Rhinoscleroma of the Larynx: First case report in the State of Qatar

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Abstract:
Rhinoscleroma is a chronic slowly progressive inflammatory condition of the upper airways mostly caused by infection with Klebsiella pneumoniae subspecies rhinoscleromatis and less frequently subspecies ozaenae. The disease is uncommon in developed countries but it still exists in Africa and South America. The disease usually takes a chronic course and diagnosis, which might be delayed, depends on the isolation of the organism from the tissue and the characteristic histopathologic features.

We report here a case of rhinoscleroma of the upper airways that presented as a laryngeal mass in a young Egyptian man living in Qatar. The methods of diagnosis and treatment are discussed and the relevant literature reviewed.

Case History:
A 23-year-old Egyptian male was admitted through the emergency room with a two-day history of stridor preceded by a dry cough and dysphagia. Two months prior to admission he noticed a gradual change of voice with repeated attacks of epistaxis associated with nasal obstruction. He had a history of repeated attacks of upper respiratory tract infections that were treated with courses of antibiotics while he was in Egypt. He had no history of allergy or asthma but had been a cigarette smoker for five years.

His physical examination in the emergency room showed respiratory distress, restlessness and biphasic stridor. His temperature was 37.8°C; blood pressure 120/75 mmHg; pulse 56/min and regular. His physical examination was otherwise unremarkable except for a crust in the right nostril. Emergency tracheostomy was performed.

Endoscopic examination under general anesthesia showed a crust in the right nostril with narrowing in both choanae and severe narrowing in the larynx. There was a granular mass under the right vocal cord and in the right subglottic area.

Laboratory investigations showed white blood cells 12,300/mm³ (75% neutrophils); hemoglobin 16 gm/dl; platelet count normal; ESR 6 mm/hour. Liver, kidney and thyroid function tests were normal. Chest radiograph and barium swallow were normal. Computerized tomographic scan (CT) of the neck (Figure 1) showed diffuse thickening of the right glottis at the level of the vestibule and true vocal cords region with narrowing of the airway as a slit-like glottis on axial images. The anterior part showed no marginal enhancement, no lymph node or vascular structure abnormalities.

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Figure 1: CT scan of the neck showing severe narrowing of the glottis with a mass in the right subglottic region.

He was given oxygen and intravenous hydrocortisone, amoxicillin-clavulanic acid for a presumed retropharyngeal abscess.

Pan-endoscopy under general anesthesia showed narrowing of both choanae with granulation in the right arytenoids and the right vocal cord with a mass on the right subglottic area that was biopsied. Fiberoptic bronchoscopy was unremarkable. Bronchial wash was negative for malignant cells and for acid-fast bacilli and cytological examination was consistent with acute inflammation.
Histopathologic examination of the biopsies showed diffuse infiltration with foamy histiocytes on a background of plasma cells with Russell bodies and lymphoid cells (Figure 2). The right and left vocal cord biopsies showed diffuse infiltration with foamy histiocytes (Mikulicz cells) that were positive for bacilli when stained by the Warthin-Starry method (Figure 3). Cultures of the sputum, bronchial wash and nasopharyngeal secretions all grew Klebsiella pneumoniae subspecies ozaenae which was susceptible to ampicillin, cefuroxime and ciprofloxacin. The patient received intravenous ciprofloxacin and amoxicillin-clavulanic acid (augmentin) for one month and was discharged on oral ciprofloxacin and augmentin to be taken for a further six months.

A CT scan after three weeks of treatment with antibiotics showed improvement in the airway space with symmetry of left and right vocal cords (Figure 4).

**Discussion:**

Rhinoscleroma is a slow, progressive chronic inflammation of the respiratory tract. The disease was first reported in Europe in 1882 by the Polish surgeon Johannes von-Radecki Mikulicz and the causative organism was identified by Anton von Frisch as a subspecies of Klebsiella pneumonia.

The disease is endemic in South America and Africa but is rare in the developed countries including Europe and the United States although more cases are being seen there due to the influx of immigrants from endemic countries. The infection is acquired by direct inhalation of droplets or contaminated material. Predisposing factors include poor hygiene, overcrowding, malnutrition, and iron-deficiency anemia. It is more common in females than males. The causal agent is usually Klebsiella subspecies rhinoscleromatas although subspecies ozaenae has been isolated in some cases. It is believed that the mucopolysaccharides of the capsular material of the organism are capable of inhibiting the phagocytic function of the macrophages and the T-lymphocytes (mainly the CD4 types) facilitating infection.

Our patient was a young male from Egypt working in Qatar; his main presentation was stridor developed over a background of gradual change in his voice which reflected an acute or chronic laryngeal involvement. He had a history of epistaxis and repeated upper respiratory tract infections with many courses of antibiotics prescribed when he was in Egypt. The crust that was noted in the right nostril probably indicated minor nasal involvement with the disease.

The nasal vestibule is the area most commonly involved but the subglottic area of the larynx and the vocal cords are involved in 15–80% of cases. In a series of 22 cases of rhinoscleroma from America, 13 cases involved the larynx, and nine had subglottic and glottic stenosis. The nasopharynx and oropharynx, paranasal sinuses and even the orbit can also be involved. Cases with major...
tracheal stenosis and deformities have been reported.\(^{(5)}\)

Loëc de Pontual, et. al. studied 11 cases of rhinoscleroma in France over 16 years from 1990 to 2005; one case was a young African male patient in whom the infection had an aggressive course; the tumor invaded the cavernous and ethmoidal sinuses causing ischemic stroke by invasion of branches of the middle cerebral artery.\(^{(6)}\)

The disease usually takes three stages in which the clinical presentations might be different; a catarrhal stage which is an early acute inflammation where the patient will be presented with repeated upper respiratory tract infection; a granulomatous stage where masses of granulation tissue develop and can be seen most commonly on the nose and lips but they can invade the paranasal sinuses. Rarely, fatal airway obstruction due to laryngeal involvement might occur.

The sclerotic stage is a late stage that leads to fibrous deformities in the nose and the trachea and the major bronchi.\(^{(8)}\)

The diagnosis of rhinoscleroma is often delayed because symptoms can be attributed to more common conditions such as viral infections of the upper respiratory tract. Confirmation of the diagnosis depends on identification of the micro-organism Klebsiella pneumoniae subspecies rhinoscleromatis or ozaenae in tissue biopsies or from samples of the lower respiratory tract that can be positive in up to 60% of cases. Positive blood cultures have also been reported.\(^{(7,8)}\)

The pathological findings of chronic inflammation and infiltration with lipid-rich vacuolated histiocytes containing the organisms (Mikulicz cells) are considered characteristic of the disease.\(^{(9)}\)

The disease should be differentiated from other chronic infections and granulomatous diseases of the upper airways such as tuberculosis, leprosy, Wegner granuloma, sarcoidosis and malignant lymphomas and carcinomas. In our patient we first ruled out nasopharyngeal tumor, parapharyngeal abscess and foreign body inhalation.

The disease is endemic in Egypt, five cases of rhinoscleroma being reported in 1954, and a few cases have been reported from Saudi Arabia and Bahrain.\(^{(2)}\) To our knowledge this is the first case of rhinoscleroma reported from Qatar.

Our patient presented with epistaxis, dysphagia, and airway obstruction due to a mass on the vocal cords and in the subglottic area. He complained of repeated upper respiratory tract infection that was mostly related to the same pathology since the organism was grown from the bronchial wash and sputum cultures. The diagnosis in our patient was established by histopathology showing the characteristic findings (Figures 2, 3) and by the isolation of Klebsiella pneumoniae subspecies ozaenae from cultures of the bronchial wash and tracheal aspirate.

The treatment of rhinoscleroma depends on the stage of the disease.\(^{(10)}\) For the catarrhal and early granulomatous stages antibiotic treatment is recommended, usually ciprofloxacin or tetracycline for 3–6 months, with some authors recommending corticosteroids as adjuvant treatment.\(^{(10)}\)

The mass of granulation tissue in our patient showed a rapid and marked improvement with antibiotic treatment; a CT scan after three weeks of treatment with intravenous ciprofloxacin and hydrocortisone showed resolution of the mass with improvements in the air space and symmetry of both vocal cords (Figure 4). Ciprofloxacin and Augmentin tablets were continued for six months after discharge from hospital. He was seen regularly in the clinic and in his last visit he was in a good general condition and had no new complaints.

Surgery to remove masses and to correct deformities in the nose and the nasopharynx might be needed in the late sclerotic stages of the disease.\(^{(11)}\) Carbon dioxide laser therapy might help in correcting some deformities such as tracheal stenosis and nasopharyngeal deformities.

In summary, this is the first reported case of rhinoscleroma in the State of Qatar that presented as acute airway obstruction due to laryngoscleroma which was treated successfully with emergency tracheostomy and long-term antibiotics.

Rhinoscleroma should be suspected in patients with long-standing chronic respiratory symptoms and appropriate risk factors. The course of the disease can be fatal if the larynx is involved and the airway obstruction is not relieved urgently. Patients with rhinoscleroma need long-term follow-up to detect and treat any late complications and rule out relapses.

### References: