Section: Otorhinolaryngology

Case Series

A CASE SERIES ON RHINOSCLEROMA AT TERTIARY CARE HOSPITAL AT HYDERABAD

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ABSTRACT

Rhinoscleroma is a long-lasting, progressive inflammatory disorder affecting the upper respiratory tract, linked to the Klebsiella rhinoscleromatis infection. This case series outlines the clinical and pathological characteristics seen in four patients diagnosed with rhinoscleroma at the Deccan College of Medical Sciences in Hyderabad from 2020 to 2024. Each patient exhibited cutaneous symptoms exclusively, and histological analysis confirmed the diagnosis. The patients were treated with a combination of antibiotics. Due to the rarity of rhinoscleroma in Hyderabad, diagnosing it can be quite challenging. This report aims to increase awareness about the disease to facilitate early diagnosis and treatment for those affected. Without proper intervention, rhinoscleroma can result in serious complications, such as involvement of the lower airways. Therefore, early detection and prompt treatment are crucial for reducing the morbidity associated with this condition.

Keywords: rhinoscleroma, nose, nasal, scleroma, klebsiella rhinoscleromatis

INTRODUCTION

Rhinoscleroma is a chronic and progressive inflammatory condition affecting the upper respiratory tract. The name "skleroma," originating from Greek in 1932, means hard swelling, highlighting its impact on respiratory health. The condition was first described by Ferdinando Von Hebra in the 1870s, with its histological characteristics detailed by Mikulicz in 1877. In 1882, Von Frisch identified Klebsiella (C.) rhinoscleromatis as the causative agent.

This disease primarily occurs in rural areas with poor living conditions, notably in parts of Africa, Southeast Asia, Mexico, Central and South America, and certain regions of Central and Eastern Europe. It spreads through airborne transmission and is exclusive to humans. Overcrowding, inadequate hygiene, and malnutrition increase the risk of developing rhinoscleroma. It is seldom seen on other continents, and when cases emerge outside endemic regions, they are typically linked to individuals from those areas. The disease mainly affects the nasal cavities (95%-100%), as well as the throat (18%-43%), sinuses, trachea, and bronchi.

Rhinoscleroma is divided into three stages: catarrhal, proliferative, and fibrotic.

In the catarrhal stage, patients commonly experience a foul-smelling, thick nasal discharge and nasal obstruction. Doctors may observe crusty, eroded areas in the nasal lining, and microscopic analysis shows alterations in nasal lining cells along with an increase in particular white blood cells.

During the proliferative stage, patients may have nosebleeds, altered nasal shape, a hoarse voice, loss of smell, and excessive tearing. On examination, physicians might find swollen, bluish-red masses in the nose, and tissue analysis reveals specific cell types indicative of this stage.

The fibrotic stage is marked by significant nasal deformity and constriction due to extensive scar tissue, with microscopic examination demonstrating considerable fibrous tissue.

This report discusses four cases of rhinoscleroma in middle-aged women from Hyderabad with limited travel history. These instances differ from previously recorded cases, especially in regard to the patients' living conditions and the disease's skin manifestations. The goal of this article is to aid physicians in early diagnosis for effective treatment and to avert serious complications. Additionally, we highlight the challenges in treating rhinoscleroma and the potential complications that may arise.

****Case Reports****

This report discusses four female patients, aged between 31 and 70, who experienced symptoms of their condition for 1 to 5 years before seeking help at the Deccan College of Medical Sciences in Hyderabad. Each patient presented only with skin symptoms and was otherwise healthy, with no swollen lymph nodes in the neck.

In all four instances, doctors confirmed the diagnosis through laboratory tests, identifying large cells known as Mikulicz cells, which contained numerous small empty spaces and either live or dead bacteria. They also discovered Russell bodies, characteristic structures in plasma cells. The bacteria's presence inside these cells was confirmed using specialized stains, namely Periodic acid-Schiff (PAS) and Giemsa stains. The specific infection, C. rhinoscleromatis, was verified using a silver stain test known as Warthin-Starry stain.

Case 1 involved a 63-year-old Indian woman with a red and yellow bump on her left nostril. A nasal swab for C. rhinoscleromatis returned negative. She began antibiotic treatment with ciprofloxacin (500 mg twice daily) and doxycycline (100 mg twice daily). After three weeks, her condition showed significant improvement.

Case 2 details a 65-year-old Indian woman with a red bump on her right cheek. She was treated with ciprofloxacin (500 mg twice daily) for a month, after which doxycycline was added (100 mg twice daily). The antibiotics proved effective, and she was continuing the treatment while this paper was written, aiming for a total of six months.

Case 3 concerns a 46-year-old Indian woman with a recurring sore on the left side of her nose, which had returned after three surgeries. Tests for infections, including pyogenic bacteria, tuberculosis, and fungi, were negative. She initially took ciprofloxacin (500 mg twice daily for six weeks), but since the sore returned, she switched to doxycycline (100 mg twice daily), which led to improvement.

Case 4 features a 31-year-old Indian woman who experienced a recurrent spot on the left side of her nose, which persisted after previous removal. She underwent electrocautery treatment and received the antibiotic cephalexin, showing some improvement.

DISCUSSIONS

Rhinoscleroma is a challenging disease to diagnose, being rare and often overlooked by physicians when evaluating nasal concerns. Most documented cases arise from regions where rhinoscleroma is endemic. However, the subjects in this case study had not traveled to such areas. Typically, rhinoscleroma affects the respiratory system, impacting the lungs and airways, but the patients in our study exhibited only skin-related symptoms without respiratory issues or needing specialized ear, nose, and throat consultations.

The precise cause of rhinoscleroma remains unclear, though it appears linked to immune system deficiencies. In these patients, there was a notable alteration in the ratio of immune cell types, specifically CD4 and CD8, which may impair the function of macrophages and facilitate bacterial growth. Certain genetic factors may predispose individuals to this condition; for instance, genetic mutations in mice have shown increased susceptibility to infections. In humans, a specific genetic marker (HLA-DQA1’03011-DQB’0301 haplotype) has been identified as a major risk factor for respiratory rhinoscleroma. Testing for these immune and genetic factors in our patients would have been beneficial.

To diagnose rhinoscleroma, doctors typically examine tissue samples microscopically. All four patients exhibited typical disease markers, including specific cell types and positive staining results. Routine blood tests can identify the bacteria (C. rhinoscleromatis) in 50%-60% of cases with the granulomatous form, which might explain the negative culture results in two patients since they only presented with skin-related issues. Disentangling various bacterial types through molecular techniques is challenging due to similar DNA sequences, and while blood tests exist, they may yield ambiguous results due to cross-reactivity. Imaging modalities such as X-rays, CT scans, or MRIs can indicate disease location but are seldom useful for definitive diagnosis.

When assessing symptoms, it’s essential to consider various infections (such as tuberculosis, actinomycosis, and leprosy) that may cause throat lumps, as well as fungal infections, mucocutaneous leishmaniasis, malignancies like lymphomas, and inflammatory conditions such as sarcoidosis and Wegener's granulomatosis.

Management of these conditions can be challenging since rhinoscleroma is difficult to treat and often relapses. Laboratory studies indicate that common antibiotics like amoxicillin-clavulanate and chloramphenicol affect the K. rhinoscleromatis bacteria. Effective real-world treatments include streptomycin, doxycycline, and rifampicin, especially since C. rhinoscleromatis resides within cells, responding best to extended treatments with rifampicin and fluoroquinolones that achieve high intracellular concentrations.

Surgical intervention may be warranted in cases with significant breathing difficulties or noticeable cosmetic changes due to the disease. Relapses are frequent in rhinoscleroma, making long-term antibiotic therapy and vigilant follow-ups essential for early detection of recurrences. Other rare complications include airway narrowing, hemorrhaging, severe infections, or potential cancer development.

CONCLUSION

In summary, rhinoscleroma is a multifaceted disease requiring further research for better comprehension and improved treatment strategies to reduce relapses and associated complications. Our case studies illustrate that rhinoscleroma can manifest solely with skin symptoms, and we aim to enhance awareness of this condition among healthcare professionals.

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