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 charecterised by typical tubercles with or without caseation, surrounded by epitheloid histiocytes and multinucleate giant cells in the superfi cial epidermis with prominent peripheral lymphocytes. **Materials and Method:** All cases of clinically and histopathologicaly 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 eaqual 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

Access this article online

Website: www.idoj.in

DOI: 10.4103/2229-5178.142497

Quick Response Code:

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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.[1] Lupus vulgaris is the most common form of cutaneous TB in adults in the Indian subcontinent and South Africa.[2-4] All age groups are equally affected, with females two to three times more commonly than males.[5] 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.[5] The most prominent histopathologic feature is the formation of typical tubercles with or without caseation, surrounded by epitheliod histiocytes and multinucleate giant cells in the superficial epidermis with prominent peripheral lymphocytes.[1] 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.[5]

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

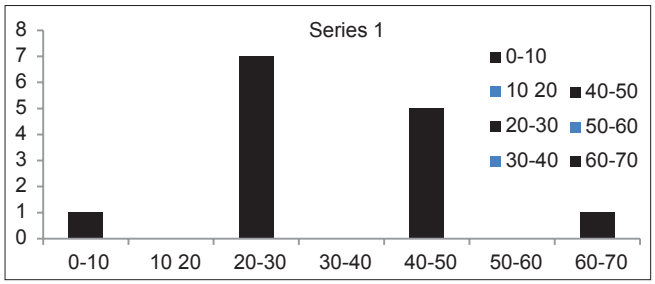

Figure 1: Incidence among various age groups

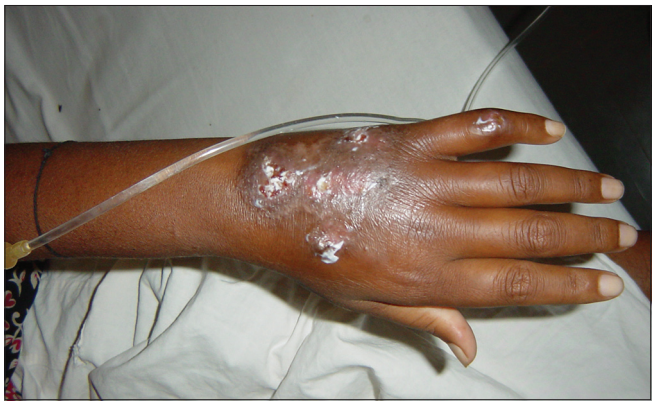

Figure 3: Multiple ulcerative lesions over the dorsum of the hand

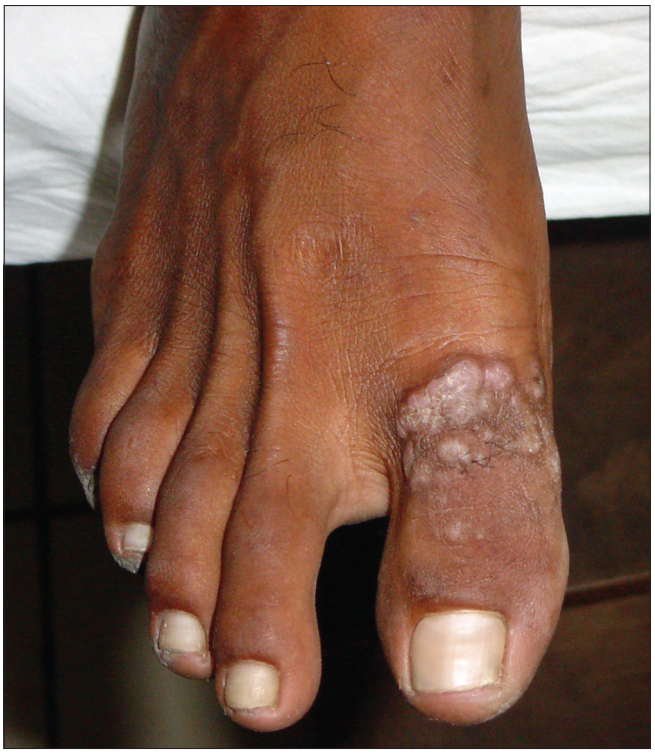

Figure 2: Plaque present over the dorsum of the great toe with minimal central scarring

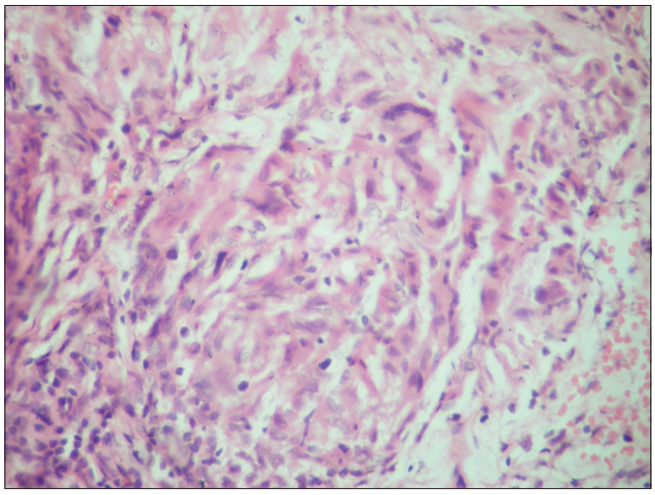

Figure 5: Reticular dermis shows marked histiocytic and lymphocytic infiltrate with few plasma cells. Epitheliod cell granulomas with Langerhans type giant cells are seen. Mixed inflammatory infiltrate is seen in the papillary dermis (H and E, ×100)

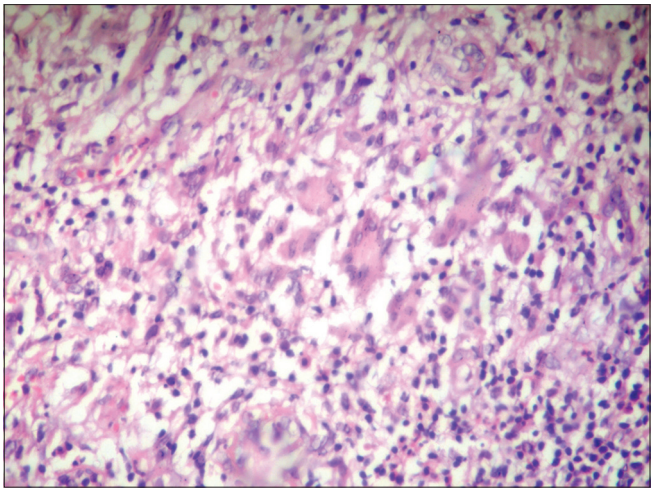

Figure 4: Epitheliod cell granulomas with Langerhans type giant cells seen in the mid dermis. Mixed inflammatory infiltrate is seen in papillary dermis (H and E, ×100)

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 (27.2%) 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]

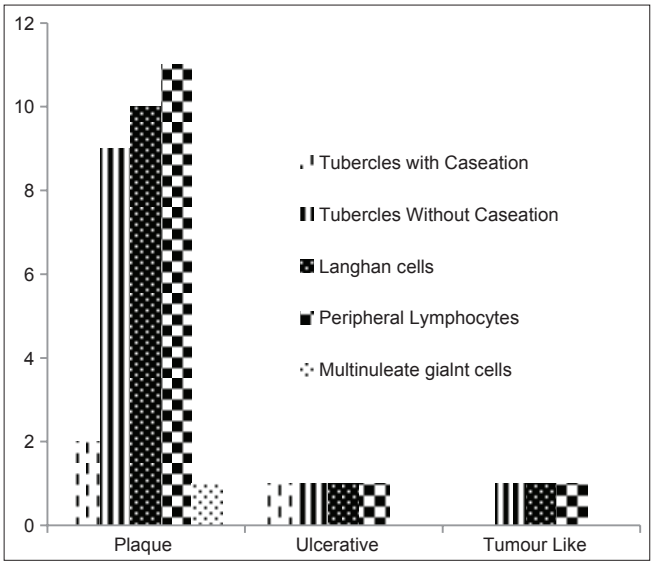

Figure 6: Primary histological changes in relation to the clinical types of lupus vulgaris

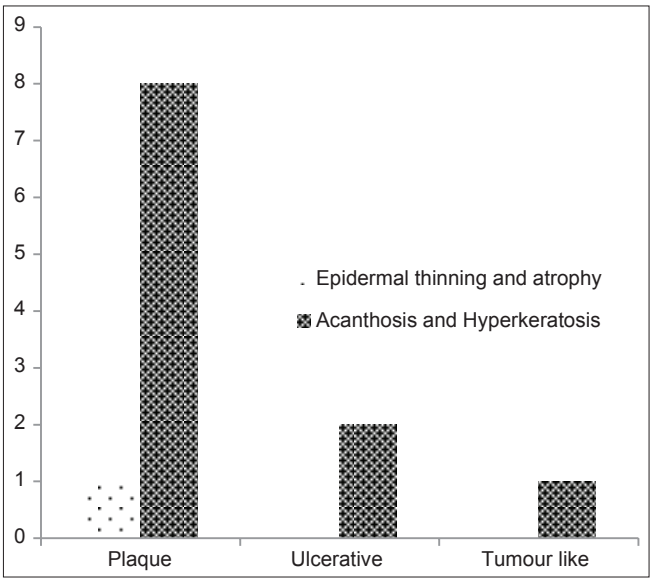

Figure 7: Secondary histological changes in relation to the clinical types of lupus vulgaris

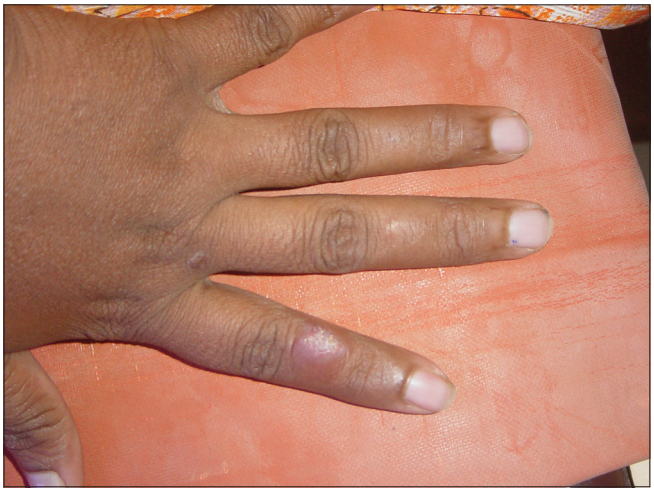

Figure 8: Solitary plaque over the index finger

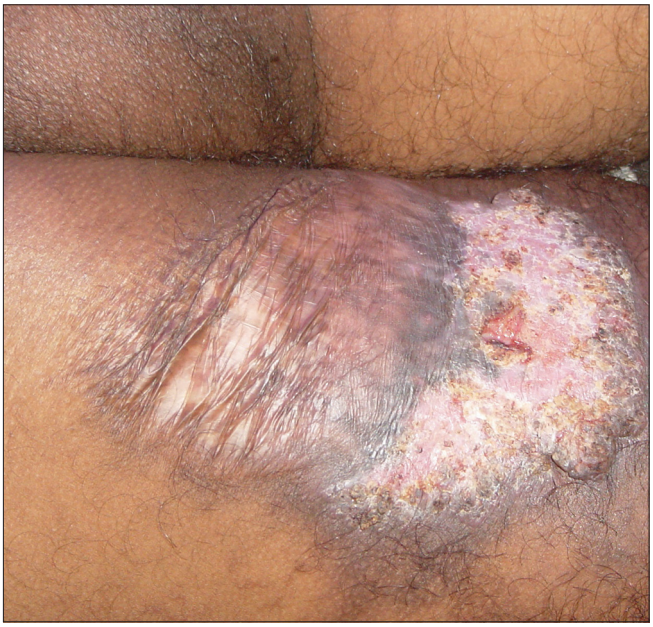

Figure 9: Scarring plaque over the gluteal region

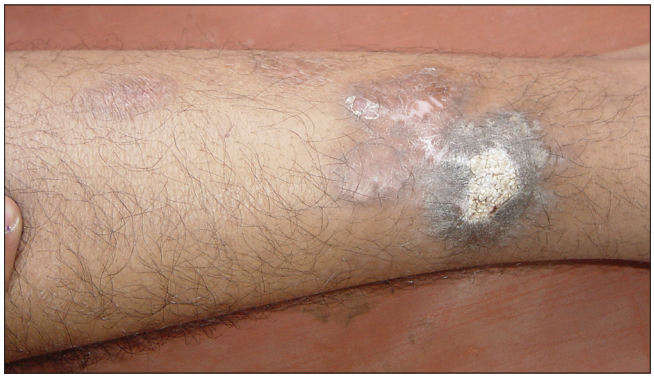

Figure 10: Plaque over the lower limb

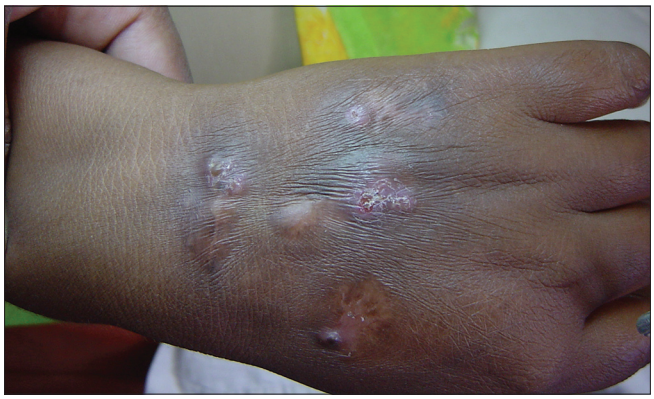

Figure 12: Healed lesion of Figure 2

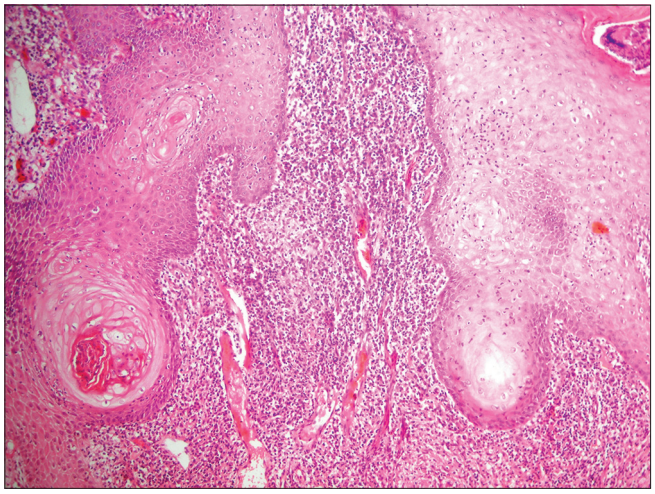

Figure 14: Section shows epidermis with marked acanthosis and neutrophilic abcess mixed with debris in keratin layers. The papillary dermis shows epitheliod cell granuloma with multinucleated gaint cell. The granuloma are surrounded by plasma cells and lymphocytes. Few eosinophils are seen amidst chronic inflammatory cell infiltrate. H and E, ×40

In India, scrofuloderma is the most common form of skin TB in childhood, whereas, lupus vulgaris is the most common form in adults.[12]

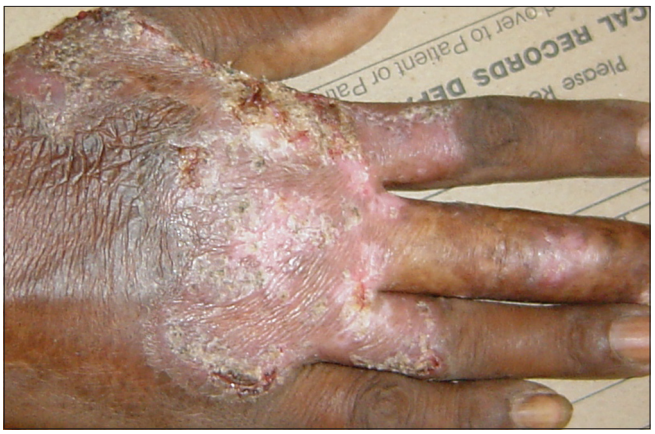

Figure 11: Plaque over the dorsum of hand

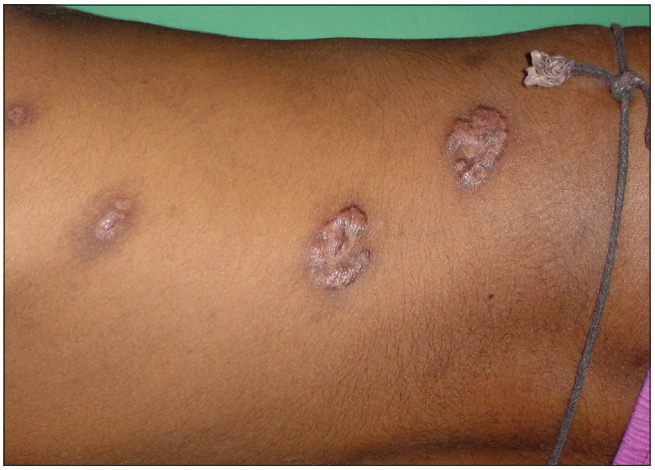

Figure 13: Multiple plaques over the body

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.

When the nasal or auricular cartilage is involved, extensive destruction and disfigurement occurs.[1]

4. Tumor-like forms, present either as soft-tumor-like nodules or as epithelial hyperplasia with the production of hyperkeratotic masses.[1] 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 immunosuppresion.

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.[1] 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.[1]

The differential diagnoses to be considered in the early stages are leprosy, sarcoidoisis, 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.[1] 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."[1]

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.[15]

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.

REFERENCES

1. Yates VM. Mycobacterial infection. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. Rook's Textbook of Dermatology. 8th ed. Oxford: Blackwell Science Ltd; 2010. p. 31.16-9.
2. Kumar B, Muralidhar S. Cutaneous tuberculosis: A twenty-year prospective study. Int J Tuberc Lung Dis 1999;3:494-500.
3. Visser AJ, Heyl T. Skin tuberculosis as seen at Ga-Rankuwa Hospital. Clin Exp Dermatol 1993;18:507-15.
4. Bhutto AM, Solangi A, Khaskhely NM, Arakaki H, Nonaka S. Clinical and epidemiological observations of cutaneous tuberculosis in Larkana, Pakistan. Int J Dermatol 2002;41:159-65.
5. Tappeiner G. Tuberculosis and infections with atypical mycobacteria. In: Wolff K, Goldsmith LA, Katz SI, Gilchrist BA, Paller AS, Leffell DJ, editors. Fitzpatricks Dermatology in General Medicine. 7th ed. New York: McGraw Hill; 2008. p. 1771-2.
6. Marcoval J, Servitje O, Moreno A, Jucglà A, Peyrí J. Lupus vulgaris. Clinical, histopathologic, and bacteriologic study of 10 cases. J Am Acad Dermatol 1992;26:404-7.
7. Tan HH, Seow CS. A review of cutaneous granulomas and lupus vulgaris following BCG vaccination in a skin hospital in Singapore. Ann Acad Med Singapore 2002;31:663-5.
8. Horwitz O. The localization of lupus vulgaris of the skin. Acta Tuberc Scand 1960;39:1-137.
9. Sehgal VN, Wagh SA. Cutaneous tuberculosis. Current concepts. Int J Dermatol 1990;29:237-52.
10. Vashisht P, Sahoo B, Khurana N, Reddy BS. Cutaneous tuberculosis in children and adolescents: A clinicohistological study. J Eur Acad Dermatol Venereol 2007;21:40-7.
11. Patra AC, Gharami RC, Banerjee PK. A profile of cutaneous tuberculosis. Indian J Dermatol 2006;51:105-7.
12. Kumar B, Rai R, Kaur I, Sahoo B, Muralidhar S, Radotra BD. Childhood cutaneous tuberculosis: A study over 25 years from northern India. Int J Dermatol 2001;40:26-32.
13. Thakur BK, Verma S, Hazarika D. A clinicopathological study of cutaneous tuberculosis at Dibrugarh district, Assam. Indian J Dermatol 2012;57:63-5.
14. Sehgal VN, Srivastava G, Khurana VK, Sharma VK, Bhalla P, Beohar PC. An appraisal of epidemiologic, clinical, bacteriologic, histopathologic, and immunologic parameters in cutaneous tuberculosis. Int J Dermatol 1987;26:521-6.
15. Ramam M, Mittal R, Ramesh V. How soon does cutaneous tuberculosis respond to treatment? Implications for a therapeutic test of diagnosis. Int J Dermatol 2005;44:121-4.

Cite this article as: Pai VV, Naveen KN, Athanikar SB, Dinesh US, Divyashree A, Gupta G. A clinico-histopathological study of lupus vulgaris: A 3 year experience at a tertiary care centre. Indian Dermatol Online J 2014;5:461-5.

Source of Support: Nil, **Conflict of Interest:** None declared.