

Lichen scrofulosorum: An Uncommon Manifestation of a Common Disease

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Abstract

Tuberculid is a cutaneous immunologic reaction to the presence of tuberculosis (TB), which is often occult, elsewhere in the body or their fragments released from a different site of manifest or past tuberculous infection. These eruptive lesions are due to hematogenous dissemination of bacilli in a host with a high degree of immunity against *Mycobacterium tuberculosis*. Although rare, these specific lesions are important diagnostic markers of TB. Lichen scrofulosorum (LS) is one of the recognized tuberculids, usually seen in children and young adults. We report a female who was diagnosed with LS and was treated appropriately. This case report highlights the uncommon, easily misdiagnosed but readily treatable case of LS and emphasizes its early diagnosis, detection, and treatment of otherwise an occult systemic TB in young patients.

Keywords: Biopsy, cutaneous tuberculosis, hypersensitivity, tuberculid

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INTRODUCTION

The disease tuberculosis (TB) is perhaps as old as humankind, with evidence of the disease being found in the vertebrae of the Neolithic man in Europe and Egyptian mummies. However, it was not until 1882 that Robert Koch discovered the causative agent as *Mycobacterium tuberculosis*. It has been estimated that the genus *Mycobacterium* probably causes more suffering for humans than all other bacterial genera combined.^[1] The current resurgence of TB is attributed to the acquired immune deficiency syndrome epidemic, emergence of drug-resistant *M. tuberculosis* strains, poverty, immigration, and inadequate TB control programs.^[2,3] The estimated TB incidence in India is 27 lakhs^[4] Cutaneous TB is a relatively uncommon, comprising 1%–1.5% of all extrapulmonary TB manifestations, which manifests only in 8.4%–13.7% of all TB cases. In another study, the prevalence of cutaneous TB was 0.26% with the most common type of cutaneous TB was lupus vulgaris. The second most common type was scrofuloderma, followed by TB verrucosa cutis, while tuberculids were rarest.^[5]

A tuberculid is a cutaneous immunologic reaction to the presence of TB, which is often occult, elsewhere in the body. Although tuberculids are rare in Western countries,

they are important disorders in developing nations, where 95% of all cases of TB occur. A wide range of skin disorders have been interpreted as tuberculids in the past. However, currently, only three entities are regarded as true tuberculids:

1. Papulonecrotic tuberculid,
2. Lichen scrofulosorum (LS), and
3. Erythema induratum.^[6]

LS is a rare tuberculid, presenting as lichenoid papules in children and young adults with TB. Patients with LS have a strongly positive tuberculin reaction; concurrent tuberculous involvement of the lymph nodes, bones, or other organs; and an excellent response to treatment with antituberculous drugs.^[6] These features demonstrate a strong link between LS and TB. LS usually accompanies active TB, which can either be in an early or late stage.

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CASE REPORT

A 30-year-old female came with the complaint of skin rashes over the trunk and loin region for past 2 years. There was no history of fever, cough, or any other systemic symptoms. Past and family history did not reveal any significant illness. The patient is a migratory laborer working in a construction site. Physical examination found bilateral enlargement of the supraclavicular nodes, <1 cm in size, nontender, firm, and matted. Cutaneous examination revealed multiple, grouped, erythematous, 2–3 mm, lichenoid follicular and extrafollicular papules over both anterior and posterior aspects of the trunk and loin [Figure 1].

Cardiovascular, respiratory, abdominal, and central nervous system examinations were unremarkable. Laboratory investigations found a normal complete blood cell count, renal and hepatic functions, urinalysis, and chest radiograph. Erythrocyte sedimentation rate was 52 mm in the first hour, and the Mantoux test was reactive, with an induration of 17 mm. Fine-needle aspiration cytology from an enlarged cervical lymph node showed a granulomatous infiltrate consisting of epithelioid cells, Langhans-type giant cells, and few lymphocytes, consistent with tuberculous lymphadenitis. Stain and culture for acid-fast bacilli were negative. Skin biopsy showed perifollicular tuberculoid granulomatous inflammation consisting lymphocyte and epithelioid cells. Another similar focus was present in mid-dermis around a vessel [Figure 2a-c]. Tubercle bacilli could not be detected on acid-fast staining, and a culture for *M. tuberculosis* was sterile.

A standard 6-month regimen of antituberculous therapy (ATT) with first-line drugs (rifampicin, isoniazid, ethambutol, and pyrazinamide for 2 months, followed by rifampicin, isoniazid, and ethambutol for the next 4 months) was instituted and the patient is currently on follow-up.

DISCUSSION

LS is a rare tuberculid, first recognized by Hebra in 1868,^[7] that occurs mostly in children and young adults. The eruption

consists of tiny, perifollicular, lichenoid papules arranged in groups. These are usually skin colored, but at times may be yellowish or reddish brown. The papules have a flat top or there might be a minute horny spine or fine scale on their surface. The size of an individual papule rarely exceeds 5 mm. They are mainly found on the abdomen, chest, and back and involute after several months, leaving no scars. Histopathology reveals noncaseating granulomas in the papillary dermis usually surrounding a hair follicle or a sweat gland. The granuloma is composed of epithelioid cells with a few Langhans giant cells and a narrow rim of lymphocytes. Tubercle bacilli are not seen in the pathologic sections nor can they be cultured from the skin biopsy material.^[8]

Differential diagnosis includes all asymptomatic follicular lesions with a tendency to group, including keratosis pilaris, lichen spinulosus, lichen nitidus, pityriasis rubra pilaris, and lichenoid sarcoidosis. Keratosis pilaris is usually generalized, noninflammatory, and there is little tendency toward grouping. Lichen spinulosus is not difficult to differentiate due to the presence of spinous projections over the lichenoid papules. Lichen nitidus has shiny, generalized lichenoid papules with characteristic involvement of male genitalia. Pityriasis rubra pilaris and lichenoid sarcoidosis can be easily differentiated by their characteristic histopathology.

The pathogenesis of tuberculids is poorly understood. All tuberculids are thought to be due to hematogenous dissemination of bacilli in a person with a moderate or high degree of immunity against *M. tuberculosis*.^[6] However, it is not possible to detect the tubercle bacilli in tuberculids, either because they are present in a fragmented form or because they have been destroyed at the site of tuberculids by immunologic mechanisms. Mycobacterial DNA can be detected in some lesions using the polymerase chain reaction technique.

Previously, LS was reported in association with some forms of extrapulmonary TB, especially that of the lymph nodes



Figure 1: Multiple, grouped, erythematous, 2–3 mm, lichenoid follicular and extrafollicular papules over loin (with biopsy suture marks)

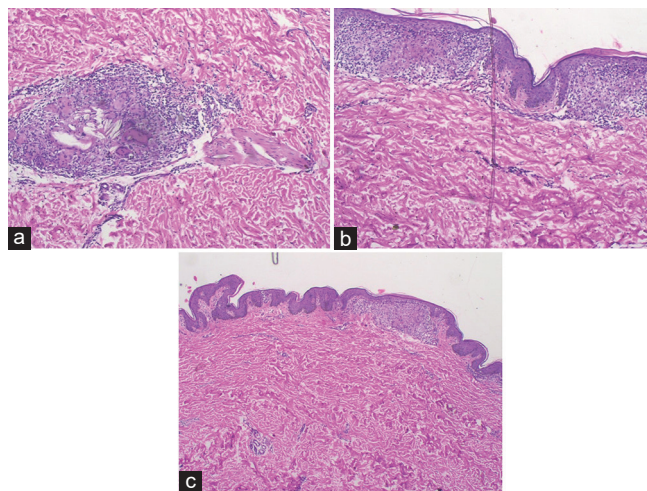


Figure 2: (a-c) Skin biopsy showing tuberculoid granulomatous inflammation consisting of lymphocyte and epithelioid cells in the perifollicular area and mid-dermis around a vessel

and bones, or both, but recently, it has also been reported with pulmonary TB and with *Mycobacterium avium* infection.^[9] It has also been reported to occur after Bacille Calmette Guerin (BCG) vaccination. These forms of extrapulmonary TB are characterized by a high tuberculin positivity, indicating a greater degree of tissue hypersensitivity. Other forms of extrapulmonary disease, such as miliary or meningeal TB, seen in very young children with a less effective host immune response, have never been reported to be associated with LS. This confirms the pathogenic basis of LS as a delayed Type IV hypersensitivity response. Similarly, LS is more common in children, probably due to a higher degree of tissue hypersensitivity to *M. tuberculosis* as compared to adults. A greater predisposition to develop extrapulmonary TB in children and a gradual decrease in delayed hypersensitivity with advancing age are the likely factors influencing the age distribution of this eruption.

In a study by Varshney and Goyal,^[10] the overall incidence of cutaneous TB was 0.7% (131 of 18,720 outpatients). The most common variants seen were scrofuloderma (36.5%), lupus vulgaris (31%), TB verrucosa cutis (12.9%), LS (11.4%), papulonecrotic tuberculids (3.8%), erythema nodosum (2.2%), and erythema induratum of Bazin (1.5%). Cutaneous TB may also be a direct manifestation of underlying TB as seen in the case reported by Al Zayyani *et al.*,^[11] in which chronic tuberculous epididymo-orchitis presented as scrotal ulcers. Among the three tuberculids, the incidence of LS was found to be the lowest (2%) in a large study conducted in Hong Kong. Singal and Bhattacharya^[12] studied 39 cases of LS, of which 72% had an underlying focus of TB, whereas 28% had no identifiable focus of TB.

CONCLUSION

A high index of suspicion and awareness about this entity is needed for diagnosis. As these lesions are subtle and asymptomatic, neither the patient nor the physician may give it enough importance and thus may miss the diagnosis. These lesions may be ignored even in patients with established TB at another site. Lack of awareness about this entity may also lead to a wrong diagnosis by the treating physician and may be a reason why this condition has been considered rare. Patients presenting primarily with LS should be extensively and thoroughly screened for a possible occult tubercular focus. These patients have an excellent response to the ATT

irrespective of the presence or absence of an associated tubercular focus. Widespread awareness and the ability to diagnose this entity can prove to be crucial in the early diagnosis, detection, and treatment of otherwise an occult systemic TB in young patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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