CASE REPORT

Childhood dermatomyositis recurring in adulthood half a century later

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SUMMARY

A 62-year-old woman presented to the emergency department with malaise, lethargy and proximal muscle weakness. She had a similar presentation in childhood. A muscle biopsy was inconclusive, however, she was treated for a presumptive diagnosis of inflammatory myositis. In a second flare in childhood, she developed proximal muscle weakness and calcinosis cutis. A muscle biopsy was consistent with juvenile dermatomyositis. She was treated with corticosteroids, however, she relapsed again 1 year later, requiring a prolonged course of corticosteroids, which led to remission and treatment was subsequently stopped. She remained well for 30 years without any relapses, however, at the age of 40 she was diagnosed with malignant melanoma, treated with local excision and radiotherapy. She had a further period of 22 years in good health, until her current presentation, with a relapse of dermatomyositis, 52 years after the last flare. She is currently in remission following successful immunosuppressive therapy.

BACKGROUND

This is a very unusual presentation of dermatomyositis, in three main ways.

First, generally patients with dermatomyositis tend to follow a monocyclic course. This patient had a polyphasic phase in childhood followed by a very prolonged phase of remission with no functional impairment.

Second, The patient had the longest remission interval of dermatomyositis reported in the literature.

Third, the link between dermatomyositis and malignancy is well-established, usually the malignancy coinciding with a recurrence of the inflammatory muscle disease. In this case, the presentation of malignancy was at least 30 years after and 21 years prior to the occurrence of the inflammatory muscle disease.

CASE PRESENTATION

A 62-year-old woman presented to the accident and emergency department with malaise, lethargy and proximal muscle weakness affecting upper and lower limbs (power 2/5 in proximal muscle groups, 5/5 distally). All other systems on presentation were unremarkable, apart from an erythematous macular rash over the anterior chest wall.

She had an interesting medical history as a child, whereby in 1956, at the age of 8, she presented with a 5-week history of proximal muscle weakness of the legs, arms and trunk, fever, malaise and arthralgia. The erythrocyte sedimentation rate (ESR) at the time was 105 mm/h. A muscle biopsy was

inconclusive, however, the patient was started on corticosteroids for a presumptive diagnosis of inflammatory myositis. The patient made a good recovery and was discharged home on a reducing regime of corticosteroids.

This initial presentation was followed by a second event in 1957 when the patient had a similar presentation to the one before, with proximal muscle weakness of upper and lower limbs, this time with associated calcinosis cutis. A muscle biopsy was undertaken, which showed "fat necrosis with reactionary lymphocytic infiltration and calcification, muscular thickening and intimal proliferation of smaller arterioles and degenerative changes in muscle fibres." The clinical and histopathology findings were thought to be consistent with a diagnosis of juvenile dermatomyositis. The patient was treated with corticosteroids, only to represent with yet another relapse in 1958. On this occasion, she was given a prolonged course of oral prednisolone, leading to remission and the treatment was subsequently stopped.

She remained completely well for 30 years, on no medical treatment and without encountering any relapses of her original disease or other medical problems. However, at the age of 40 she was diagnosed with malignant melanoma, treated with local excision and radiotherapy. She had a further period of 22 years, which she spent in good health, until her recent presentation in 2009, when she had a relapse of dermatomyositis (DM) occurring over 2–3 weeks.

INVESTIGATIONS

During this admission, initial blood tests showed a creatine phosphokinase (CPK) level of 18 319 U/L, ESR 95 mm/h, C reactive protein (CRP) 36.5 mg/L. Antinuclear antibody and antineutrophil cytoplasmic antibody were negative. The patient developed prominent symptoms of dysphagia to solid food during her admission and in view of the known associations of DM with underlying malignancy, an oesophagogastroduodenoscopy was undertaken, which was reassuringly normal. A CT of the thorax, abdomen and pelvis, which formed part of the malignancy screen investigations, was also normal.

The muscle biopsy confirmed inflammatory myositis, with the distribution of the lymphocytic infiltrate (largely perivascular and perifascicular with B-lymphocytes) being suggestive of DM.

TREATMENT

The patient was treated with three pulses of intravenous methylprednisolone (500 mg) followed by oral prednisolone (30 mg) and methotrexate (20 mg weekly).

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OUTCOME AND FOLLOW-UP

The CPK normalised within 3 weeks of treatment (CPK 50 U/L, ESR 20 mm/h), dysphagia improved and there was gradual improvement in muscle strength. She is currently in remission on low-dose prednisolone, and methotrexate.

DISCUSSION

DM and polymyositis are autoimmune myopathies characterised by symmetric proximal muscle weakness, muscle inflammation, extramuscular manifestations such as lung and skin involvement and frequently, the presence of autoantibodies.¹ A small but significant proportion of DM and polymyositis is associated with malignancy, the precise link remaining incompletely understood.^{2–3} Generally, DM patients tend to follow a monocyclic course.⁴

DM has been known since 1887. Unverricht⁵ was the first to recognise the association of skin and muscle lesion and in 1891 gave the disease its present name.

We report a case of DM relapsing after 51 years of remission. The association between malignancy and DM is well recognised, with malignancy often detected within the first year of DM.¹ This case is unusual not only due to the late recurrence of the myositis, but also in the presentation of malignancy at least 20 years after and prior to the recurrence of the inflammatory muscle disease.

Our patient had a polycyclic phase with three relapses over 3 years in her childhood. She then had a very prolonged phase of remission with no functional impairment. A report by Touton⁶ describes two cases of DM in childhood with recurrence in adulthood after 30 and 21 years of disease-free period. In another case reported by Martini,⁷ the patient had a polyphasic clinical course with recurrences separated by long periods of remissions. We report the longest recurrence period of DM 51 years after complete remission. This is the longest remission interval of DM reported in the literature, which supports that relapses can occur at late stage, raising issues concerning maintenance therapy.

In addition, the case described raises a further issue. Despite the juvenile-onset of DM, the most recent relapse in our patient was during the postmenopausal period.

Hormones play an immunomodulatory role in the development of autoimmune disease and similar to other rheumatological conditions such as systemic lupus erythematosus (SLE), rheumatoid arthritis (RA) and systemic sclerosis, DM also predominantly affects females, with a considerable proportion of patients being of reproductive age. It is believed that the female predilection for these conditions could be due to oestrogens playing a role in their pathogenesis.⁸ Oestrogen has been shown to potentiate the phagocytic and antigen-presenting activity of macrophages, promote the maturation of T-helper cells and activate polyclonal B cells.⁹ However, there still remains controversy in their role in the pathogenesis and expression of common rheumatological conditions. The menopause seems to

lessen the severity of conditions like SLE but coincides with the appearance of many of the common arthritic conditions, including osteoarthritis and RA.¹⁰ Here we present yet another rheumatological condition from the inflammatory myositis family, representing after five decades of remission and during the postmenopausal period, in a patient with normal menarche and puberty. This raises questions and further controversy around the role of oestrogens in the pathogenesis of this condition and highlights the need for further research to establish the hormonal links between pathogenesis and disease expression.

Learning points

- ▶ Dermatomyositis is an important differential diagnosis in patients presenting with proximal muscle weakness, and the condition can present in childhood and continue into adult life.
- ▶ A high index of suspicion for malignancy is required in patients with dermatomyositis.
- ▶ Dermatomyositis can relapse at any stage even following a very long period of remission.
- ▶ Maintenance immunosuppressive therapy should be carefully considered in these patients.

Contributors AY was involved in patient care. All the authors have contributed significantly towards the case report.

Competing interests None.

Patient consent Obtained.

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