

**CASE REPORT**

Lupus Vulgaris of Nose

Saurabh Varshney, Pratima Gupta*, S.S Bist, RK Singh, Nitin Gupta

Abstract

Lupus vulgaris represents a rare manifestation of infection by *Mycobacterium tuberculosis*. It is important to consider tuberculosis in the differential diagnosis of all nasal lesions and take biopsy samples for histological and bacteriological studies. Antitubercular chemotherapy is satisfactory with good result. We hereby report 3 cases of Lupus vulgaris of nose.

Key Words

Lupus Vulgaris, *Mycobacterium Tuberculosis*, Nose

Introduction

Lupus Vulgaris is a progressive form of cutaneous tuberculosis. Nasal tuberculosis (TB) comes mainly from the haematogenic or lymphatic extension of pulmonary TB; the nasal mucosa is not usually affected, despite being a point of entry for the *Mycobacterium tuberculosis*. They are usually secondary to inoculation by scratching (1). Other rarer localizations, including the sinuses, rhinopharynx, Pina (2) and nose, the latter being characterised by its infrequency and its clinical polymorphism. The variable ways in which it manifests itself mean that it is sometimes confused with granulomatous or neoplastic processes for which reason diagnostic suspicion is important (3). Nasal tuberculosis has been reported in past by few authors (4-10) but it remain a rare. In this study we present one case of Lupus vulgaris of nose and discuss the diagnosis and treatment modalities of this rare entity.

Case-1

A 18-year-old man with no relevant medical history came to the clinic, with a six month history of ulceroproliferative lesion on the nasal dorsum, along with nasal disfigurement. The rhinoscopic examination showed ulceroproliferative lesion (3cm x 4 cm) over nasal dorsum, ala and columella, nasal mucosal edema and abundant thick mucous that blocked both nasal fossae with normal nasal septum. (Fig 1) A simple radiological study of nose and paranasal sinuses, and a culture of the exudation were carried out without any pathologies found and empirical antibiotic treatment was begun for 10 days. On

subsequent visit worsening of the clinical symptoms and the appearance of granulomatous lesion was observed, for which a biopsy was carried out, that suggested chronic granulomatous inflammation with epithelioid granulomas including giant multinucleate cells and caseous necrosis (Fig 4), the analysis showed an increase in the rate of sedimentation. Acting on a diagnostic suspicion of TB, the Tuberculin test (Mantoux) was carried out which was positive, V.D.R.L. test for syphilis and Elisa for HIV were negative. Ziehl-Nielsen staining revealed acid-alcohol resistant bacilli and a culture on Lowenstein-Jensen medium showed a growth of *Mycobacterium tuberculosis*. The findings of the remaining ENT areas, neck, lung and general examination were normal, and thus the problem was diagnosed as primary nasal TB (Lupus vulgaris) and the case was treated with isoniazid, rifampicin and pyrazinamide for 3 months, followed by rifampicin and isoniazid for 6 months. The response to treatment was favourable, resulting in a complete resolution of the nasal lesion, except for contracture scar.

Case 2

A 24 years old male, reported with slowly progressive ulcerative lesion over nasal dorsum for the last 4 months, (Fig 2) without any history suggestive of pulmonary tuberculosis. General E.N.T. & Head neck examination was normal, with raised lymphocytes and ESR. Biopsy from the lesion was reported as Lupus vulgaris. Patient was successfully treated with 9 months treatment of antitubercular therapy.

From the Department of ENT & Head Neck Surgery and * Microbiology HIMS, Jolly Grant Dehradun UA India

Correspondence to : Dr Saurabh Varshney Professor & Head Dept. of ENT, HIMS, Jolly Grant Dehradun UA 248140 India



Figure 1-3. Showing Ulcero-Proliferative Lesion Over Nasal Dorsum, Involving Ala and Columella

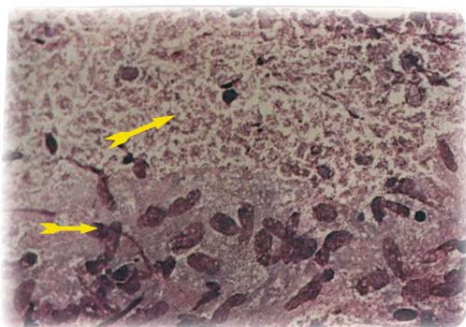


Figure 4. H-E (40X) The Epithelioid Granuloma Includes Giant Multinucleate Cells and Caseous Necrosis

Case 3

A 7+ years old boy came to E.N.T. OPD with a plaque lesion over nasal tip and supratip area for the last 7 months (Fig 3), without any history suggestive of pulmonary or cervical nodal tuberculosis. General E.N.T- Head neck examination with radiological examination of lungs was normal. On diascopy, lesion showed `apple jelly` type of nodules. Biopsy from the lesion suggested chronic granulomatous inflammation with epithelioid granulomas including giant multinucleate cells and caseous necrosis, hence a diagnosis of lupus vulgaris was made. Patient was treated with antitubercular therapy for 9 months with complete cure.

Discussion

Lupus vulgaris is a progressive form of cutaneous tuberculosis. The characteristic lesion is a plaque composed of nodules of apple jelly colour, which extends irregularly in some areas, while in others scarring occurs, causing considerable tissue destruction over many years (1).

Although the incidence of lupus vulgaris has steadily declined during 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 TB in developed countries. Reports from Indian subcontinent have shown incidence from 0.11% to 2.5%. (11). There is a greater prevalence of the disease in regions with a cool, humid climate, but the incidence is not influenced by rural versus urban residence. Females appear to be affected two or three times as often as males. In children also has been reported in India (11).

Lupus vulgaris is a post primary form of skin tuberculosis, arising in previously sensitized individuals with only moderate immunity. The lesions progress steadily and although spontaneous involution does occur, new lesions arise within old scars. Complete healing is rarely observed without therapy. Lupus vulgaris originates from tuberculosis elsewhere in the body by hematogenous, lymphatic or contiguous spread, rarely it may follow primary inoculation tuberculosis, or BCG vaccination. Frequently it develops from a tuberculous condition beneath the surface of the skin, and in about 30 percent of the cases it is preceded by scrofuloderma. Most often, lupus vulgaris develops after cervical lymphadenitis or pulmonary tuberculosis (4).

The lesions are usually solitary, but two or more sites may be involved simultaneously. In about 80% of the patients, the head and neck are involved. In general, lupus vulgaris is asymptomatic. The initial lesion is the lupus macule or papule, characterized by a brownish red colour and a soft consistency. Upon diascopy the infiltrate shows a diagnostic "apple jelly" colour and if the lesion is probed, the instrument breaks through the overlying epidermis. Early lesions measure only a few millimeters in diameter, they are rather ill defined, slightly raised or within the level of the skin, and may reveal a smooth surface or may be covered by a scale. Larger patches are formed by peripheral enlargement and coalescence of smaller papules, and further progression is characterized by an elevation of the lesions and a deeper brownish colour. The course of this disease is marked by ulceration and scarring, thus, its clinical manifestations



are diverse and a number of complications may ensue. `Plan forms` manifest as flat plaques with a serpiginous or polycyclic configuration, a smooth surface, or psoriasis form scaling. `Hypertrophic forms` may present as tumorous growths of soft consistency that exhibit a nodular surface or as epithelial hyperplasia with the production of hyperkeratotic masses. Edema, lymphatic stasis, recurrent erysipelas, elephantiasis thickening, and vascular dilatation may lead to gross deformity. In `ulcerative forms` the underlying tissue may be affected by progressive necrosis, and if the nasal or auricular cartilage is involved, extensive destruction takes place. Granulations at the floor of the ulcers lead to vegetating papillomatous lesions. Scarring is a prominent feature of lupus vulgaris. Atrophic scars occur subsequent to or independent of ulceration, and new `apple jelly` nodules may develop within the cicatricial areas. Sometimes scarring with excessive deformation and mutilation is very pronounced & keloid like fibrosis leads to contracture (3).

The typical histological lesion is the "lupus nodule" and it manifests itself as itchiness and bleeding nasal scabs. The most prominent feature is the formation of typical tubercle with sparse caseation necrosis. (Fig 4) 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 of the foreign body type may develop. Longstanding quiescent lesions are composed chiefly of epithelioid cells. Nasal lesion is paucibacillary for which reason the nasal cultures tend to be negative in 50% of the cases. DNA identification of the Mycobacterium tuberculosis by means of PCR (Polymerase Chain Reaction) is also recommended (4-10).

Typical lupus vulgaris plaques do not present diagnostic problems, they have to be distinguished from lesions of sarcoidosis, lymphocytoma, discoid lupus erythematosus, tertiary syphilis, leprosy, blastomycosis or other deep mycotic infections and chronic vegetative pyoderms. Criteria helpful in the diagnosis are the softness of the lesions, the brownish red colour, and the slow evolution. The `apple jelly` nodules revealed by diascopy are highly characteristic finding. Histologic examination is mandatory, and in some cases sparse acid fast bacilli can be demonstrated. Tuberculin test is strongly positive .

Lupus vulgaris is chronic and without therapy its course usually leads to considerable disfigurement. The most serious complication of long-standing lupus vulgaris is the development of carcinoma (12). Early in this century this complication was estimated to be almost 10%. Squamous cell carcinoma outnumber basal cell carcinoma.

Standard antitubercular therapy should be given. Fortunately, there is a satisfactory response to medical treatment of nasal TB, which is the same as that prescribed for its classical pulmonary form: isoniazid (5-7 mg/kg/day) rifampicin (10-20 mg/kg/day) and pirazinamide (15-30 mg/kg/day) for 2-3 months, followed by isoniazid and rifampicin for 4-6 months. In the case of suspected resistance to isoniazid (in developing countries) ethambutol is prescribed. Therefore medical treatment is satisfactory and free from resistance in all cases. Surgery is left exclusively for the treatment of residual lesions. Nodular areas persisting after treatment can be destroyed by diathermy (1,6).

References

1. Hup AK, Haitjema T, de Kuijper G. Primary nasal tuberculosis. *Rhinology* 2001; 39(1): 47-48
2. Varshney S, Prasad D. Lupus vulgaris of pinna. *Indian Journal of Otolaryngology*. 1998;4(4):191-92
3. Sim DW, Crowther JA. Primary nasal tuberculosis masquerading as a malignant tumour. *J Laryngol Otol* 1988; 102(12): 1150-52.
4. Choi YC, Park YS, Jeon EJ, Song SH. The disappeared disease: tuberculosis of the nasal septum. *Rhinology* 2000; 38(2): 90-92.
5. Gentic A, Garre M. Nasal tuberculosis, two cases in elderly patients. *Clin Infect Dis* 1992; 15(1):176-77.
6. Medina A, Robleda E, García M. Tuberculosis nasal. *Anales Otorrinolaringológicos Iberoamericanos* 1982;9(4): 251-58.
7. Paredes C, Del Campo F, Zamarrón C, Del Valle A. Nasal tuberculosis: a rare entity. *Anales de Medicina Interna* 1990; 7(11): 602.
8. Ennouri A, Hajri H, Bouzoua N. Tuberculosis de las cavidades nasales y paranasales. *Encyclopédie Médico-Chirurgicale Otorhinolaryngologie* 2000; 20- 375-A-10
9. Serra J, Moliner M, Molinero A, Ramírez J. Tuberculosis nasal. *Med Clin* 1990; 95(10): 396.
10. Aguilles MJ, Fornes. Primary nasal tuberculosis. *Acta Otorrinolaringol Esp* 2004; 55: 240-243
11. Sethuraman G, Ramesh V, Ramam M, Sharma VK. Skin tuberculosis in children: Learning from India. *Ind Dermatol Clin* 2008; 26(2) 255-95
12. Haller D, Reisser C. Lupus Vulgaris manifestation as a destructive nose and facial tumor. *HNO* 2008, (Ahead of Print).