Acquired generalised lipodystrophy as an uncommon complication of anti-Mi-2 dermatomyositis: a case report

Sirs,

Acquired lipodystrophy is an uncommon extra-muscular manifestation of dermatomyositis (DM), particularly in adult patients. Here, we describe a 60-year-old woman with anti-Mi-2-positive DM who developed acquired generalised lipodystrophy (AGL). A chronological timeline of the clinical course is presented in Figure 1. Written informed consent from the patient was obtained for the publication of this case and accompanying images.

The patient had no significant past medical history and initially presented with erythematous, scaly cutaneous lesions misdiagnosed as contact eczema. Despite topical corticosteroids and emollients, the lesions progressed, and she subsequently developed Gottron's papules, heliotrope rash and the shawl sign, accompanied by upper limb myalgias without overt muscle weakness. Laboratory evaluation showed mildly elevated ESR and CRP, normal CK and aldolase, ANA 1:1280 (speckled), and positive anti-Mi-2α/β antibodies. The remainder of the myositis panel was negative. Nailfold capillaroscopy revealed megacapillaries, tortuosity and avascular zones. Hydroxychloroquine, azathioprine and oral prednisone were initiated.

One year later, the patient presented with worsening fatigue and progressive proximal weakness. Laboratory tests revealed markedly elevated CK (3980 U/L) and LDH (1166 U/L), persistent ANA 1:1280 (speckled), and positive anti-Mi-2 antibodies. She received intravenous methylprednisolone pulses, followed by oral prednisone, intravenous immunoglobulin, and azathioprine, achieving normalisation of muscle enzymes and symptomatic improvement.

Two years after the initial diagnosis, progressive, symmetrical subcutaneous fat loss in a cephalocaudal pattern became evident during outpatient follow-up and corticosteroid tapering, as shown in Figure 2. Soft-tissue ultrasound and magnetic resonance imaging (MRI) confirmed the lipoatrophy, demonstrating marked reduction in subcutaneous fat without evidence of muscle involvement. Multidisciplinary evaluation, including Dermatology, Endocrinology, and Plastic Surgery, was performed. Metabolic complications, including insulin resistance, dyslipidaemia and hepatic steatosis, were excluded through fasting glucose, HbA1c, lipid profile, liver function tests, and HOMA-IR assessment. Reconstructive procedures, including autologous fat grafting, were considered but deferred due to limited expected benefit. At last follow-up, the patient remained clinically stable, with no new fat loss or metabolic derangements. Acquired generalised lipodystrophy is rare, predominantly affects women, and is frequently associated with autoimmune diseases, particularly systemic lupus erythematosus and DM (1). Diagnosis relies on clinical evaluation, with characteristic symmetrical fat loss in a cephalocaudal distribution (2). Metabolic complications may arise (3) but were absent in this patient.

In DM, lipodystrophy has most commonly been associated with anti-TIF1γ antibodies, chronic disease course, or calcinosis. (4); few reports describe its occurrence in anti-Mi-2-positive adult patients (5). This case expands the spectrum of DM-associated lipodystrophy, suggesting it may be underrecognised. There is no specific treatment for DM-related lipodystrophy; management focuses on controlling underlying myopathy and monitoring metabolic complications. Leptin-replacement therapy (metreleptin) may be considered in severe cases (6), while reconstructive options provide variable cosmetic benefit (7).

This case highlights that AGL can develop even in adult patients with anti-Mi-2-positive DM, a subgroup typically associated with favourable prognosis. Awareness of this rare complication is essential for timely recognition, metabolic evaluation, and individualised management.

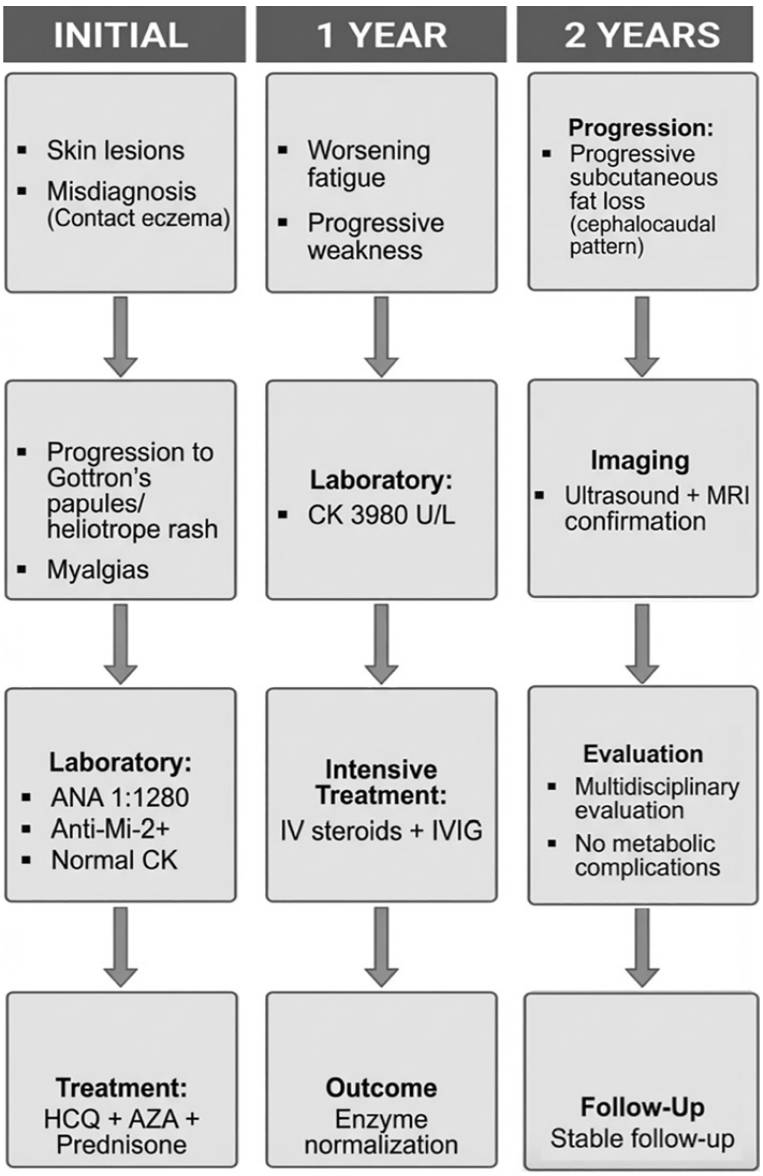

Fig. 1. Timeline illustrating the chronological sequence of clinical manifestations, laboratory findings, treatments, and onset of acquired generalised lipodystrophy in a patient with anti-Mi-2-positive dermatomyositis.

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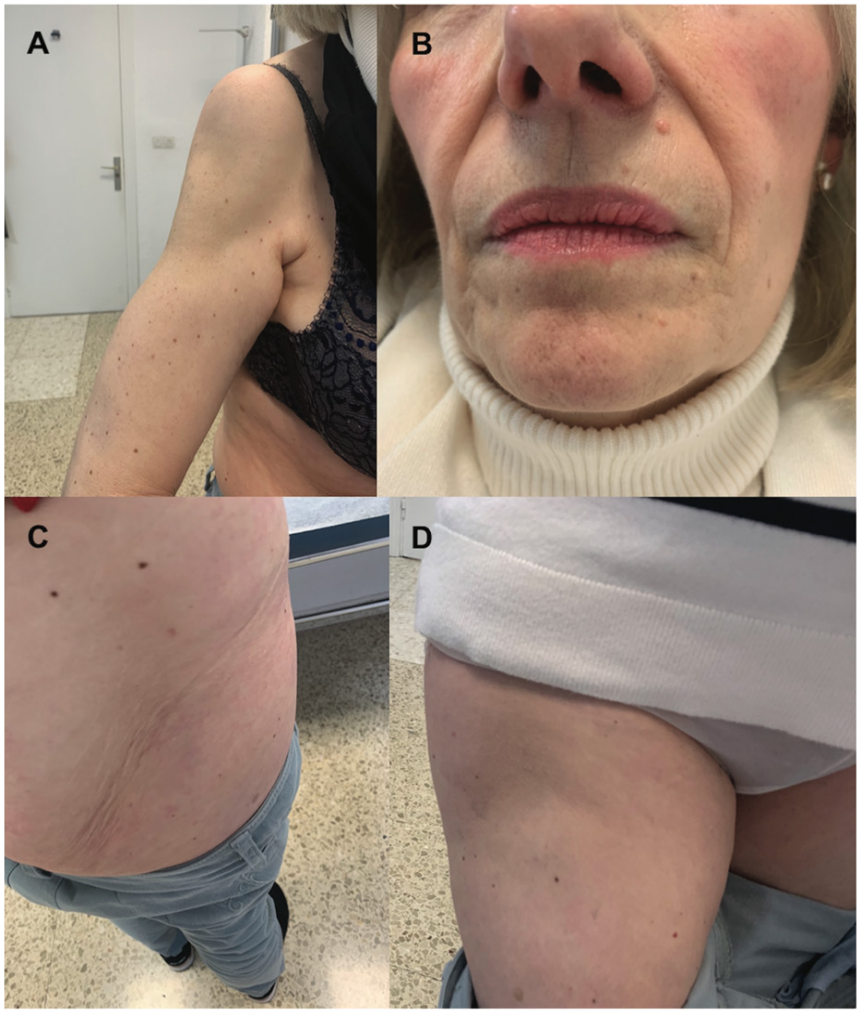

Fig. 2. Clinical manifestations of acquired generalised lipodystrophy in an anti-Mi-2-positive dermatomyositis patient. **A**: Proximal upper-arm adipose atrophy with prominence of muscular and bony contours. **B**: Facial lipoatrophy with loss of volume in the cheeks, perioral region, and jawline, accentuating nasolabial folds. **C**: Symmetrical reduction of subcutaneous adipose tissue in the femoral region. **D**: Bilateral gluteal lipoatrophy with decreased contour and loss of adipose prominence. The distribution pattern is symmetrical and cephalocaudal, typical of acquired lipodystrophy in inflammatory myopathies.

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