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Case Report

Lupus Vulgaris : Unusual Presentation on Face

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Lupus vulgaris is a variant of cutaneous tuberculosis. As the disease has potential to mutilate when left untreated, leaving deforming scars and disfigurement, an early diagnosis is of paramount importance. Though the common type is plaque type, rarely mutilating and vegetative forms also are found. A 28 year old female, labourer presented with progressive annular plaque over right side of cheek extending upto right lower lid and ala of nose. There were two satellite plaques near the right side of giant lesion. On diascopy apple jelly nodule was seen. There was no regional lymhadenopathy. Histopathological examination showed many granulomas in upper dermis extending to deep dermis comprising of epitheloid cells with langhans' type of giant cells, lymphocytic infiltration & focal necrosis suggestive of lupus vulgaris. The consequences of failing to make an early diagnosis can be disastrous for the patients, as the progression of the disease can lead to necrosis, destruction of bones and cartilage leading to permanent deformity. Thus it is vital for clinicians to have a high index of suspicion of such atypical forms and take biopsy samples for histological and bacteriological studies.

Keywords : *Mycobacterium tuberculosis*, *Cutaneous Tuberculosis, Lupus Vulgaris*

Introduction

Lupus vulgaris represents a rare manifestation of skin infection by *Mycobacterium tuberculosis.* There is a resurgence of tuberculosis everywhere because of a combination of factors including immigration from endemic countries, HIV pandemic, poverty, etc. as a result, cutaneous tuberculosis remains a clinical and diagnostic problem. *Cutaneous tuberculosis* forms a small proportion of extrapulmonary tuberculosis and out of which, *Lupus vulgaris* is the most common morphological variant of cutaneous tuberculosis accounting for approximately 59% of cases of cutaneous tuberculosis in India. (Khandpur and Reddy 2003, Chakravarti et al 2006, Yates 2010, Ramesh et al 1999, Jain et al 2009). It is the bizarre clinical presentation and involvement of atypical sites, which often lead to inappropriate diagnosis causing significant morbidity. Thus it is vital for clinicians to have a high index of suspicion of such atypical forms and take biopsy samples for histological and bacteriological studies. Anti-tubercular chemotherapy is satisfactory with good result.

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Case

A 28 year old female, labourer presented with progressive non-healing lesion over right side of cheek involving corresponding side of lower lid and side of the nose since 2 months. Lesion was associated with itching & burning sensation.

There was no family or past history of tuberculosis.

On examination a single well defined erythmatous edematous annular plaque of size 7*8 cm was present over right side of cheek extending upto right lower lid and same side of ala of nose with central crusting. There were two satellite plaques near the right side of giant lesion. On diascopy apple jelly nodule was seen. There was no regional lymhadenopathy. Ear Nose and Throat examination was normal. Neurological examination was normal. Montoux test was positive with 24*10 mm of induration. Routine hematological and biochemical examination was

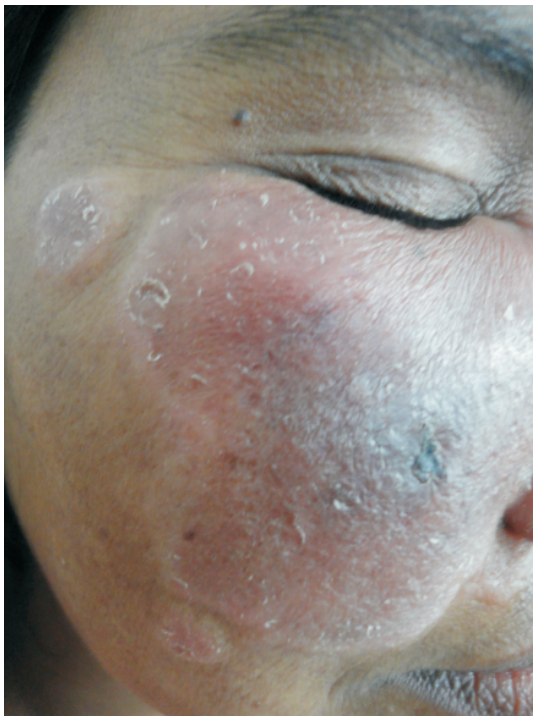

Fig 2 : Two satellite plaques

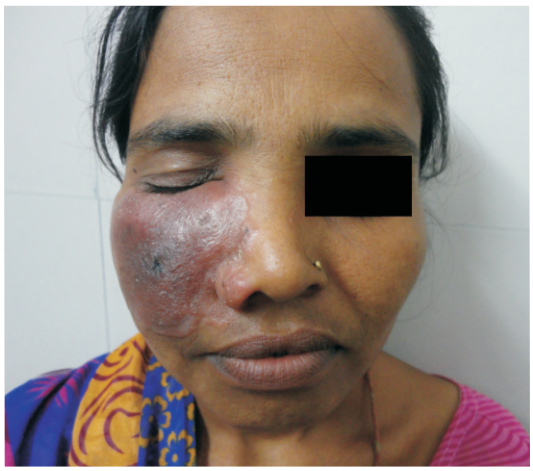

Fig 1 : Well defined erythmatous edematous annular plaque of size 7*8 cm was present over right side of cheek extending upto right lower lid and same side of ala of nose with central crusting.

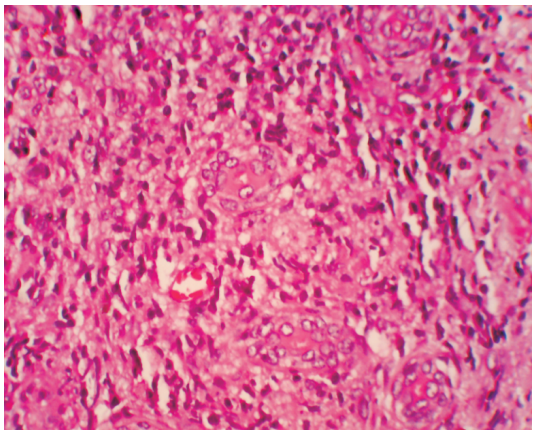

Fig 3 : Many granulomas in upper dermis extending to deep dermis comprising of epitheloid cells with langhans' type of giant cells, accumulation of lymphocytes & focal necrosis suggestive of lupus vulgaris.

within range except raised ESR which was 84mm/hr. Patient was sero-negetive and the chest radiograph didn't reveal any abnormality. Histopathological examination showed many granulomas in upper dermis extending to deep dermis comprising of epitheloid cells with langhans' type of giant cells, accumulation of lymphocytes & focal necrosis suggestive of lupus vulgaris. AKT was started (rifampicin 450 mg/day, isoniazid 300 mg/day, ethambutol 800mg/kg/day and pyrazinamide 1500mg/kg) given for initial 2-months in the intensive phase ollowed by treatment with rifampicin and isoniazid during the continuation phase for next 4-months with no adverse effects noted. Excellent response to treatment was noticed clinically.

Discussion

Lupus vulgaris is acquired exogenously by direct inoculation of the bacilli or endogenously via hematogenous or lymphaticspread from associated tuberculosis of other organs. *Mycobacterium bovis*, atypical Mycobacteria and the BCG Vaccine can cause tuberculosis involving the skin (Kinra et al 2009). The diagnosis of cutaneous tuberculosis is challenging and requires the correlation of clinical finding with diagnostic testing. Sarcoidosis and rosacea may sometimes simulate early lupus vulgaris (Kinra et al 2009). Other conditions like basal cell carcinoma, DLE, leprosy, deep fungal infection can also be considered for differential diagnosis. The determinants of what happens in tuberculosis infection, includes, the virulence of the organism, the size of the inoculum, the route of infection, and the immune status of the patient.

It begins as asymptomatic, infiltrating papules and plaques. The lesions slowly spread and can affect a large area of the body. Typical lesion is a well-demarcated, skin-colored or erythematous plaque. Healing and scarring in one area and activity in another is the hallmark of lupus vulgaris. In children, the lower extremities and gluteal region are the commonly affected sites (Sebastian 2005). Many clinical variants have been described: the classic plaque or the keratotic type, the hypertrophic form, the ulcerative form, the atrophic form and the mutilating form. Of these, the plaque or the keratotic type is the most common form reported.

In our case erythematous plaque with central crusting was present.

In India, lower extremities, especially buttocks are most frequently affected, while the face is the most commonly affected site in Western countries with a frequent affliction of the nose and cheeks. In our case there was facial presentation which is rare in Indian scenario. Frequent localization of the lesions around the nose and on cheeks is attributed to the rich and porous venous plexuses with stasis, cold and hypoxia, impaired fibrinolysis and host defense at a lower temperature, as prevailing in western countries (Findlay 1987). Higher temperatures prevailing in most parts of India may be responsible for relatively low frequency of the facial lesions in comparison to the Western world.

Lupus vulgaris is completely curable. The consequences of failing to make an early diagnosis can be disastrous for the patients, as the progression of the disease can lead to necrosis, destruction of bones and cartilage leading to permanent deformity (Khandpur and Reddy 2003, Chakravarti et al 2006, Kakakhel 1989, Ramesh et al 1999).

The special test like polymerase chain reaction (PCR) for TB can be done to substantiate the diagnosis or wherever the histopathology is inconclusive (Arnold et al 1990). Polymerase chain reaction (PCR) provides rapid, specific and sensitive testing for *M. tuberculosis*. A strongly

positive test is significant. It is now possible to detect drug resistance by molecular means or by using the light-producing enzyme Luciferase, the gene for which has been added to mycobacteriophage (Chakravarti et al 2006), instead of more time consuming culture and sensitivity techniques.

In view of emerging drug resistant strains only multi-drug chemotherapy is now recommended for all forms of tuberculosis, and a strict compliance to the regimen is needed.

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