Lupus miliaris disseminatus faciei with complete response to isotretinoin

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Abstract

Lupus miliaris disseminatus faciei is an uncommon type of granulomatous rosacea characterized by a papular eruption in the central regions of the face. A 43-year-old woman presented with an asymptomatic papular eruption on the face that had developed over a period of five months. Physical examination revealed multiple, small, reddish-brown papules, distributed symmetrically on the central area of the face. A biopsy was taken, showing dermal epithelioid cell granulomas with central necrosis and surrounding lymphocytic infiltrate with multinucleate giant cells. No foreign bodies were found in granulomas and no mycobacterial or fungal components were detected. On the basis of these findings, the diagnosis of lupus miliaris disseminatus faciei was made. The patient was given oral isotretinoin 20mg/day with initial slow response. After 6 months' treatment the lesions completely disappeared. Many authors consider this entity to be a variant of granulomatous rosacea. It is a chronic condition that primarily affects young adults. Treatment is usually unsatisfactory. Therapies with corticosterois, tetracyclines, retinoids, clofazimine or topical tacrolimus have been described but there is a lack of controlled studies and convincing results. Our success with a 6-month course of low dose isotretinoin suggests consideration of a longer trial prior to abandoning this as treatment.

Keywords: lupus miliaris disseminates faciei, granulomatous rosacea, isotretinoin, granuloma

Introduction

Lupus miliaris disseminatus faciei (LMDF) is an uncommon dermatosis of unknown etiology, characterized by the presence of reddish-brown, dome-shaped papules involving the central face. It is a chronic inflammatory disorder which affects young adults of both sexes. Histopathological features include dermal epithelioid cell granulomas with central necrosis. Treatment is frequently unsatisfactory. Several treatments have been reported, however no treatment has consistently been able to prevent the desfiguring scars. We report a new case of LMDF successfully treated with isotretinoin without residual scaring.

Case Synopsis

A 43-year-old woman presented to the dermatology clinic with an asymptomatic papular eruption on the face that had developed over a period of five months. Physical examination revealed multiple small reddish-brown, papules distributed symmetrically on the central face, namely the eyelids and perioral area (**Figure 1**). There was accompanying scaling. A complete blood count with renal and hepatic panel were normal. A biopsy was taken, showing dermal epithelioid cell granulomas with central necrosis and surrounding lymphocytic infiltrate with multinucleate giant cells. No foreign bodies were found in the granulomas. No mycobacterial or fungal components were detected in dermal tissues by Ziehl-Neelsen or periodic acid-Schiff staining (**Figure 2**). On the basis of these findings, the diagnosis of LMDF was made. The patient was given oral isotretinoin 20 mg/day with initial slow response. By three months, a moderate improvement had been achieved with flattening of the papules. After 6 months of treatment with

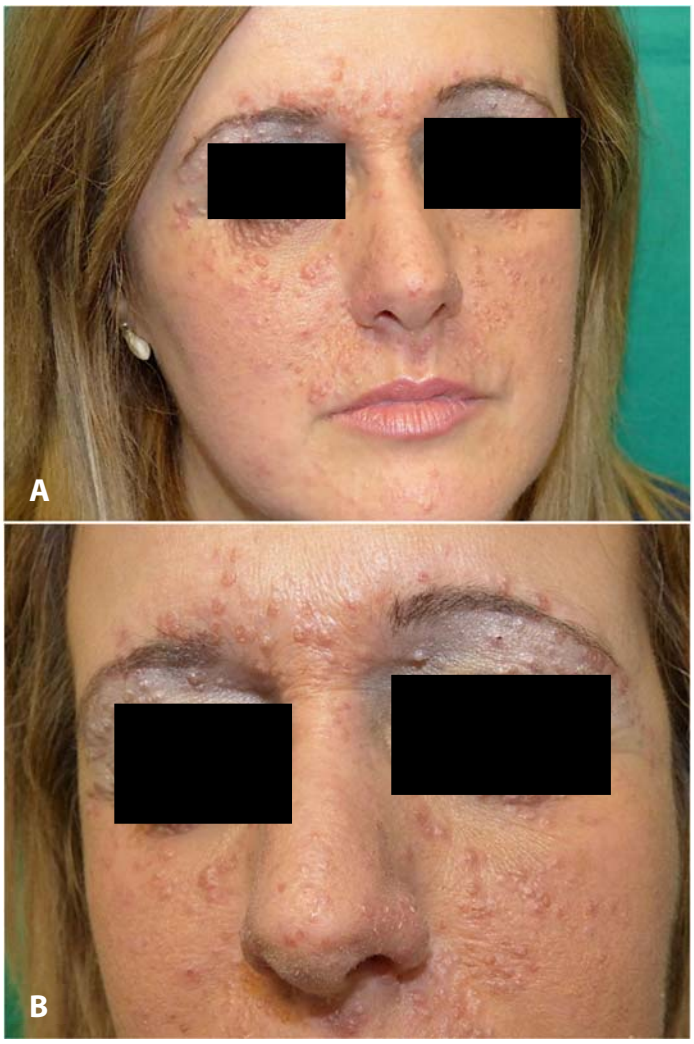

Figure 1. ***A)*** *First visit. The patient presented with multiple small reddish-brown, papules distributed symmetrically on the central face.* ***B)*** *On closer view, multiple papules around the eyelids can be appreciated.*

isotretinoin the lesions had resolved, leaving no disfiguring scars on the face (**Figure 3**). The patient maintained isotretinoin treatment 6 more months with a lower dosis (5mg/day) and treatment was then discontinued. At present the patient has no active treatment and has maintained a complete response after 12 months of medical monitoring.

Case Discussion

Lupus miliaris disseminatus faciei is a rare inflammatory dermatosis first described by Fox in 1878 [1]. The etiology is unknown. At first, it was associated with *Mycobacterium tuberculosis* infection owing to the presence of central caseating necrosis on histopathologic studies, but the use of polymerase chain reaction and culture have shown no evidence of this bacterium [2]. Similarly, LMDF

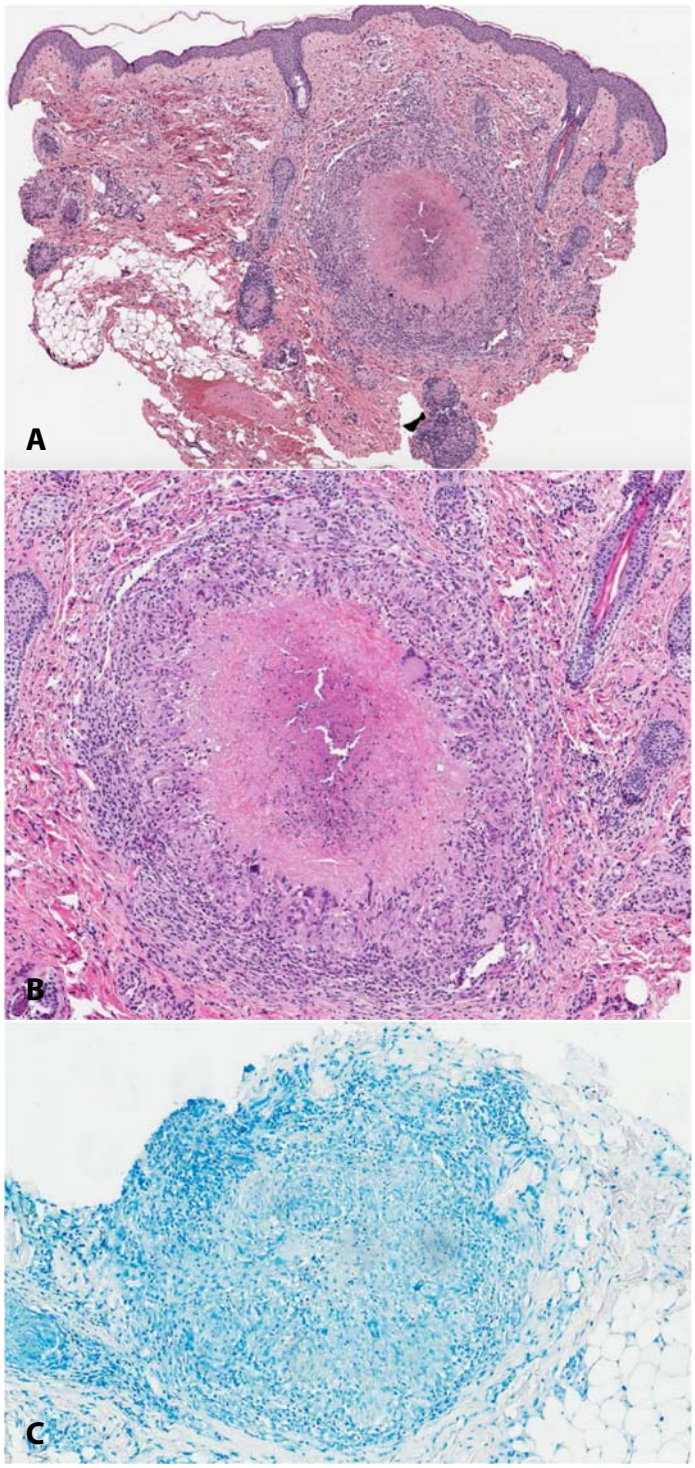

Figure 2. ***A)*** *Panoramic image of dermal granuloma with central necrosis. H&E, 10×.* ***B)*** *Epithelioid cell granuloma with central necrosis and surrounding lymphocytic infiltrate on higher magnification. H&E, 100×.* ***C)*** *No mycobacterial or fungal components were detected in dermal tissues by Ziehl-Neelsen technique, 100×.*

Figure 3. ***A)*** *Front view. After 6 months of treatment with isotretinoin, examination revealed a complete resolution of lupus miliaris disseminatus faciei. The patient presented neither pitted scars nor residual pigmentation.* ***B)*** *Lateral view.*

was also believed to be a form of sarcoidosis. However, LMDF is currently considered as a type of granulomatous rosacea, or in some cases a granulomatous periorificial dermatitis. The incidence of this disease is unknown but is considered to be uncommon [3]. It occurs most commonly in young adults, between the second and the fourth decades of life [3]. It affects both genders, although most cases have been reported in men [2]. Rare cases have been described in children [4] and the elderly [5,6].

Clinically, it is characterized by an asymptomatic yellow-brown or brown-redish papular eruption mainly involving the central face, especially the eyelids [2,7]. Extrafacial m anifestations have been described in other localizations such as axillae [8] or hands [9]. Lupus miliaris disseminatus faciei usually involutes spontaneously within 12 to 24 months [2,10] often leaving numerous pitted scars and pigmentation. Despite the usual spontaneous resolution, no treatment has been able to prevent the disfiguring scars.

Histopathological studies usually show dermal epithelioid cell granulomas with central necrosis and surrounding lymphocytic infiltrate with multinucleate giant cells [10]. Foreign bodies, mycobacteria, and fungi should be eliminated by Ziehl-Neelsen and periodic acid-Schiff staining. Histopathological findings in granulomatous rosacea and LMDF frequently show overlap, but t he LMDF subset particularly exhibits granulomas with central necrosis.

Many therapies have been reported in the literature but there is no established treatment for this entity. Unfortunately, despite treatment or spontaneous involution, residual scars are a frequent outcome. Reported treatments include oral glucocorticoids [11], dapsone [12], tetracyclines (doxycycline, minocycline) [13], isotretinoin [14,15], and clofazimine [16]. Successful treatment of LMDF with combined oral metronidazole or oral dapsone combined with topical tacrolimus also have yielded good results [17]. More recently, laser therapy also may have a role in the treatment of LMDF with a 1,450nm diode laser [18] and a non-ablative fractionated 1,565nm laser [19].

Conclusion

Lupus miliaris disseminatus faciei is considered as an uncommon variant of granulomatous rosacea. Some cases might also be an example of granulomatous periorificial dermatitis. It is necessary to rule out other granulomatous diseases. Treatment of LMDF is usually unsatisfactory. We report a new case of LMDF in an adult woman successfully treated with low dose oral isotretinoin. Slow improvement was noted at first, but eventually she achieved and maintained a complete response without recurrences after 12 months of monitoring. Pitted or disfiguring scars did not result. Oral isotretinoin should be considered a good option for treatment of LMDF and should be initiated as soon as posible in hopes of preventing scarring.

Potential conflicts of interest

The authors declare no conflicts of interests.

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