Lichen Scrofulosorum with Atypical Presentation: A Case Report and Literature Review

Supawee Phanmamuang MD, Poonnawis Sudtikoonaseth MD.
Institute of Dermatology, Department of Medical Services, Ministry of Public Health, Bangkok, Thailand.

ABSTRACT:
Lichen scrofulosorum is a rare form of cutaneous tuberculosis caused by an immune hypersensitivity reaction to *Mycobacterium tuberculosis* infection. The common locations are the trunk and proximal extremities. This study reported atypical presentation of lichen scrofulosorum. A 36-year-old Thai male presented with perifollicular papules on both periorbicular areas and cheeks. The diagnosis was confirmed by histopathology, which showed perifollicular non-caseous granulomatous formation, and tissue polymerase chain reaction (PCR) was positive for *Mycobacterium tuberculosis*. He was treated with standard antituberculosis therapy. The clinical symptoms completely resolved after six months of treatment.
Key words: Lichen scrofulosorum, Cutaneous tuberculosis, Tuberculid, Antituberculosis therapy

Introduction

Lichen scrofulosorum (LS) is a rare form of cutaneous tuberculosis. The common presentations are perifollicular lesions on the trunk or proximal extremities. We reported an interesting case of the atypical location in a middle-aged Thai male who presented with perifollicular papules on the face.

Case report

A 36-year-old Thai male presented with multiple erythematous papules on the face for four months. No other systemic symptoms such as fever, chronic cough, or weight loss. He denied a history of trauma or facial procedure. He has no current underlying disease and no history of contact with tuberculosis. Physical examination revealed multiple bilateral well-defined non-scaly erythematous perifollicular papules with some coalescing into plaques on both periorbicular areas and cheeks (Figure 1A, 1B, 1C, 1D). No lymphadenopathy or abnormal breath sound was noted. The skin biopsy showed mild acanthosis of the epidermis and mixed inflammatory cell infiltration composed of lymphocytes, neutrophils, plasma cells, and histiocytes. There was a perifollicular non-caseous granulomatous formation, especially adjacent to the hair follicle (Figure 2A, 2B). No microorganism was seen. AFB, PAS, and GMS special stains were all negative. Tissue PCR was positive for *Mycobacterium tuberculosis*. Culture for bacteria, fungus, and mycobacteria were all negative as well as other laboratory examinations including quantiferon-TB Gold test, and anti-HIV. chest X-ray demonstrated no abnormality. The patient was diagnosed with lichen scrofulosorum. His clinical improvement was noted after a month of antituberculosis treatment and a complete resolution after six months.

Corresponding Author: Poonnawis Sudtikoonaseth MD, email: usopyon@hotmail.com

Received: 21 November 2022 Revised: 24 April 2023 Accepted: 2 May 2023

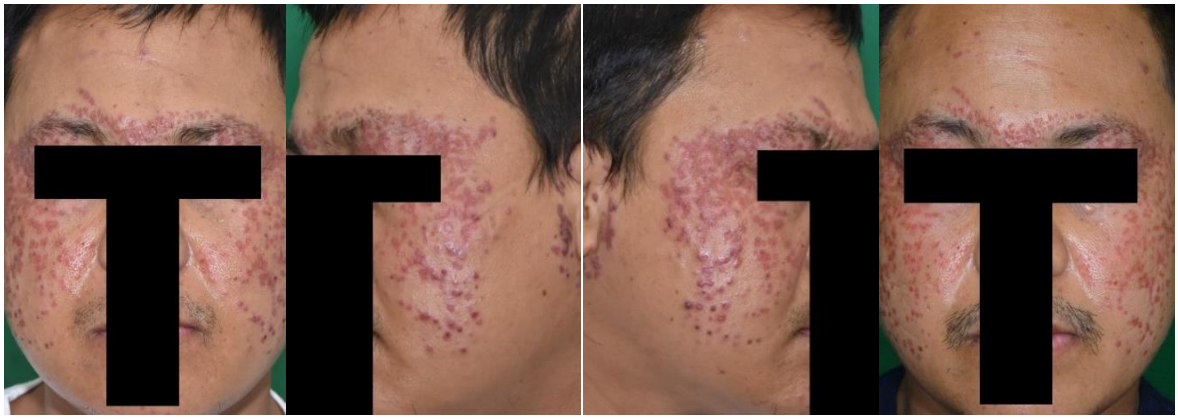

Figure 1A Figure 1B Figure 1C Figure 1D

Figure 1 Multiple perifollicular papules on both periorbital areas and cheeks (1A, 1B, 1C). Lesions improve after a month of treatment (1D).

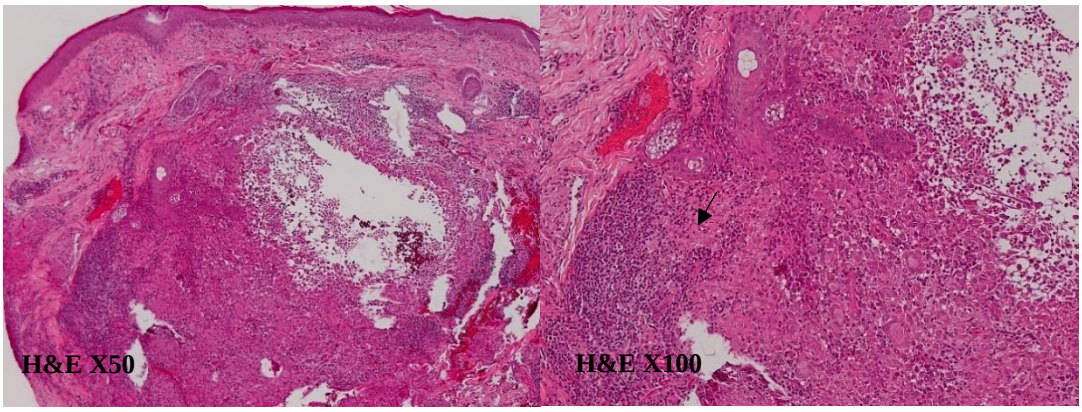

Figure 2A Figure 2B

Figure 2 Mild acanthosis of the epidermis and mixed inflammatory cell infiltration. There was a perifollicular non-caseous granulomatous formation (arrow), adjacent to the hair follicle. (2A,2B)

Discussion

Lichen scrofulosorum (LS) is a rare form of cutaneous tuberculosis. It is classified in the tuberculids group which is caused by an immune hypersensitivity reaction to *Mycobacterium tuberculosis* infection at other sites[1,2]. The incidence of LS is not known due to a few studies. A single-centered, retrospective study in India found 15.2% of this condition in patients with cutaneous tuberculosis[4] and more than 80% in patients younger than 15 years old[2,3]. The cutaneous manifestations are asymptomatic monomorphic groups of tiny follicular, perifollicular, or lichenoid papules with fine scales. The common locations are the trunk and proximal extremities[1,3,4]. The less common sites are the face and external genitalia which were found at only 5% and 3.6% respectively[4]. The lesions of LS mimic follicular disorders. Thus, the differential diagnosis includes keratosis pilaris, lichen spinulosus, lichen nitidus, and lichenoid sarcoidosis[1,5]. Lupus miliaris disseminatus faciei is also considered especially for those

with face and eyelids lesions[1,6]. We discussed the key differences in Table 1.

Although histopathology and culture are needed for diagnosis[7], dermoscopy can also help to differentiate LS from other follicular lesions. One of the characteristics is monomorphic perifollicular round dots with central brown follicular plugs. Peripheral hyperpigmentation and fine scales can also be seen[8]. A histopathological examination shows perifollicular epithelioid cell granulomas with giant cells in the upper dermis. The caseous necrosis is usually absent. Mycobacterial culture is a gold standard for the diagnosis of cutaneous tuberculosis but its sensitivity is very variable. It requires a few months to interpret, so molecular technique like polymerase chain reaction (PCR) is very useful. The tuberculin skin test (TST) has a sensitivity and specificity of 58.9% and 62.5% respectively. TST is strongly positive in tuberculids[7,9]. Quantiferon-TB Gold test which measures the T-cell release of IFN-γ is beneficial in a patient with a history of previous BCG vaccination. Interferon-Gamma Release Assays have a high sensitivity of about 80%. Screening for HIV infection is recommended by the Centers for Disease Control and Prevention (CDC) in all TB patients. Furthermore, an investigation for the underlying primary organ of mycobacterial infection which is common in pulmonary, lymph nodes, and bone should be done in all patients who have symptoms [4,7,10]. Chest X-ray is suggested for all cutaneous tuberculosis patients [7].

Table 1 Comparison of LS and LMDF

	LS	**LMDF**[6,11]
Clinical manifestations	Multiple tiny follicular, perifollicular, or lichenoid papules with fine scales. Trunk, proximal extremities	Discrete, erythematous dome-shaped papules Central face, eyelids
Histopathology	Perifollicular epithelioid non-caseous granulomas with giant cells in the upper dermis.	Dermal epithelioid granuloma with caseous necrosis
Dermoscopic finding	Perifollicular round dots with central brown follicular plugs. Peripheral hyperpigmentation and fine scale	Follicular plugs, telangiectasias, and white structures in late lesions
Age	Children and adolescent	Young adult
Symptom	Asymptomatic	Asymptomatic
Investigation	skin biopsy AFB: negative skin biopsy PCR: usually negative TST: positive Interferon-gamma release assay: usually positive	skin biopsy AFB: negative skin biopsy PCR: negative TST: negative Interferon-gamma release assay: negative
Treatment	Antituberculosis therapy	Tetracycline, systemic corticosteroid
Systemic symptom	Tuberculosis infection: lung, lymph nodes, bones	-
Prognosis	Chronic Heal with no scar	Self-limited, Resolved within 15 months Generally present

Abbreviations: LS, Lichen scrofulosorum, LMDF, Lupus miliaris disseminatus faciei.

The first-line treatment of LS is antituberculosis therapy. The Infectious Diseases Society of America (IDSA) and CDC recommend two months for rifampin, isoniazid, ethambutol, and pyrazinamide, followed by four months of isoniazid and rifampin. The clinical responds to therapy approximately 4 - 6 weeks with a complete resolution by 1 - 5 months. The lesions usually resolve without scars[2,7].

Our patient presented with multiple perifollicular papules on both periorbital areas and cheeks for four months without systemic symptoms. The skin biopsy showed mixed inflammatory cell infiltration with foci of non-caseous granulomatous formation. AFB turned out to be negative. Mycobacterial culture appeared negative for the entire three months. However, the PCR test was positive for mycobacteria. There are a few cases of lichen scrofulosorum that also had positive PCR mycobacterial in skin biopsy. Because LS is a paucibacillary infection, molecular diagnosis techniques can be beneficial to identify mycobacterial DNA in the tissues[12]. The TST is useful for diagnosing tuberculids, but was not tested in this case. Interferon-gamma release assay is another laboratory test that has high sensitivity but is reported in the literature with both positive and negative results[12]. The quantiferon-TB Gold test in the patient is negative. Chest X-ray found no abnormality. The patient was, then, diagnosed with lichen scrofulosorum. Clinical improvement was noted after a month and resolved after six months of treatment.

References

1. Kaul S, Kaur I, Mehta S, Singal A. Cutaneous tuberculosis. Part I: Pathogenesis, classification, and clinical features. J Am Acad Dermatol 2022;S0190-9622.
2. Rajendiran R, Bolia R, Khuraijam S, Singh A. Lichen Scrofulosorum: Cutaneous Manifestation of Tuberculosis. J Pediatr 2021;239:246-7.
3. Singal A, Bhattacharya SN. Lichen scrofulosorum: a prospective study of 39 patients. Int J Dermatol 2005;44:489-93.
4. Singal A, Kaur I, Pandhi D, Gandhi V, Jakhar D, Grover C. Clinico-epidemiological profile of lichen scrofulosorum: a 22-year, single-center, retrospective study. Int J Dermatol 2021;60:1278-84.
5. Kajal NC, Prasanth P, Dadra R. Lichen scrofulosorum: An uncommon manifestation of a common disease. Int J Mycobacteriol 2020;9:313-5.
6. Seo J-I, Shin MK. Lupus miliaris disseminatus faciei versus granulomatous rosacea: A case report. Case Reports in Dermatology 2021;13:321-9.
7. Kaul S, Jakhar D, Mehta S, Singal A. Cutaneous tuberculosis. Part II: Complications, diagnostic workup, histopathological features, and treatment. J Am Acad Dermatol 2022:S0190-9622.
8. Jassi R, Yadav A, Chander R. Dermoscopy of Lichen Scrofulosorum. Indian Dermatol Online J 2020;11:876-7.
9. Joshi HS, Zacharia A, Warrier A. Lichen scrofulosorum. BMJ Case Rep 2014;2014.
10. Vashisht P, Sahoo B, Khurana N, Reddy BS. Cutaneous tuberculosis in children and adolescents: a clinicohistological study. J Eur Acad Dermatol Venereol 2007;21:40-7.
11. Dudani P, Mehta N. Dermoscopy of Lupus Miliaris Disseminatus Faciei Lesions in Different Stages of Evolution. Dermatol Pract Concept 2022;12:e2022017.
12. Predescu T, Mărgăritescu I, Giurcăneanu C, Mihai MM, Forsea A-M. Lichen scrofulosorum - A rare form of cutaneous tuberculosis-case report. Dermatovenerologia 2015;60:19-30.