IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 21, Issue 8 Ser.6 (August. 2022), PP 34-36
www.iosrjournals.org

Lupus miliaris disseminatus faciei: Pathologist's perspective of a rare entity

Divya Tiwary[1], Babai Halder[2*], Ratandeep Singh[3]
1&3- PG Trainee, 2- Associate Professor, Department of Pathology, MGM Medical college & LSK hospital, Kishanganj, Bihar-855107.

Abstract
Lupus miliaris disseminatus faciei (LMDF) is a rare chronic inflammatory dermatoses without an established etiology, was first described by Fox in 1878 [1]. Owing to the close morphological resemblance between Rosacea, LMDF. Here we are documenting a a case of a healthy 16-year-old male patient who presented to our dermatology outpatient clinic and was diagnosed with Rosacea. Skin biopsy was performed and histopathological examination revealed it to be LMDF. Thus, this report focuses on misleading clinical features which may raise a doubt of rosacea during initial visits when clinicians solely depend on clinical symptoms and hence, emphasises on the importance of clinicians working in collaboration with pathologists to diagnose and start the disease specific treatment.

Key words:*Lupus miliaris disseminatus faciei, rosacea, histopathological examination*

Date of Submission: 01-08-2022 Date of Acceptance: 14-08-2022

I. Introduction

Lupus miliaris disseminatus faciei (LMDF) is a rare chronic inflammatory disease which can be seen across a wide range of ages but mostly seen in young adults and affects both sexes equally. Approximately 200 cases have been reported till date [2]. Nosologically, it is on a spectrum of facial granulomatous dermatoses and shares overlapping features with rosacea and sarcoidosis. In most cases, this disorder resolves spontaneously within several years but can leave potentially disfiguring scars if deprived of appropriate treatment. Most recently it has been proposed by authors to adopting the term Facial idiopathic granulomas with regressive evolution (FIGURE) [3] . Older terms for a similar facial granulomatous dermatosis include micropapular tuberculid, Lewandowsky's eruption, and lupoid rosacea.[4] Acne agminata has been used to refer to similar lesions in the axilla.

It was once thought to be a tuberculid because of its histology, however many authors now consider LMDF to be an extreme variant of granulomatous rosacea (GR). Many are of the opinion that it is a distinct entity because of its characteristic histology and occasional involvement of the noncentral facial areas.

The earliest reports presumed that lesions of LMDF were related to tuberculosis based on similar clinicopathologic findings to other tuberculids. The term tuberculid was historically used for reactive conditions associated with tuberculosis, in which the actual infectious agent is present elsewhere and is not found in the skin lesions. However, nowadays, LMDF does not arise in association with pulmonary tuberculosis and does not generally respond to anti-tuberculous medications. Tuberculin skin testing is often negative in these patients.[5] Moreover, investigations, including histochemical staining for mycobacteria and tissue cultures, as well as PCR based studies, have consistently failed to demonstrate evidence of *M. tuberculosis* organisms within the LMDF granulomas. A possible relationship with an unknown non-tuberculous mycobacterium has not been completely excluded.Similar to other facial granulomatous eruptions, it is often postulated that initial immune-mediated damage to the hair follicle leads to subsequent rupture of the follicle and an allergic or foreign body granulomatous response to keratin, sebum, or microbial components in the dermis [6] This hypothesis is supported by observations that the granulomatous infiltrate is often centered around a follicular structure, which can sometimes be seen on serial histologic sectioning if not initially evident.

II. Case-Report

A healthy 16-year-old male patient who presented to our dermatology outpatient clinic with asymptomatic multiple reddish brown papular lesions over right and left infraorbital region, forehead and more intensely over central face involving nasal bridge, persisting for more than 3 months and gradually increasing in number [Figure 1]. There was no history of fever, weight loss in our patient.Past history was insignificant.

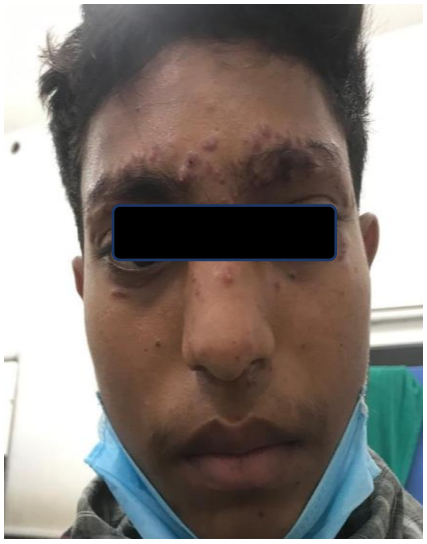

Figure 1: Multiple reddish brown papular lesion in our patient

A punch biopsy from a papular lesion over central face measuring (0.5x0.4x0.3) cm was received in the Department of Pathology.
Histopathological examination revealed epithelioid cell collection centered by large area of caseous necrosis and surrounded by dense granulomatous infiltrates.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 findings were consistent with the diagnosis of Lupus miliaris disseminatus faciei.

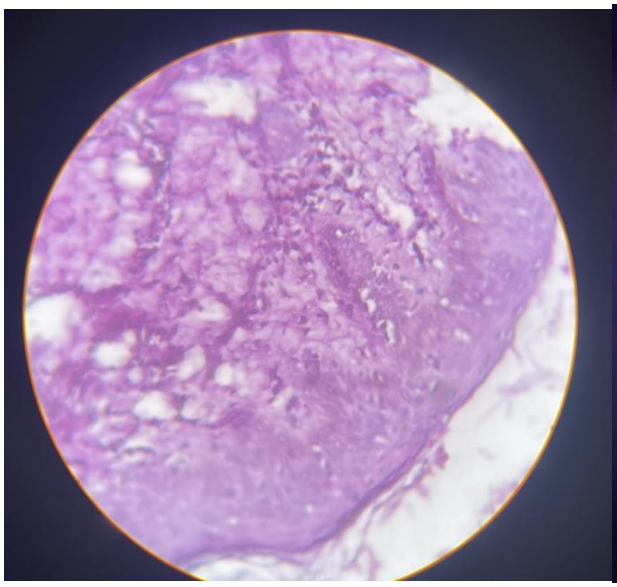

Figure 2: Atrophic epidermis (Hematoxylin & eosin)40X

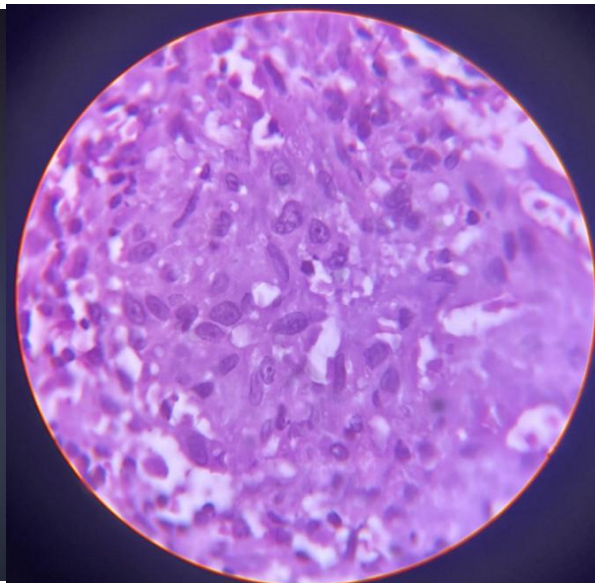

Figure 3: Dermis shows presence of epithelioid cell collection forming granuloma (Hematoxylin & eosin)100X

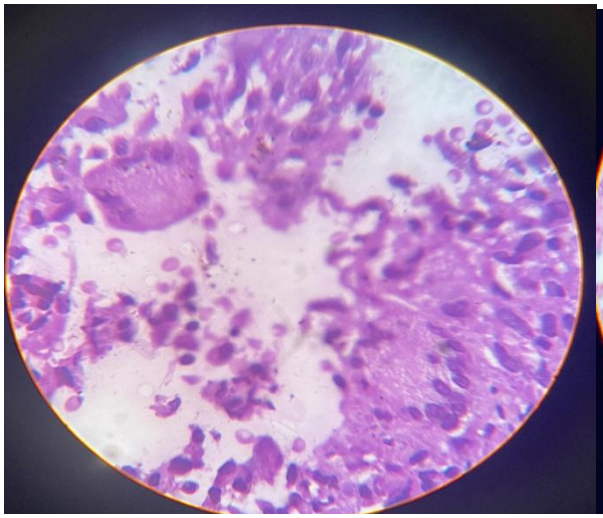

Figure 4: Langhan's giant cell (Hematoxylin & eosin) 100X

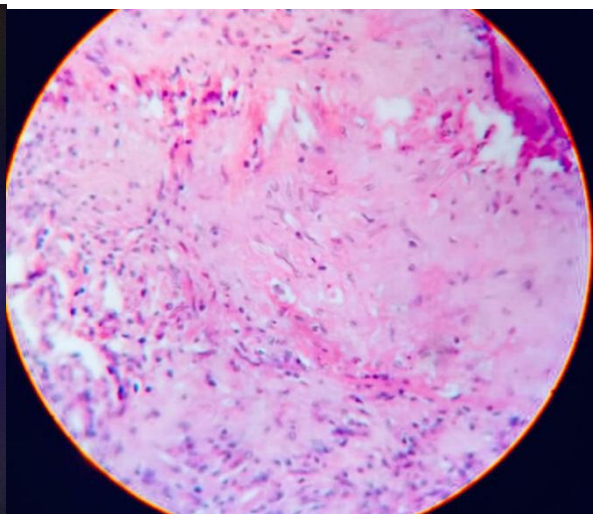

Figure 5: Caseous necrosis (Hematoxylin & eosin) 40X

III. Discussion

LMDF is particularly is characterized by multiple reddish-yellow or reddish-brown primarily over central face, periorbital and perioral area. Extrafacial zones can be rarely affected, including the axillae, neck, scalp, legs, trunk and genitalia.

The histopathological hallmark of the disease is an epithelioid cell granuloma with central necrosis. However, the histological pattern can vary according to the timing of the biopsy. Early lesions may show superficial perivascular infiltrates of lymphocytes, histiocytes, and occasional neutrophils. Late lesions have these changes together with dermal fibrosis, particularly around hair follicles. Established lesions may show tuberculoid or suppurative granulomas.

Like LMDF, granulomatous variants of rosacea can present with bilateral, symmetric facial papules with similar morphology. There is considerable histologic overlap, including relationship to pilosebaceous units, although LMDF may have larger granulomas that are more prone to caseation necrosis, and is not expected to have actinic damage, vascular dilation, or the presence of *Demodex* mites.[7] The clinical features and course may help discriminate these entities. Notably, LMDF occurs in the absence of phymatous changes, ocular involvement, or vascular manifestations of rosacea such as background erythema, flushing, or telangiectasia.[8] Compared to rosacea, LMDF tends to affect adults at a younger age, including some cases in adolescents and exceptional cases in children and may affect men more commonly.[9]The involvement of eyelids, upper lips, and neck is more common in LMDF, and extra facial involvement can occur. [10]

IV. Conclusion

The clinical presentation in the index case may raise a doubt of rosacea during initial visits when clinicians solely depend on clinical symptoms. Therefore, it is very important for the clinicians to work in collaboration with pathologists to start the disease specific treatment.

References

[1]. Fox T. Disseminated follicular lupus (simulating acne) *Lancet* 1878;112:75–6.
[2]. Esteves T, Faria A, Alves R, Marot J, Viana I Vale E. Lupus miliaris disseminatus faciei: a case report. *Dermatol Online J*2010;16(5):10.
[3]. van de Scheur MR, van der Waal RI, Starink TM. Lupus miliaris disseminatus faciei: A distinctive rosacea-like syndrome and not a granulomatous form of rosacea. Dermatology 2003;206:120-3.
[4]. Hodak E,Trattner A,Feuerman H,Feinmesser M,Tsvieli R,Mitrani-Rosenbaum S,David M.Lupus miliarisdisseminatus faciei- The DNA of Mycobacterium tuberculosis is not detectable in active lesions by polymerase chain reaction. Br J Dermatolol 1997; 137: 614-9.
[5]. Chougule A,Chatterjee D,Yadav R,Sethi S,De D,Saikia UN, Granulomatous Rosacea Versus Lupus Miliaris Disseminatus Faciei-2 Faces of Facial Granulomatous Disorder: A Clinicohistological and Molecular Study. Am J Dermapathol2018; 40(11): 819-23.
[6]. Al-Mutairi N. Nosology and therapeutic options for lupus miliaris disseminatus faciei. J Dermatolol 2011; 38(9): 864-73.
[7]. Makkar R,Ramesh V. On the diagnosis of facial granulomatous dermatoses of obscure origin. Int J Dermatolol 2005; 44(7): 606-9.
[8]. Beleznay K, Friedmann DP, Liolios AM, Perry A, Goldman MP. Lupus miliaris disseminatus faciei treated with 1,565nm nonablative fractionated laser resurfacing: A case report. *Lasers Surg Med* 2014;46:663–5.
[9]. Kang BK, Shin MK. Scarring of lupus miliaris disseminatus faciei: treatment with a combination of trichloroacetic acid. *Dermatol Ther* 2014;27:168–70.
[10]. 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.