ORIGINAL RESEARCH PAPER

Volume-7 | Issue-9 | September-2018 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

# RHINOSCLEROMA: A CASE REPORT

Otolaryngology

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## ABSTRACT

Rhinoscleroma is rarely seen these days. We report a case of unilateral rhinoscleroma in 62 years old female, she was diagnosed in proliferatral stage and put on medical treatment, however, the disease still progressed to fibrotic stage and required surgery for unilateral nasal obstruction.

## KEYWORDS

## INTRODUCTION

Rhinoscleroma (Hard Nose) is chronic slowly progressive, granulomatous bacterial disease that usually affects the respiratory tract mucosa and other organs nearby. It is rarely seen in developed countries. Several factors have been associated with rhinoscleroma like – more frequent among second or third decades of life, living in overcrowded conditions, rural population, poor hygiene, iron deficiency anemia and female sex ( female: male ratio is 13: 1). There is no racial predisposition and mostly there is bilateral involment [1, 2]. Clinical course is slow and insidious, usually over many years and patient's general health is unaffected.

It is caused by Klebsiella rhinoscleromatis (subspecies of Klebsiella pneumoniae) a gram negative, encapsulated, non lactose fermenting, non motile, rod shaped bacillus (diplobacillus), which is a member of enterobacteriaceae family [3]. The disease process start at the vestibule of the nose and spreads by means of direct inhalation of droplets or contaminated airborne material which are otherwise usually expelled by coughing or sneezing. The bacterium is not a typical resident in the respiratory tract and skin microbiota, however, it has affinity for nasal mucosa [4].

We report an unusual case of unilateral rhinoscleroma in a female with very short history which was refractory to medical treatment.

## CASE REPORT

A 62 years old female presented with a history of left side nasal discharge, blockage, facial pain on same side and low grade fever for last three months. She was treated by general practitioners as a case of chronic rhinosinusitis and vestibulitis with some relief. She had recurrence of symptoms after discontinuation of the antibiotics and she developed narrowing of the left nostril with disfiguring and hardening of skin of alar region.

General physical examination was essentially normal and she was immuno-competent. Otorhinolarynological examination revealed mild DNS to right side, watery discharge and narrowing of left nostril in the alar region. Airflow on left side was also reduced. X ray PNS OM view showed mild DNS and clear sinuses .Diagnostic endoscopy was performed and no abnormality noted except mild DNS.Biopsy was taken and reported as rhinoscleroma. Biopsy showed pseudostratified ciliated columnar to squamous epithelium with marked infiltrations by foamy histiocytes and other inflammatory cells in subepithelial zone.

The foamy histiocytes and macrophages (Mikulicz cells) (Fig-1).

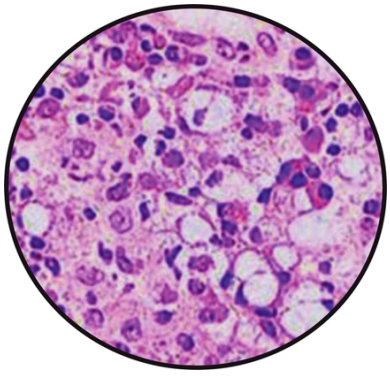

**Fig.1 Lesion showing foamy histiocytes and macrophages (Mikulicz cells). H&E, X400.**

She was put on tablet ciprofloxcin 500 mg BID and capsule rifampicin 450 mg OD for four months, however, the narrowing and hardening of alar region rather increased. Surgical debridement of woody hard tissue from the alar region was done and grafted with post auricular free split skin graft. A piece of endotracheal tube was placed in the left nostril for three weeks (Fig-2).

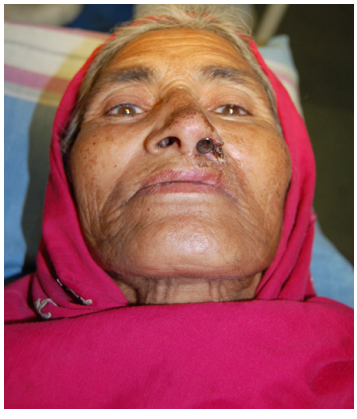

**Fig.2 Post- operative clinical Photograph showing tube in left nostril.**

She was put on ciprofloxcin 500mg BID and rifampicin450mg OD for two weeks. On follow after six years she was symptoms free with normal nasal airway (Fig-3).

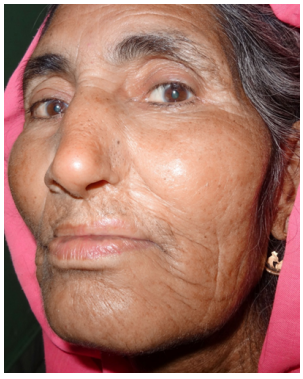

**Fig.3 Clinical photograph after six years.**

## DISCUSSION

Von Hebra (1870) described the rhinoscleroma as ancient disease. The Polish surgeon Johann von Mikulicz[2] (1877) reported the histological feature; von Frisch identified the organism in 1882 [5]. The occurrence in families suggests that genetic control of the host response to Klebsiella rhinoscleromatis may be important factors in endemic areas. The disease is rarely seen in United States, however, most cases have been reported from poorer regions such as Central Africa, Central and South America; Eastern & Central Europe, middle east, India and Indonesia. In recent years some cases in non- endemic areas were reported, which was explained by the increased migration of population over the world [6].

The disease commonly affects the nasal cavity (95-100%), but it can also affect the nasopharynx (18-43%), larynx (15-40%), trachea (12%) and the bronchi (2-7%) [7,8]. The oral cavity, paranasal sinuses and soft tissue of lips and nose can also be involved. In rare cases, rhinoscleroma spreads to orbit. Pathologically it has three histologic stages of development of disease. In the catarrhal stage, the mucosa has non specific inflammatory changes with neutrophils, cellular debris and granulation tissue; proliferative stage has an intense infiltrate of plasma cells and large foamy histiocytes termed 'Mikulicz cells' which contain numerous rod shape bacteria within their cytoplasm. In last stage, lesion undergoes fibrosis developing large firm intramucosal nodules which can lead to bony invasion and airway obstruction. Fully developed rhinoscleroma, can lead on to external expansion of the nose known as 'Hebra nose'or woody nose [9].

The differential diagnosis of rhinoscleroma includes leprosy, sarcodosis, lymphoma, tuberculosis, mucocutaneous leishmaniasis, various mycotic infections and histiocytosis with invasive lymphadenopathy[10]. Diagnosis is usually made by the identification of Mikulicz cells in tissue biopsy. The bacteria may be seen in H&E stained section, but are more easily identified with PAS, Geimsa or Warthin- Starry stains [6]. Tissue biopsies in latter stages are often non diagnostic. Immunoperoxidase stains for Klebsiella capsule antigen sensitivity have increased sensitivity. The bacteria may also be identified by culture of mucosal lesions [5].

Rhinoscleroma is treated medically with prolonged antibiotic therapy to eradicate the bacteria, because the relapse rate is very high. In vitro, these bacteria are inhibited by clinically achievable concentrations of amoxicillin- clavulanate, chloremphenicol, trimethprim- sulfameth axazole, cephalosporins, streptomycin, tetracyclines and ciprofloxin . However, in vivo antibiotics with demonstrated efficacy are streptomycin, doxycline, tetracycline, rifampicin, second & third generation cephalosporin, sulphonamides, clofazimine, ciprofloxacin and ofloxacin [1,2, 11]. Many workers published studies on duration of therapy from six weeks to six months, until repeated cultures and histological examinations are negative[12]. Topical antibiotics such as acriflavin 2 % or rifampicin have been used with significant results [13], however, we did not use topical antibiotic in this case. We used ciprofloxacin and rifampicin in the present case.

Granulomatous tissue, crusting and fibrotic scarring are lesions that produce various degree of airway obstruction and cosmetic deformity which may need surgical intervention. Carbondioxide surgical laser has been shown to be of great value in few cases [14]. Prophylaxis is based on the improvement of sanitary conditions and general living conditions of poor people of endemic area.

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