



Case Report

Lupus Vulgaris of the Eye and Nose in a 10 Year Old Child: A Clinicopathological Diagnosis

JA. Ngbea¹, MO. Ochoga², KN. Malu³, BA. Eke⁴, R. Vhritherhire¹, BA. Ojo¹

¹Department of Histopathology, ²Department of Paediatrics, ³Department of Ophthalmology,
⁴Department of Surgery,
Benue State University Teaching Hospital, Makurdi, Nigeria.

Corresponding Author: JA. Ngbea

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ABSTRACT

Tuberculosis is still a major health problem in developing countries. Despite a high frequency of tuberculosis, Lupus Vulgaris which is the most frequent cutaneous form of tuberculosis is still uncommon. Lupus vulgaris originates from latent tuberculosis focus in the body and spreads by hematogenous or lymphatic routes, and by direct or exogenous inoculation. It is the most common morphological variant of cutaneous tuberculosis.

The most common site of involvement is the head and neck. It is therefore important to consider tuberculosis in the differential diagnosis of all eyes and nasal skin lesions and biopsies taken for histological studies.

The diagnosis was confirmed on histopathology of the biopsy taken from ulcerated part of the lesion. The response to three months of anti tuberculous drugs was dramatic. We hereby report an unusual case of vegetative Lupus vulgaris in a 10 year old Nigerian female involving the nose, upper lip and the right eye.

Keywords: Tuberculosis, Lupus vulgaris, nose, upper lip, right eye.

INTRODUCTION

Tuberculosis has recently reemerged as a major health concern. Each year, approximately 2 million persons worldwide die of tuberculosis and 9 million people are infected. [1]

The prevalence of tuberculosis is increasing because of the high number of patients infected with Human Immunodeficiency Virus (HIV), the resistance of Mycobacterium tuberculosis to medications, increased international travel and immigration from countries with high

prevalence, and the growing number of homeless and drug abusers. [2]

Tuberculosis is an infectious disease caused by the rod-shaped, non-spore forming aerobic intracellular bacilli measuring 0.5mm by 0.3mm. It is an acid fast bacilli with a unique cell wall which contains a fatty acid called mycolic acid providing an extraordinary lipid barrier. [3] This barrier is responsible for many of its medically challenging characteristics which include resistance to antibiotics and host defense mechanisms. The composition and quantity of cell wall components affect the

bacteria's virulence and growth rate. Another important component of the cell wall is lipoarabinomannan, a carbohydrate structural antigen on the outside of the organism that is immunogenic and facilitates the survival of mycobacteria within the macrophages. [4] Lupus vulgaris is a progressive form of cutaneous tuberculosis.

Nasal tuberculosis comes from haematogenous or lymphatic extension of pulmonary tuberculosis, the nasal mucosa is not usually affected, despite being a point of entry for the organism, inoculation is by scratching. [5] Other rare locations include sinuses, rhinopharynx, pinna and nose. [6]

Cutaneous tuberculosis constitutes approximately 1.5% of all extrapulmonary tuberculosis cases in childhood. [7]

The clinical and pathological lesions are varying from scrofuloderma to lupus vulgaris in cutaneous tuberculosis but the most common form of cutaneous tuberculosis is Lupus Vulgaris. [8]

Pathogenesis is hinged on entry of the organism into macrophages by endocytosis. Once inside the macrophage, the organism block fusion of the phagosome and lysosome by inhibiting Ca^{2+} signals.

T-helper 1 (TH-1) cell activates macrophages to become bactericidal and also stimulates the production of interferon- γ which is a cytokine that enables macrophages to contain Mycobacterium tuberculosis infection. INF- γ stimulates formation of phagolysosome within the macrophage that destroys the bacillus. It also stimulates expression of inducible nitric oxide synthase (iNOS) which produce nitric oxide (NO) capable of destroying the organism from its cell wall to DNA. TH-1 response also orchestrates granuloma formation and caseous necrosis.

CASE REPORT

A 10 year old female child presented at the pediatric outpatient department with a

7-month history of pappilomatous ulcerated lesions of the nose, upper lip and the right eye with eye discharge. There was no history of cough or weight loss, but a history of low grade fever usually at night.

On examination, she was a young female child that was not pale or in respiratory distress, but febrile ($37.8^{\circ}C$). Her respiratory rate was 16 beats per minute. There was no peripheral lymphadenopathy. A fungating, ulcerated skin lesion on the nose and the upper lip extending to the right eye with purulent conjunctivitis was found.

An initial impression of subcutaneous mycosis was made to rule out lupus vulgaris. A Mantoux test was done which revealed $> 15mm$ induration after 48 hours. The patient was not previously immunized with BCG. Full blood count revealed packed cell volume (PCV) as 30%. Erythrocyte sedimentation rate (ESR) was 14mm/hour. Retroviral screening was non-reactive. On subsequent follow-up visits, it was observed that the skin lesions were getting worse with the antifungal and antibiotics' (Fluconazole capsules 50mg twice daily per oral for two weeks, amoxicillian 500mg three times daily for ten days, fluconazole skin cream topically, ciprofloxacin eye drops four times daily for two weeks) medication so a biopsy of the lesion was carried out and a diagnosis of lupus vulgaris was made.

Histologic section showed a stratified keratinizing squamous epithelium overlying a fibrocollagenous dermis granulomata consisting of epithelioid cells in the dermis. Langhans giant cells with a rim of lymphocytes were also observed.

A short-course chemotherapy of three antituberculous drugs for the first two months (isoniazid 10mg/kg, rifampicin 10mg/kg pyrazinamide 30mg/1kg) followed by four months of isoniazid and rifampicin was started. Marked improvement of the lesions was noticed after three months on

treatment, but the patient was lost to follow-up visits.



Fig. I Ulcerated and fungating skin lesions involving the upper lip and nose



Fig. II Skin lesion involving the nose and the right eyelids with purulent discharge.

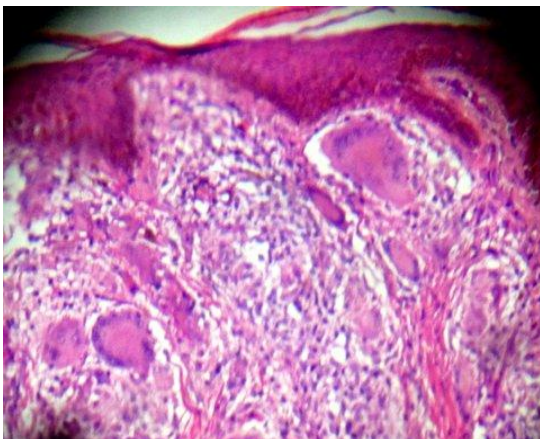


Fig. III Histologic section showed stratified squamous epithelium overlying a fibrocollagenous dermis with various sized granulomata consisting of epithelioid cells in the dermis. Langhan's giant cells with a rim of lymphocytes is also observed.(H and E X20)

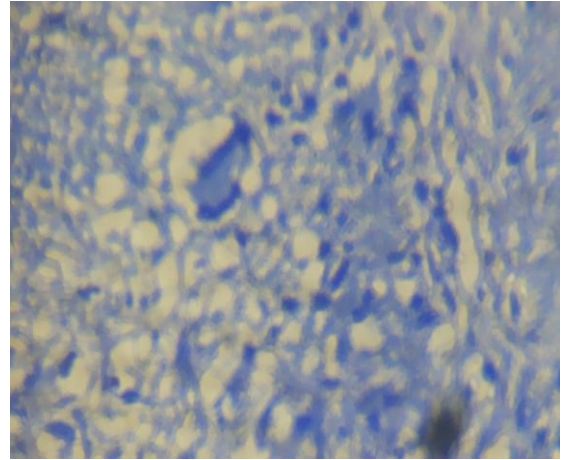


Fig IV Z/N stain positive for slender rods x 20.

DISCUSSION

Lupus vulgaris is a progressive form of cutaneous tuberculosis characterized by macules or papules with a brownish-red colour. Common areas of lupus vulgaris are the face, ears and neck may heal with scarring. [9] In Europe over 80% of lesions are on the head and neck, particularly on the nose and cheek. [10,11]

The incidence of lupus vulgaris has steadily declined over the past decades. The appearance of the infection due to HIV (Human Immunodeficiency Virus) has contributed to an upturn in the incidence of cases of tuberculosis in developing countries. Reports from Indian subcontinent have shown incidence of about 25%. [12]

Studies in Nigeria show that cutaneous tuberculosis such as lupus vulgaris is rare. [13] Another study showed that the incidence is on the increase due to prevalence of HIV/AIDS in Nigeria. [14]

The typical histological lesion is the lupus nodule and manifest with itching and bleeding nasal scabs. The most prominent feature is the formation of typical tubercle with sparse caseating necrosis. Secondary changes are often superimposed. Epidermal changes include thinning and atrophy or acanthosis with excessive hyperkeratosis and occasionally, pseudoepitheliomatous hyperplasia. Necrosis and ulceration are

usually accompanied by non specific inflammatory reactions that may partially conceal the tuberculous structures. Granulomatous reactions may develop with epithelioid cells. Nasal lesion is paucibacillary for which reason the nasal cultures tend to be negative in 50% of cases. DNA identification of the mycobacterium tuberculosis by means of polymerase chain reaction (PCR) is also recommended. [15]

Although the present case reports a common form of tuberculosis, its clinical appearance is unique. Usually lupus vulgaris presents as an erythematous plaque and/or nodules that grow with scarring. In this particular case the clinical appearance of the lesion is vegetative, which led to the misdiagnosis of a fungal infection. This appearance of a lupus vulgaris lesion is not commonly reported in a medical literature.

The differential diagnosis in lupus vulgaris includes sarcoidosis, deep fungal infections, tertiary syphilis, leprosy, Leishmaniasis, lupus erythematosus and lichen simplex chronicus. [16]

The Mantoux test is positive in most cases of Lupus vulgaris. When it is negative, however, it should be regarded as a sign of possible visceral involvement.

CONCLUSION

In conclusion, tuberculosis is still an important health problem in underdeveloped and developing countries due to the poor hygiene, low socio-economic level and accessibility of health care with increase morbidity.

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