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Images in Clinical Rheumatology

Extensive Calcinosis in Adult Dermatomyositis[☆]

Calcinosis extensa en dermatomiositis del adulto

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A female aged 48 who presented at the surgery with photosensitivity, heliotrope erythema and cervical and upper trunk level erythematous injuries, in addition to myalgias and proximal muscle weakness, 3/5 in upper and lower limbs, and weakness of the flexor tendons of the neck. Laboratory tests revealed erythrocyte sedimentation of 54 mm/h, CPK 9.870 U/l, significant elevation of transaminase levels and lactate dehydrogenase levels, and ANA 1:1.280 mottled pattern. Magnetic resonance of the thighs showed exaggerated inflammatory changes. Electromyography had potentials of polyphasic motor units of short duration and low amplitude, mainly at gluteus level. Muscular biopsy was compatible with dermatomyositis. Treatment was initiated with 40 mg/day prednisone, 20 mg/week methotrexate, 5 mg/week folic acid, 200 mg/12 h hydroxychloroquine and muscular physiotherapy. Twenty days later, the patient presented with progression of weakness of 2/5 in lower limbs and 1/5 in upper limbs and was incapacitated and prostrate. Three pulsatile administrations of methylprednisone, cyclophosphamide and gammaglobulins were given. Treatment continued with pulsatile administrations of cyclophosphamide and she received infusions of 1 g/kg/2 days in three consecutive stays every 30 days. Improvement occurred with intensive treatment and rehabilitation, with the patient being able to stand up and to walk after 3 months of treatment. There was an improvement in her extensively compromised cutaneous barrier after initiating treatment with 1 g rituximab. Cutaneous calcifications progressed in triceps, gluteus, thighs and legs despite treatment with gammaglobulins, rituximab, pamidronate and colchicine (Fig. 1).

Calcinosis is a common finding in inflammatory myopathies, especially in juvenile dermatomyositis (20%–70%), but it is infrequent when the disease presents in the adult (20%).¹ It is associated with the disease activity and progression, a highly compromised cutaneous barrier and a delay or lack of response to treatment.² It is more common in regions of persistent stress and trauma. Its pathogenesis is unclear.² Several recent reports associate this complication with the presence of anti-NXP2 antibodies in disseminated form and with anti-PM/ScI^{1,3,4} forms.

This complication is difficult to treat. Several strategies which have demonstrated improvement in case reports are: warfarin (not currently recommended due to risk of bleeding), thalidomide, diltiazem or colchicine (with tolerance-dependence dose adjustment), bisphosphonates (cyclical infusions of amidronate or alendronate with oral administration of maintenance treatment), immunoglobulins, and biologic agents, including infliximab and abatacept.^{5–10} Some recent case reports suggest that rituximab could be a favourable therapeutic option in severe and refractory cases.^{5,6} Surgery is reserved for discrete lesions due to the risk of infection.^{6–10}

Ethical Liabilities

Protection of people and animals. The authors declare that no experiments using human beings or animals have been carried out for this research study.

Data confidentiality. The authors declare they have followed the protocols of their centre of work on patient data publication.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

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Figure 1. Extensive lineal calcifications of reticular appearance in dermis and hypodermis: (A, B) upper limbs; (C–F) lower limbs.

Conflicts of Interest

The authors have no conflicts of interest to declare.

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