



Rhinoscleroma , endemic in Egypt with atypical presentations

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Introduction

Scleroma or Rhinoscleroma is a progressive granulomatous disease affecting the nose and later extending into the nasopharynx and oropharynx, the larynx and sometimes the trachea and bronchi may be affected. Laryngeal involvement may occur in almost 50% of cases and the disease is better designated as respiratory scleroma, rather than rhinoscleroma.

Pathology : The disease is caused by the Gram-negative bacillus, *Klebsiellarrhinoscleromatis*, also known as Frisch bacillus.

The organism predominantly resides intracellularly and can be difficult to isolate in the laboratory. The characteristic histological features include granulomatous tissue infiltrates in the submucosa, characterized by the presence of plasma cells, lymphocytes and eosinophils. Besides, there are scattered large foam cells

(Mikulicz cells) which have a central nucleus and a vacuolated cytoplasm containing bacilli and Russel bodies.

Epidemiology :

It may occur at any age and in either sex, but is more common in females and in the middle aged. It is seen mainly in central and south-eastern Europe, North Africa, the Indian subcontinent, Indonesia and Central and South America.

It is endemic in Egypt and especially in Fayoum.

Clinically :

The disease has three stages :

1. The atrophic stage: The features resemble that of atrophic rhinitis, including crust formation and foul smelling discharge.
2. Granulomatous or proliferative (or nodular) stage: Non-ulcerative nodules develop, bluish red nasal mucosa and the development of intranasal rubbery nodules or polyps. Epistaxis, nasal deformity, and destruction of the nasal cartilage are also noted (Hebra nose). The damage may result in anesthesia of the soft palate, enlargement of the uvula, dysphonia and various degrees of airway obstruction. Regional lymph node involvement is extremely rare
3. Fibrotic stage: Adhesions and stenosis distorting the normal anatomy.

Diagnosis : Diagnosis in the atrophic stage is extremely difficult and the disease is usually recognized only in the granulomatous stage, it is usually established by a biopsy demonstrating the characteristic histological features. A positive culture in MacConkey agar is diagnostic, but cultures are only positive in 50-60% of cases. The diagnosis may be further aided by a complement fixation test .

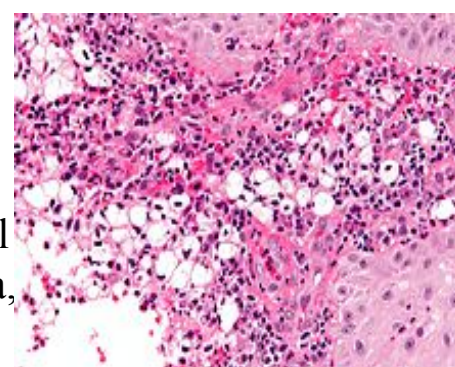


Fig. (1) : microscopic pict.

Treatment of rhinoscleroma requires a combination of appropriate antibiotics and surgical debridement if there is significant airway obstruction. The results of current treatment are unsatisfactory and recurrence often occurs. Bactericidal antibiotics in large doses are given for a minimum of four to six weeks and are continued until two consecutive cultures from biopsy material are proven negative. The traditional antibiotics used are streptomycin and tetracycline. Some reports have emphasized the good results achieved with oral therapy with rifampicin, sulphamethoxazole-trimethoprim combination, levofloxacin and ciprofloxacin..

We reported two cases of atypical presentation among patients presented to (Fayoum university hospitals) in the form of :

- 1) A large vocal fold polyp with normal subglottis (the most common site for laryngeal affection) and free nasal cavity in a female patient aged 32 yrs. old presented with hoarseness of voice with mild stridor and shocking attacks



Fig. (2) : pre op. view

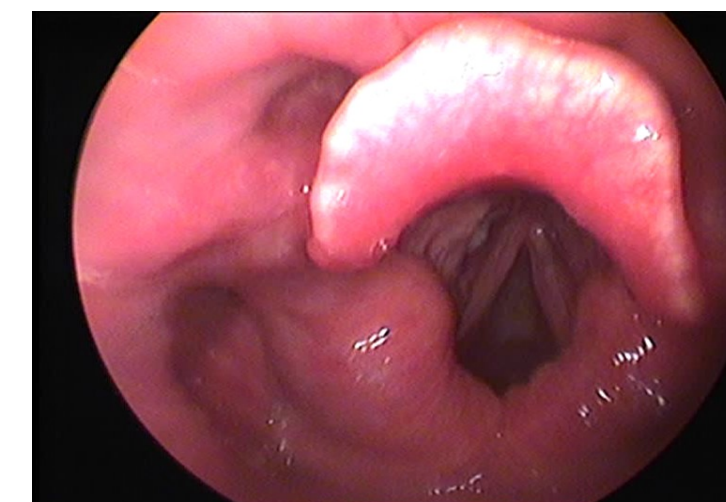


Fig. (3) : post op view

- 2) Large nasopharyngeal mass looks like lymphoma with normal nasal cavity in a male patient aged 38 yrs old presented with bilateral nasal obstruction and bilateral middle ear effusion without palpable lymph nodes.

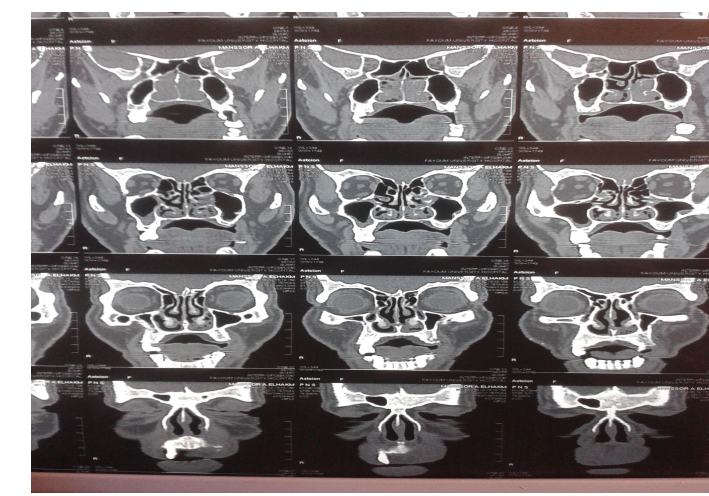
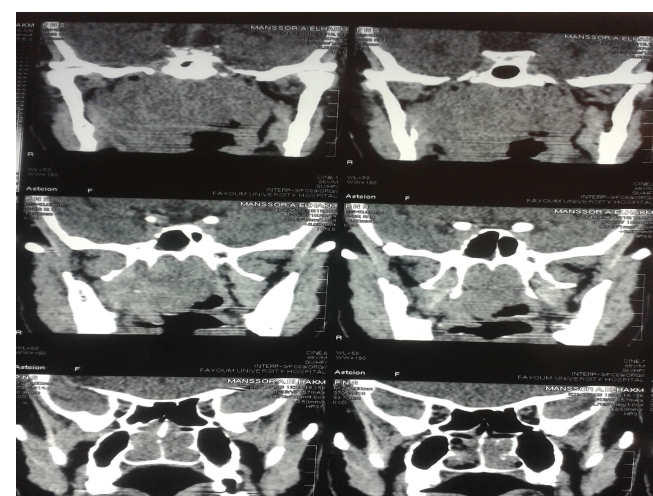


Fig. (4) : CT of the nose and nasopharynx