Case Report

Lupus Vulgaris and Lichen Scrofulosorum with Disseminated Tuberculosis

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Abstract

Cutaneous tuberculosis (CTB) represents only 1%–2% of extrapulmonary forms of tuberculosis (TB). CTB can present in the isolation or coexist with pulmonary and disseminated forms of TB. Pathologically confirmed lupus vulgaris (LV) coexisting with lichen scrofulosorum with disseminated TB is presented and discussed. A 12-year-old boy presented with 2 years history of slowly progressive facial plaque and multiple skin colored papules on the neck. Tuberculin skin test was positive with a reading of > 20 mm. Histopathology revealed ulcerated areas with impetiginisation and keratopurulent debris. The entire skin showed pseudoepitheliomatous hyperplasia with numerous granulomas in the superficial dermis consisting of Langhans-type multinucleated macrophages. Lesions responded to anti-tuberculosis therapy (ATT) with residual facial scar. LV and lichen scrofulosurum are two forms of CTB which rarely occurs together. This case is being presented to highlight the occurrence of this rare presentation and the need to institute ATT to prevent scarring.

Keywords: Cutaneous tuberculosis, lichen scrofulosurum, lupus vulgaris

Introduction

Cutaneous tuberculosis (CTB) is an infectious disease caused mainly by *Mycobacterium tuberculosis* (MTB) rarely by *Mycobacterium bovis* or Bacille Calmette-Guérin (BCG) vaccine. The route of transmission could be exogenous, endogenous, or from autoinoculation from contiguous sites.[1]

CTB occurs infrequently, despite a high and increasing the prevalence of tuberculosis (TB) worldwide and it accounts for approximately 1%–2% of the total number of extra-pulmonary cases of TB.[2,3] The diagnosis of these skin disorders is challenging, as they simulate many otherwise more common dermatological disorders. Definitive diagnosis by microbiological confirmation is difficult, there is also paucity of histopathology and polymerase chain reaction (PCR). Lupus vulgaris (LV) is a chronic, progressive, pauci-bacillary form of CTB occurring in patients with a moderate immunity and high degree of tuberculin sensitivity.[4] Lichen scrofulosorum (LS) is a rare tuberculid (immune hypersensitivity response to the presence of MTB antigens) commonly affecting children and young adults with underlying mycobacterial disease.[5]

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Case Report

A 12-year-old boy presented to the dermatology clinic of the Federal Teaching Hospital, Abakaliki with slowly progressive, reddish-brown, scaly, crusted plaque involving the nose and the upper lip of 2 years duration. There was history of intermittent low-grade fever; there was no history of cough or weight loss. There was no preceding history of trauma. About 6 months before presentation, he developed nonitchy papules on the neck and trunk. The presence of the facial plaque had affected his performance/attendance to school, as he was embarrassed by it. There was no history of contact with anyone with TB, though his father had worked as a technician in a microbiology laboratory of a hospital.

General examination showed that he was mildly pale and had few nontender, discrete submental lymph node enlargement;

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the largest measured 2 cm × 2 cm. Cutaneous examination revealed erythematous to flesh-colored, thick, and crusted annular plaque over the face extending from the nose to the upper lip [Figure 1] there were numerous flesh-colored papules on the neck and trunk [Figure 2].

An initial diagnosis of cutaneous TB consisting of plaque type LV on the face and LS was made. The differential diagnoses considered were lupus pernio (sarcoidosis) and deep fungal mycoses (for LV); miliaria rubra, lichen nitidus, and keratosis pilaris for LS.

Mantoux test showed a >20 mm induration after 48 h as shown in Figure 3. Patient had, however, received BCG vaccination at birth as part of the National Program of Immunization. The erythrocyte sedimentation rate was 104 mm in the 1st h. Full blood count showed packed cell volume of 34%; white blood cell count of 6.1 × 10^9/L with differential of neutrophil 53%, lymphocyte 41%, eosinophil of 3%, and monocytes 4%. Retroviral screening test was negative. The chest radiograph revealed bilateral hilar opacities [Figure 4]. A skin biopsy was taken from the facial lesions with the aid of a 5 mm punch set for histopathology. Histopathology showed ulcerated areas with impetiginisation and keratopurulent debris. The entire skin showed pseudoepitheliomatous hyperplasia with numerous granulomas in the superficial dermis made up of Langhans-type multinucleated macrophages. There was also dense infiltrate of neutrophils and eosinophils in the dermis [Figures 5 and 6].

Following results of investigations, a diagnosis of disseminated TB (cutaneous TB, TB lymphadenitis, and pulmonary TB) was made and patient was referred to the directly observed treatment short centre of the hospital for anti-tuberculosis therapy (ATT). He received intensive phase of therapy consisting of 600 mg of rifampicin, 300 mg of isoniazid, 1 g of pyrazinamide, and 800 mg of ethambutol and the continuation phase was with 600 mg of rifampicin and 300 mg of isoniazid. He also received 50 mg of pyridoxine to ameliorate isoniazid-induced peripheral neuropathy.

Follow-up visit after 2 months of intensive therapy showed remarkable interval improvement in the appearance of lesions of LS and LV, though with residual scar tissue

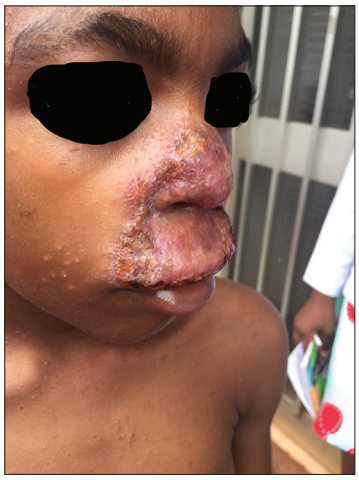

Figure 1: Lesions of lupus vulgaris before treatment

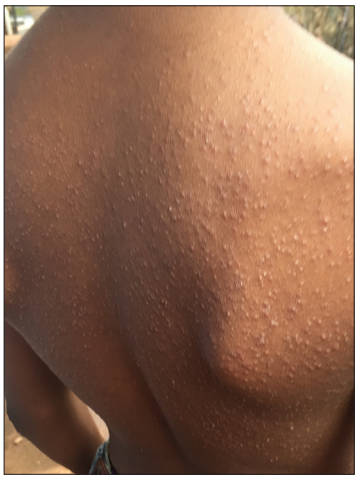

Figure 2: Flesh coloured peri-follicular papules on the posterior aspect of the trunk (lichen scrofulosorum) on the posterior aspect of the trunk

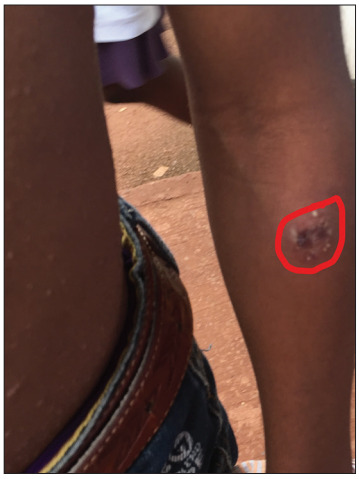

Figure 3: Site of tuberculin skin (Mantoux) test

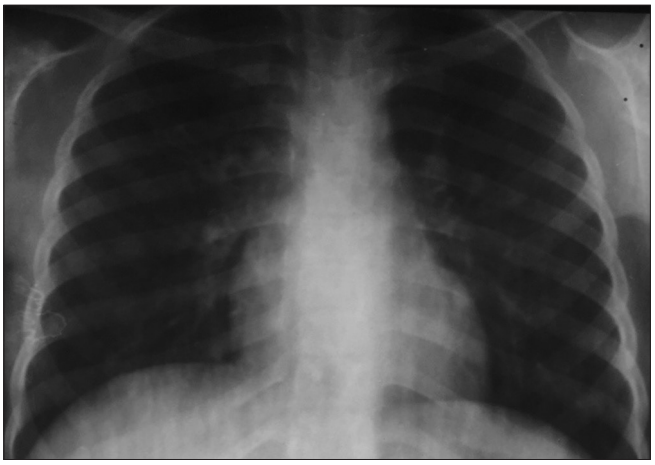

Figure 4: Chest radiograph with bilateral hilar opacities

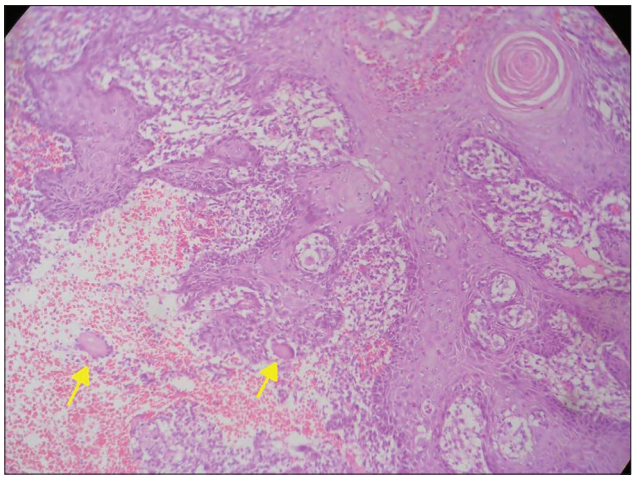

Figure 5: Numerous Langhan's giant cells (multi-nucleated with numerous peripherally located nuclei) in the dermis (yellow arrows)

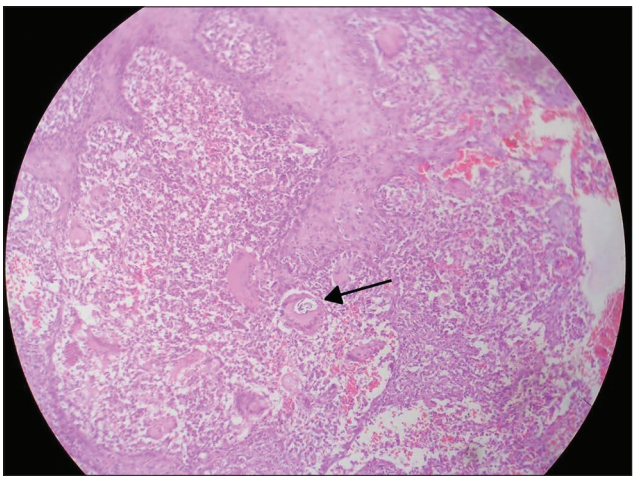

Figure 6: Langhan's cells containing calcified material (schaumann bodies) in their cytoplasm (black arrow)

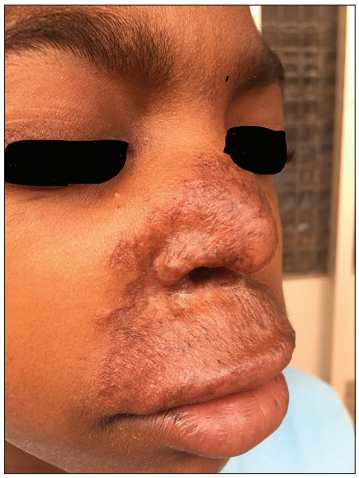

Figure 7: Lupus vulgaris after intensive phase of treatment

on the affected site of LV lesion [Figure 7]. There was also a significant resolution of regional lymph node swellings and minimal improvement of the chest radiographic findings.

Discussion

The prevalence of CTB ranges from 0.07% (Hong Kong), 0.1%–0.9% (India) and 2% in North Africa.[1,2,6-8] A 5-year review of clinic records from Ibadan, South Western Nigeria revealed a rare occurrence of CTB.[9] CTB can be classified into localized disease: primary tuberculous chancre, TB verrucosa cutis, LV, and smear negative scrofuloderma; disseminated disease which includes disseminated TB, tuberculous gumma, orificial TB, miliary CTB, and tuberculids comprising papulonecrotic tuberculids, erythema nodosum, and LS.[6]

LV is the most common clinical form of CTB in adults and the second most common form in children, (after scrofuloderma), with a female-to-male ratio of 2–3:1.[5] LV is a chronic form of CTB seen in those with high degree of tuberculin sensitivity.[1] Eighty percent of the lesions occur on the head and neck; other sites include lower extremities and the gluteus.[10-12] LV starts as a soft, brownish-red papule, or nodule that gradually expands by involution in one area with expansion in another, gradually progressing over a period of many years to form a well-defined skin-colored to erythematous plaque (as was seen in this patient), invariably he had disseminated TB involving the lymph nodes, the lungs in addition to localized CTB (LV), and tuberculid (LS).

The morphological variants of LV are classical plaque type, ulcerative or mutilating, vegetating, tumor-like, and papulonodular.[13] Histopathology of LV lesions reveals typical epithelioid granulomas in the upper dermis, with lymphocytes and Langhans giant cells in up to 80% of the cases, with the remainder showing nonspecific changes. Histology in this case was in keeping with the classical plaque type of LV.

LS, on the other hand is a tuberculid, initially described by Hebra in 1860.[14] A large study carried out in Hong Kong revealed a low incidence of LS; it was found to be the lowest (2%), while the other tuberculids had incidences of 93.3% for erythema induratum of Bazin and 4.7% for papulonecrotic tuberculid.[8] LS is clinically characterized by tiny, skin-colored, and perifollicular papules arranged in groups usually with a smooth surface, occasionally spiny projections with fine scales may be seen. Histology shows noncaseating, epithelioid cell granulomas in the upper dermis and around dermal appendages. Other disorders to consider include keratosis pilaris, lichen spinulosis, lichen nitidus, pityriasis rubra pilaris miliaria, and lichenoid sarcoidosis.[15,16] Tuberculin skin test (TST) is strongly positive, thereby aiding in the diagnosis of LS, the index patient had TST reading of >20 mm. In addition, PCR can detect mycobacterial antigens in lesional skin.[17]

Despite the prevalence of TB, especially pulmonary TB and other types of extrapulmonary TB, cutaneous TB is not a frequent finding in such patients. In addition, LV and LS rarely coexist. The only recorded report of LS and LV occurring together was by Dash *et al*. in India occurring in one of the 21 patients with tuberculids.[18] LS and LV show excellent

response to ATT, though the sequelae of scarring of LV lesion may ensue requiring cosmetic surgery.

Conclusion

LV and LS are two forms of CTB which rarely occurs together. This case is being presented to highlight the need for an increase in the index of suspicion as LV and LS both have several mimicking differential diagnoses and can occur in isolation or as part of disseminated TB. Prompt identification of these conditions (especially the facial plaque type of LV) and early treatment will alleviate psychosocial morbidity associated with scarring. Early therapy will invariably prevent the loss of self-esteem which negatively impacts other aspects of social and economic life including loss of school time with attendant poor performance in school, as occurred in the index case.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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