A CASE REPORT: LUPUS MILIARIS DISSEMINATUS FACIEI

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SUMMARY

Lupus miliaris disseminatus faciei (LMDF), an uncommon inflammatory granulomatous dermatosis with possible immune mechanisms, affects young adults. We report a typical case of LMDF on a 28-year-old male patient diagnosed by clinical and histopathology. The patient was successfully treated with a dual therapy of oral minocycline 100mg/day and methylprednisolone 4mg/day.

1. INTRODUCTION

Lupus miliaris disseminatus faciei (LMDF) is a rare inflammatory skin disease with unknown pathogenesis. Papules of the midface and around the eyelids are the main characteristics of this disease. Typical histopathological features are dermis epithelial granulomatosis, central necrosis, and surrounding lymphoid infiltration with giant multinucleated cells. The disease may regress spontaneously and may leave scarring. Treatment of the disease reamins difficult due to a lack of clinical studies. We report a typical case of Lupus miliaris disseminatus faciei diagnosed at the Vietnamese National Hospital of Dermatology and Venerology.

2. CASE REPORT

A 28-year-old male patient presented to our hospital with some red papules on his face for 7 months. The patient was primarily diagnosed with acne vulgaris and treated with isotretinoin 20mg/day but after 2 months of treatment, the lesions did not improve.

Physical examination revealed multiple small papules 1 - 3mm in size, dome-shaped with skin-colored and slightly red, symmetrically distributed on the face, mainly on the forehead, cheeks, and chin (Figures 1, 2, 3, 4). In addition, scar lesions and hyperpigmentation in the cheek were also observed. No vasodilatation, scaling, or flushing were observed. The patient had no past medical history of topical or oral corticosteroid use.

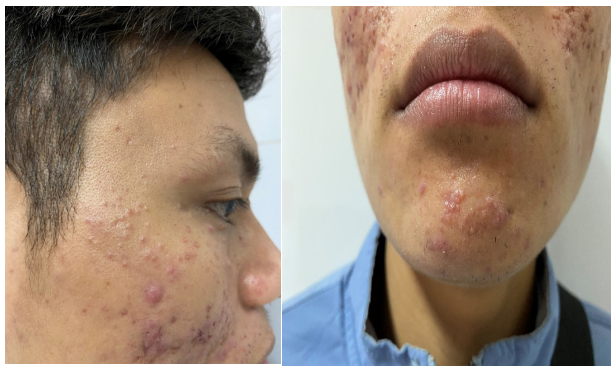

Figure 1, 2. Multiple small red papules, symmetrically distributed on the forehead, cheeks, under the chin

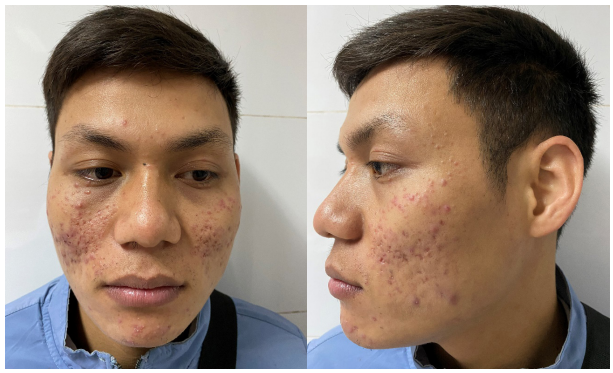

Figure 3, 4. Skin lesions are skin-colored, slightly red, dome-shaped papules

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Histopathological picture of biopsied lesions on the chin showed epithelial granulomatous with central necrosis surrounded by lymphocytes, histocytes, and multinucleated giant cells. (Figure 5 and 6). No foreign body was found in the granuloma, and PAS and Ziehl-Neelsen staining were negative. Chest X-ray and other tests showed no abnormalities.

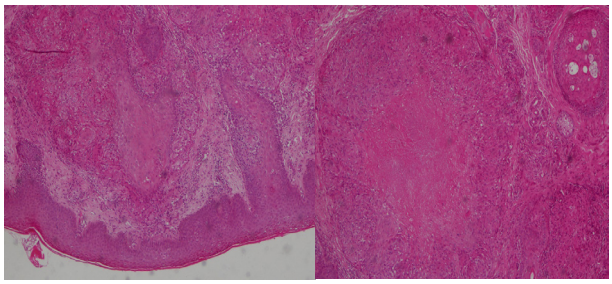

Figure 5. The epidermis is normal, the dermis has a granulomatous reaction (HE X10)

The confirmed diagnosisof Lupus miliaris disseminatus faciei (LMDF) was made. The patient was first treated with oral doxycycline 100 mg/day in combination with methylprednisolone 4mg/day and topical tacrolimus 0.03% daily for 1 month. Lesions improve little and new lesions appear. The patient was switched to oral minocycline 100 mg/day in combination with methylprednisolone 4 mg/day for 3 months. The pustular papules disappeared and no new lesions appeared (Figure 7, 8). Currently, the main lesions are scars, and we plan to continue to monitor and treat to improve the patient's cosmetic problems.

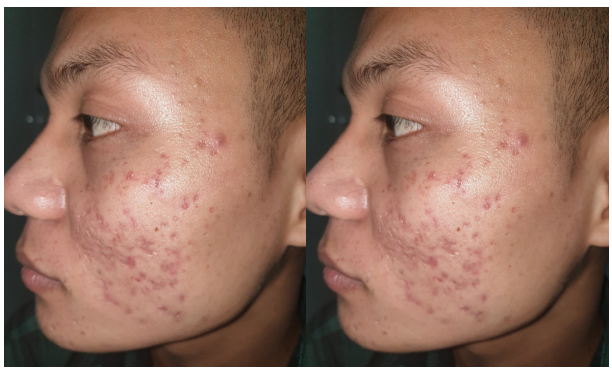

Figure 7, 8. After 4 months of treatment, papules and pustules completely disappeared, leaving scars

3. DISCUSSION

Lupus miliaris disseminatus faciei (LMDF) was first described by Fox in 1878 as "disseminated follicular lupus" because this author assumed that the lesions were diffuse hair follicle papules. In 1903, Radcliffe-Crocker coined the term "acne agminata" or "acnitis" to describe this condition because the lesions had many features similar to acne.

LMDF is a rare skin disease with just over 200 cases reported in the literature. The most common age group is 20 - 40 years old with some rare cases in children and the elderly. The disease occurs in both sexes and women often have a later onset than men did. The pathogenesis of LMDF is still unknown. Initially, the disease was considered as a form of skin tuberculosis, however, *Mycobacterium tuberculosis* was not found in the lesions, so this hypothesis was not accepted. In 1980, many authors considered LMDF to be a form of granulomatous rosacea, but there are some differences, such as self-limiting, scarring, histopathological necrosis, and not accompanied by erythema lesions, vasodilation, flushing. Another hypothesis related to *Demodex folliculorum* was also rejected when no association was found. *Propionibacterium acne* was also found in LMDF lesions, suggesting that the pathogenesis might relate to some bacteria affecting hair folliclesbut its mechanism remains unclear.

The typical clinical presentation is multiple small, skin-coloredor reddish papules on the face, typically around the eyelids, with no itch or pain. Papules may or may not be in the hair follicles. In some cases, pustular lesions are seen. Lesions appear abruptly , progress chronically and leave scars after 12 - 24 months. A review on 26 Japanese patients of Dianawaty Amîruddîn et al found that the median time to regression was 15.7 months in men and 19.7 months in women and only 3 cases

persisted for more than 4 years, all in women. In addition to facial lesions, lesions might be found in other locations such as neck (33%), chest (29%), armpits (24%), hands, feet....

Image of LMDF on dermoscopy is apple jelly-colored nodules. Linear and hairpin-shaped blood vessels surround the flame or elliptical hair follicle.

Histopathological features vary according to the stage of the lesion. In the early stages, infiltration of lymphocytes, perivascular histiocytosis, and adnexa can be seen. In the middle stage, the lesion is characterized by epithelial granulomatous with central necrosis surrounded by lymphocytes and giant multinucleated cells. Late-stage lesions may present with extensive perifollicular fibrosis with a nonspecific inflammatory response.

The clinical features of LMDF can be mistaken with some skin diseases presented with small papules on the face, especially in the early stages. Differential diagnoses include acne, syringoma, nodular sarcoidosis, rosacea,… but they have difference features on histopathology, clinical and laboratory signs.

Studies and reports on treatment of LMDF are currently limited. Because the disease can be in remission after 1 - 2 years, it is difficult to accurately assess the effectiveness of treatment. The priority is tetracycline antibiotics (doxycycline and minocycline). Other treatments include isotretinoin, dapsone, corticosteroids, clofazimine, tranilast, anti-tuberculosis drugs, metronidazole and cyclosporin. Several case reports showed that tranilast (used in atopic dermatitis, allergic rhinitis due to its effect on mast cells, its ability to inhibit collagen synthesis and fibroblast proliferation) was also effective in LMDFs previously failed with tetracycline and macrolide. Topical medications such as tacrolimus, or laser therapy may be effective in LMDF. Early treatment and corticosteroids can help limit scarring. Treatment of LMDF-induced scars is similar to that in acne, including 100% trichloroacetic, CO_2 laser...

Al-Mutairi's report on 29 patients showed that among 11 patients treated with minocycline and doxycycline, only 2 patients had a good response. Two out of 4 cases were treated with isotretinoin had a good response. In the group of prednisolone treatment, 3 patients had excellent response, 2 patients had good response and only 1 patient had a poor response. Notably, 7 patients were treated with topical dapsone and tacrolimus and all of them responded very well.

Helena Toda-Brito et al treated a 43 female patient with oral minocycline 100 mg/day and prednisolone 5 mg/day. Flattening of the papules was observed within 3 weeks of therapy. A moderate improvement had been achieved by 16 weeks and no recurrence was noted over a 12-month observation period.

Tanvi Dev et al reported a 35-year-old male patient with LMDF who was treated with minocycline 50 mg twice daily and tacrolimus 0.1% for 2 months. However, the lesions did not improve and new lesions continued to appear. Betamethasone was added in dosage of 5mg/day for 2 consecutive days/week and the patient had a rapid response after 6 weeks.

4. CONCLUSION

Lupus miliaris disseminatus faciei is a rare granulomatous dermatitis that occurs mainly in young adults and its pathogenesis remains unknown. LMDF is easily misdiagnosed as acne. The disease is characterized by bilateral symmetric papules mainly in the face and granulomatous, with central necrosis in histology. Currently, there is no consensus on treatment of LMDF. We successfully diagnosed and treated a case of LMDF with oral minocycline 100mg/day and methylprednisolone 4mg/day.

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