Lupus miliaris disseminatus faciei*

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Abstract: Lupus miliaris disseminatus faciei is a rare inflammatory dermatosis of unknown etiology that primarily affects young adults. Clinically, it is characterized by an asymptomatic papular eruption mainly involving the central face, typically on and around the eyelids. Characteristic histopathological features include dermal epithelioid cell granulomas with central necrosis and surrounding lymphocytic infiltrate with multinucleate giant cells. Lupus miliaris disseminatus faciei has a spontaneously resolving course, yet can be cosmetically debilitating given the location and potential for scarring. Treatment is difficult and there is a lack of controlled studies. We report a new case of lupus miliaris disseminatus faciei successfully treated with minocycline and systemic steroids, and briefly discuss its nosology and therapeutic options.
Keywords: Minocycline; Granuloma; Prednisolone

INTRODUCTION

Lupus miliaris disseminatus faciei (LMDF), first described by Fox in 1878, is a rare granulomatous inflammatory dermatosis that mostly affects young adults. [1,2] It is characterized clinically by a bilaterally symmetrical papular eruption located on the central area of the face, and histopathologically by epithelioid cell granulomas with caseous necrosis.[3] Despite the characteristic clinical-pathological features, its etiopathogenesis remains unknown and the treatment is often unsatisfactory. [2]

CASE REPORT

A previously healthy 43-year-old woman was admitted to our department with an asymptomatic micropapular eruption on the face that had evolved over a period of five months. Physical examination revealed multiple, small (1 to 3 mm), dome-shaped, reddish-yellow and yellowish-brown papules, distributed symmetrically on the central area of the face, namely the forehead, eyelids, nose, cheeks, perioral area, and chin (Figure 1). There was no accompanying scaling, telangiectasia, or flushing. No other body areas were affected. The patient had previously been treated with oral isotretinoin, 40 mg/day for four months, with no improvement of the lesions. She denied taking oral steroids or applying topical steroids on the face at any time in the past.

Histopathological examination of a skin biopsy taken from a representative lesion on the chin revealed dermal epithelioid cell granulomas, some with central areas of necrosis, and surrounding moderate lymphohistiocytic infiltrate with multinucleate giant cells, mostly of the Langhans type (Figure 2). No foreign bodies were found in the granulomas, and no mycobacterial or fungal components were detected in dermal tissues by Ziehl-Neelsen staining or periodic acid-Schiff (PAS) staining. The DNA of *Mycobacterium tuberculosis* was not detectable in the active lesions when submitted to polymerase chain reaction (PCR). Chest X-ray and routine laboratory studies (including serum levels of calcium and angiotensin-converting enzyme) were within normal limits; a tuberculin skin test proved to be negative.

On the basis of these findings, the diagnosis of LMDF was made, and the patient was given oral minocycline 100 mg/day, together with oral prednisolone 5 mg/day. Flattening of the papules was observed within 3 weeks of therapy. By 16 weeks, a moderate improvement had been achieved, despite the residual depressed scars, and minocycline was tapered to 100 mg every other day and prednisolone to 5 mg every other day, which she maintained for an additional eight weeks (Figure 3). No recurrence was noted over a 12-month observation period.

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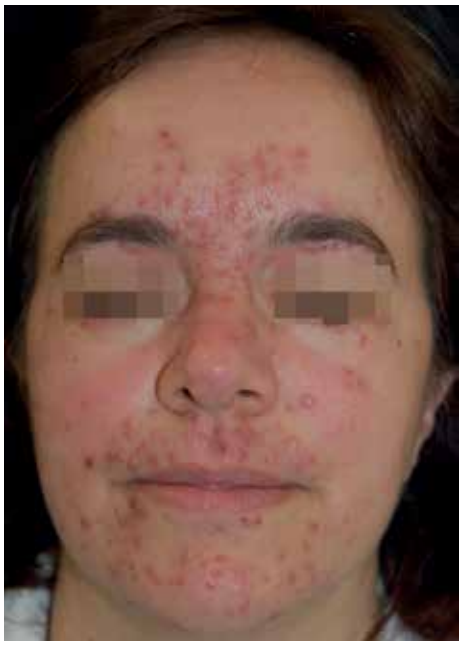

Figure 1: Multiple, small, reddish-brown papules scattered over the forehead, eyelids, nose, cheeks, perioral area, and chin

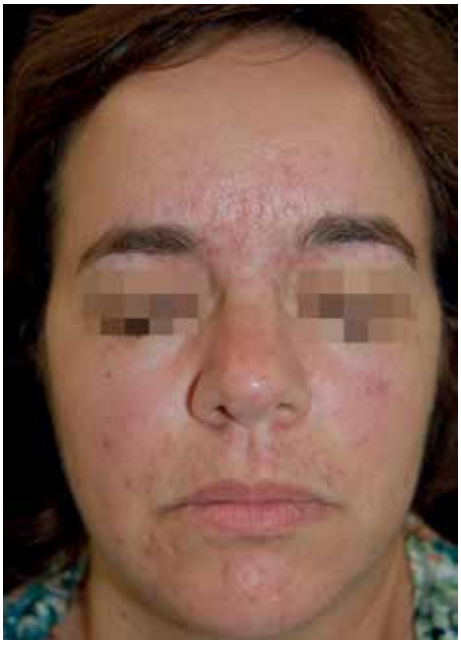

Figure 3: Significant improvement of the lesions after 16 weeks of combined treatment with minocycline and prednisolone

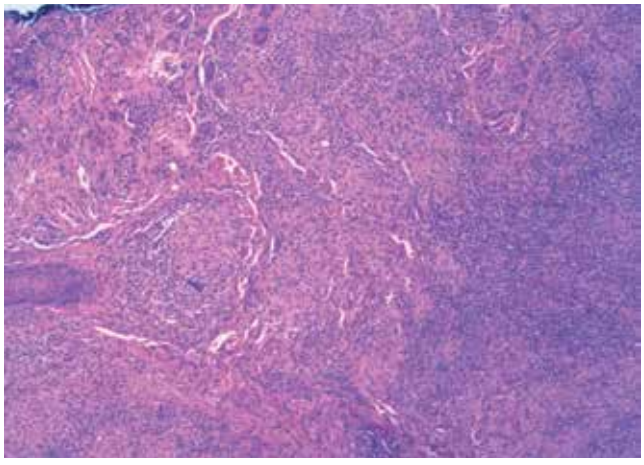

Figure 2: Histopathological examination of a skin biopsy demonstrating epithelioid cell granulomas in the dermis, some with central necrosis, surrounded by lymphohistiocytic infiltrate with multinucleate giant cells (Hematoxylin & eosin, X100)

DISCUSSION

LMDF is an uncommon dermatosis, with about 200 cases reported to date.[2] This is most commonly observed in adults between the second and the fourth decades of life, although cases have been reported among children and the elderly. [2]

Clinically, this dermatosis appears as small, discrete, reddish-yellow or yellowish-brown asymptomatic papules involving primarily the central face, typically on and around the eyelids, although there are some reports of extrafacial involvement.[2,4] The papules may occur singly or in crops, can be follicular or nonfollicular and, in many cases, show a pustular top.[3] The eruption develops rapidly, running a chronic course, and usually involutes spontaneously within 12 to 24 months, often leaving small pitted scars.[3]

The histopathological hallmark of LMDF is a dermal epithelioid cell granuloma with central necrosis, but the histological pattern can vary according to the stage of the lesion.[3] Early lesions may show a perivascular and periadnexal lymphohistiocytic infiltrate. In the fully developed stage, the following spectrum of changes can be seen: epithelioid cell granuloma with and without central necrosis, epithelioid cell granuloma with an abscess, and nongranulomatous nonspecific inflammatory infiltrate. Late lesions may show extensive perifollicular fibrosis with nonspecific cell infiltrate. [2]

The exact etiopathogenesis of LMDF remains unknown.[2,4] Originally it was thought to be a tuberculid, but studies have failed to demonstrate *Mycobacterium tuberculosis* in LMDF lesions, and this theory is no longer accepted.[3,4] In the 1980s, many authors considered LMDF to be a variant of granulomatous rosacea, but there are many aspects that differentiate the two diseases, such as the self-limited course with scarring, equal gender distribution, caseation necrosis in the histology, as well as an absence of erythema, flushing, and telangiectasia. [2,3] Other authors proposed *Demodex folliculorum* as the causative organism, but this association has not been confirmed [3]. Based on the frequent association with the hair follicle, LMDF has been proposed to represent an immune response to the pilosebaceous units, triggered by hair follicle destruction or ruptured epidermal cysts.[2-4] *Propionibacterium acne* signatures have recently been detected in LMDF granulomas, suggesting a pathogenic role for these bacteria, generally present as a commensal agent in the hair follicles. [5] Currently, most authors believe LMDF is a distinct and independent entity. A name change to facial idiopathic granulomas with regressive evolution (FIGURE) was proposed in 2000, but this nomenclature does not appear to have been widely accepted to date.[4]

Other granulomatous disorders that should be differentiated from LMDF include Facial Afro-Caribbean Childhood Eruption (FACE) syndrome, sarcoidosis, non-tuberculous mycobacterium infection and deep fungal infection. [2,6] Histopathologically speaking, LMDF can be differentiated from these diseases by the presence of caseous necrosis. Additionally, sarcoidosis can be distinguished by physical examination, chest X-ray, and laboratory tests, and infectious disorders by the absence of microorganisms detected through histochemical stains (PAS, Ziehl-Neelsen). [6]

Treatment of LMDF is usually unsatisfactory, and there is lack of controlled studies in the literature.[7] Because LMDF spontaneously resolves within 1-2 years, the impact of therapy on the course of the disease is difficult to assess.[3] Tetracyclines (doxycycline and minocycline) are a usual first-line treatment, but they are not consistently effective.[3,4] Many other systemic treatments have been reported to be effective in some patients, including isotretinoin, dapsone, corticosteroids, clofazimine, tranilast, anti-tuberculous drugs (alone or in association with tetracyclines),[8] and metronidazole.[2,3,6-8] Additionally, topical tacrolimus (in association with oral dapsone or metronidazole) and laser therapy using a 1450 nm diode laser and a 1565 nm non-ablative fractionated laser resurfacing have improved LMDF.[4,9,10] Prevention of scarring may be possible with early intervention, using low dose corticosteroids.[3] The successful management of LMDF scars has been reported using a combination of 100% trichloroacetic acid and carbon dioxide lasers.[8] ❑

REFERENCES

1. Fox T. Disseminated follicular lupus (simulating acne). Lancet. 1878;112:75-6.
2. Rocas D, Kanitakis J. Lupus miliaris disseminatus faciei: Report of a new case and brief literature review. Dermatol Online J. 2013;19:4.
3. Esteves T, Faria A, Alves R, Marote J, Viana I, Vale E. Lupus miliaris disseminatus faciei: A case report. Dermatol Online J. 2010;16:10.
4. Al-Mutairi N. Nosology and therapeutic options for lupus miliaris disseminatus faciei J Dermatol. 2011;38:864-73.
5. Nishimoto J, Amano M, Setoyama M. The detection of Propionibacterium acnes signatures in granulomas of lupus miliaris disseminatus faciei. J Dermatol. 2015;42:418-21.
6. Koike Y, Hatamochi A, Koyano S, Namikawa H, Hamasaki Y, Yamazaki S. Lupus miliaris disseminatus faciei successfully treated with tranilast: Report of two cases. J Dermatol. 2011;38:588-92.
7. Nomura H, Egami S, Kasai H, Yokoyama T, Fujimoto A, Sugiura M. A patient with lupus miliaris disseminatus faciei treated successfully with a combination of oral metronidazole and topical tacrolimus. J Dermatol. 2014;41:645-6.
8. Kang BK, Shin MK. Scarring of lupus miliaris disseminatus faciei : treatment with a combination of trichloroacetic acid. Dermatol Ther. 2014;27:168-70.
9. Jih MH, Friedman PM, Kimyai-Asadi A, Friedman ES, Hymes SR, Goldberg LH. Lupus miliaris disseminatus faciei. Treatment with the 1450-nm diode laser. Arch Dermatol. 2005;141:143-5.
10. Beleznay K, Friedmann DP, Liolios AM, Perry A, Goldman MP. Lupus miliaris disseminatus faciei treated with 1,565 nm nonablative fractionated laser resurfacing: A case report. Lasers Surg Med. 2014;46:663-5.

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