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Case Report

Lupus vulgaris of centrofacial area – a peculiar presentation

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Abstract Lupus vulgaris is a cutaneous manifestation of *Mycobacterium tuberculosis* infection. It assumes various clinical forms including plaque, ulcerative, hypertrophic, vegetative, papular and nodular type. We here present an unusual case of a ten-year-old girl diagnosed as hypertrophic lupus vulgaris of centrofacial area including nose. Histopathological examination and bacteriological studies were carried out to establish the diagnosis. Unlike plaque type, hypertrophic variety is quite uncommon and only few cases have been reported. Its peculiar appearance on the face which is hardly described in previous literatures and well response to antitubercular therapy prompted us to report it.

Key words
Lupus vulgaris, centrofacial, hypertrophic, ATD therapy

Introduction

Cutaneous tuberculosis is caused by *Mycobacterium tuberculosis*, *M. bovis* or rarely *Bacillus Calmette – Guerin* (BCG), an attenuated strain of *M. bovis* employed for vaccination.[1] It has a wide array of clinical spectrum depending upon the mode of infection, immune status of the host or history of previous sensitization to tubercle bacilli.[2] Lupus vulgaris is a chronic, progressive form of cutaneous tuberculosis affecting persons having moderate to high immune status.[2] It may arise as contiguous extension from an underlying tubercular focus (like bone, joint, lymph node), by hematogenous or lymphatic spread. Sometime it develops following primary exogenous inoculation like trauma or tattooing,[3] in the scar of scrofuloderma[4] or at the site of BCG vaccination.[5,6] Classical plaque type lupus vulgaris is not always seen in facial or nasal area and seldom it assume different unusual forms with disfigurement. This hypertrophic variety is pretty uncommon and few cases have been reported so far. Hence we present such an unusual case of lupus vulgaris of nose and central face with grotesque appearance.

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Case Report

A 10-year-old girl from rural Bengal presented with a painless, greyish-yellow, stony hard, cauliflower like, bizarre shaped, tumoriform keratotic mass over mid portion of the face including the nose for last five years. The lesion had started as a small asymptomatic plaque over the nose and was treated by different doctors by different therapies without proper diagnosis and specific treatment. The lesion had gradually progressed to involve the entire nose and encroached towards both cheeks and mid-forehead area to assume the present state. On examination, the patient had a hypertrophic and verrucous plaque with some horn like projections on its surface. The plaque obscured the entire nose extending towards both cheek and mid-forehead area. The surface had several deep fissures and reddish-yellow crusts over the nasal tip and root of the nose. The border showed atrophy in some areas (**Figure 1**). The affected nose led to partial

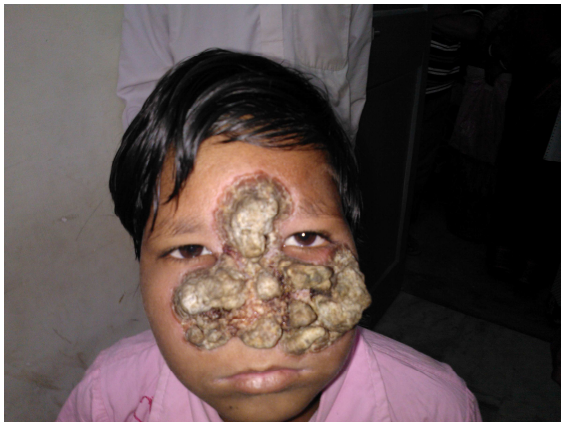

Figure 1 Pretreatment photograph with large thick yellowish protuberant plaque in central area of the face.

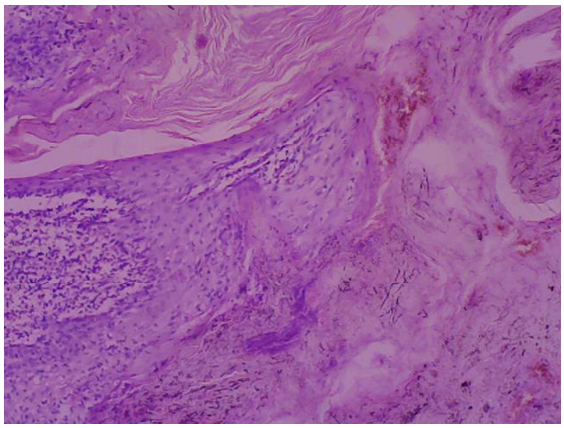

Figure 2 Skin biopsy showing epidermal hyperkeratosis, acanthosis and papillomatosis (Original magnification H & E × 40).

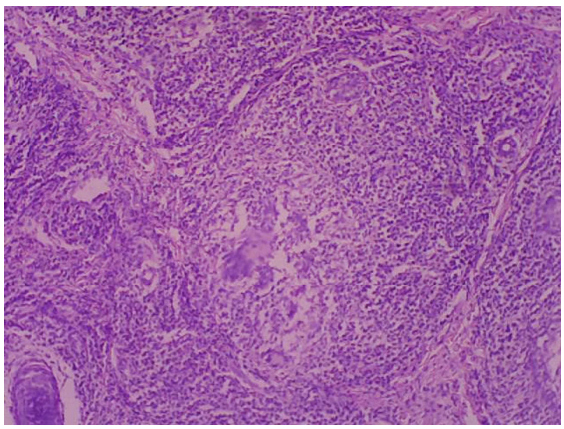

Figure 3 Dermis shows epithelioid cell granuloma and Langerhans cell giant cell (original magnification H & E × 40).

obstruction of the nostrils causing difficulty in breathing. Oral cavity showed no growth on the hard palate and upper gingival area. She did not have any cervical lymphadenopathy. There were no other significant cutaneous findings and general examination was normal.

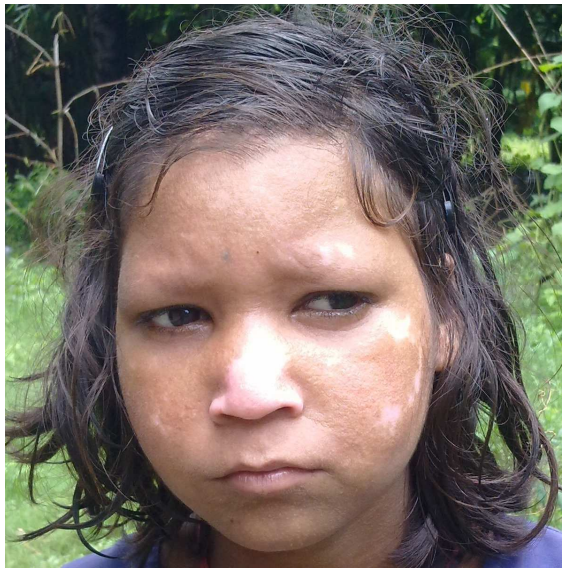

Figure 4 Posttreatment photograph with mild scarring and depigmentation.

There was no personal or family history of pulmonary tuberculosis or any earlier history of trauma at same area. Based on these clinical features we considered the differential diagnosis of cutaneous tuberculosis (lupus vulgaris), chromoblastomycosis or deep fungal infection.

Radiological examination of paranasal sinuses showed no bony erosion or deformity. On chest X-ray no abnormality was detected. Laboratory tests were also carried out. Sputum for acid-fast bacilli (AFB) was negative. Mantoux test was positive with erythema and induration of 18 mm and 17 mm, respectively. VDRL and ELISA for HIV were non-reactive. PAS stain was negative. Fungal culture from the lesional tissue showed no growth. Ziehl-Neelsen stain demonstrated no tubercle bacilli. Culture on Lowenstein-Jensen medium showed no growth. Histopathological examination of the lesion revealed epidermal hyperkeratosis, acanthosis and papillomatosis. Dermis showed epithelioid cell granuloma and Langhans type giant cells mainly in the upper dermis with chronic inflammatory cells infiltration, predominantly lymphocytes. No caseation necrosis was found. No AFB was found by Ziehl-Neelsen stain nor any fungal elements were demonstrated in the tissue section (**Figures 2** and **3**).

Based on the clinical features and histopathological findings, diagnosis of hypertrophic lupus vulgaris was established. The patients was treated with antitubercular drug therapy (rifampicin + INH + ethambutol + pyrazinamide for 2 months and then rifampicin + INH for 4 months). Dramatic response to treatment was noted with complete clearance. Only mild depigmentation and scarring were present. No significant adverse drug reaction was seen during the therapy (**Figure 4**).

Discussion

Lupus vulgaris is a chronic, progressive form of cutaneous tuberculosis which occurs in persons having moderate to high level of immunity. Various clinical types depending upon the local tissue response to the mycobacterial infection have been described including plaque, ulcerative and mutilating, vegetative, hypertrophic or tumor-like, papular and nodular type.[2] A classical lesion develops as a small, reddish-brown, soft plaque which gradually enlarges and extends peripherally with evidence of healing in one side and appearance of new lesions on other side. The surface is studded with multiple erythematous nodules. On diascopy, these nodules show as diagnostic apple jelly color. The lesions are usually solitary but two or more sites may be involved at a time. Disseminated form with associated active pulmonary tuberculosis[7] or sporotrichoid variety of lupus vulgaris has also been reported.[8] Patients not always present with classical plaque type but atypical forms are also pretty common.

The classical type manifests as plane plaque having serpiginous border with polycyclic configuration and smooth or psoriasiform scaling surface. It often shows areas of atrophy in one site and active lesions in other site. The edge is hyperkeratotic. Ulcerative and mutilating forms present as predominant ulceration with crusting and scarring. Deep tissues and cartilage may be invaded causing deformity. Vegetating forms are characterized by gross infiltration with ulceration, necrosis and scarring. When mucous membrane and cartilage are involved, extensive destruction and disfigurement occurs. Tumor-like or hypertrophic forms are characterized by soft tumoriform nodules or epithelial hyperplasia with formation of hyperkeratotic mass. It is often associated with marked lymphedema and vascular dilatation. Multiple lesions occurring in disseminated pattern characterize papular and nodular form of lupus vulgaris.

Our case resembled hypertrophic variety of lupus vulgaris, but stony hard consistency with horny projections was the peculiarity and gave it an unusual appearance. This stony hardness may be due to continuous exposure of sunlight for a long duration. Moreover it did not have any associated lymphedema or vascular dilatation.

Lupus vulgaris has varied histopathological features according to the clinical type of presentation. The epidermis may be atrophic, acanthotic, ulcerated or grossly hyperkeratotic showing pseudoepitheliomatous hyperplasia. Upper dermis shows tuberculoid granulomas consisting of epithelioid cells and multinucleated giant cells mainly of Langhans type with sparse or no central caseation within the tubercle[9] along with abundant lymphocytic infiltration. Z-N stain hardly demonstrates small number of tubercle bacilli and polymerase chain reaction (PCR) is needed for detection of mycobacterial DNA in such cases.[10]

Lupus vulgaris is slowly progressive in nature and if left untreated causes scarring, keloid formation, fibrosis and contracture, extensive tissue destruction leading to considerable disfigurement.[11] In long-standing cases development of squamous cell carcinoma has also been reported.[12] A trial with antitubercular therapy is considered for confusing cases and desired response is usually attained within four to six weeks. Standard four drug treatment with rifampicin, isoniazid, pyrazinamide and ethambutol for first two months followed by

rifampicin and isoniazid for next four months fortunately shows satisfactory result. Surgical intervention and electrocauterization are reserved for residual lesions or correction of deformities.

Fortunately our case responded well to ATD and the tubercular mass cleared completely after 6 months of therapy with minimal scarring and depigmentation.

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