Rhinoscleroma: A Look at an Unusual Fascinating Disease from the Tropics

Review of the Literature and Report of 2 New Cases

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Abstract Rhinoscleroma is an unusual disease that is seen in many parts of the tropics. It is caused by a bacteria called Klebsiella rhinoscleromatis and can leave the patient with a chronic inflammatory lesion of the upper respiratory airway and much disfigurement. In the age of global migration and travel physicians must learn to recognize this disease especially in immigrants and patents that come from countries endemic of the disease. It is endemic in parts of Mexico, Central and South America, Central and Eastern Europe, Southeast Asia and Africa. Humans are the only identifiable host and the transmission is thought to be airborne. This disease is mostly seen in the poor rural areas of the world where overcrowding and poor hygiene are common.

Keywords Rhinoscleroma, Klebsiella Rhinoscleromatis, Respiratory Involvement

1. Introduction

Rhinoscleroma is an unusual disease that is seen in many parts of the tropics. It is caused by a bacteria called Klebsiella rhinoscleromatis and can leave the patient with a chronic inflammatory lesion of the upper respiratory airway and much disfigurement. In the age of global migration and travel physicians must learn to recognize this disease especially in immigrants and patents that come from countries endemic of the disease. It is endemic in parts of Mexico, Central and South America, Central and Eastern Europe, Southeast Asia and Africa. Humans are the only identifiable host and the transmission is thought to be airborne. This disease is mostly seen in the poor rural areas of the world where overcrowding and poor hygiene are common.

Ferdinando von Habre first described the disease in the 1870s (2) and then the histological features were more fully described by Mickultz in 1877 (3). The causative agent Klebsiella rhinoscleromatis was described by Von Frisch in 1882. (4) This paper describes the epidemiology, clinical features, pathology and treatment of this unusual disease in addition to presenting two new cases.

Case report 1

A 44-year-old Guatemalan male who presented with a four-year history of a progressively enlarging mass in his nasal cavities. (Fig 1) He had no associated fevers, chills or weight loss. On examination he presented with a large mass that had a rubbery consistency occluding both his nasal passages. These were non-tender, firm and did not bleed on touch. The rest of the physical examination on admission was unremarkable. He underwent a biopsy of the nasal mass, which revealed Mickultz cells, and the cultures eventually grew Klebsiella rhinoscleromatis. The CT scan showed no evidence of bone involvement. He was treated with a 6 months of ciprofloxacin and rifampin with resolution of the masses.

Case Report 2

A 24-year-old female from Guatemala with a mass in both her nasal cavities, which she had for almost 2 years. She had no associated fevers, chills or night sweats. Clinical examination revealed occlusion of both the nasal cavities by a large rubbery mass. The rest of the examination was normal. The labs were unremarkable at the time of the admission. Biopsy revealed Mickultz cells and tissue cultures were positive for Klebsiella rhinoscleromatis. She

Figure 1. Foamy Macrophages-mikulics Cells
was treated with a six-month course of trimethoprim-sulfamethoxazole with resolution of the masses.

2. Discussion

Rhinoscleroma is an unusual disease and rather difficult to diagnose and are typically found in Central and South America, some parts of Europe and Asia. With the advent of global travel and immigration the expansion of these endemic diseases from one country to another may not become sight.

The typical presentation sites include the nasal mucosa (approx. 95 %), pharynx in 18 -43%, paranasal sinuses, trachea and bronchi in the remaining (5,6). Very rarely is there involvement of the adjacent cartilage and bone structures. In a study published in 2008, 11 cases where evaluated. The median age at the time of diagnosis was 35.7 years; three of the 11 patients had a familial history with an early onset. The probable duration of exposure in these endemic areas seems to vary widely between 0 - 28 years. The clinical features and outcomes also varied considerably among the cases. The biopsies revealed granulomas containing Mickultz cells. In addition cultures of the biopsy tissue were positive for K.R in five of the 11 cases (7). Russell bodies are structures within the cytoplasm of the plasma cells and are found characteristically during the granulomatous stage.

The pathophysiology of Rhinoscleroma is not well understood at this time. It may or may not have an underlying immune deficiency problem, a genetic problem or none of the above. *K. rhinoscleromatis* is a subspecies of *Klebsiella pneumonia*, which is a gram-negative, non-motile coco-bacillus that belongs to the *Enterobacteriaceae* as subgroup (*Klebsiella, Enterobacteriaceae, Serratia.*) Russell bodies are structures within the cytoplasm of the plasma cells and are found characteristically during the granulomatous stage.

The differential diagnosis includes other granulomatous, neoplastic and other conditions such as sarcoidosis, Wegener's granulomatosis, vasculitis, lymphoma, basal cell cancer, carcinoma, *actinomycetes, para - coccidiomycosis*, *Leishmania*, leprosy and tuberculosis.(12,13)

The diagnosis can be made with the brush biopsy of the mass or incisional biopsy sent for cytology and culture. A positive culture for *Klebsiella rhinoscleromatis* in the blood or McConkey agar is diagnostic of *K. rhinoscleromatis* in about 50 to 60% of the patients. The bacteria can also be seen using the periodic acid Schiff, Giemsa or Warthin Starry stains. (14,15)

Imaging studies are not very helpful in the diagnosis. CT scans occasionally show a homogeneous non-enhancing mass with distinct margins; occasional bony or cartilaginous erosion can be seen. MRIs may occasionally show bony destruction and show high signal intensity on the T-1 and T2 weighted images in the hypertrophic stages.

6. Treatment

Rhinoscleroma remains a very difficult condition to treat and relapses are common. Treatment consists of a combination of surgical debridement and long-term antibiotic therapy. Treatment is also stage dependent. In the early years, streptomycin was widely accepted as the drug of choice however other antibiotics such as the tetracycline group, trimethoprim sulfamethoxazole and the quinolones gradually became useful in the treatment of these patients. It appears that six to nine months of TMP-SMX or a quinolone
along with rifampin tends to prevent relapse and effects a more permanent cure. (15), There have been combinations of trimethoprim and sulfamethoxazole with rifampin used in endemic areas that have shown improved results in cure and treatment. The quinolones appear to achieve much higher tissue penetration and seem to concentrate within the macrophages results in resulting in bactericidal activity against *Klebsiella rhinoscleromatis* (15). In our patients the outcome appeared to be the same regardless of the antibiotic used.

Indications for surgery include relief of airway obstruction and reconstruction of defects (16). Occasionally tracheostomy should be considered a laryngeal obstruction in either the granulomatous or the sclerotic stage. Occasionally reconstructive surgery may be needed for nasal or lower airway stenosis from scarring or perforation. Carbon dioxide laser therapy is currently the most effective surgical method for eradication of clinical or histological disease. This form of therapy has been used to treat hypertrophied turbinates and in many cases used to treat cases of Rhinoscleroma that are not amenable to surgery.

**REFERENCES**


