CASE REPORT

Rhinoscleroma causing upper airway obstruction

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Rhinoscleroma is a chronic granulomatous condition of the respiratory tract, and is not uncommon in tropical regions; particularly, Mexico, Central America and the Middle East. A few cases have been reported in North America, primarily involving immigrants from endemic countries. The causative organism is Klebsiella rhinoscleromatis, a Gram-negative coccobacillus. Diagnosis is made on the basis of culture of the organism and the characteristic pathology of Mikulicz cells on light microscopy.

The condition primarily affects the upper airway, and frequently presents with nasal discharge, nasal obstruction or frontal facial pain. Despite the term 'rhinoscleroma', there may be involvement of the entire respiratory tract. Although the condition is slowly progressive, its natural course portends extensive destruction. Laryngotracheal involvement occurs in approximately 15% to 80% of cases, but patients rarely present with isolated laryngotracheal disease. In the present paper, a case of rhinoscleroma presenting with symptoms of upper airway obstruction is described.

Key Words: Laryngotracheal scleroma; Rhinoscleroma; Upper airway obstruction

A 56-year-old male pharmacist, originally from Egypt, presented with a two-month history of shortness of breath on exertion. He complained of a foreign-body sensation in his throat with associated wheezing but denied chest pain, cough, hemoptysis or hoarseness. He had not experienced fever, weight loss or night sweats.

His past medical history was significant for chronic sinusitis since childhood, characterized by long-standing malodorous secretions, crusting and intermittent nasal blockage. He had seen an otolaryngologist and received a tentative diagnosis of immotile cilia syndrome 12 years before his current presentation, although this diagnosis was not proven by biopsy and he was the natural father of two children. He had both hypertension and hyperlipidemia, and no known family history of respiratory illness or malignancy. He quit smoking six weeks before presentation.

On physical examination, he appeared well with no evidence of respiratory distress at rest. His blood pressure was 140/90 mmHg. His trachea was midline and he did not have cervical or supraclavicular adenopathy, cyanosis or clubbing.

Examination of his chest revealed symmetric chest expansion with normal breath sounds throughout all lung fields, and specifically, an absence of stridor or wheezing. The remainder of his physical examination was normal.

A chest x-ray taken at the time of presentation was normal, with no evidence of a mass or parenchymal disease. He had an x-ray six years previously which appeared the same. He underwent spirometry and produced a flow-volume loop demonstrating classical features of fixed upper airway obstruction (Figure 1).

A computed tomography scan of the patient’s thorax revealed normal lungs with wall thickening of the proximal trachea and an irregular-appearing endoluminal surface at the level of the thyroid. A subsequent computed tomography scan of the head and neck showed moderate circumferential narrowing of the subglottic larynx and superior trachea, as well as nodular thickening at the level of the inferior cricoid resulting in 50% to 60% endoluminal narrowing (Figure 2).

The patient underwent bronchoscopy, the findings of which were suggestive of a proximal tracheal tumour involving

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the glottis and subglottis. The patient was referred to an oto-
laryngologist for evaluation of a potential malignancy. 
Laryngoscopy confirmed that there was a 3 cm tumour of the 
tracheal lumen, with evidence of cartilaginous destruction. The 
mid- and distal trachea were clear, as were the supraglottic and 
glottic larynx. Endoscopic resection was performed to improve 
the patient’s airway and biopsies were sent for pathology.

Pathology revealed a packed mucosal infiltrate of histio-
cytes admixed with inflammatory cells (mainly plasma cells). Silver methenamine and Gram staining revealed numerous 
Gram-negative bacilli within the histiocytes. The low-power 
view of the laryngeal biopsy showed an infiltrate of large, vac-
uolated histiocytes or Mikulicz cells (Figure 3).

Culture of the tumour specimen confirmed 
*Klebsiella rhinoscleromatis* was the causative organism. The patient was 
started on ciprofloxacin hydrochloride 500 mg twice a day for 
six weeks. He remained well after resection with no symptoms 
of upper airway obstruction. Bronchoscopic follow-up at 
six months demonstrated no recurrence of disease.

**DISCUSSION**

The objective of the present case discussion is to highlight a disease process that is generally identified predominantly as a cause of nasal disease, although it can involve the lower respiratory tract. Because it is not endemic to North America and infrequently presents to respiratory specialists, the diagnosis may be missed. However, with increasing immigration from endemic countries, this cause of upper airway disease should be familiar to Canadian respirologists. In the present case, the patient described a long-standing history of nasal obstructive symptoms and rhinorrhea before his presentation to a respirologist. He remained misdiagnosed and untreated until his dis-

ease progressed to cause proximal tracheal obstruction. Ultimately, the diagnosis was not made on the basis of patient 
presentation or appearance of the lesion, but rather on the basis 
of the pathological findings and culture. If suspicion of malignancy is sufficiently high and the characteristic pathology is missed, Gram staining may not be performed and the causative 
organism may not be identified.

Typically, rhinoscleroma progresses through three clinical 
stages. Initially, a catarrhal phase is characterized by purulent 
and fetid rhinorrhea, which may progress to nasal mucosal atrophy and crusting. The second phase is a granulomatous phase, in which nodules or masses may lead to obstructive 
symptoms in the respiratory tract. The final stage is a cicatri-
cial stage of scarring which may cause external deformity or 
tenosis of nasal passages or laryngeal airway (1).

The differential diagnoses for respiratory scleroma include 
nasal tumours and other tumours of the respiratory tract, fun-
gal infections or leprosy. Wegener's granulomatosis, T cell lymphomas, sarcoidosis and basal cell carcinomas may also have similar clinical presentations.

Alfaro-Monge et al (2) described a case of scleroma present-
ing as an upper tracheal mass causing stridor and respiratory dis-
tress. The patient was managed with urgent resection and an 
eight-week course of oral ciprofloxacin hydrochloride. In their 
literature review of lower respiratory tract scleroma, there were 
only two cases of isolated laryngotracheal scleroma, both of 
which required urgent tracheotomy (3,4). In a Los Angeles 
study of 22 patients with scleroma (5), 59% of patients had laryngotracheal scleroma. All patients in this study had nasal mucosal involvement. Patients presented with dysphonia, exertional dyspnea and/or stridor. Patients in this series were 
initially managed with intravenous antibiotics and dexamethasone, and three required tracheotomies. All patients were in 
the granulomatous phase of infection, and it appeared that granuloma formation was more common in the glottis than in
the subglottis. Yigla et al (6) described a case of a severe form of laryngotracheal rhinoscleroma, which resulted in the death of the patient.

The mainstay of treatment is surgical debridement, when indicated for preservation of the airway, and prolonged antibiotic therapy (7-9). Agents of choice include quinolones, doxycycline or tetracycline, and less frequently, streptomycin and rifampicin (8,9). Based on reported cases (8), several weeks of therapy are indicated because the relapse rate is high. The present patient had a six-week course of ciprofloxacin hydrochloride and has exhibited a satisfactory clinical response after six months of follow-up. Long-term follow-up is indicated to ensure remission on the basis of both clinical and endoscopic findings.

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REFERENCES