



A Rare Case of Lupus Vulgaris in a Private Tertiary Health Facility in South-Western Nigeria

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Authors' contributions

This work was carried out in collaboration between all authors. Author M. A. Akinola designed the study, wrote the first draft of the manuscript. Author M. A. Ajani did the literature search, worked on the histology slide and supplied the photomicrograph. Author TOS gave histopathology report and was involved in the written of the discussion. Author OPY wrote the case history, got the informed consent from the patient and the mother and did the initial discussion writing and author RAA wrote the abstract and supplied the clinical photograph. All authors read and approved the final manuscript.

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Case Report

ABSTRACT

Background: Lupus Vulgaris is the most common form of cutaneous tuberculosis. It represents a reactivation of infection in people with moderate to high immunity against mycobacterium tuberculosis. Lesions can develop in all ages, as a collection of discrete, red-brown papules that subsequently coalesce to form an indolent asymptomatic plaque on the affected site. Diagnosis constitutes a major challenge as the most definitive investigative modality, mantoux test is not always positive. Histological examination and initiation of antitubercular therapy usually leads to diagnosis and clinical cure respectively.

Objective: We report a case of Lupus Vulgaris of external nose, a rare, underreported and commonly misdiagnosed condition in Nigeria.

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Case Report: A 13-year-old female presented with proliferative, raised multinodular lesion over the external nose. Clinical features, histopathological examination and a strongly reactive tuberculin skin test were all supportive of a diagnosis of Lupus Vulgaris. Antitubercular therapy (9-month regimen) was started and later followed by serial debridement of necrosed tissue. The lesion significantly resolved.

Keywords: Antitubercular therapy (ATT); cutaneous tuberculosis (CTB); Gene Xpert; lupus vulgaris (LV); nose.

1. INTRODUCTION

Lupus vulgaris (LV) is the most common form of cutaneous tuberculosis in Europe. It is a chronic and progressive form of cutaneous tuberculosis that represents a reactivation of infection in people with moderate to high immunity against mycobacterium tuberculi [1]. It may occur either as a result of direct extension from an underlying focus such as tuberculous lymphadenitis which is specifically referred to as scrofuloderma or via lymphatic or haematogenous spread. LV occurs in individuals of all ages and women are two to three times more likely than men to be affected. This disease usually begins as a collection of discrete, red-brown papules that subsequently coalesce to form an indolent, asymptomatic plaque; the plaque gradually reaches a size of 0.5-10cm and develops central clearing and atrophy. The borders may acquire a serpiginous or verrucous quality. Hypertrophic, ulcerative and vegetative forms of LV may also occur [2].

Cutaneous tuberculosis is an uncommon form of extrapulmonary tuberculosis largely occurring in developing countries. The recent rise in the incidence of tuberculosis especially due to Human Immunodeficiency Virus infections, has led to a resurgence of its extrapulmonary forms [3]. Cutaneous tuberculosis constitutes 1-2% of all tuberculosis cases [4,5]. There is a diversity of morphological forms and an unusual clinical presentation with involvement of the face, nose, conjunctiva and buttocks [6,7,8].

Diagnosis poses a major challenge as results of microscopy are usually negative due to sparse bacilli load in the lesion. Clinical presentation is ambiguous as facial lesions of sporotrichosis, chromoblastomycosis, cutaneous leishmaniasis and rosacea may mimic cutaneous tuberculosis [6,7,9]. This accounts for under-estimation and under-reporting of cases as the diagnosis may be overlooked. However, molecular diagnostic techniques such as Gene Xpert and Polymerase Chain Reaction (PCR) followed by a positive response to antitubercular therapy (ATT) may resolve the diagnostic dilemma.

In Nigeria, with increasing prevalence of HIV/AIDS, there has been an associated increase in the prevalence of pulmonary tuberculosis (PTB), consequently, atypical presentation such as extrapulmonary/cutaneous forms of the disease may be on the increase as well [10].

We therefore, report a case of LV to further promote awareness of the clinical resurgence of cutaneous tuberculosis, the importance of histopathological examination (although this is not always diagnostic) and the role of positive response to antitubercular therapy in the clinical diagnosis especially in resource-poor countries.

2. CASE REPORT

A 13 year old female who presented to Ear, Nose and Throat (ENT) clinic with non-healing, raised, gradually progressing asymmetrical multinodular lesion over the tip of the nose extending to the bridge, spanning over four years. It developed as a non-discharging solitary papule which subsequently became multinodular on the nasal tip, progressing to involve the adjacent alae nasi. It was described as a moderately painful, pruritic and occasionally hemorrhagic lesion. There was no associated fever, cough, haemoptysis or chest pain. No similar lesion in other parts of the body. She had no history of trauma or insect bites to the site of lesion. No history of prior contact with patients with tuberculosis. The patient showed no improvement on antibiotic treatment. Review of systems was essentially normal.

General physical examination showed a moderately malnourished child, afebrile and well hydrated. Examination of the nose and nasal cavities showed irregular multiple nodular lesions, dome-like, measuring about 6cm by 4cm at its widest points distributed over the tip and alae nasi. The lesion was characteristically raised with erythematous borders and adherent brownish crusts over the surface, causing complete distortion of the nasal anatomy. The

cavities were essentially patent with areas of hyperemia on the septum and inferior turbinates bilaterally.

Examination of the ears, oral cavity and oropharynx were essentially normal.

Differential diagnoses of Acne Rosacea, Lupus Vulgaris and Basal cell Carcinoma were considered.

A skin biopsy was taken. Other laboratory workups to rule out background immunosuppressive state and fungal diseases were negative.

Although the Ziehl-Neelsen smears were negative for acid-fast bacilli and Gene Xpert was also negative for mycobacterium, Mantoux test was significantly reactive, showing the exaggerated response of 25-30 mm which is strongly suggestive of cutaneous tuberculosis.

A tissue sample was obtained by punch biopsy and histology done showed multiple granulomas composed of aggregates of epithelioid cells which were surrounded by lymphocytes, plasma cells, histiocytes and few eosinophils. Few foreign body and Langhan types' multinucleated cells were also seen. The overall features were that of chronic granulomatous inflammatory lesion most likely cutaneous tuberculosis (Lupus Vulgaris) as seen in Fig. 1.

Fungal infection was clinically, histologically and histochemically excluded and a final diagnosis of Lupus Vulgaris was established.

A regimen for treatment of Cutaneous tuberculosis (TB) was commenced using quadruple antitubercular therapy (ATT) with rifampicin 450 mg, isoniazid 300 mg, ethambutol 800 mg and pyrazinamide 1500 mg daily for 2 months, followed by isoniazid and rifampicin in the same doses for 7 months. This was combined with serial debridement following significant necrosis of the lesion within 2 months of commencement of therapy. See Picture before treatment (Fig. 2) and Picture after treatment (Fig. 3).

3. DISCUSSION

LV is a chronic, progressive form of cutaneous tuberculosis (TB) that is contacted either primarily by direct inoculation of the bacilli or at the site of previous Bacille Calmette-Guérin (BCG) vaccination in individuals with moderate immunity. It could also be contacted secondarily by haematogenous and lymphatic spread from underlying distal focus [11,12]. The characteristic lesion is a plaque composed of nodules of apple jelly colour which extends irregularly to the contiguous structures while in some areas, scarring occurs, causing considerable tissue destruction over many years [11].

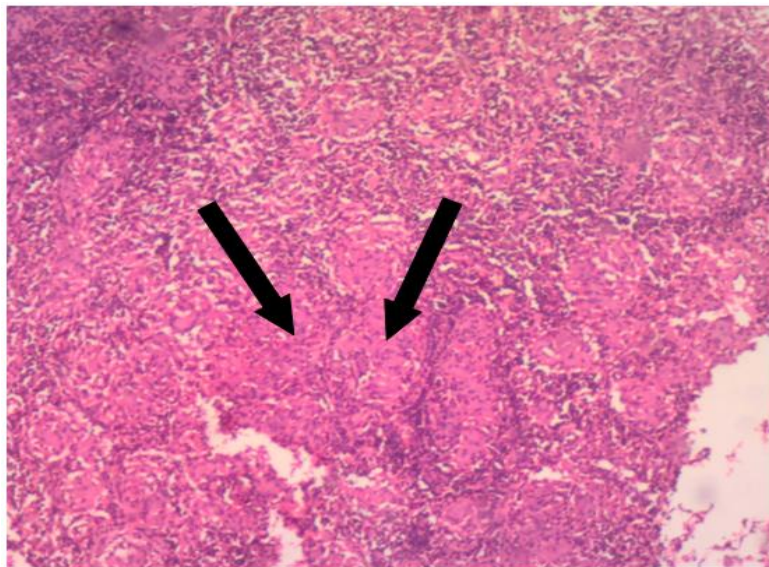


Fig. 1. Photomicrograph showing multiple granulomas (arrows). (Haematoxylin and eosin, X100)



Fig. 2. Photograph before treatment showing irregular multiple plaques forming a dome-like lobulated lesion distributed over the tip and alae nasi



Fig. 3. Photograph after treatment

Although LV is reckoned to be the most common variant of cutaneous TB in India accounting for up to 74% of cases with incidence of 5.9 cases per 1000 population [13], it is still considered a rare disease. The incidence decreased in recent decades [11]. However, it is significantly under-reported in Nigeria probably as a result of the challenges of making a definitive diagnosis. The pathogenesis and localisation of nasal tuberculosis comes mainly from haematogenous or lymphatic extension of pulmonary TB. The nasal mucosal is not usually affected despite being a point of entry for the *Mycobacterium tuberculosis*. It is usually secondary to inoculation following digital trauma to the skin. Rarer sites include the pinna, sinuses, nasal cavities, and external nose, the later being characterised by its infrequency and clinical polymorphism [11]. This is similar to our case in that its variability usually confuses nasal LV with other chronic granulomatous inflammatory processes and for this reason, diagnostic suspicion is important [13].

Moreover, nasal tuberculosis, specifically, has been reported in the past only by a few authors. Although it remains a rare disease condition, it has been largely misdiagnosed or grossly underreported because of its resemblance to nasal neoplasm or other cutaneous condition such as rocasea, cutaneous leishmaniasis among others [13].

LV lesions commonly, are flat, multinodular apple jelly in appearance which usually involves orifices extending to the face and orbital skin. It rarely affects only the nose as seen in this case report. It also distinguishes this case from tuberculosis cuties as reported by Fatusi et al. [11].

Making a diagnosis of cutaneous TB requires absolute criteria: Positive culture of *Mycobacterium tuberculosis* or DNA identification by PCR. However, a negative PCR result in formalin-fixed paraffin- embedded tissue block does not rule out cutaneous TB because formalin

fixation has been found to decrease the PCR amplification signal. So, a fresh specimen for PCR has been recommended [14].

In this case, Gene-Xpert (PCR study) on formalin-fixed paraffin- embedded tissue block was negative, a result in keeping with many earlier studies where PCR is hardly done or rarely positive [9,11,12,14].

In general, an association between LV and chronically undiagnosed systemic tuberculosis is not uncommon [14]. Thus, our patient was commenced on standard antitubercular therapy (ATT) after obtaining a voluntary, informed consent from the mother. Two months following initiation of treatment, there was significant necrosis of the lesion and no new lesion was observed. We subsequently introduced minimal serial debridement of the necrosed tissue. While, ATT was ongoing, the lesion had almost completely cleared (Fig. 3).

In spite of the exhaustive laboratory studies, the diagnosis remained an enigma. With an exaggerated Mantoux test, commencing ATT resulted in convincing favourable response supporting the diagnosis of lupus vulgaris. It resulted in necrosis of the lesion within 8 weeks, an experience similar to findings by Verma et al [9]. ATT can help to achieve an early cure and prevent irreversible destructive lesions [12].

4. CONCLUSION

In low resource settings, a clinical and histological examination may not be adequate but are strongly helpful in diagnosing cutaneous tuberculosis. PCR (Gene Xpert) is strongly indicated although not always diagnostic. However, a strongly reactive Mantoux test and response to anti-tubercular therapeutic trial as observed in our patient is another strong diagnostic tool. There is the need to make a highly sensitive molecular diagnostic tool like PCR available and affordable for confirmation in all cases and discourage presumption as in the case of older modalities [9].

CONSENT

Our patient was commenced on standard antitubercular therapy (ATT) after obtaining a voluntary, informed consent from the mother.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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