Case Report

Rhinoscleroma with laryngotracheal involvement: a case report

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ABSTRACT

Rhinoscleroma is a chronic, slowly progressive, granulomatous inflammatory disease of the upper respiratory tract. It is more common in developing countries and rural areas and endemic in Asia, Africa and South and Latin America. We report a case of rhinoscleroma in a young male presented to our Outpatient Department with complaint of hoarseness since one and half years. He had history of dyspnoea on exertion. Patient also had history of bilateral nasal discharge and obstruction. A nasal specimen was taken for histological diagnosis which confirmed rhinoscleroma. Patient was put on medical management and was assessed weekly for improvement by nasal endoscopy and laryngeal examination for one year. A high degree of suspicion is required by clinician to diagnose the disease and prompt treatment should be given to avoid the progression of disease and complications. Patient requires long term follow up for proper management.

Keywords: Rhinoscleroma, Hebra nose, Klebsiella rhinoscleromatis, Mikulicz cells, Larynx, Trachea

INTRODUCTION

Rhinoscleroma or scleroma is a chronic granulomatous disease caused by gram negative bacillus called Klebsiella rhinoscleromatis or Frisch bacillus which is a gram negative, non-motile bacterium. The disease was first described by the dermatologist Ferdinand Von Hebra in 1870. 1 The name of the disease can be derived from the greek term skleroma which denotes a hard nodular change. In India it is endemic in Madhya Pradesh, Uttar Pradesh, Rajasthan and Karnataka. The endemic regions are characterised by common environmental factors, such as low level of hygiene, malnutrition, and overcrowded housing. Women are more frequently affected (1.3:1). The inflammatory process usually starts in the nasal cavity and then spreads on to nostrils, pharynx and larynx. Here we are presenting a case of rhinoscleroma with laryngotracheal involvement in a young patient.

CASE REPORT

A 20-year old male patient, resident of Uttar Pradesh, farmer by occupation, presented to our OPD with complaint of hoarseness since one and half years. He had history of dyspnoea on exertion. Patient also had history of bilateral nasal discharge and obstruction and nasal bleed. There was no history of substance abuse, cough, weight loss, voice fatigue or vocal abuse. Diagnostic nasal endoscopy showed bilateral posterior choanal stenosis along with foul smelling mucopurulent discharge and crusting (Figure 1).

On indirect laryngoscopy, bilateral true vocal folds were edematous and hemorrhagic, bilateral arytenoids were congested and edematous and only posterior chink around 5 mm was present (Figure 2).

X-ray soft tissue neck (STN) was done which showed significant airway obstruction (Figure 3).
Keeping in mind the strong suspicion of granulomatous pathology, contrast enhanced computed tomographic (CECT) scan neck was done which showed diffuse mucosal and submucosal thickening with mild post contrast enhancement at level of false vocal cords, glottis and subglottic region and upper part of trachea up to level of T2 vertebra with significant lumen narrowing (Figure 4 and 5).

Nasal biopsy was taken and histopathological examination (HPE) came out to be rhinoscleroma (Figure 6).

Patient was admitted and kept under observation. He was started on oral ciprofloxacin 500 mg twice a day and tetracycline 500 mg four times a day. He was assessed weekly by nasal endoscopy and laryngeal examination. We noted significant improvement in symptoms after two weeks of treatment.

Serial X-ray STN lateral view done at one month interval (Figure 7).

Repeat nasal biopsy was done at 4th week and 8th week, both of which showed reduction in inflammation. Patient
Rhinoscleroma is a chronic infectious disease caused by Klebsiella rhinoscleromatis, a gram-negative, facultative intracellular, non-motile encapsulated bacillus identified by Von Frisch. The disease primarily involves mucosa and submucosa of nasal cavity but may involve larynx, especially the subglottic region and trachea. Laryngeal involvement has been reported in approximately 15 to 80% of cases. In our case patient presented with rhinoscleroma of nose with involvement of larynx. Cases of rhinoscleroma of lower respiratory tract, maxillary sinuses, orbital cavity, lacrimal ducts and cervical lymph nodes have also been reported.

The disease can be divided into three morphological stages: catarrhal and atrophic, granulomatous or proliferative and sclerotic. Disease starts with acute exudative inflammation of the nasal mucus membrane with serous or mucus secretion. At second stage, characteristic granulation tissue is seen with nodules in the nasal cavity, which clinically can imitate cancerous tumors. Deformities of tip of nose are sometimes observed. At third stage, inflammatory tissue is replaced by connective tissue with thick cicatricial strips.

Microscopically, large macrophages with light, foamy cytoplasm known as Mikulicz’s cells, plasma cells and lymphocytes are seen. Plasma cells with hyaline degeneration called Russel bodies can also be seen.

Rhinoscleroma spreads from person-to-person by airborne transmission. The pathogenesis of rhinoscleroma is still unclear. An alteration in CD4:CD8 population in blood has been postulated as a cause of chronicity of this disease. The altered proportion of CD4+ and CD8+ lymphocytes in the lesion may produce disabled macrophages, allowing bacterial multiplication inside them and an ineffectual delayed type hypersensitivity response. A positive culture in MacConkey agar is diagnostic of rhinoscleroma, but it is positive in only 50-60% of patients. The diagnosis is confirmed by histology. Differential diagnosis includes other granulomatous conditions, e.g., tuberculosis, leprosy and fungal infections. Treatment consists of oral or intravenous antibiotics like tetracycline and cephalosporins. Surgical procedures like laryngeal dilatation, endoscopic resection and tracheostomy may be required during sclerotic phase. Untreated rhinoscleroma may cause death owing to airway obstruction.

CONCLUSION

A high degree of suspicion is required by clinician to diagnose the disease and prompt treatment should be given to avoid the progression of disease and complications. Patient requires acute as well as long term follow up for proper management.

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