

CASE 7

Patient: A 63-year-old Thai female from Pathumthani

Chief Complaint: Progressive facial eruption in 1 month

Present Illness: The lesions involved only facial area without itching or pain. No associated systemic symptoms and precipitating factors including sunlight, spicy foods were noted. The eruption was not improved with topical steroids.

Past History: None

Family History: Unremarkable

Dermatological Examination (Figure 7.1-7.3): Multiple discrete dome-shaped erythematous papules and plaque on face, Rt. Eyelid



Histopathology (S10-5379) (Figure 7.4-5): Perifollicular nodular infiltrate of histiocytes, multi nucleated langerhan giant cells and some lymphocytes with the features of tuberculoid granuloma and central necrosis

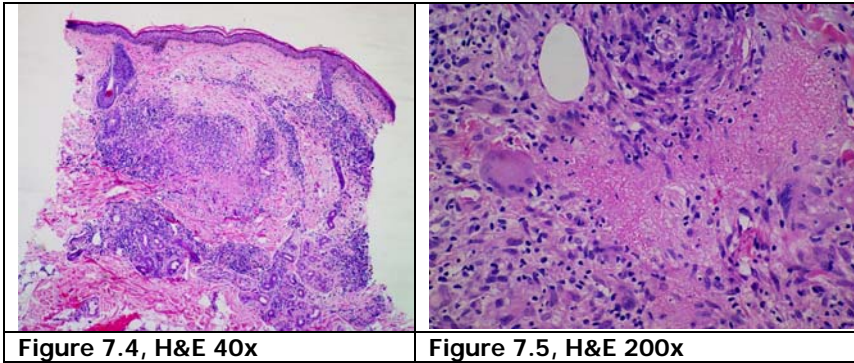


Figure 7.4, H&E 40x

Figure 7.5, H&E 200x

Investigation: CXR-no infiltration, PPD-negative

Diagnosis: Lupus miliaris disseminatus faciei (LMDF)

Treatment: Doxycycline (100) 1*2

Presenter: Premjit Vaiyavatjamai

Consultant: Natta Rajatanavin

Discussion:

Lupus miliaris disseminatus faciei (LMDF) or acne agminata is a rare asymptomatic granulomatous skin eruption which is characterized by multiple discrete red-brown, dome-shaped papules on the medial and lateral areas of the face especially on and around the eyelids, and often extends on to head and chin¹. Extrafacial manifestations² are not uncommon and may affect axillae, shoulders, arms, hands, groins and legs. Lesions have the appearance of an 'apple jelly' color on diascopy. With the chronic progressive course of disease, LMDF is usually developed spontaneous involution in about 1-2 years and typically leaves small pitted scars.

Epithelioid granuloma with central necrosis is the histopathological hallmark³. However, the histological pattern can vary according to the timing of the biopsy⁴. In the early

lesions, superficial perivascular infiltration of lymphocytes, histiocytes and occasional neutrophils are noticed while dermal fibrosis especially around the hair follicles is reported in late lesions. In the fully established lesions, tuberculoid or suppurative granulomas might be shown. As a result, LMDF histologically resembles tuberculosis, sarcoidosis, granulomatous rosacea and other granulomatous disorders. Therefore, LMDF has been termed Lewandowsky's rosacea-like tuberculid, micropapular tuberculid, lupoid rosacea⁵ and Facial Idiopathic GranUlomas with Regressive Evaluation (F.I.GU.R.E)⁶.

Pathogenesis of LMDF is unclear and controversial. Possible involving etiologies include granulomatous rosacea, tuberculid, papular form of sarcoidosis and demodex folliculorum. Regarding to the histological association with pilosebaceous units and granulomas, granulomatous rosacea was issued⁵. Nevertheless, granulomatous rosacea is commonly presented in a 20 to 50 year-old female with erythema, telangiectasia, flushing, blepharitis and conjunctivitis with no lower eyelids involvement then, the resolution occurs without scar. Alcohol, ultraviolet and spicy food are the precipitating causes of rosacea. Topical metronidazole and oral tetracyclines are drugs of choice. On the other hand, LMDF is generally found in young adult male with multiple singly or crops of erythematous to brownish papules on face and eyelids. Clinical course shows chronic but spontaneous resolution with scars. No aggravating factor and no consistent treatment are shown. Though both LMDF and tuberculid demonstrate similar histopathology of tuberculoid granuloma⁷, LMDF does not present the evidence of tuberculosis-no concomitant TB, no bacilli and negative PCR for TB in lesions and variable response to both PPD and antituberculosis. Also, both LMDF and sarcoidosis can reveal

naked granuloma; however, LMDF do not have systemic symptoms especially lung involvement, like sarcoidosis. Previously, this granuloma formation was regarded as a delayed-type hypersensitivity reaction to demodex folliculorum⁸; nonetheless, the association has not been confirmed. Later, LMDF is assumed as a granulomatous reaction to hair follicle destruction or ruptured epidermal cysts.

It is difficult to control the progression of LMDF. Numerous studies of various treatments with variable responses have been published. Medications previously reported are low-dose prednisolone⁹, dapsone¹⁰, tetracycline¹¹, doxycycline¹², minocycline, clofazimine¹³, antimalarial, antituberculosis and isotretinoin¹⁴. Lastly, 1450-nm diode laser¹⁵ has been used to disrupt sebaceous glands and hair follicles in order to decrease sebum production and reduce inflammation. Generally, tetracycline has been proposed as the first line of treatment. However, the controlled study to establish the best treatment is lacking.

In conclusion, doxycycline has been prescribed to this patient. No progressive eruption and slight improvement have been shown.

References

1. Kim DS, Lee KY, Shin JU, Roh MR, Lee MG. Lupus miliaris disseminatus faciei without facial involvement. *Acta Derm Venereol* 2008;88:504-5.
2. Hillen U, Schroter S, Denisjuk N, Jansen T, Grabbe S. Axillary acne agminata (lupus miliaris disseminatus faciei with axillary involvement). *J Dtsch Dermatol Ges* 2006;4:858-60.
3. Esteves T, Faria A, Alves R, Marote J, Viana I, Vale E. Lupus miliaris disseminatus faciei: a case report. *Dermatol Online J* 2010;16:10.
4. el Darouti M, Zaher H. Lupus miliaris disseminatus faciei--pathologic study of early, fully developed, and late lesions. *Int J Dermatol* 1993;32:508-11.
5. 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.

6. Skowron F, Causeret AS, Pabion C, Viillard AM, Balme B , Thomas L. F.I.G.U.R.E.: facial idiopathic granulomas with regressive evolution. is 'lupus miliaris disseminatus faciei' still an acceptable diagnosis in the third millennium? *Dermatology* 2000;201:287-9.
7. Nino M, Barberio E , Delfino M. Lupus miliaris disseminatus faciei and its debated link to tuberculosis. *J Eur Acad Dermatol Venereol* 2003;17:97.
8. Ruffli T , Buchner SA. T-cell subsets in acne rosacea lesions and the possible role of *Demodex folliculorum*. *Dermatologica* 1984;169:1-5.
9. Uesugi Y, Aiba S, Usuba M , Tagami H. Oral prednisone in the treatment of acne agminata. *Br J Dermatol* 1996;134:1098-100.
10. Kumano K, Tani M , Murata Y. Dapsone in the treatment of miliary lupus of the face. *Br J Dermatol* 1983;109:57-62.
11. Moloney FJ , Egan CA. Case 3. Acne agminata (lupus miliaris disseminatus faciei). *Clin Exp Dermatol* 2003;28:685-6.
12. Goh BK , Tan HH. Doxycycline in the treatment of acne agminata. *Clin Exp Dermatol* 2003;28:677-9.
13. Seukeran DC, Stables GI, Cunliffe WJ , Sheehan-Dare RA. The treatment of acne agminata with clofazimine. *Br J Dermatol* 1999;141:596-7.
14. Berbis P , Privat Y. Lupus miliaris disseminatus faciei: efficacy of isotretinoin. *J Am Acad Dermatol* 1987;16:1271-2.
15. Jih MH, Friedman PM, Kimyai-Asadi A, Friedman ES, Hymes SR , Goldberg LH. Lupus miliaris disseminatus faciei: treatment with the 1450-nm diode laser. *Arch Dermatol* 2005;141:143-5.