DISSEMINATED LUPUS VULGARIS WITHOUT UNDERLYING ACTIVE PULMONARY TUBERCULOSIS: A CASE REPORT OF AN ATYPICAL PRESENTATION

**Key Words**: LUPUS VULGARIS, CUTANEOUS TUBERCULOSIS

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**Conflict of interest**: None

**Key clinical message:**

Lupus vulgaris is a paucibacillary form of cutaneous tuberculosis occurring in those with high degree of immunity. Given the high degree of immunity that underlies this chronic disease, lesion is usually localized and disseminated forms are uncommon. Also disseminated forms without underlying active pulmonary tuberculosis is even rarer. Because of the rarity of the disseminated disease, non-specific presentation and lack of bacilli in Acid Fast Bacilli stains, its diagnosis is often delayed and prompt treatment is deferred, which can lead to complications such as scarring, contracture, tissue destruction and malignancy. Given the limitation of available diagnostic modalities in a resource poor setting, diagnosis can be confusing. As most of the lupus vulgaris lesion present in an isolated manner, our case of disseminated disease without underlying active pulmonary tubercular foci was an interesting phenomenon.

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Case Report:

BACKGROUND

Lupus Vulgaris is the most common form of cutaneous tuberculosis which is endemic in Nepal.1 Lupus vulgaris is a paucibacillary form of cutaneous tuberculosis occurring in those with high degree of immunity. Given the high degree of immunity that underlies this chronic progressive disease, lesions are mostly solitary and localized to face, neck and extremities.2 In Asian subcontinent, predominant site of involvement are thighs, legs and buttocks.3 Presence of disseminated lupus vulgaris is a rare finding and its occurrence in those without underlying active pulmonary tuberculosis is even more rarer.4 Patients present with slowly progressive plaque preceded by apple jelly nodule, commonly over normal skin but also over sites of previous scar.5 Lupus vulgaris usually occurs from an underlying primary foci of infection such as lymph node or bone. This is even more prominent in disseminated forms where presence of active pulmonary tubercular foci is a frequent finding.6 This etiological association with the underlying tubercular foci has led to the systemic screening for underlying infection in those with cutaneous manifestation.Given the high immunity coupled with decreased number of bacilli that occurs in this disease, diagnosis is often a challenge.5 As there’s lack of availability of Polymerase Chain Reaction (PCR) for diagnosing lupus vulgaris in resource poor setting like ours, priority should be given towards a vigilant clinical eye and good histopathological section with AFB staining.7 The histopathology of lupus vulgaris which is usually helpful shows tubercular granuloma with scanty or absent central caseation, surrounded by multinucleated giant cells and epithelioid histiocytes with absent or a very low number of tubercular bacilli.5 Disseminated lupus vulgaris without active pulmonary tubercular foci is difficult for the clinicians to diagnose and treat because of the rarity of the disease, non- specific presentation and lack of bacilli in the AFB stain.8 Here, we report a rare case report of disseminated lupus vulgaris without active pulmonary tuberculosis and significant improvement and prevention of complications after start of Anti Tubercular Therapy (ATT).

OBSERVATION

A 75-year-old male patient from rural village of Nepal, farmer by occupation, presented with asymptomatic annular, brownish, scaly elevated plaque over buttocks, thighs and forearm. Initial lesion was a soft reddish papule over left forearm that spread peripherally over period of years to involve bilateral buttocks, lateral thighs and bilateral groin. Papulo-nodular lesions became confluent to form plaque with brownish hue with induration, infiltration, peripheral extension, irregular rough surface and whitish adherent scales. On examination, largest annular plaque with serpiginous peripheral extension and central sparing was noted over extensor aspect of buttocks with disto-lateral extension, along with confluent plaques over bilateral lateral thigh with distal extension, groin and left antero-lateral region of distal forearm. (Figure 1-4). There was no associated local tissue destruction, no symptoms of persistent dry cough, hemoptysis, chest pain or any other stigmata of active pulmonary tuberculosis or recent vaccination. Biopsy was done which showed well formed epithelioid granuloma with scattered Langham’s type multinucleated giant cell and moderate to dense lymphohistiocytic infiltrate in the papillary dermis. Stain for AFB was negative compatible with lupus vulgaris. Chest X-ray showed emphysematous change in bilateral lung without patchy consolidation, poorly defined nodule or calcification. Patient was started on Anti Tubercular Therapy (ATT), with significant improvement in lesional morphology after completion of initiation phase treatment of two months. Category-I ATT as per Nepal government ATT protocol was started and the extension of lesion was halted with only residual pigmentation without active disease post treatment. There was complete cure of the disease post treatment with no recurrence till date. New lesions have not evolved for 1 year now and the lesion has healed with no symptoms and minimal pigmentation.

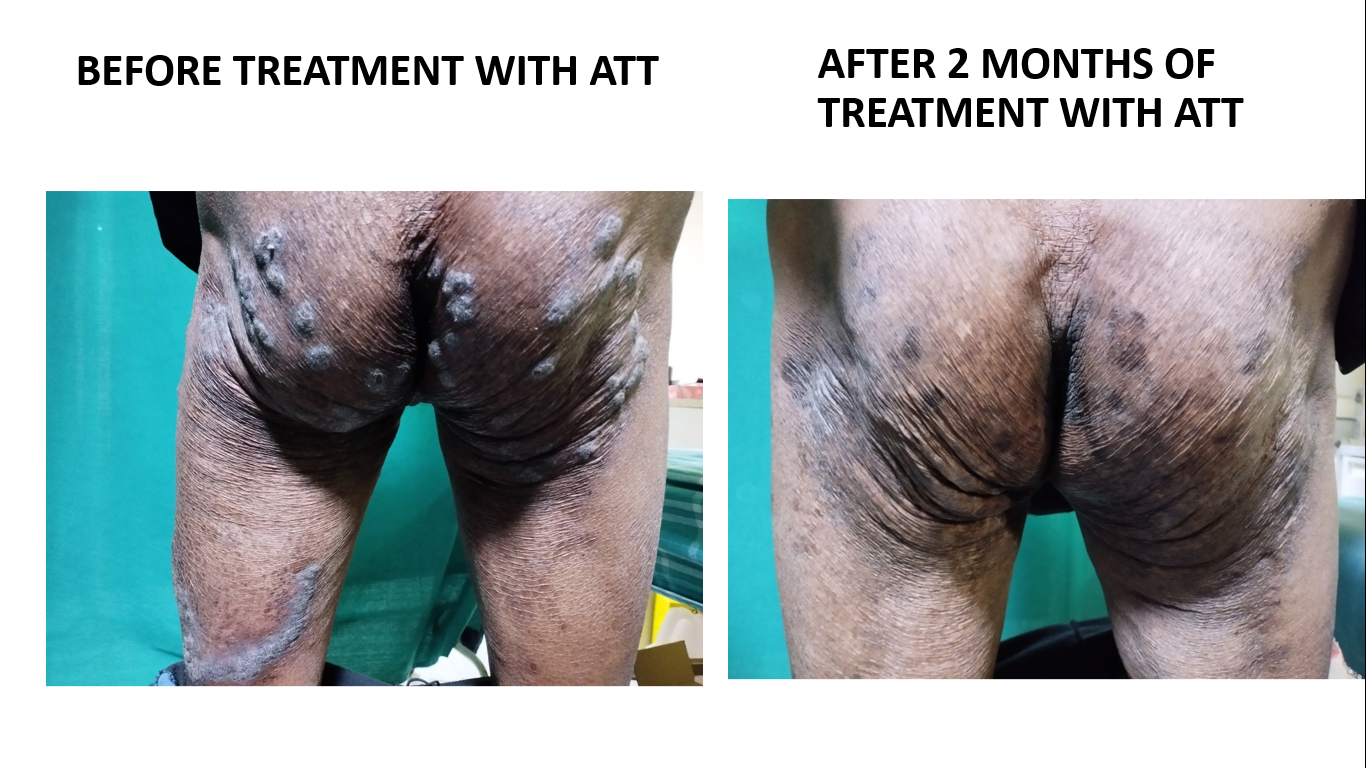


Figure 1: Largest annular plaque with serpiginous peripheral extension and central sparing over extensor aspect of buttocks before and after 2 months of ATT



Figure 2: Multiple plaques with adherent white scales and keratotic surface over extensor aspect of buttocks with lateral extension- before and after 2 months of ATT, residual post inflammatory hyperpigmentation seen post therapy.



Figure 3: Disseminated Annular plaque with serpiginous expanding edge and central clearing over extensor buttock with posterolateral extension



Figure 4: keratotic irregular plaque with adherent white scales and central sparing, with confluent nodules over anterolateral part of distal forearm

DISCUSSION

Lupus vulgaris is a gradually progressive, paucibacillary form of cutaneous tuberculosis occurring in individuals with high immunity, usually presenting as a solitary plaque over extremities and trunk in Asian subcontinent.3 Disseminated lupus vulgaris involving multiple sites over trunk and extremities is a rare finding and even rarer is its occurrence in those without active pulmonary foci of tubercular infection.6 However, lupus vulgaris can occur in those with re-infection due to re-inoculation, after BCG vaccination or in those with tubercular involvement of bone and lymph node.9 Patients mostly present with slow growing reddish brown papule that increase in size with peripheral extension, serpiginous edge and keratotic scaly surface.5 The reddish brown papule will tend to be more brownish in color over time with induration, rough surface, serpiginous peripheral spread with central sparing and irregular scarring.9 Pigmentation, scarring, contractures, local tissue destruction and occasionally cutaneous Squamous Cell Carcinoma (SCC) are dreaded complications.10 There’s increased gender predilection for females in this chronic cutaneous disease.9 As disseminated lupus vulgaris, without active pulmonary tubercular foci is rare, given the limitation of available diagnostic modalities like PCR, in a resource- poor setting, diagnosis can be confusing.7 Diagnosis can be made with good clinical acumen, histopathology and PCR. Histopathology of Lupus Vulgaris will show well-formed epithelioid granuloma with scattered Langhan’s type multinucleated giant cell and moderate to dense lymphohistiocytic infiltrate in the papillary dermis. Stain for AFB can be negative, given the paucibacillary nature of the disease.5 With proper diagnosis, overall prevalence of the disease can be estimated and clinical therapeutic trials can be performed with timely prevention of dreadful complications like malignancy.

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**Data availability statement**

The data that support the findings of this study are openly available in Clinical Case Reports

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**Detailed author’s contribution**:

PP ,SP and SG contributed to the collection of data and the management of the patient. PP and SP wrote the initial draft of manuscript. PP, SP, SA, SG and PP revised and prepared the final version of the manuscript. All authors have read and approved the final manuscript and agree to take full responsibility for the integrity and accuracy of the work.

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