

CASE REPORT

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Lupus Miliaris Disseminatus Faciei: A Case Report

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Abstract

Lupus miliaris disseminatus faciei (LMDF) is an inflammatory disease with an unknown etiology. It is characterized by the presence of several dome shaped monomorphic papules in the centro-lateral facial areas. Diascopy of relatively larger lesions often reveals an apple jelly like nodular appearance. Based on the typical clinical manifestations (reddish yellow papules localized in the central and lateral areas of the face, especially the eyelids) and histopathological findings (giant epithelioid granuloma with varying caseating necrosis) a diagnosis of LMDF can be established. Herein, we would like to present a typical case of LMDF in a 43-year-old Chinese female with a brief insight on the clinical features. The patient had used before antibiotic creams without obvious improvement. Based on the clinical and histo-pathological findings, a diagnosis of LMDF was made and use of medicines was suggested.

Keywords Acnitis, acne agminata, facial idiopathic granulomas, lupus miliaris.

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Introduction

Lupus miliaris disseminatus faciei (LMDF) is an uncommon granulomatous, inflammatory disease of unknown etiology. It is also known as acne agminata, or Facial Idiopathic Granulomas with Regressive Evolution (FIGURE) [1]. It clinically manifests as reddish yellow papules localized to the central and lateral areas of the face, especially involving the eyelids. It usually manifests in adults (especially males) and can also be seen in elderly and pediatric age groups [1, 2]. The etio-pathogenesis of LMDF is currently unknown [3]. To date, about 200 cases have been reported worldwide [4] and we would like to present a typical case of LMDF in a 43-year old Chinese female patient.

Cash report

A 43-year-old Chinese female presented at our outpatient department with an asymptomatic facial rash for about a year. Physical examination revealed multiple monomorphic reddish yellow papules distributed prominently on the central and lateral areas of the face (forehead, eyebrows, eyelids, naso-labial gap, and perioral areas), most obviously on the lower eyelids (Fig. 1A) while the trunk and extremities remained unaffected. In the past three months, the patient had occasionally used some topical antibiotic creams without any obvious

improvement. The patient was otherwise healthy and denied any significant past medical history. The patient also denied the presence of a similar skin rash or that of Tuberculosis in her family. Chest X-ray and other routine laboratory tests were within normal limits. The result for T-Spot-TB test was also negative. For further evaluation a skin biopsy of the facial papule was performed (Fig. 1B). Histo-pathological analysis of the biopsy specimen revealed a large clear-boundary epithelioid cell granuloma with central necrosis surrounded by a lymphocytic infiltrate (Fig. 2A and 2B). Based on the clinical and histo-pathological findings, a diagnosis of lupus miliaris disseminatus faciei (LMDF) was made. The patient was counseled and educated regarding the course of the disease and the possible outcomes. The patient was started on doxycycline 100 mg bid and topical tacrolimus ointment 0.1% bid for several months. However, the patient did not visit for further follow up.

Discussion

Lupus miliaris disseminatus faciei (LMDF) usually manifests as multiple reddish brown or yellowish brown dome shaped translucent papules that symmetrically involve the central and lateral areas of the face, especially the areas around the eyelids [4]. Diascopy of relatively larger lesions often reveals an apple jelly nodule like appearance. LMDF may also involve other sites such as neck,

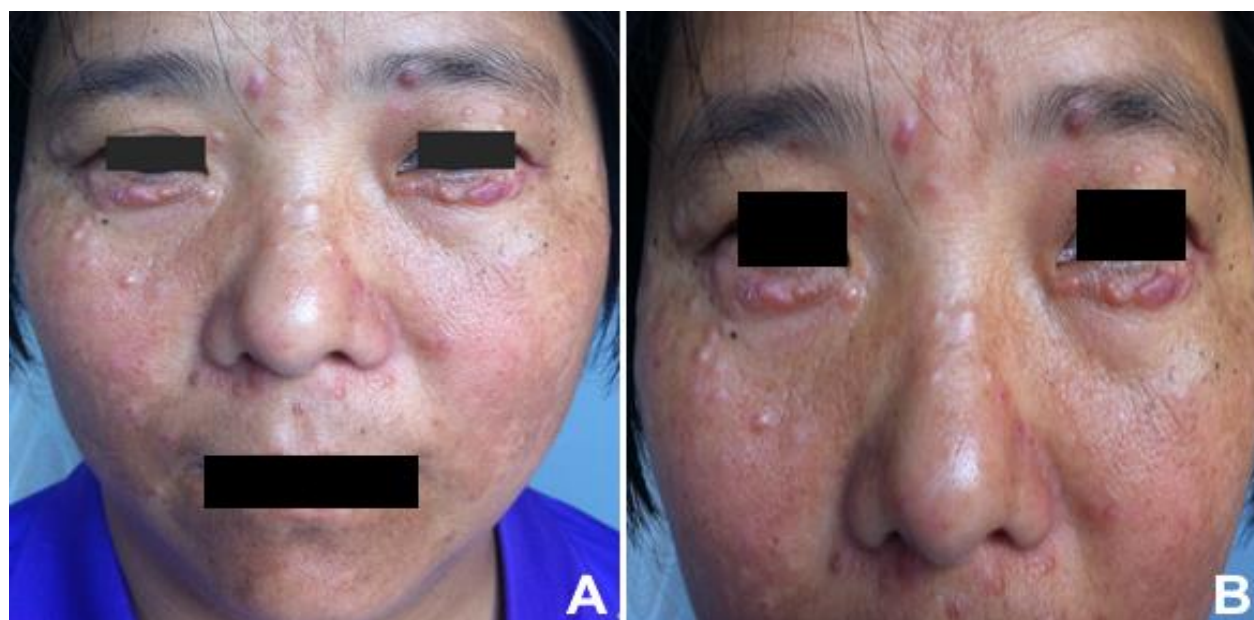


Fig. 1 Multiple reddish yellow papules over the eyelids, nose, cheeks and peri-oral area (A) and symmetrically distributed multiple reddish brown translucent papules over the central and lateral areas of the face, especially the areas around the eyelids (B).

chest, scalp, axillae, trunk and genitalia making the diagnosis a little bit trickier [5].

As far as the etio-pathogenesis is concerned, there are various theories. Earlier LMDF was considered to be a tuberculid reaction due to the presence of epithelioid cell granuloma with central necrosis. However, the bacterial cultures from LMDF lesions were negative (didn't reveal any Bacilli) and polymerase chain reaction didn't detect any *Mycobacterium tuberculosis* DNA. Hence, this hypothesis is no more accepted [2]. Some considered LMDF to be a variant of granulomatous rosacea due to the localization of the lesions in the central areas of the face and some of the histo-pathological findings that may resemble the ones seen in granulomatous rosacea. However, in comparison, the LMDF lesions don't contain demodex mites nor do the patients complain of flushing and moreover, the lesions run a chronic course and are usually resistant to treatment. Hence, this hypothesis is no more accepted [1, 2].

Typical histo-pathological findings of LMDF include the presence of large epithelioid cell granuloma with caseating necrosis surrounded by a lymphocytic infiltrate. However, these findings are not always consistent and may vary with the age of lesions. Early stage lesions are characterized by a perivascular and periadnexal lymphocytic infiltrate. Shitara had classified the histo-pathological findings of a fully developed lesion into four groups:

epithelioid cell granuloma with central necrosis, epithelioid granuloma without central necrosis (sarcoidal-type granuloma), epithelioid cell granuloma with abscess and non-granulomatous non-specific inflammatory infiltrate. Late stage lesions show extensive peri-follicular fibrosis with non-specific cell infiltrates [2, 3, 6].

Over the years, anti-tuberculous drugs, tetracycline, isotretinoin, dapsone, corticosteroids, tranilast, metronidazole, topical tacrolimus have been used either as a solo therapy or in combination with variable efficacy [2, 5, 7]. Since the skin lesions of LMDF usually resolve with scarring, the use of various lasers has been reported. Kang et al. [1] reported the use of 100% trichloroacetic acid and carbon dioxide laser with a satisfying outcome in treating the scars of LMDF. Beleznyay et al. [8] reported the use of 1,565 nm non-ablative fractional laser resurfacing in the treatment of LMDF with a gradual improvement in the overall appearance of the lesions.

In the end, we would like to conclude that LMDF is a rare chronic granulomatous disease of unknown etiology. Based on the typical clinical manifestations (reddish yellow papules localized in the central and lateral areas of the face, especially the eyelids) and histopathological findings (giant epithelioid granuloma with varying caseating necrosis) a diagnosis of LMDF can be made. The differential diagnosis includes sarcoidosis, granulomatous rosacea, syringiomas and FACE

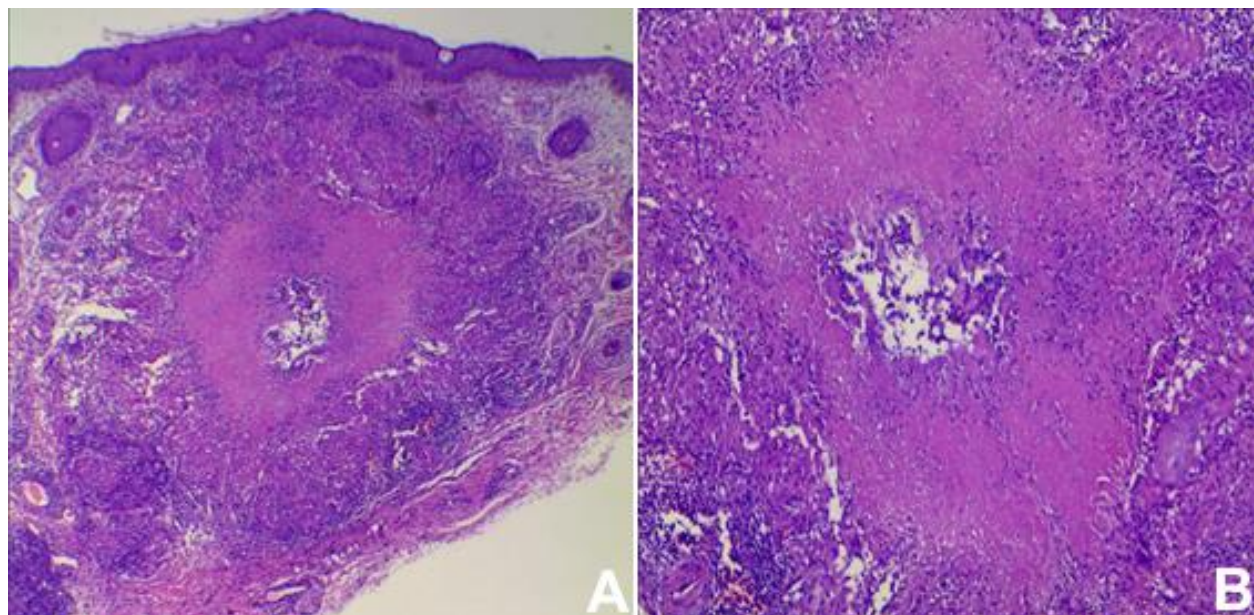


Fig. 2 Histo-pathological analysis of a lesion revealed a large clear-boundary epithelioid cell granuloma with central necrosis (HE stain x 40) (A) and higher magnification shows the centralized necrosis and surrounding epithelioid cellular infiltrate (HE stain x 100) (B).

(Facial afro-Caribbean childhood eruption) syndrome. As far as treatment is concerned, further studies are required to find a suitable therapeutic regimen (combination of drug and laser therapy) with a relatively satisfying outcome.

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