

## Lupus Miliaris Disseminatus Faciei- A Case Report and Literature Review

Dr. Priyanka Anand<sup>1\*</sup>, Dr. Namrata Sarin<sup>2</sup>, Dr. V.K Khurana<sup>3</sup>

<sup>1</sup>M.D.Pathology, Senior Resident, Department of Pathology, NDMC and Hindu Rao hospital, India

<sup>2</sup>M.D. Pathology, Head of Department, Department of Pathology, NDMC and Hindu Rao hospital, India

<sup>3</sup>M.D Dermatology, Head of Department, Department of Dermatology, NDMC & Hindu Rao hospital, India

### Case Report

#### \*Corresponding author

Dr. Priyanka Anand

#### Article History

Received: 03.07.2018

Accepted: 13.07.2018

Published: 30.07.2018

#### DOI:

10.21276/sjpm.2018.3.7.2



**Abstract:** Lupus Miliaris Disseminatus Faciei is an idiopathic granulomatous skin disease affecting the face. It is also called as Acne agminata or Facial idiopathic granulomas with regressive evolution. It has a predilection for lower eyelids, forehead, nasolabial folds and perioral areas. Caseating as well as non- caseating epithelioid cell granulomas are identified on histopathological examination therefore differentials such as tuberculosis, sarcoidosis, granulomatous rosacea and other granulomatous disorders should be ruled out. A thirteen year old female presented with multiple papules perinasally and periorally around chin. A skin biopsy was performed from the lesion and sent for histopathological examination. On microscopic examination, a granulomatous reaction pattern was seen in the dermis. X-ray chest was unremarkable. Mantoux test was negative. Apple- jelly nodule like appearance was observed on diascopy. A diagnosis of LMDF was rendered on histopathology after clinical correlation. The diagnosis of LMDF should be kept in mind in case of papular eruptions on face not responding to antitubercular treatment. Mainstay of treatment is tetracycline and oral isotretinoin.

**Keywords:** Lupus miliaris disseminatus faciei, Acne agminata, caseous necrosis.

### INTRODUCTION

Lupus Miliaris Disseminatus Faciei (LMDF) is an uncommon granulomatous skin disease of unknown etiology affecting the face. It was first described by Tilbury Fox *et al* in 1878 [1].

It is also called as acne agminata or Facial idiopathic granulomas with regressive evolution. The patient usually presents as small, discrete reddish-brown, dome shaped papules singly or in crops, rapidly developing, follicular or non- follicular lesion with a pustular top in majority. It has a predilection for lower eyelids, forehead, nasolabial folds and perioral areas [2]. Extrafacial involvement has been rarely documented in literature.

Caseating as well as non- caseating epithelioid cell granulomas are identified on histopathological examination therefore differentials such as tuberculosis, sarcoidosis, granulomatous rosacea and other granulomatous disorders should be kept in mind.

Spontaneous involution occurs in 12-24 months leaving small pitted scars [3]. Pathogenesis of the disease is still not known. It usually affects adults in second and fourth decade.

It was considered before to be a tuberculid, many authors now believe that it is an extreme variant of granulomatous rosacea. Moreover others have the opinion that it is a distinct entity. Mainstay of treatment is tetracycline and oral isotertoinin but the response is

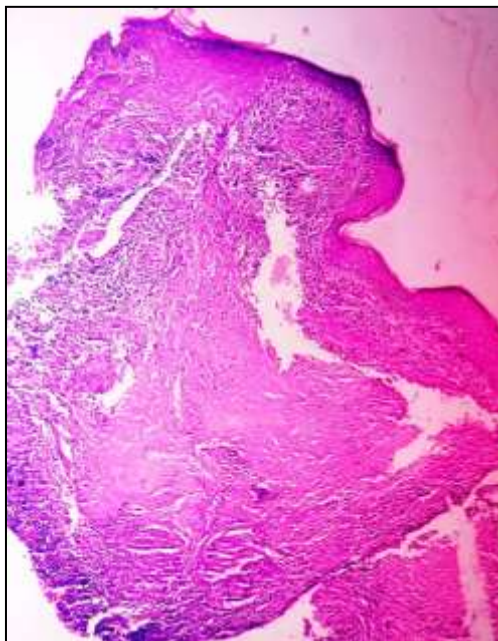
not very satisfactory. We herein report a rare case of LMDF and briefly review literature about its salient clinicopathologic features.

### CASE PRESENTATION

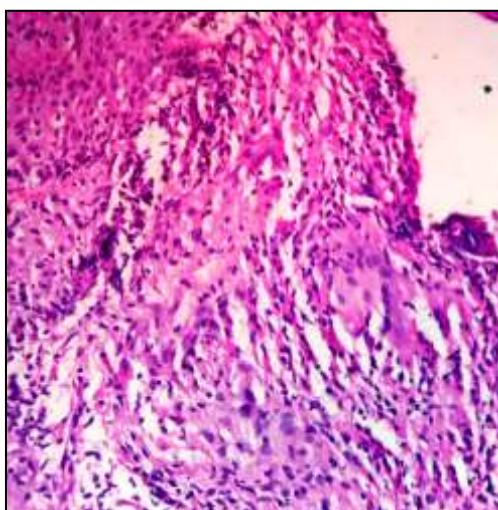
A thirteen year old female presented with asymptomatic, reddish- brown, discrete, multiple papules, varying in size from 2-4mm in size. Papules were present perinasally and around chin. These lesions had evolved over about one year. On examination, there were no other cutaneous lesions. History of photosensitivity was absent. There was no history of topical application of steroids on face. She denied history of any similar lesions in family. On examination, there was no flushing, redness or telangiectasia present. There was no cervical lymphadenopathy. A skin biopsy was performed from the lesion and sent for histopathological examination. On microscopic examination, epidermis showed mild acanthosis. In the dermis, a granulomatous reaction was seen. Granulomas comprised of central area of necrosis surrounded by epithelioid cells, multinucleated giant cells and a lymphocytic cuff (Fig-1 & 2). Stains for acid- fast bacilli and fungi were negative. X-ray chest was unremarkable. Routine laboratory investigations were within normal limits. Mantoux test was negative.

Apple- jelly nodule like appearance was observed on diascopy. Clinically, differentials given were LMDF, popular sarcoidosis, syringoma and trichoepithelioma.

A diagnosis of LMDF was rendered on histopathology after clinical correlation. The patient was put on isotretinoin for four months.



**Fig-1: Microphotograph shows epithelioid cell granuloma in dermis with central caseous necrosis surrounded by epithelioid cells, giant cells and lymphocytes. (H and E stain, 10x)**



**Fig-2: High power view of epithelioid cell granuloma showing giant cells and lymphocytic infiltrate. (H and E stain, 40x)**

## DISCUSSION

Lupus miliaris disseminates faciei is a rare, chronic inflammatory dermatosis. It is also called as “acne agminata” or “acnitis”. The patients present with yellowish- red asymptomatic papules over the central part of face typically involving periorbital area, nasolabial folds, cheeks and perioral areas. On histopathological examination, infiltrate resembles that of lupus vulgaris, granulomas with central necrosis that are in close proximity to follicular structures but sebaceous glands are unusually involved. Perivascular

infiltrate of lymphocytes, histiocytes in superficial dermis may be evident in early lesions. In established lesions, perifollicular fibrosis is seen and granulomas may be seen. Previously LMDF was supposed to be a variant of lupus vulgaris or tuberculid because of histological resemblance but there has been no evidence regarding its relation to tuberculosis [4]. Some studies suggest that it is a granulomatous reaction to follicle destruction. It has variable mantoux positivity in response to tuberculin. Majority of patients do not respond to antitubercular treatment. Lesions fail to

demonstrate acid fast bacilli and neither culture nor the polymerase chain reaction techniques for detection of *M.tuberculosis* DNA is helpful.

According to the literature some authors believe that LMDF is a form of granulomatous rosacea [5] as the granulomas are in association with pilosebaceous units in LMDF [6, 7].

But in rosacea, mainly extrafacial sites are involved and there is sparing of lower eyelids which goes against LMDF being a type of rosacea. Also absence of erythema, flushing and telangiectasia does not favour LMDF to be a form of rosacea. Therefore, LMDF should be considered as a distinct entity [8].

Skowron *et al.*, in 2000, changed the name from LMDF to Facial idiopathic granulomas with regressive evolution (FIGURE) however it is not been widely accepted. Differential diagnosis includes clinical entities such as acne, lupus vulgaris, granulomatous rosacea, sarcoidosis and histoid leprosy [9].

The diagnosis of LMDF should be kept in mind in case of papular eruptions on face not responding to antitubercular treatment. Lesions usually have a spontaneous resolution with scarring. Mainstay of treatment is tetracycline and oral isotertinoiin but the response is variable and not very satisfactory.

#### REFERENCES

1. Fox, T. (1878). Clinical Lecture on disseminated follicular lupus (SIMULATING ACNE). *The lancet*, 112(2864), 75-76.
2. Esteves, T., Faria, A., Alves, R., Marote, J., Viana, I., & Vale, E. (2010). Lupus miliaris disseminatus faciei: a case report. *Dermatology online journal*, 16(5).
3. Marks, R., & Wilkinson, D. S. (1992). Acne agminata. *Textbook of Dermatology (Champion RH, Burton JL, Ebling FJG, eds). Oxford: Blackwell Scientific Publications*, 1862-3.
4. Hodak, E., Trattner, A., Feuerman, H., Feinmesser, M., Tsvieli, R., Mitrani-Rosenbaum, S., & David, M. (1997). Lupus miliaris disseminatus faciei—the DNA of *Mycobacterium tuberculosis* is not detectable in active lesions by polymerase chain reaction. *British Journal of Dermatology*, 137(4), 614-619.
5. Helm, K. F., Menz, J., Gibson, L. E., & Dicken, C. H. (1991). A clinical and histopathologic study of granulomatous rosacea. *Journal of the American Academy of Dermatology*, 25(6), 1038-1043.
6. Shitara A. Clinicopathological and immunological studies of luous miliaris disseminatus faciei. *J Dermatol*. 1982;9:383-95.
7. El Darouti, M., & Zaher, H. (1993). Lupus miliaris disseminatus faciei-pathologic study oe early, fully developed, and late lesions. *International journal of dermatology*, 32(7), 508-511.

8. van de Scheur, M. R., van der Waal, R. I., & Starink, T. M. (2003). Lupus miliaris disseminatus faciei: a distinctive rosacea-like syndrome and not a granulomatous form of rosacea. *Dermatology*, 206(2), 120-123.
9. Skowron, E. A. (2000). The role of differentiation of self in marital adjustment. *Journal of counseling Psychology*, 47(2), 229.