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Lupus Vulgaris - With Elbow Deformity

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Abstract: ***Lupus vulgaris is a form of cutaneous tuberculosis. It occurs in previously sensitized individuals with a moderate degree of immunity against tubercle bacilli. The lesions are acquired by hematogenous spread from an underlying tuberculosis focusor by direct inoculation. [1] Cutaneous tuberculosis is a rare infection that represents 1 - 1.5% of extrapulmonary tuberculosis whose etiological agents are Mycobacterium tuberculosis, Mycobacterium bovis, attenuated form of Bacillus Calmette - Guerin (BCG vaccine). Here, we present a case of a 76 - year - old male who presented to the OPD with Lupus vulgaris.***

Keywords: Lupus vulgaris, cutaneous tuberculosis (CTB), mycobacterium tuberculosis

1. Introduction

Lupus vulgaris is a chronic, progressive, post - primary, paucibacillary form of cutaneous tuberculosis (TB). It represents reactivation of infectionin people with moderate to high immunity. It is the most common form of cutaneous TB in adults in the Indian subcontinent and South Africa. [2] Females are two to three times affected more than males. It is characterized by groups of 'apple jelly'- colored nodules which is due to the deep dermal infiltration with tuberculous granulomas. Nodules coalesce into large plaques and lead to scarring and distortion. Lupus vulgaris may occur either because of direct extension from an underlying focus or via lymphatic or hematogenous spread.

Clinically lupus vulgaris presents in five patterns: plaque form, ulcerative and mutilating form, vegetating form, tumor like form and papular and nodular form. Plaque form presents in the form of flat plaques with irregular, serpiginous edge. Large plaques show irregular areas of scarring. In ulcerating and mutilating form, scarring and ulceration are present. Crusts form over area of necrosis. Vegetative form is characterized by marked ulceration, extensive infiltration, and necrosis. Tumor like form presents with deep infiltration in the form of soft and smooth grouped nodules or a reddish - yellow plaque. Papular and nodular form presents with multiple lesions occur simultaneously in disseminated form. [3] The complications and comorbidities include scarring, contracture, tissue destruction& malignant changes.

2. Case Report

A 76 - year - old man presented to the outpatient department with multiple scaly plaques over the left elbow for 40 years. Initially it was small and gradually progressed to the present size. He had a history of tuberculosis six years ago and was on anti - tuberculosis therapy but did not complete the treatment. The patient was on ayurvedic treatment 5 years ago and progressed to the present size. There was no history of trauma prior to the onset of the lesion. No history of fever, cough, no loss of sensation, pain over the lesion.

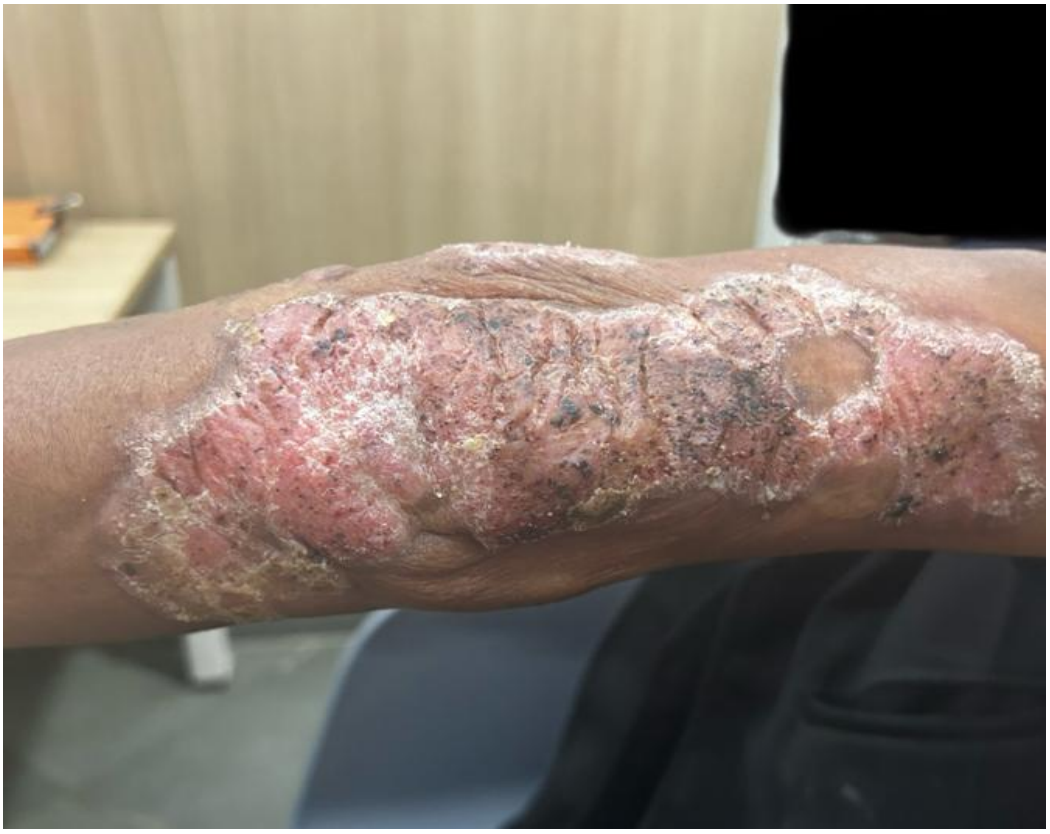

Figure 1: Multiple hypopigmented scaly plaques present over the extensor aspect of left forearm

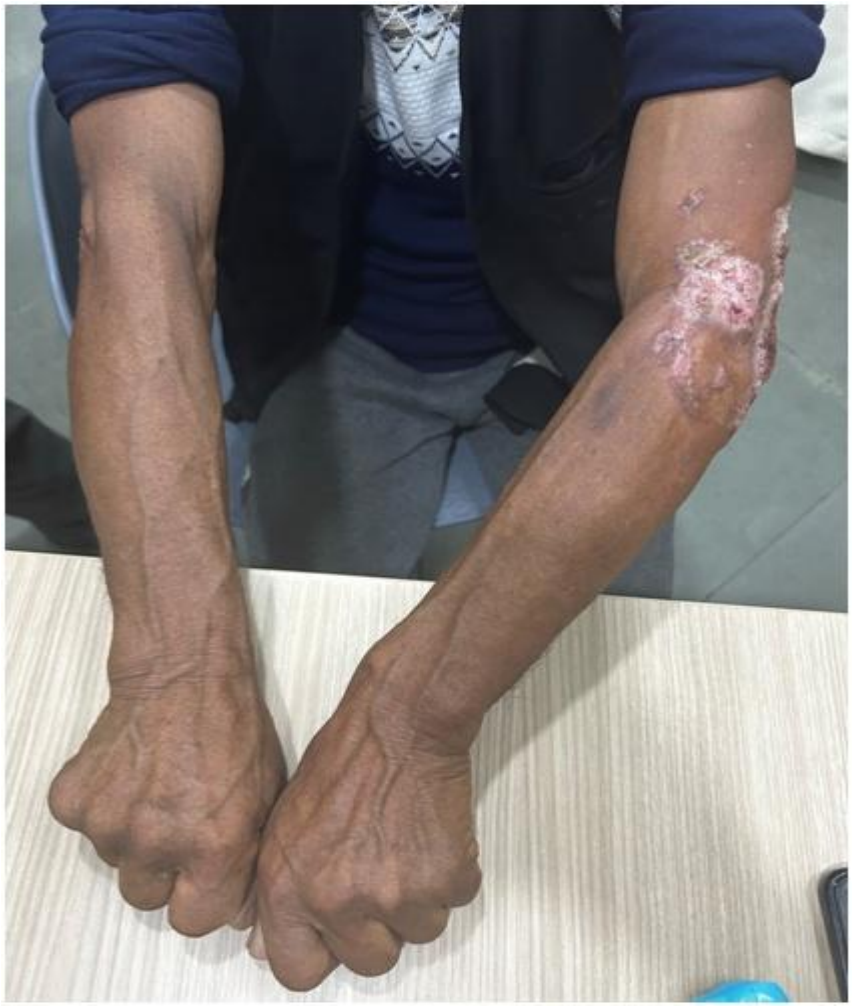

Figure 2: Left elbow deformed compared to right elbow

3. On Examination

Multiple erythematous to hypopigmented scaly plaques with raised borders and central atrophy over the left arm, forearm and elbow. The left elbow was deformed compared to the right elbow as depicted in Figure 2. To confirm the diagnosis a 4mm punch biopsy was done from the left forearm lesion and histopathological examination shows Epithelioid granuloma in the upper dermis, composed of epithelioidhistiocytes, and Langhan giant cell. There is diffuse infiltrate of neutrophils with lymphocytes plasma cells and histiocytes in upper dermis as depicted in Figure 3 Epidermis shows irregular hyperplasia suggestive of Lupus Vulgaris. There was no regional or generalized lymphadenopathy. Systemic examination was normal.

Haematological investigations revealed decreased hemoglobin, raised erythrocyte sedimentation rate, HIV ELISA was non reactive, HBsAg ELISA and HCV ELISA was non reactive. The Mantoux test was positive. The patient was started on anti tubercular treatment.

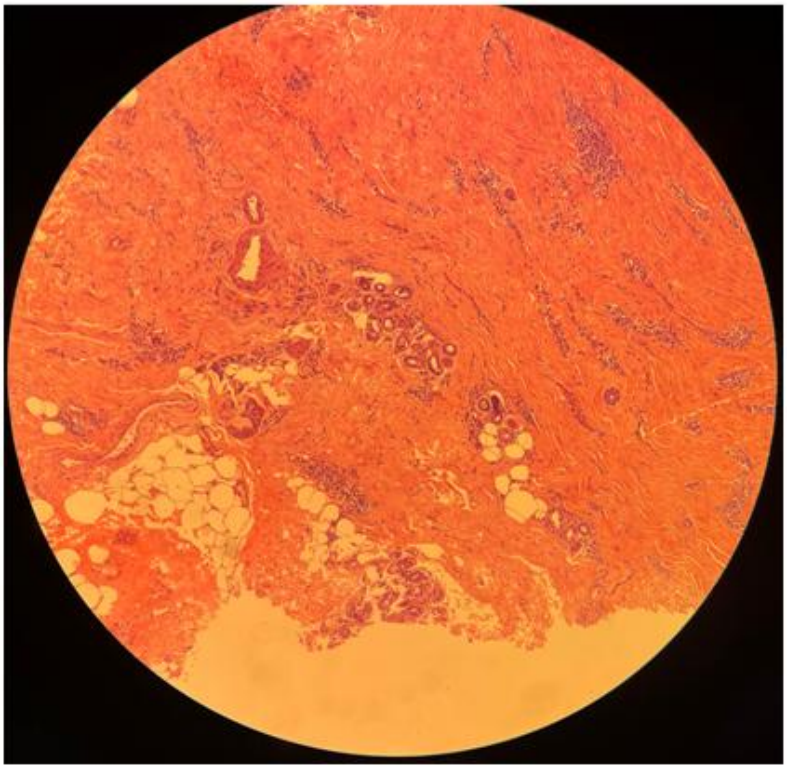

Figure 3: Under 10x magnification -. Diffuse infiltrate of neutrophils with lymphocytes, plasma cells and histiocytes in upper dermis.

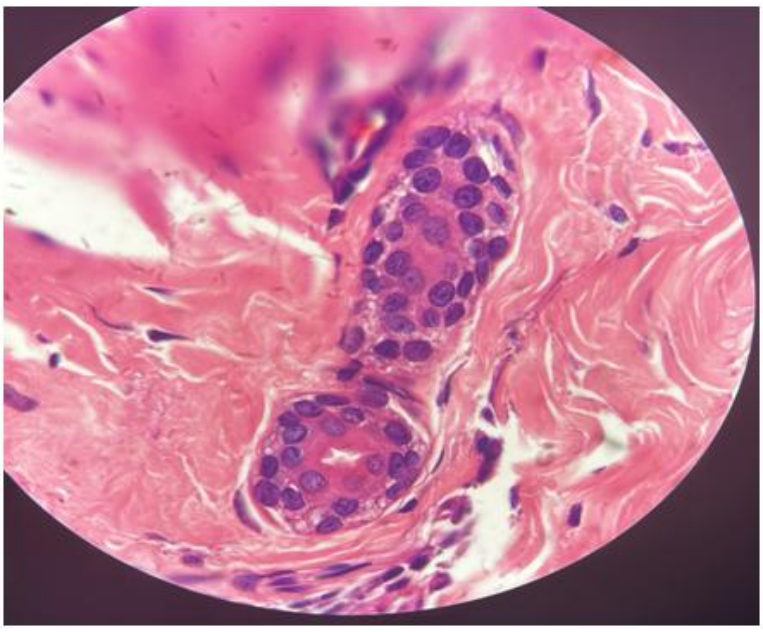

Figure 4: Under Oil immersion shows Langhan Giant Cell

4. Discussion

Lupus vulgaris is cutaneous tuberculosis seen in individuals who previously encountered tuberculosis bacilli, and those with moderate, and high tuberculin sensitivity, while it can develop as an exogenous infection on verrucous tuberculosis, scrofuloderma scar, and BCC vaccination site. CTB accounts for 1.5 - 3% of extrapulmonary cases. Classification of CTB based on route of infection and host immunity - True CTB and Tuberculids. True CTB classified based on route on infection includes - Inoculation: Tuberculous chancre, TB verrucosa cutis Hematogenous: Lupusvulgaris, Acute miliary TB, TB gumma, Autoinoculation: Orificial TB, Contiguous: Scrofulofuloderma. Tuberculids are classified into Lichen Scrofulosorum, Papulonecrotic tuberculid and Erythema Induratum of Bazin. [4] In Lupus Vulgaris initially the lesion is small, reddish brown, flat plaque, soft, gelatinous consistency. Gradually the lesion becomes elevated and grows by peripheral extension with areas of centralatrophy, apple jelly nodules on diascopy. Typical histopathological findings of lupus vulgaris is tuberclesor tuberculoid granuloma consisting of epithelioid histiocytes, and Langhans - type giant cells surrounded by lymphocytes, and monocytes. [5] The flexion deformity of the elbow is usually due to tenosynovitis particularly of flexor tendons. Musculoskeletal ultrasound and MRI scans are more sensitive at detecting joint abnormalities compared with conventional radiography. Other relevant investigations include medical history, clinical examination findings, complete hemogram with erythrocyte sedimentation (ESR) and C - reactive protein (CRP), liver function test, Mantoux test, and chest X - ray, Additionally, patients undergo enzyme - linked immunosorbent assay (ELISA) screening for HIV. Differential diagnosis of Lupus vulgaris includes Sarcoidosis, Psoriasis, Granuloma annulare, Discoid lupus erythematosus, Squamous cell carcinoma.

The standard multidrug antituberculosis therapy is followed which includes the 6 months regimen ofusing 4 drugs in two phases.

5. Conclusion

In this article we presented a long - standing case of lupus vulgaris with elbow deformity in an elderly man. This is to emphasize the early diagnosis and treatment of lupus vulgaris to prevent the complications such as contractures, scarring, tissue destruction and malignant changes. Malignant changes are known to arise such as squamous cell carcinoma which takes 20 - 30 years. [6] The treatment involves combination of isoniazid, rifampicin, pyrazinamide, and ethambutol given over a period of six months for a standard course. Multi - drug resistant tuberculosis has become a significant problem worldwide for which new anti - tuberculous drugs are being developed.

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