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SHORT COMMUNICATION

Successful Treatment of Lipedematous Alopecia using Mycophenolate Mofetil

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Lipedematous alopecia (LA) is a rare form of non-scarring alopecia described by Coskey in 1961. To date, 78 cases have been reported in the literature (1). LA generally starts from the vertex, with a centrifuge extension that may affect the whole scalp. Patients may present pain or pruritus along the affected area. Palpation reveals a smooth, diffuse and cotton-wool-like induration (1, 2). Currently there is no effective treatment to halt or delay progression. The use of topical and intralesional steroids has, in general, had a negative response (3). High & Hoang reported no response to hydroxychloroquine in a female patient with LA and discoid lupus erythematous (DLE) (4). Yip et al. (5) reported surgical treatment with a successful debulking of the scalp after achieving inactivity of the disease, with no recurrence after a 1-year follow-up. We describe here a patient with LA who responded successfully to oral mycophenolate mofetil (MMF), an immunosuppressive drug not previously used to treat this condition.

CASE REPORT

A 51-year-old man with no history of comorbidities or medication intake presented with diffuse and progressive hair loss, increased scalp volume, and pruritus in the last 5 months. Physical examination revealed non-cicatricial alopecia with erythema on the vertex and bilaterally on the parietal zone (Fig. 1A). Palpation revealed a cotton wool-like induration on the affected area. Dermoscopy showed multiple linear telangiectasias with conservation of the follicular openings. Ultrasound revealed thickening of the hypodermis in the affected areas, with marked effacement of the dermal-hypodermic interphase.

Laboratory tests showed a normal complete blood count (CBC), antinuclear antibody (ANA) and extractable nuclear antigen (ENA). Thyroid, kidney, and hepatic function tests were also normal. Biopsy highlighted a superficial perivascular lymphocytic infiltrate with marked thickening of the subcutaneous cellular tissue. There was also loss of normal lobular architecture at the hypodermis due to interstitial oedema. Ectatic lymphatic vessels were seen in the deep dermis and hypodermis (Fig. 2). Direct immunofluorescence was negative.

Treatment began with intralesional steroid infiltration every 14 days, totalling 8 sessions in 4 months. Follow-up showed refractoriness and even worsening of the condition, characterized by centrifugal progression of alopecia and increased signs of inflammation. After new clinical and laboratory tests that demonstrated normal function of other organs, treatment with 1 g/day of MMF was initiated, obtaining the first satisfactory results after the third month (Fig. 1B). MMF was well tolerated by the patient, with no side-effects. The same dose was therefore maintained for 10 months.

After this period, a clinical hair examination evidenced a marked increase in growth and hair density, with a clear decrease in telangiectatic vessels under dermatoscopy. Ultrasound showed a decrease in hypodermic thickening with less effacement of the dermohypodermic interphase. Treatment was discontinued after 10 months when complete recovery of the hair was achieved. After 6 months of follow-up the patient still has total hair recovery with no inflammatory signs on the scalp (Fig. 1C).

DISCUSSION

MMF is an immunosuppressant drug that is increasingly used in the management of various inflammatory dermatoses. MMF affects activated T and B lymphocytes (6).

Although the main microscopic feature of LA is thickening of the subcutaneous layer, other features include perivascular and periadnexal lymphocytic infiltrate associated with hyperkeratosis, follicular plugging and follicular fibrosis. All these factors have predisposed some authors to speculate that some cases of LA could represent an unusual late sequela of DLE (4). It has also been demonstrated that MMF inhibits certain fibroblast functions involved in tissue fibrosis and it has been

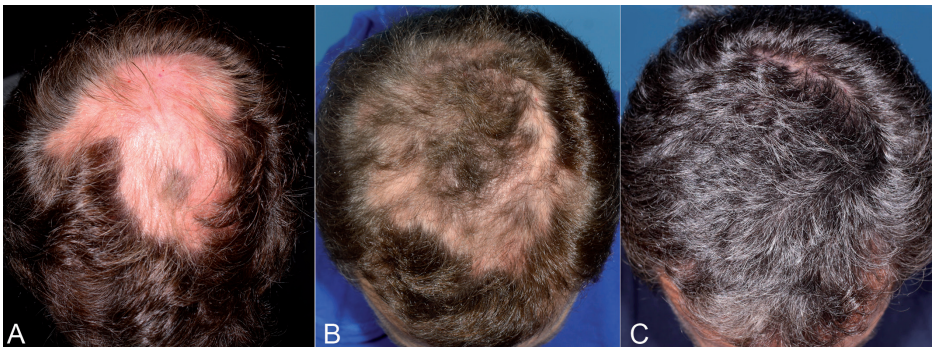

Fig. 1. Clinical features and treatment course. (A) Before treatment. (B) After 3 months of treatment with mycophenolate mofetil. (C) Complete recovery after 10 months.

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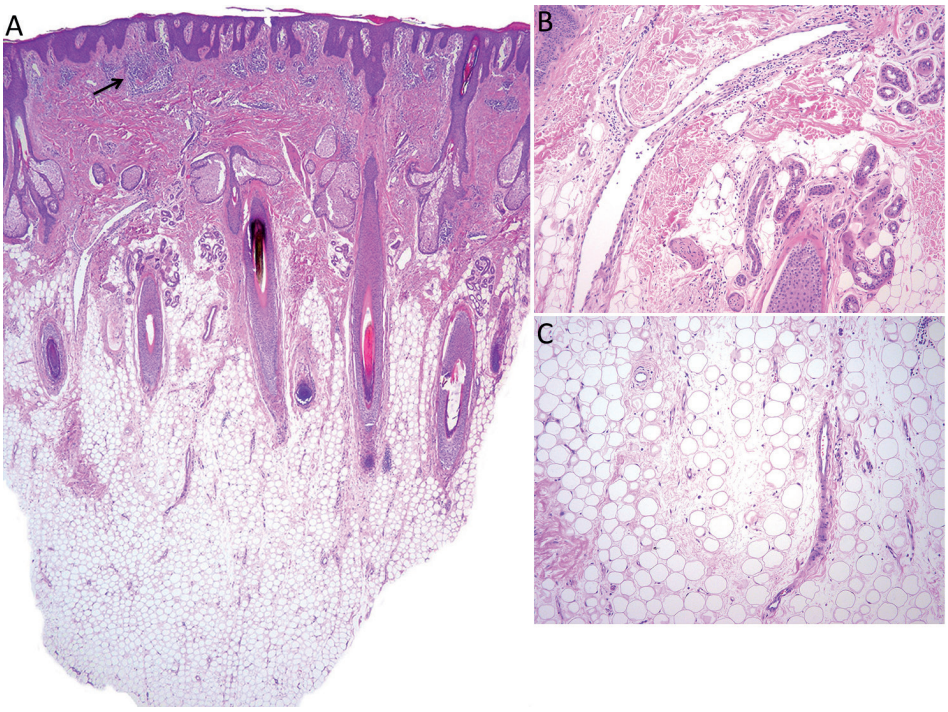

Fig. 2. Histopathology with haematoxylin-eosin staining. (A) Superficial perivascular lymphocytic infiltrate is visible in the dermis (arrow) with thickening of the subcutaneous cellular tissue (20 × magnification). (B) Dilated lymphatic vessels are seen in the deep dermis (100 × magnification). (C) Mature adipocytes with loss of normal architecture due to interstitial oedema (100 × magnification).

used in the treatment of pathologies such as morphea and lichen planopilaris (7–9). The slight perifollicular fibrosis or fibrous tracts adjacent to some hair follicles in LA may also be inhibited by the action of MMF (10).

No comprehensive explanation of the pathophysiology of alopecia in LA exists in the literature (1–4). But if we consider that MMF is a broad-spectrum immune suppressor that works on antigen-presenting cells, adhesion molecules and inhibits the migration of lymphocytic cells producing cytokines related to inflammation and fibrosis (11) – and has been used in the treatment of lichen planopilaris and hair DLE (9, 12) – we can assume that its use in LA has clear anti-inflammatory effects, as demonstrated in the case described here. However, further clinical research is required into the efficacy of MMF in the treatment of LA.

The authors declare no conflicts of interest.

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