A clinico-histopathological study of lupus vulgaris: A 3 year experience at a tertiary care centre

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ABSTRACT

Background: Lupus vulgaris is the most common form of cutaneous tuberculosis in adults. Lupus vulgaris is caused by hematogenous, lymphatic, or contiguous spread from elsewhere in the body. Histologically it is characterized by typical tubercles with or without caseation, surrounded by epitheloid histiocytes and multinucleate giant cells in the superficial epidermis with prominent peripheral lymphocytes. Materials and Method: All cases of clinically and histopathologically diagnosed lupus vulgaris over the previous five years were included in the study. Results: Fourteen cases of lupus vulgaris cases reported during the study period with equal incidence among males and females. Discussion: Plaque type of lupus vulgaris was the most common type. Histopathologically tubercular granulomas were seen in all cases as compared to other studies. Conclusion: Different patterns of lupus vulgaris are reported.

Key words: Lupus vulgaris, cutaneous, tuberculosis

INTRODUCTION

Lupus vulgaris is a chronic, progressive, post-primary, paucibacillary form of cutaneous tuberculosis (TB), occurring in a person with moderate or high-degree of immunity. Lupus vulgaris is the most common form of cutaneous TB in adults in the Indian subcontinent and South Africa. All age groups are equally affected, with females two to three times more commonly than males. Lupus vulgaris is caused by hematogenous, lymphatic, or contiguous spread from elsewhere in the body. Spontaneous involution may occur, and new lesions may arise within old scars. Complete healing rarely occurs without therapy. The most prominent histopathologic feature is the formation of typical tubercles with or without caseation, surrounded by epitheloid histiocytes and multinucleate giant cells in the superficial epidermis with prominent peripheral lymphocytes. Secondary changes like epidermal thinning and atrophy or acanthosis with excessive hyperkeratosis or pseudoepitheliomatous hyperplasia can also be noted. Acid-fast bacilli are usually not found.

MATERIALS AND METHODS

The study was retrospective and was conducted in those patients who attended the out-patient clinic in the Department of Dermatology, SDM Medical College and Hospital, Dharwad over a period of 3 years. Patients of all age group were included in the study. A total of 14 cases of lupus vulgaris were diagnosed during the study period. The demographic details, history, clinical presentation, routine investigations and chest X-ray findings were recorded from the patients' case sheet. Patients were also screened with venereal disease research laboratory (VDRL) and enzyme linked immunosorbent assay for human immuno deficiency virus. The diagnosis was based on clinical features, histopathology (Hematoxylin and Eosin and Ziehl-Neelsen stains) and microbiology of the tissue smears and in case of discharging sinuses the tissue exudate. Chest X-ray was performed prior to therapy to exclude active pulmonary involvement.

RESULTS

Fourteen cases of lupus vulgaris reported during the study period. The incidence in males and
females was almost equal. Most patients belonged to the third to fifth decade [Figure 1]. Among the various patterns of presentation of lupus vulgaris, plaque type (11 of the 14) was the most common type of presentation (78.5% of all lupus vulgaris cases) followed by ulcerative type (two cases) and one case of tumor like presentation [Figures 2 and 3]. Among the 11 patients who presented with plaque type of lupus vulgaris, eight of them (72.7%) presented with only one plaque [Figures 8 and 11]. History of pulmonary TB was positive in 2 out of 14 cases [Figure 13] and family history was positive in one case.

Lower limb was involved in seven patients and upper extremity in five [Figures 10 and 12]. Two patients had lesions on the face. Neck and gluteal region was involved in one case only [Figure 9].

Histopathological examination revealed epitheliod granulomas with caseation in 3 out of 14 cases (21.4%) and granulomas without caseation in 11 out of 14 cases (78.5%), lymphocytic infiltrates and Langerhans cells [Figures 4-6]. Secondary changes like acanthosis and hyperkeratosis were seen in 11
out of 14 cases (78.5%) and epidermal thinning and atrophy was noted in 1 case [Figures 7]. No bacilli were seen. Epithelial dysplasia and normal epithelium each were noted in a case. Dermal fibrosis was noted in 3 cases of plaque type and foreign body giant cells and calcific bodies in giant cells were noted in 2 cases of plaque type.

**DISCUSSION**

Lupus vulgaris originates from an underlying focus of TB, typically in a bone, joint or lymph node, and arises by either contiguous extension of the disease from underlying affected tissue or by hematogenous or lymphatic spread.[1] In patients where the underlying focus is not apparent it has been postulated that it may be due to the reactivation of a latent cutaneous focus secondary to previous silent bacteremia.[6] It can also arise after exogenous inoculation or as a complication of Bacillus Calmette–Guérin (BCG) vaccination.[7]

In Europe, over 80% of lesions are on the head and neck, particularly around the nose.[8] Next in frequency are the arms and legs, but involvement of the trunk is uncommon. In India, cutaneous TB more commonly affects the buttocks and extremities rather than the face.[9] Such a pattern is usually due to re-inoculation and may relate to playing without clothing or shoes.[10] Similar findings were seen in our study.

The overall prevalence of cutaneous tuberculosis (TB) in various Indian studies is 0.25-0.6%. [11]
The initial lesion is a brownish red, soft or friable macule or papule with a smooth or hyperkeratotic surface. On diascopy the infiltrate exhibits a typical apple jelly color.\(^5\)

The clinical forms fall into five different general patterns, depending on the local tissue response to the infection:
1. In plaque form, the lesions have irregular or serpiginous edge and large plaques show irregular areas of scarring with islands of active lupus tissue. The edge often becomes thickened and hyperkeratotic.\(^1\) This was the most common presentation in our study with most having single plaque lesions.
2. In ulcerative and mutilating forms, scarring and ulceration predominate with crusts forming over areas of necrosis. The deep tissues and cartilage are invaded and contractures and deformities can occur.\(^1\) Two ulcerative type of lesions were seen in our study with scarring but deformities were not noticed.
3. Vegetating form is characterized by marked infiltration, ulceration and necrosis with minimal scarring. Mucous membranes are invaded and cartilage is slowly destroyed.

In India, scrofuloderma is the most common form of skin TB in childhood, whereas, lupus vulgaris is the most common form in adults.\(^12\)
When the nasal or auricular cartilage is involved, extensive destruction and disfigurement occurs.\[^{11}\]

4. Tumor-like forms, present either as soft-tumor-like nodules or as epithelial hyperplasia with the production of hyperkeratotic masses.\[^{11}\] Single case of tumor variety was seen in our study.

5. In papular and nodular forms, multiple lesions occur simultaneously in disseminated lupus – true "miliary lupus" and usually occur after immunosuppression.

Histopathologically, tubercular granulomas with scanty or absent central caseation, surrounded by epithelioid histiocytes and multinucleate giant cells are present in the superficial dermis with prominent peripheral lymphocytes.\[^{11}\] Though our study did not reveal much difference in the histopathological finding with respect to the type of clinical presentation [Figure 1], tubercular granulomas were seen in all cases as compared to other studies where granulomas were noted in only 70% of the patients.\[^{13,14}\]

There was not much difference with respect to caseation necrosis. Secondary changes like epidermal hyperkeratosis was seen in 78% as compared to 41% in other studies.\[^{14}\]

The natural course of an untreated lesion is progressive leading to scarring, contractures and tissue destruction. The scars are usually thin, white and smooth, but are unstable and may break down or become keloidal. Active lupus vulgaris frequently reappears in scar tissue. Squamous cell and basal cell carcinomas or sarcomas can occur insidiously in upto 8% of patients and may be confused with renewed activity of the lupus itself.\[^{11}\]

The differential diagnoses to be considered in the early stages are leprosy, sarcoidosis, lymphocytoma, Spitz naevus and lupus erythematosus and in older patients' syphilis must be excluded. The histopathologic and culture reports help to differentiate lupus vulgaris from the deep mycoses, which closely resemble the vegetating and crusted type.\[^{11}\] Leprosy and sarcoidosis are the chief causes of diagnostic difficulty. The nodules in leprosy are firmer with other positive signs of leprosy and the nodules of sarcoidosis resemble grains of sand rather than "apple jelly."\[^{11}\]

Standard four drug anti-tuberculosis therapy must be given in histopathologically confirmed cases for six months. A therapeutic trial of triple anti-tuberculosis therapy: Isoniazid, rifampicin and pyrazinamide may be considered in cases where the diagnosis is difficult. A clinical response would be expected within 4-6 weeks.\[^{13}\]

In conclusion, lupus vulgaris is most common among the middle aged patients, commonly presenting as plaque type, with frequent involvement of the extremities. Diagnosis is based on clinical features and histopathological examination. On histopathological examination, granulomas without caseous necrosis are commonly reported with acanthosis with hyperkeratosis. All patients should be treated with the standard anti-tubercular regimen.

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