Lupus Vulgaris of Nose: A Case Report

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Abstract: Lupus Vulgaris is a nodular form of cutaneous tuberculosis. Nasal tuberculosis is almost invariably caused by Mycobacterium tuberculosis. Frequent contamination of nasal fossae by Mycobacterium tuberculosis is common in smear positive pulmonary TB. Clinical disease however is extremely rare and in Head and Neck TB, involvement of cervical lymph nodes, larynx, pharynx and ear is more common than nasal involvement. Lupus Vulgaris is rare manifestation of Mycobacterium tuberculosis infection. Furthermore, skin and nasal manifestation of lupus vulgaris makes it difficult to differentiate this clinical entity from other granulomatous condition of nose. But once diagnosed, response to standard six month AKT treatment is satisfactorily with good results.

Objective: Hereby we report rare case of Lupus Vulgaris of Nose and demonstrate clinical analysis in differential diagnosis of Lupus Vulgaris.

Case Report: A male patient presented with ulceroproliferative lesion over nose and upper lip. On clinical features, histopathological examination and highly positive tuberculin test, diagnosis of Lupus Vulgaris was made. AKT (6 months regimen) was started and ulcerated plaque was completely resolved.

Key Words: Lupus Vulgaris, Nose, Anti-Koch’s Treatment (AKT)

I. Introduction

Lupus Vulgaris is chronic, progressive and tissue destructive form of cutaneous tuberculosis seen in patient with moderate or high immunity. The lesion is confused with granulomatous and neoplastic diseases for which diagnostic suspicion is important. In this study we present a case of Lupus Vulgaris of Nose and discuss the Investigation, diagnosis and management modalities of this rare entity.

II. Case Study

A 16 year old male patient came to ENT OPD of our hospital with H/o ulcerative lesion over the external nose gradually increasing since 2 months. It started as papule over alar region, then extending over the Nasal tip, Vestibule, Bilateral Naso-labial fold, Philtrum and Upper lip in span of 2 month forming Ulceroproliferative lesion. It was associated with crusting, foul smelling discharge and bleeding after removal of crust. He was treated with antibiotics by family physician without any symptomatic improvement. On Examination, the tip, alar region, Bilateral naso-labial area, philtrum and upper lip showed ulceroproliferative lesion covered with Brown crust with nasal mucosal edema, thick mucous that blocked both nasal fossae with normal septum. Diagnostic Nasal Endoscopy revealed thick crusting, mucosal edema with normal septum. Diascopy showed "Apple Jelly Nodules". There was no regional or distant lymphadenopathy, general, physical and systemic condition was normal. There was no personal or family history of tuberculosis. All the haematological and bio-chemical investigation were normal except Raised Erythrocyte sedimentation Rate (ESR – 50 mm/1st hr.). The Mantoux test (intradermal tuberculin test) positive 20 x 22 mm. Liver function test, Urine Analysis and Chet X-ray were normal. Biopsy of Ulcerated plaque showed non-caseating epithelioid cell granuloma with langerhans cells in dermis, surrounded by lymphocytes. Lowenstein-Jenson culture showed Mycobacterium tuberculosis bacilli. Involvement of nerve fibers by granuloma or features of vasculitis were not observed. Special stains for fungi, bacterial, acid fast bacilli, mycobacterial and fungal cultures were negatives. Serology VDRL for syphilis, ELISA for HIV 1 & 2, sputum for acid fast bacilli and ANCA (Anti-nuclear
cytoplasmic antibody), LD (Leishman Donavan) Bodies were negative, 3 sputum examination for AFB were negative.

However based on clinical feature, Histology, Highly positive tuberculin test, Lupus Vulgaris was diagnosed.

AKT (Anti-Koch's Therapy) (6 month regimen) 2 HRZ + 4 HR was started. Clinical improvement was observed in 15 days. At the end of six month of therapy, the ulcerated plaque had completely resolved. (Fig III)

III. Discussion

Lupus vulgaris is progressive form of cutaneous tuberculosis, caused by Mycobacterium tuberculosis, acquired either endogenously by haematogenous or lymphatic spread from underlying distant focus or exogenously by direct inoculation of the bacilli or at the site of Bacille calmette-Guerin (BCG) Vaccination in previously sensitized individual with moderate immunity.

The condition is more common in males than females. Clinically it is characterized by soft, reddish-brown plaques with "Apple Jelly nodules" in diascopy.

The course of this disease is marked by ulceration and scarring, thus its clinical manifestation are diverse and number of complication may ensure.

The diverse clinical forms of Lupus Vulgaris include papular, nodular plaque, ulcerative, vegetative and tumour like lesion.

In India, buttocks, trunk and extremities are predominant sites affected, in the West the lesions favor Head and neck. The disease commonly affects the nose and nasal cartilage. The nasal bones are usually spared. Ulceration, Necrosis and scarring occur with destruction of deeper tissues and cartilage leading to gross deformities and contractures. Direct extension or lymphatic spread from the nasal focus may also involve the soft and hard palate, gingiva, larynx and pharynx.

A Deep tissue Biopsy is essential for histological examination.

The most common feature is the formation of typical tubercle with sparse caseation necrosis. Necrosis and ulceration are usually accompanied by non specific inflammatory reaction that may partially conceal the tuberculosis structures.

Long standing quiescent lesions are composed chiefly of epitheloid cells and langhan's giant cells in papillary and upper reticular dermis (Fig I).

Figure I Haematoxylin & Eosin Stain (40 X magnification)

IgG Antitubercular Antibodies and PCR (Polymerase Chain Reaction) DNA of mycobacterium are also recommended when histopathology is inconclusive. Typical Lupus Vulgaris plaques are confused and have to be distinguish from various infective & non infective disorders like sarcoidosis, lymphocytoma, discoid lupus erythematosis, tertiary syphilis, blastomycosis, leshmaniasis, Rhinoscleroma, Wegener’s Granulomatosis, Midline granuloma, deep mycotic infection and chronic vegetative pyoderma.

Lupus Vulgaris criteria helpful in diagnosis are softness of lesions, the brownish red colour and slow evolution. The "Apple Jelly" nodules on diascopy are highly characteristic finding. Histological examination is mandatory. Tuberculin test is highly positive & ESR is raised.

In atypical mycobacterial infection, culture characteristic help to reach correct diagnosis. In Leishmaniasis, Leishman Donovan bodies representing parasite is grown on culture. In Rhinoscleroma caused by Klebsiella rhinoscleromatis, shows characteristic Miculikz cells in deep dermis.
In Hansen's disease loss of lateral eyebrows and diffuse infiltration of face and ear lobes are unique to leprosy. Syphilis rule out by serological & special histological stains.

Among non-infective disorder, nasal septal defects can be observed in Wegener’s granuloma (WG), Midline granuloma & Sarcoidosis. Wegener’s granuloma, a systemic vasculitis, primarily involves upper and lower respiratory tract and kidney, shows Antineutrophil Cytoplasmic antibody (ANC) test positive and presence of necrotizing granulomatous vasculitis on histology.

Cutaneous sarcoidosis i.e. Lupus Pernio may be confused with Lupus Vulgaris. There may be intra thoracic involvement along with bone and eye lesions. Histology and radiology help in diagnosis.

Thus Lupus Vulgaris is correctly diagnosed by high index of clinical suspicion helped by morphology and histology and appropriate investigation to rule out other simulating disorder.

Standard Anti-Koch’s treatment given is
- Isoniazid (H) (5-7 mg/kg/day)
- Rifampicin (R) (10-20 mg/kg/day)
- Pyrazinamide (Z) (15-30 mg/kg/day) for 2-3 months followed by, Isoniazid & Rifampicin for 4-6 months.

AKT can help to achieve an early cure and can prevent the irreversible destructive lesions.

Summary
- Lupus Vulgaris of nose is rare condition.
- Histopathological Examination of Deep Biopsy from Ulcerated lesion is key to diagnosis.
- Confused with other granulomatous and neoplastic disease for which diagnostic suspicion is important.
- Medical Management by Anti Koch’s therapy gives excellent prognostic Result.
References