ARP Rheumatology 2024;3:237-239
https://doi.org/10.63032/WGLX8122

IMAGES IN RHEUMATOLOGY

Hard as stone: an exuberant form of calcinosis cutis

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BACKGROUND

Dermatomyositis (DM) is an immune-mediated myopathy characterized by proximal skeletal muscle weakness, muscle inflammation, and distinct skin manifestations. Calcinosis cutis, the deposition of insoluble calcium salts in soft tissues, affects approximately 8% of DM patients[1] and poses significant treatment challenges. It can complicate with inflammation, ulceration, pain, and local and systemic infections, resulting in considerable morbidity.

CASE REPORT

A 69-year-old woman presented to our Rheumatology department for follow-up. She was diagnosed with dermatomyositis at the age of 51, after experiencing generalized myalgias, proximal muscle weakness, dysphagia, weight loss, and erythema of the face and upper trunk. Laboratory tests revealed increased creatine kinase (952 U/L), myoglobin (595 ng/mL), aldolase (10.7 U/L) lactate dehydrogenase (863 U/L), and mildly elevated liver enzymes (aspartate transaminase 109 U/L; alanine transaminase 62 U/L). Antinuclear antibodies were positive, with specificity for anti-SSA52. Anti-Jo1 was negative. The remaining laboratory findings, including C-reactive protein, erythrocyte sedimentation rate, and thyroid function, were normal. Electromyography study of the deltoid and iliopsoas muscles revealed abnormal myopathic low-amplitude, short-duration polyphasic motor unit potential. Deltoid muscle biopsy showed normal muscle fibers with preserved interfibrillar structure, and marked perimysial and perivascular mononuclear inflammatory infiltrate, without evidence of necrotic fibers. Esophageal manometry and transit study indicated gastrointestinal involvement. Other target organ and cancer screening studies were normal. She was initially treated with prednisolone 60mg daily and azathioprine 50mg twice daily. Despite good laboratory response, her symptoms remained refractory.

Five years after the diagnosis, she developed paresthesia in the buttocks and proximal thighs, with numerous firm papules on examination. X-ray showed multiple subcutaneous calcifications around the hip, thighs, and upper limbs, consistent with calcinosis cutis. She was treated with intravenous immunoglobulin (IVIg) 400mg/kg/day for 5 days (6 cycles) and pamidronate 60mg monthly (1 year), with little benefit. Over the years, she developed multiple ulcers with frequent extrusion of chalky material that often became infected (Figure 1). Currently, she is clinically stable but contin-

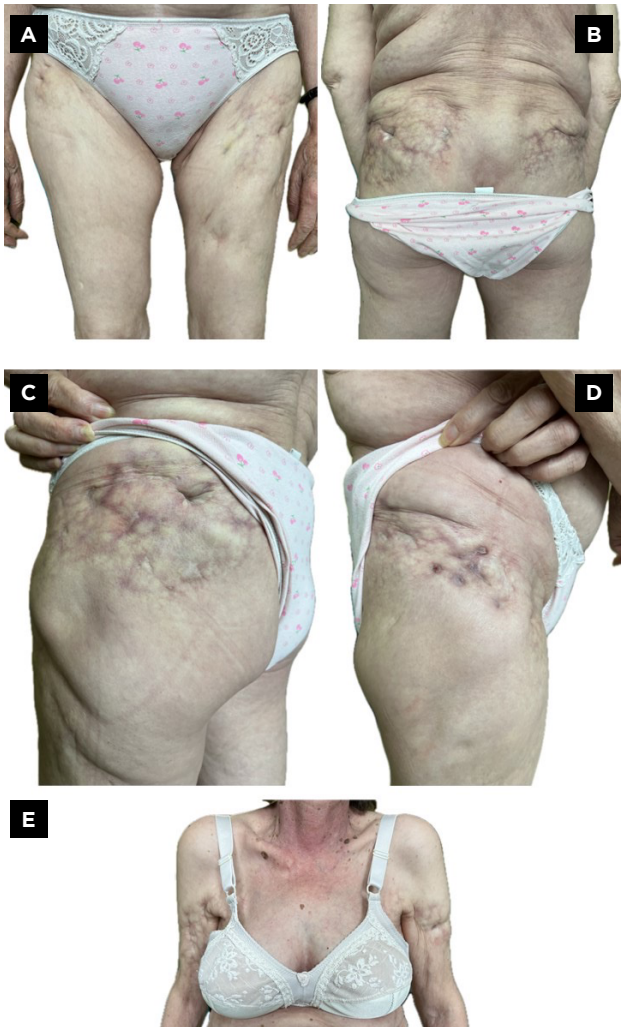

Figure 1. Ulcers on the patient's thighs and buttocks (A, B, C, D) and arms (E).

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Submitted: 22/07/2024
Accepted: 24/08/2024

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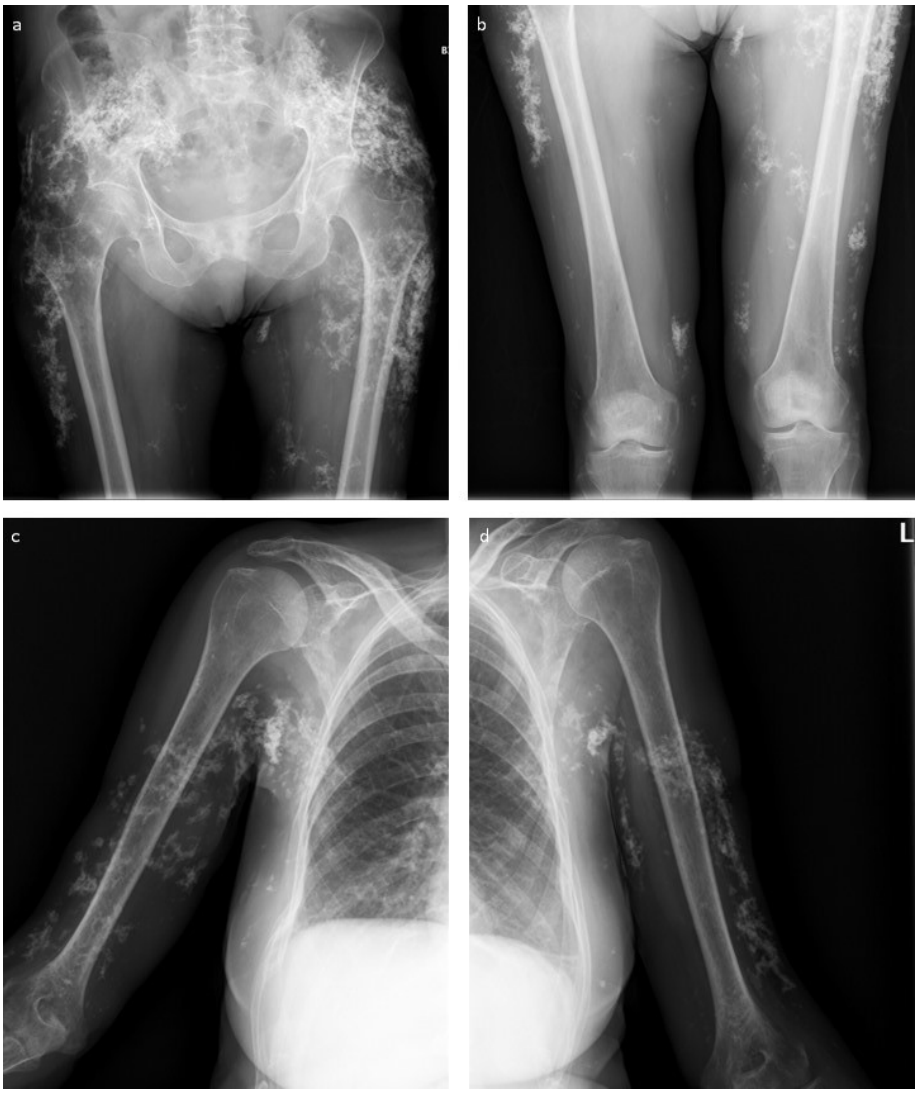

Figure 2. X-ray of the hip (A), thighs (B), right (C) and left (D) arms with multiple calcifications.

ues to experience local pain and paresthesia, and recent X-ray revealed progression of lesions (Figure 2).

DISCUSSION

Dystrophic calcinosis cutis is a serious form of skin involvement in DM, typically presenting 7.8 years after initial diagnosis[2]. It can be intracutaneous, subcutaneous, fascial, or intramuscular, commonly affecting the extremities, primarily elbows, knees, shoulders, and buttocks[3], leading to pain and limited joint motion. Diagnosis is suspected based on physical findings and imaging, namely X-ray, which also provides the depth and extent of calcification. Skin biopsy demonstrating calcium deposits in the dermis or subcutis confirms the diagnosis. Treatment is challenging and often requires a multimodal approach, including suppression of the underlying autoimmune disease. For symptomatic patients with progressive disease, interventions aim to improve lesions and associated symptoms. Therapeutic options include topical or intralesional sodium thiosulfate or surgical removal for limited presentations. In more extensive cases, systemic therapy with bisphosphonates, IVIg, diltiazem, colchicine, minocycline or rituximab may be used[4,5]. Physical therapy helps maintain joint mobility and function. Despite treatment, prognosis is variable.

This case of refractory calcinosis cutis underscores the complexity of this entity, highlighting the necessity for comprehensive and aggressive treatment. Early intervention is critical to prevent severe complications and

improve outcomes. Persistent symptoms despite various treatments, as seen in our patient, emphasize the need for ongoing research and personalized therapeutic strategies.

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