Rhinoscleroma of the Tracheobronchial Tree: Bronchoscopic, PET, and CT Correlation

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ABSTRACT

Rhinoscleroma is a chronic granulomatous condition caused by Klebsiella rhinoscleromatis endemic to many countries. There is a growing incidence of this disease in non-endemic countries in settings in which it was previously never seen. Presented is a case report of such a patient presenting from Mexico to the United States who was found to have rhinoscleroma. Treatment with a long course of antimicrobials and surgical intervention as needed is generally sufficient. Interestingly, this patient also received argon plasma coagulation of the diffuse lesions with significant improvement in his symptoms. The differential diagnosis of such a presentation is reviewed. In addition, a complimentary approach to diagnosis and management with computerised tomographic scan (CT scan), positron emission tomographic scan (PET scan), and bronchoscopy is discussed. This demonstrates the difficulty in diagnosis of such a condition, especially in non-endemic settings due to the chronicity of presentation and lack of consideration.

Key words: Rhinoscleroma, Trachea, Tracheal stenosis.

INTRODUCTION

Endemic in other parts of the world, rhinoscleroma is a chronic infectious condition affecting the respiratory tract. There is, however, an increasing incidence of this disease in non-endemic countries. We report here a case of a patient who migrated to the United States from Mexico 10 years earlier who presented with a long history of haemoptysis, dyspnoea, and dysphonia. Prior evaluations had not led to a specific diagnosis. Computerised tomographic scan (CT scan) of the chest revealed diffuse nodularity and thickening of the lining of the tracheobronchial tree. Positron emission tomographic scan (PET scan) showed diffuse uptake in the trachea. Bronchoscopic evaluation led to the recovery of tissue which, ultimately led to the correct diagnosis.

This case epitomises the difficulty in diagnosing infectious diseases in a non-endemic setting. Initially, due to the rarity of the disease in the United States, rhinoscleroma was not included in the differential diagnosis.

CASE REPORT

A 30-year-old Hispanic male presented with dyspnoea at rest, dysphonia, and intermittent haemoptysis of approximately nine years duration. He emigrated from Mexico 10 years prior to presentation and complained of recurrent episodes of sinusitis and upper respiratory infections. Treatment to date had been non-specific and consisted mainly of bronchodilators. He denied alcohol abuse or intravenous drug use, but did report a history of second-hand smoke exposure since childhood. Additional history was significant for the treatment of testicular cancer 12 years prior to presentation. For this, the patient underwent a left orchiectomy and external beam radiation without evidence of recurrence.

On examination, the patient was alert and well oriented. He was not in obvious distress. His vital signs were stable. Oxygen saturation on room air was 98 percent. The only significant finding on examination of the chest was mildly reduced breath sounds at the lung bases with prolongation of the expiratory phase. There was no stridor or wheezing. Laboratory data including rheumatologic work-up was unremarkable.

Computerised tomographic scan of the chest revealed diffuse thickening and nodularity of the lining of the trachea, right mainstem bronchus, and right lower lobe bronchus (Figure 1). A malignant process was considered in the differential diagnosis, and fluorodeoxyglucose PET scan (FDG-PET scan) was also obtained. The examination demonstrated diffusely
increased metabolic activity throughout the trachea and mainstem bronchi bilaterally. No hypermetabolic foci were evident in the lungs (Figure 2).

In view of these findings, bronchoscopy was carried out and confirmed the presence of diffuse nodularity of the mucosa extending from the level of the vocal cords, to the carina and into the right mainstem bronchus as far as the bronchus intermedius with relative sparing of the left bronchus (Figure 3). The nodular lesions were extremely friable and bronchoalveolar lavage as well as multiple endobronchial biopsies were performed.

The bronchoalveolar washings and endobronchial tissue specimens were negative for malignant cells. The material recovered was found to contain predominantly plasma cells. In addition, Mikulicz cells were present. Cultures were positive for Klebsiella rhinoscleromatis confirming the diagnosis of rhinoscleroma. Treatment was instituted with oral doses of levofloxacin 500 mg daily. In addition, the patient underwent a repeat bronchoscopy one week later and resection of the diffuse polyoid endobronchial lesions with Argon plasma coagulation performed.

A follow-up bronchoscopy three months later revealed only mild erythema of the tracheobronchial tree with a significant decrease in the extent of the lesions as compared to the prior findings.

At six months, repeat endotracheal cultures were negative and CT scan of the chest demonstrated a dramatic reduction in the thickening and nodularity of the mucosa of the tracheobronchial tree (Figure 4). The patient is currently feeling well. He completed eight months of treatment with levofloxacin 500 mg daily. Because of the fear of relapse, he has been treated periodically with antimicrobials for recurrent respiratory symptoms.

DISCUSSION

Rhinoscleroma or respiratory scleroma is a chronic progressive granulomatous infection that predominantly affects the upper airways. The infection is caused by an encapsulated gram-negative diplobacillus, Klebsiella rhinoscleromatis. The disease has been widely reported from countries in the Middle-East, India, South-east Asia, Central and South America. Tropical Africa accounts for 5% of all worldwide cases. In the developed countries, in temperate parts of the world, only sporadic cases have been reported. As in our case, most of the reported cases involve immigrants from endemic areas. Females, most commonly of child bearing age, are affected more frequently than males by a 13:1 ratio, with presentation usually in the second and third decades of life.
The causative agent, *Klebsiella rhinoscleromatis*, is a subspecies of *Klebsiella pneumoniae* with humans being the only known host. Neither the mode of transmission is understood nor is the mechanism by which it spreads and becomes established in the airways to produce disease. Specific susceptibility of the host plays an important role in the development of disease.6

**Rhinoceroma can affect the entire respiratory tract.** The nose is affected in 95%-100% of cases and the pharynx in up to half. Other affected areas include the eustachian tubes, sinuses, mouth, orbit, and larynx in 15%-40% of cases. Tracheal involvement occurs in only 12% of cases and disease in the bronchi in 2-7% of cases. The skin in proximity to affected mucosa may be involved, for example on the upper lip or around the nose. There has also been reports of skin lesions on the back and spread of the organism to the brain.7

The disease is divided clinically and pathologically into three stages: catarrhal or atrophic, granulomatous, and sclerotic.8

The differential diagnosis includes infections such as tuberculosis, actinomycosis, syphilis, leprosy, histoplasmosis, blastomycosis, paracocccidiodomycosis, sporoarthritis, rhinosporidiosis and zygomyosis, as well as mucocutaneous leishmaniasis. Non-infectious processes leading to a similar clinical presentation include foreign body granulomas, sarcoidosis, amyloidosis, basal cell carcinoma, verrucous carcinoma and lymphoma. Wegener’s granulomatosis, or extranodal Rosai-Dorfman disease should also be considered. Additionally, inflammatory bowel disease has also been reported to lead to endobronchial thickening. Post-intubation stenosis may also cause circumferential tracheobronchial thickening similar to rhinoscleroma.29

Typically the diagnosis requires recovery of affected tissue and demonstration of characteristic Mikulicz cells, Russell bodies, and at times, electron microscopic demonstration of “Type A” or “Type B” granules. Type A granules are antibodies accumulated on the bacterial surface appearing as electron-dense granular and fibrillary substances. Type B granules are less electron-dense substances representing bacterial antigens composed of mucopolysaccharides surrounded by antibodies, *i.e.* type A granules.9 Bacterial culture is positive in 50-60% of cases. Immunocytochemical examination performed with antibodies to the O2K3 antigen on *Klebsiella rhinoscleromatis* may be used to further confirm the diagnosis.10 Endoscopy during the earlier stages of the disease may demonstrate adherent unique white-green plaques in the airway. Bronchoscopy may also have a role in long-term follow-up to monitor treatment response, in monitoring for disease complications, and management of endobronchial obstruction as in this case.10

When the trachea is involved, CT scan may demonstrate crypt-like irregularities of the tracheal wall, nodular deformity of the tracheal mucosa, subglottic stricture, and concentric irregular narrowing of the trachea and central bronchi.31 Magnetic resonance imaging may be useful in staging of disease in the hypertrophic stage where there is a characteristic mild to moderate high signal intensity of both T1- and T2-weighted magnetic resonance images.12

Initially, because of our patient’s history of haemoptysis, lung cancer or metastatic disease was considered in the differential diagnosis and an FDG-PET scan was obtained. The study demonstrated avid uptake of the tracer in the trachea. As is well known, uptake of this tracer is seen in both infectious and granulomatous disease as well as in malignancy. Standardised uptake values (SUVs) have been used in an attempt to differentiate benign from malignant diseases. However, high SUVs have been seen in both mycotic infections and tuberculosis. This suggest the greatest utility of PET scan in rhinoscleroma, may be in assessing extent of disease and response to treatment.

Untreated rhinoscleroma progresses slowly through the three stages over many years and is characterised by periods of remission and relapse. Appropriate treatment regimens usually involve a combination of surgical debridement and prolonged antibiotic therapy. Even with optimal therapy the relapse rate is high. *In vitro*, *Klebsiella rhinoscleromatis* is sensitive to a wide range of antimicrobial agents, however, the mainstay of treatment for rhinoscleroma are the fluoroquinolones. Additional potentially efficacious antibiotics are third generation cephalosporins, clindamycin, amoxicillin-clavulanate, rifampin, and the anti-leprotic agent clofazimine.

Prolonged antibacterial therapy, for at least six months, has produced the best clinical results with the lowest relapse rates.7 Optimally, treatment should be continued until follow-up nasal biopsies are negative for the disease. Balloon dilation or mechanical dilation via rigid bronchoscope may be required to treat significant fibrotic stenosis of the trachea or bronchus. Tracheostomy is occasionally required for severe involvement of the larynx or subglottic space.10 We used argon plasma coagulation to treat the endobronchial lesions with significant improvement in patients of dyspnoea. We believe this procedure should be considered when there is airway obstruction secondary to significant endobronchial disease.

Death from rhinoscleroma in a properly treated patient is rare and occurs only in patients diagnosed in late stages of the disease who present with marked airway obstruction. After treatment induced remission, prolonged follow-up is essential to monitor for relapse and permit re-institution of treatment before complications arise.13

Our patient is of particular interest for a number of reasons. First, he was a male, a rarer presentation. Secondly, was his presentation for diagnosis in a non-
endemic region. Although he was an immigrant from an endemic country, he had been a resident in the United States for 10 years. In part, the delay in diagnosis may also have been related to his inability to obtain proper medical care, but initial exclusion of rhinoscleroma from the differential diagnosis was also a factor. Additionally, the patient presented with disease only involving the trachea and bronchial tree, which, as noted above, is atypical. Also, the use of a PET scan for evaluating the activity of the disease process is demonstrated.

To the best of our knowledge, this is the first report that define the use of PET scan in rhinoscleroma. CT scan, PET scan, and bronchoscopy in this condition are complimentary tools. One test exhibits the anatomical relation, the other demonstrates the activity of the disease process, and bronchoscopy aids in diagnosis and management of complications of the disease.

The use of non-surgical interventional bronchoscopic correction with argon plasma coagulation of lesions has not been previously reported and led to a significant decrease in the patient’s symptoms. This modality should be considered in the treatment options for rhinoscleroma when airway narrowing is a concern.

Migration of large populations from endemic regions to countries where infection with *Klebsiella rhinoscleromatis* is little known, is a new challenge in global medicine. Rhinoscleroma should be considered in the differential diagnosis for immigrant patients with chronic upper respiratory illnesses.

### REFERENCES