

Case report

Rhinoscleroma in Karbala

-- Case Report --

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Abstract

A 14 year old boy presented with a history of progressive nasal obstruction with frequent bleeding episodes, for the last three months. The otolaryngological examination revealed slight dilated and flattened nose with grey red lobulated mass filling only the right side of the nose with no extension to the postnasal space. The mass excised completely and send for histopathology and the result was highly suggestive of chronic granulomatous inflammation of sinonasal cavity. The condition recurs six months later and the patient presented with the same lesion filling both sides of the nose. The patient underwent complete excision of the mass. The pathological report consistent with rhinoscleroma. The patient received ciprofloxacin with good improvement. He has remained asymptomatic up to the last visit six months following treatment and has shown no evidence of recurrence. This case report to raise awareness of this condition among physicians and aid in early diagnosis so as to reduce morbidity.

Key words: Klebsiella rhinoscleromatis, Mikulicz cells, Russell bodies, rhinoscleroma.

Introduction

Rhinoscleroma (RS) or respiratory Scleroma is a chronic, progressive, granulomatous infectious disease of the upper respiratory tract. It was first reported in Europe, but it is now rarely diagnosed on that continent. The term “rhinoscleroma” was first coined in 1870 by the Viennese dermatologist Ferdinando Von Hebra ⁽¹⁾, who described a nasal lesion that they classified as a form of sarcoma. In 1877, Mikulicz ⁽²⁾ described the histological features of this disease in detail and established its nonneoplastic, inflammatory nature. Von Frisch ⁽³⁾ identified the causal agent of this lesion in 1882 as a gram-negative coccobacillus, now known as *Klebsiella rhinoscleromatis* (KR). Rhinoscleroma is found predominantly in rural areas with poor socioeconomic conditions. Acquisition of the disease is facilitated by crowding, poor hygiene and malnutrition. Females are

more frequently affected than males (ratio 13:1) and disease tends to present in the second and third decades of life. There is also a suggestion that iron deficiency may predispose to disease acquisition ⁽⁴⁾. The disease is endemic to regions of Africa, Southeast Asia, Mexico, Central and South America, as well as Central and Eastern Europe. The transmission of this disease is via air-borne routes, and humans are the only identified host. It is rarely found in other continents, and infection in non-endemic regions is usually attributed to migration of patients. The sites of involvement commonly include the nasal mucosa (95%–100%), pharynx (18%–43%), paranasal sinuses, trachea and bronchi^(5,6). Rhinoscleroma is usually classified clinically and pathologically into three stages: the catarrhal (or atrophic) stage, the proliferative (or granulomatous) stage, and the fibrotic (or sclerotic) stage ⁽⁵⁾. In the catarrhal stage, patients present with foul

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smelling purulent nasal discharge and nasal obstruction. There is crusting and atrophy of the nasal mucosa on examination. Histologically, epithelial squamous metaplasia with subepithelial infiltrate of polymorphonuclear cells and granulation tissue are seen. In the proliferative stage, there are usually complaints of epistaxis, nasal deformity, hoarseness, anosmia and epiphora. On examination, bluish red, rubbery granulomatous lesions are seen. In the fibrotic stage, increased deformity and stenosis, with large amounts of fibrous and scarring tissue on histology, are noted.⁽⁷⁾ Histological analysis at the granulomatous stage reveals more characteristic features, in the form of Mikulicz cells and Russell bodies, Mikulicz cells are large phagocytes with a single shrunken nucleus, displaced to the cell periphery because of the presence of multibacillary cytoplasmic vacuoles⁽⁸⁾, while Russell bodies (reddish violet elliptical structures, slightly bigger than plasma cells and thought to represent degenerated plasma cells). The Mikulicz cell is a major histologic feature of scleroma. However, the plasma cell, which undergoes some hyaline degeneration to become the larger Russell body, is not specific. Russell bodies may also be seen in any inflammation in which plasma cells are a prominent element. Such is the case in leprosy, malignant bubo and venereal granulomatosis⁽⁹⁾. Bacterial culture from biopsy specimens remains the most useful method of diagnosis (positive in 50% of patients at the granulomatous stage)⁽¹⁰⁾. Antibiotic treatment is effective but must be prolonged^(8,11). Person to person transmission of KR probably occurs via airborne secretions⁽¹²⁾. However, healthy individuals in contact with a patient for many years do not necessarily develop infection⁽¹³⁾.

Case Report

A 14 year old boy presented with a history of progressive nasal obstruction with frequent bleeding episodes, for the last three months. His family denied weight loss and night sweats, he lived in poor socio-economic condition with crowding, poor hygiene, and poor nutrition. The otolaryngological examination revealed slight dilated and flattened nose with grey red lobulated mass occupying the right nostril, the endoscopic examination showed that the mass filling only the right side of the nose with no extension to the postnasal space, computerized tomography of the paranasal sinuses revealed enhanced soft tissue mass involving only the right side of the nose with no extension to the postnasal space and clear other sinuses. The remaining physical exam was within normal limits. Simple intranasal polypectomy was done for him and the mass excised completely and send for histopathology and the result was highly suggestive of chronic granulomatous inflammation of sinonasal cavity, no definite malignancy is seen and further immunological tests (c-ANCA & p-ANCA) for Wegener's granulomatosis were negative and a panel of immunohistochemical markers were applied and showed reactive pattern positivity of infiltrating cells for CD20, CD3 (excluding B & T cell non-Hodgkin's lymphoma), negative for CD68 (excluding histocytic tumour) and CK (cytokeratin excluding epithelial tumours) and Ki67 index (proliferative marker) was less than 2% confirm the low proliferative activity of the infiltrate that goes with benign condition. The condition recurs six months later and the patient presented with the same lesion but on both sides of the nose, computed tomography of the paranasal sinuses revealed enhanced soft tissue mass in both nasal cavities with posterior extension into ethmoidal air cells and slightly to the postnasal space, clear other sinuses (Fig.1).

The patient underwent complete excision of the mass under general anesthesia trans-

nasally and the specimen send for histopathology. The pathological report showed polypoid tissue, densely and diffusely infiltrated by polymorphic lymphoplasmocytic cells and very rich in foamy macrophages with scattered Mikulicz cells and plasma cells with Russell bodies, picture which is highly suggestive of rhinoscleroma, the presence of intracellular organisms was not demonstrated by Giemsa stain. Fig. 2

Treatment was commenced with ciprofloxacin 500 mg twice a day. Definitive clinical improvement was seen after three weeks of antibiotic. Ciprofloxacin was reduced to 250 mg twice daily after three months, and the patient went on to complete a total duration of six months of antibiotic with good resolution of clinical lesion. Upon completion of antibiotic therapy, he remained asymptomatic with no clinical evidence of recurrence after six months.

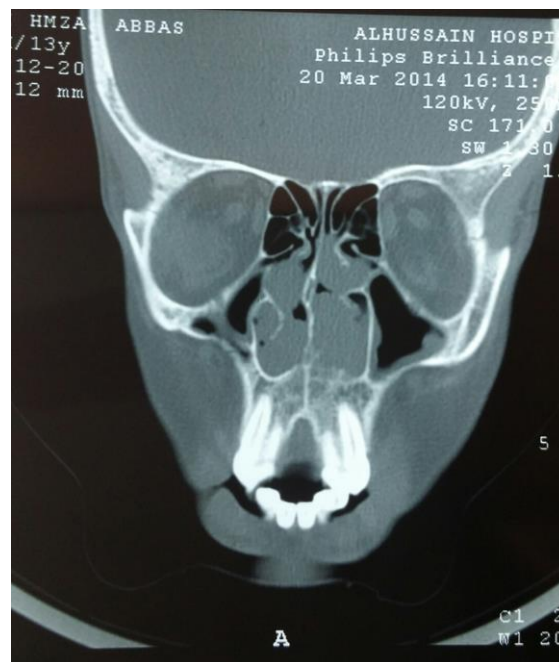


Fig 1. Computed tomography of the paranasal sinuses (coronal view) : Bilaterally enhanced soft tissue mass in both nasal cavities, ethmoidal air cell, and nasopharynx.

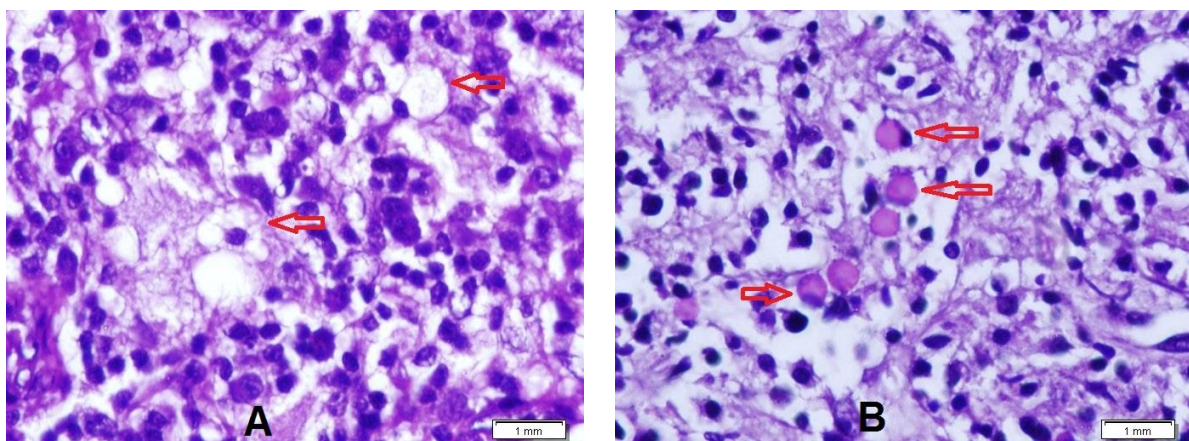


Fig 2. (A) Photomicrograph shows typical histology of Rhinoscleroma with subepithelial infiltration of Mikulicz cells and mild stromal fibrosis (arrows), X400, H&S stain. (B) Photomicrograph shows plasma cells with Russell bodies (arrow), X400, H&S stain.

Discussion

Scleroma is a specific granulomatous infection that affects the nose and, less frequently, other structures of the respiratory tract. It usually affects people with lower social and economic status, and is associated with lack of hygiene and prolonged contact with infected patients^(14,15).

Scleroma is an opportunistic disease in immunosuppressed patients and has many differential diagnoses, such as specific granulomatous diseases caused by bacteria (tuberculosis, actinomycosis, syphilis and leprosy), by fungi (histoplasmosis, blastomycosis, paracoccidioidomycosis), by other parasites (mucocutaneous leishmaniasis), systemic diseases (sarcoidosis and Wegener's granulomatosis), and neoplasms (verrucous carcinoma)⁽¹⁶⁾. Rhinoscleroma is a rare disease, Iraq is a nonendemic area where rhinoscleroma occurs sporadically⁽¹⁷⁾, and a low index of suspicion in nonendemic areas could explain the extreme delay in the diagnosis, as seen in our patient. A high degree of suspicion is warranted when patients present with persistent, unremitting rhinitis or nasal obstruction unexplained by other causes⁽¹⁸⁾. Humans are the only identified hosts of *K. rhinoscleromatis*. Because the bacteria is not found in normal nasal secretions, demonstration of *K. rhinoscleromatis* through culture is diagnostic. However, routine cultures in MacConkey agar reveal positive results among 50-60% of the patients^(19,20). Intracellular *K. rhinoscleromatis* sometime cannot be identified by the use of special stains as in our case. Although the microorganisms were sometimes easily seen in the haematoxylin and eosin-stained slides, they were best demonstrated using the Warthin-Starry stain as it stained the organisms black, leading to easier detection⁽⁷⁾. Serologic tests such as complement fixation tests,

immunofluorescence staining assays with high titers of IgA have been described in the literature, but these tests have cross reactions and are therefore difficult to interpret⁽²¹⁾. Imaging studies can be used to delineate the extent of involvement. Radiography, computed tomography or magnetic resonance imaging rarely lead to a diagnosis but would be able to demonstrate lesions and affected sites⁽²²⁾. Histopathology defines the diagnosis based on finding Mikulicz cells and degenerated plasmocytes in Russel bodies. The Mikulicz cells are characteristic of rhinoscleroma but not pathognomonic. They are also seen in leprosy and bubonic plague and are thought to represent the cells nonspecific response to toxins produced by the causative organism⁽²³⁾. Thus, it is key to have high clinical suspicion in conjunction with positive histopathologic evidence to confirm the diagnosis⁽¹⁸⁾, as we diagnosed our patient.

Antibiotic treatment is used as a single treatment to eradicate the infection mostly in the catarrhal stage, or as ancillary treatment in other stages of the disease to reduce mortality and avoid complications. Drug treatment may be combined with surgery in cases with granulomatous lesions or scarring stenosis. Many antibiotics have been used to treat rhinoscleroma. Streptomycin has severe side effects, especially on the vestibular system; also resistance to this drug has developed in a number of countries. Tetracycline requires a prolonged course of treatment and also has significant adverse effects. Rifampicin provides good results in the treatment of rhinoscleroma, but requires effective monitoring of toxicity. Trimethoprim-sulphamethoxazole may be effective and is widely used in Third World countries. Recent studies have shown that the quinolones, ciprofloxacin and fluoroquinolone readily penetrate the tissues and are clinically effective⁽¹⁴⁾. However, a prospective study done in the

Mayo Clinic, USA, by Andraca *et al.* in 1993 demonstrated the efficacy of fluoroquinolones⁽²⁴⁾. Treatment with fluoroquinolones also confers the benefit of a lower side-effect profile. Dosing of the antibiotic is variable between different studies, but most agree that long-term therapy for months and sometimes years is necessary to adequately treat the infection⁽²⁴⁻²⁶⁾. Despite treatment, recurrence has been reported in up to 25% of cases at 10 years^(8,27). Consideration should be made when addressing whether a patient requires surgical de-bulking of the scar in rhinoscleroma formed during the cicatricial stage. Indications for surgical de-bulking include airway patency, treatment of bulky disease, and cosmesis⁽¹⁸⁾. The presented case demonstrates that rhinoscleroma is very rare disease and this report to raise awareness of this condition among physicians and aid in early diagnosis so as to reduce morbidity.

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