous diseases. The final diagnosis appears to rest on any associated abnormal features. The absence of such features leads us to believe that the published cases of granulomatous myositis presenting with contractures had idiopathic granulomatous myositis. Our case 1 was published previously as having sarcoidosis although this diagnosis was not felt to be substantiated at the time (Dr D G James, personal communication). The table lists the features of patients with contracturing granulomatous myositis reported to date, including our two. Contracturing of limb muscles has been reported in otherwise typical sarcoidosis and in polymyositis, but not to our knowledge in Wegener’s granulomatosis.

Our case 1 has had a remitting systemic vasculitic illness for the past nine years over which period the contractures of the legs have developed and those of the arms have increased, suggesting a close relationship between the pathological processes. This illness has included a crescentic glomerulonephritis and a positive ANCA, both features of vasculitic diseases. Our case 2 had no clinical features of a systemic vasculitis, although azathioprine appears to have induced a remission. Of the previously published cases of contracturing granulomatous myositis one had nephritis. In no case, including our own, was vasculitis proven in the muscle biopsy, although vasculitis has been seen in association with granulomatous muscle disease in Wegener’s granulomatosis.

There is no report of contracturing or of vasculitic disease in the cases of idiopathic granulomatous myositis reported to date. The clinical course in our first patient leads us to believe that the two processes may be related and that assay of ANCA may prove useful in confirming this relationship in similar cases in the future.

We are grateful to Dr D G James for permission to publish details of case 2.
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