Case Report
Olgu Sunumu

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Unusual histopathological findings of lupus miliaris disseminatus faciei: A case report

Lupus miliaris disseminatus fasiei'in olağandışı histopatolojik bulguları: Bir olgu sunumu

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Abstract

Lupus miliaris disseminatus faciei (LMDF) is a chronic granulomatous disorder affecting the face in young adults with sudden eruption of asymptomatic skin colored to red-brown papules on cheeks, eyelids and few extrafacial sites including ears and neck. There has been disputation about the origin of the disease, but the consensus is that it is an immune response to the pilosebaceous unit. Various histopathological studies have found granulomas in association with pilosebaceous units. The present case discusses the rare occurrence of foreign body granuloma surrounding a ruptured infundibular cyst in the case of LMDF. The role of a ruptured infundibular cyst in the induction of an inflammatory reaction resulting in the development of granuloma in LMDF cannot be ruled out.
Keywords: Unusual, case report, ruptured infundibular cyst

Öz

Lupus miliaris disseminatus fasiei (LMDF), genç erişkinlerde yüzü etkileyen, yanaklarda, göz kapaklarında ve kulaklar ve boyun da dahil olmak üzere birkaç ekstrafasiyal bölgede ani asemptomatik deri renginden kırmızı kahverengiye kadar değişen renkte papüllerin ortaya çıktığı kronik granülomatöz bir hastalıktır, ancak genel fikir birliği bunun pilosebase üniteye karşı bir immün yanıt olduğu yönündedir. Çeşitli histopatolojik çalışmalar granülomların pilosebase ünitelerle ilişkili olduğunu bulmuştur. Mevcut olguda, bir LMDF olgusunda rüptüre infundibüler kisti çevreleyen nadir görülen yabancı cisim granülomu tartışılmaktadır. Rüptüre infundibüler kistin, LMDF'de granülom gelişimi ile sonuçlanan enflamatuvar reaksiyonun indüklenmesindeki rolü göz ardı edilemez.
Anahtar Kelimeler: Olağandışı, olgu sunumu, rüptüre infundibüler kist

Introduction

Lupus miliaris disseminatus faciei (LMDF) is an idiopathic chronic granulomatous disease usually affecting young adults of both sexes. It is characterized by a sudden eruption of multiple, asymptomatic red to brown papules or nodules in the central and lateral regions of the face with involvement of lower eyelids and ears[1]. The complexity in its etiopathogenesis accounts for variations in the histopathology of LMDF. The perifollicular localization of granuloma is an indicator of the immune response to the pilosebaceous unit which is an important cause of LMDF[2]. We herein report a case of LMDF with a ruptured infundibular cyst in the center of granuloma.

Case Report

A 30-year-old male patient presented to the dermatology outpatient clinic with non-itchy, painless, and progressively proliferating papules on bilateral ear lobes for approximately one year. There was no history of aggravation on exposure to a hot environment or intake of spicy foods. There was no history suggestive of weight loss, breathlessness,

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photosensitivity, long-standing cough, fever, hypopigmented patches, loss of sensations, alcohol or steroid abuse. The general physical examination was normal. There was no history of tuberculosis. There was no occupational exposure to aromatic hydrocarbons. On cutaneous examination, multiple, symmetrical, firm, non-tender pale-colored to erythematous papulo-nodular lesions were noted on bilateral earlobes (Figure 1a, b). There was no associated telangiectasia, involvement of the nose or perinasal area, or eyelids. Laboratory investigations were insignificant. The slit skin smear and Mantoux test were negative. Chest X-ray was essentially normal. A 3 mm punch biopsy sample was taken from the right ear lobe and sent for histopathological analysis. The tissue sample was negative for CB-NAAT. Histopathology revealed a diffuse dense granuloma of lymphocytes, plasma cells, epitheloid cells, and numerous foreign-body giant cells. In the center of the granuloma, a ruptured infundibular cyst was seen. The cyst consisted of laminated keratin squames, and the lining of the cyst was made up of stratified squamous epithelium (Figure 2a, b). After clinical and histopathological correlation, a definitive diagnosis of LMDF was made. Informed consent was obtained.

Discussion

LMDF is a controversial entity with an unclear etiology and pathogenesis. Earlier, it was thought to be caused by *Mycobacterium tuberculosis*, but there is no evidence to support it[3]. Some authors considered it as a micropapular form of sarcoidosis, but lately it is considered a variant of rosacea[3]. The multifactorial etiopathogenesis speculates over a possible role of Demodex folliculorum infection or a granulomatous reaction to hair follicle destruction[3]. The characteristic of LMDF on histology is the presence of perifollicular epitheloid cell granuloma in a fully developed lesion[4]. The classical histopathology of LMDF includes an epithelioid cell granuloma with or without central necrosis, an epithelioid cell granuloma with abscess, or non-specific non-granulomatous inflammatory infiltrate[5]. Granuloma formation in LMDF can be caused by an immune response to the pilosebaceous unit. An unusual reaction to Demodex mite or follicular contents can trigger perifollicular epitheloid cell granuloma, or it can be a reaction to an unknown infectious agent. Another theory is that it is a granulomatous reaction to a ruptured epidermal cyst[6]. A granulomatous reaction can also be triggered by the rupture of acne nodules, but in these situations, the inflammation is typically purulent and linked to appendage rupture, deformity, or destruction[7]. In a literature search, very few reports of LMDF with ruptured infundibular cysts are reported. Sanz-Sánchez et al.[8] reported a patient with lupus miliaris disseminatus faciei with a co-existing epidermoid cyst in a patient with two independent lesions of varied morphologies. A case report from Japan mentions multiple intact and ruptured epidermal cysts in association with LMDF[9]. Interestingly, in both of these case reports, the lesions were on the face and eyelids, while in our case, only ear lobe lesions were observed.

In the literature, though there is mention of rupture of the epidermoid cyst as a trigger for LMDF, there are still only occasional reports of the coexistence of both. The present case presentation and histopathological picture of a ruptured infundibular cyst surrounded by epitheloid cell granuloma reprise the theory that, along with other factors, the rupture of the infundibular cysts may initiate an inflammatory cascade leading to the formation of granuloma in LMDF.

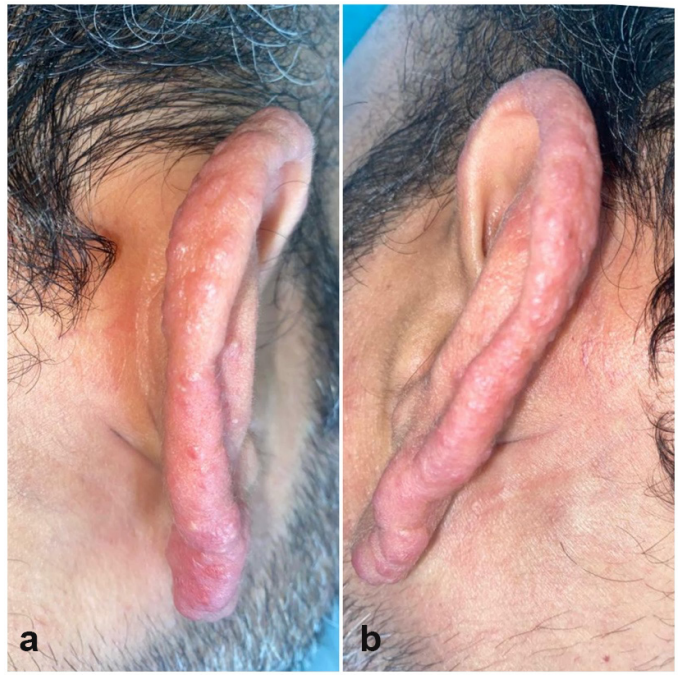

Figure 1. (a,b) Multiple small skin-colored to erythematous papules located on bilateral earlobes

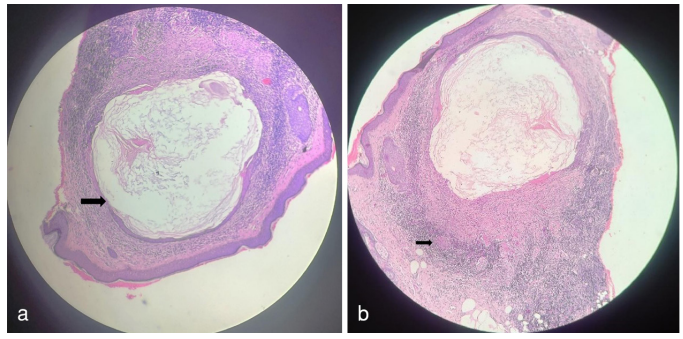

Figure 2. (a) Biopsy specimen from a lesion showing a ruptured infundibular cyst (indicated with a black arrow mark) with keratinous content surrounded by granuloma (haematoxylin- eosin stain, x10), **(b)** A diffuse, dense granuloma of lymphocytes, plasma cells, epitheloid cells (indicated with a black arrow mark) and numerous foreign-body giant cells (haematoxylin-eosin stain, x10)

Ethics

Informed Consent: It was obtained.

Authorship Contributions

Surgical and Medical Practices: P.B., N.K., Concept: P.B., N.K., Design: P.B., N.K., Data Collection or Processing: P.B., N.K., Analysis or Interpretation: P.B., P.K., N.K., Literature Search: P.B., P.K., N.K., Writing: P.B., N.K.

Conflict of Interest: No conflict of interest was declared by the authors.

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