CASE REPORT

Unusual sequelae of adult-onset dermatomyositis

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SUMMARY

A 44-year-old woman diagnosed with dermatomyositis 5 years ago based on progressive proximal muscle weakness, elevated creatine kinase, typical findings on electromyography and muscle biopsy. Despite the treatment, in contrast to improvement in her muscle symptoms, the heliotrope rash of her eyelids persisted. After several years, the patient developed multiple limited skin retraction lesions with hyperpigmentation on both lower limbs. Palpation of these lesions revealed dry, cold and very firm skin on both thighs and calves, particularly in the distal areas. X-ray and ultrasound imaging of the calves showed multiple subcutaneous calcifications in the distal muscles.

BACKGROUND

Calcinosis is a relatively common disease manifestation in juvenile dermatomyositis (DM). In adults, calcinosis is less common and usually appears later in the course of the disease. The typical involvement includes proximal muscles.

The presented case describes a unique case that behaves unlike typical cases.

CASE PRESENTATION

A 44-year-old woman developed DM 5 years ago. The diagnosis of DM was based on progressive proximal muscle weakness, elevated creatine kinase, typical findings in electromyography (EMG), and muscle biopsy. Silent underlying malignancy work-up was unrevealing. Mixed connective tissue disease and overlap syndromes were ruled out. Systemic sclerosis was very unlikely due to the absence of skin involvement, Raynaud's phenomenon and negative serology. Treatment with pulses of methylprednisolone 1 g a day for 3 days was initiated, followed by high-dose prednisone with concomitant oral weekly methotrexate (15 mg/week), and high doses intravenous immunoglobulins. Slow and gradual improvement in the well-being status was noted, and muscle strength was restored along with normalisation of creatine kinase levels. Despite the treatment, and in contrast to improvement in her muscle symptoms, the eyelids heliotrope rash persisted (figure 1). Repeated malignancy workup 2 years later was negative again. After several years, the patient developed multiple limited skin retraction lesions with hyperpigmentation on both lower limbs. Palpation of these lesions revealed dry, cold and very firm skin on both thighs and calves, particularly distal areas. X-ray (figure 2A) and ultrasound imaging (figure 2B) of calves showed multiple subcutaneous calcifications in distal muscles.

Figure 1 Heliotrope rash on eyelids.

INVESTIGATIONS

Elevated creatine kinase, typical findings in EMG, and muscle biopsy. Underlying malignancy work-up was unrevealing. Mixed connective tissue disease and overlap syndromes were ruled out. Systemic sclerosis was very unlikely due to the absence of skin involvement, Raynaud's phenomenon and negative serology.

DIFFERENTIAL DIAGNOSIS

- ▶ Systemic sclerosis
- ▶ Overlap syndromes
- ▶ Mixed connective tissue disease

TREATMENT

Treatment with pulses of methylprednisolone 1 g a day for 3 days was initiated, followed by high-dose prednisone with concomitant oral weekly methotrexate (15 mg/week), and high doses intravenous immunoglobulins.

OUTCOME AND FOLLOW-UP

After several years, the patient developed multiple limited skin retraction lesions with hyperpigmentation on both lower limbs. Palpation of these lesions revealed dry, cold and very firm skin on both thighs and calves, particularly distal areas. X-ray (figure 2A) and ultrasound imaging (figure 2B) of calves showed multiple subcutaneous calcifications in distal muscles.

DISCUSSION

Calcinosis is a relatively common disease manifestation in juvenile DM and occurs in up to 30% of

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Figure 2 (A) Soft tissue calcifications seen on X-ray. (B) Subcutaneous calcifications seen on ultrasound.

patients. It usually occurs 1–3 years after diagnosis and it is associated with considerable morbidity, as at times it may cause skin ulceration and secondary infections.¹ In adults, calcinosis is less common and usually appears later in the course of the disease.² The exact mechanism underlying the development of calcinosis is not well established. However, persistent inflammatory response seems to play a central role in the development of calcinosis. Increased local production of the tumour necrosis factor α has been suggested to be associated with prominent inflammation and calcinosis appearance.³ Shizumi *et al*⁴ demonstrated that activated macrophages and inflammatory cytokines play an important role as well. Calcinosis usually involves proximal rather than distal limbs. It has been supposed that early aggressive treatment of DM may prevent the musculoskeletal sequelae of the disease, including calcinosis itself.⁵ Terosso *et al*⁶ reported a case in which, calcinosis lesions decreased and the patient regained full range of movement and quality of life with no recurrence has occurred after 8 years of follow-up. We

presented a case of muscle atrophy and calcinosis in distal limb muscles that developed in an adult patient despite intensive treatment including bisphosphonates. It is interesting to note that the heliotrope rash persisted as well, despite intensive medical treatment. It deserves further investigation whether persistent heliotrope rash may predict the future development of calcinosis.

Contributors MEN and AB-G prepared the manuscript, obtained the informed consent and have chosen the clinical images. RB and YB-M revised the manuscript.

Competing interests None.

Patient consent Obtained.

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Learning points

- Calcinosis is a relatively common disease manifestation in juvenile dermatomyositis.
- In adults, calcinosis is less common and usually appears later in the course of the disease.
- Calcinosis usually involves the proximal rather than the distal limbs.

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