THE MEDICAL BULLETIN OF
SISLI ETFAL HOSPITAL

DOI: 10.14744/SEMB.2017.39306
Med Bull Sisli Etfal Hosp 2018;52(2):142–144

Case Report

Lupus Miliaris Disseminatus Faciei: A Case Report and Brief Literature Review

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Abstract

Lupus miliaris disseminatus faciei (LMDF) is a rarely seen, granulomatous disease of the face with an unknown etiology. Clinically, the disease is characterized by monomorphic, reddish-brown, dome-shaped papules symmetrically distributed on the face. Histopathologically, a perifollicular caseating granuloma is the hallmark.

A 20-year-old male patient was referred to our outpatient clinic with multiple papules distributed on his face, and he was diagnosed with LMDF based on histopathological examination. The patient was unresponsive to the oral tetracycline treatment; however, he was successfully treated with systemic dapsone and topical tacrolimus. This is a rare case of LMDF, which showed rapid improvement with dapsone therapy.

Keywords: Dapsone; granuloma; lupus miliaris disseminatus faciei.

Please cite this article as "Aksu Çerman A., Aktaş Karabay E., Yalçın Ö., Kıvanç Altunay İ. Lupus Miliaris Disseminatus Faciei: A Case Report and Brief Literature Review. Med Bull Sisli Etfal Hosp 2018;52(2):142–144".

Lupus miliaris disseminatus faciei (LMDF), first described by Fox et al.[1] in 1877, is a rarely seen dermatosis, and approximately 200 cases have been reported in the literature so far.[2] Previously mycobacterium tuberculosis and later on demodex folliculorum were considered as the etiological agents of caseification necrosis in LMDF papules; however, both agents could not be demonstrated in granulomas that ruled out the validity of these hypotheses.[3]

In young adults, LMDF, which is mostly seen on face and less frequently on the extrafacial regions, tends to be a self-limiting pathology.[4]

Case Report

A 20-year-old male patient was referred to our outpatient clinic with asymptomatic rashes persisting for 2 months. The patient had no history of any known disease or drug use. On dermatological examination, monomorphic, solid, reddish-brown papular lesions with a smooth surface were observed on the nasolabial sulci and right infraorbital region and more intensely on the perioral region (Fig. 1). The patient had no subjective complaints such as pruritus, pain, and burning sensation. Histopathological examination of the biopsy material excised from the papule on his chin revealed superficial hyperkeratosis; mild dermal acanthosis; dermal granulomas; an area of focal caseification necrosis surrounded by epitheloid histiocytes, multinuclear giant cells, and lymphocytes; and a perivascular, perifollicular, and interstitial lymphocytic infiltration area (Fig. 2). Ziehl–Neelsen or Periodic acid–Schiff (PAS) staining could not demonstrate any fungal or mycobacterial agents.

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Submitted Date: February 08, 2017 **Accepted Date:** April 03, 2017 **Available Online Date:** May 28, 2018

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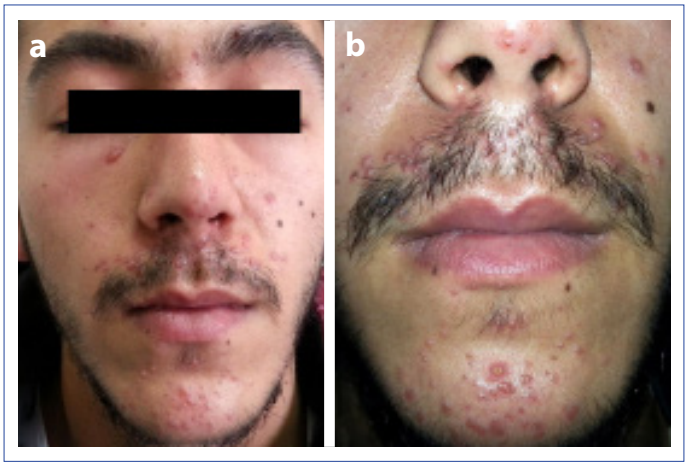

Figure 1. Lupus miliaris disseminatus faciei. **(a)** Multiple, reddish-brown papules can be seen on the patient's face. **(b)** Papular lesions were more intensely seen on the perioral region.

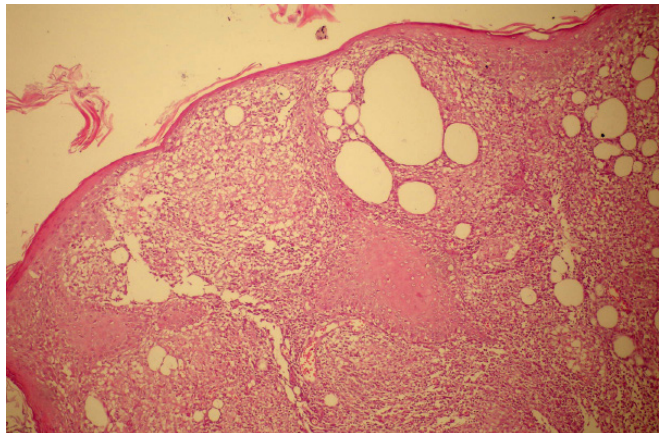

Figure 2. Histopathological examination of the papule (HE; ×100).

Complete blood counts and biochemical test results of the patient were within normal limits, and chest X-rays showed no abnormal findings. Based on clinical and histopathological findings, he was diagnosed with LMDF.

The patient was started on oral doxycycline at daily doses of 200 mg and did not show any improvement at the end of the first month. After 1 month, the lesions markedly regressed with treatment with systemic dapsone (50 mg/day) and topical tacrolimus.

Discussion

LMDF is a granulomatous, inflammatory chronic disease, which is mostly seen in young adults and affects both sexes equally.[5, 6] It particularly affects the central part of the face, and multiple, reddish-yellow or reddish-brown, solid papules are frequently seen on the malar, submandibular, and periorificial regions. The eyelid is typically involved.[2, 7] In cases with LMDF coursing with extrafacial involvement, the axilla, shoulders, arms, hands, neck, legs, and inguinal region are frequently affected.[8] Also in our case, the LMDF lesions were localized on the face, chin, nasolabial sulci, and infraorbital region and more intensely on the perioral region. Although the histopathological findings may change according to the stage of the lesion,[9] perifollicular granulomas formed by epitheloid cells with a central caseification necrosis are characteristic features of LMDF. Typically, chronic infiltrates accompany the granulomas seen in the vicinity of adnexal structures.[7] Lymphoid tissue accumulation surrounding the giant cells, which may be contained in the granulomas, may be observed.[10]

For the etiopathogenesis of LMDF, various theories have been suggested; however, the most frequently accepted one is that LMDF is a subtype of granulomatous rosacea.[11] In the differential diagnosis, sarcoidosis, nontuberculous mycobacterial infections, deep fungal infections, acne vulgaris, milia, and syringomas should be considered.[12]

Frequently, the lesions in LMDF, a self-limiting dermatosis, spontaneously regress within 12–24 months, leaving a scar tissue as a sequela.[9] For the management of LMDF, systemic (tetracycline, low-dose isotretionine, dapsone, corticosteroids, and immunosuppressive drugs) and topical (corticosteroids, tacrolimus, and psoralen combined with UV-A radiation) treatment alternatives are available.[9, 12] Treatment initiation at an early stage shortens the treatment duration and decreases scar formation.[6]

In our patient, who was refractory to doxycycline, a rapid response was obtained using systemic dapsone (50 mg/day) and topical tacrolimus. From the first month of the treatment, the lesions markedly regressed, and scar formation was seen after healing of the lesions. Our patient with rarely seen LMDF provided a rapid response to dapsone; therefore, we emphasize that dapsone and topical calcineurin inhibitors should be considered among treatment alternatives.

Disclosures

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

Authorship contributions: Concept – A.A.Ç., E.A.K.; Design – A.A.Ç.; Supervision – İ.K.A.; Materials – A.A.Ç., E.A.K.; Data collection &/or processing – A.A.Ç., E.A.K., Ö.Y.; Analysis and/or interpretation – E.A.K., Ö.Y.; Literature search – E.A.K.; Writing – E.A.K.; Critical review – A.A.Ç.

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