Rhinoscleroma of Nose Extruding into Oral Cavity

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ABSTRACT
Rhinoscleroma (RS) is a rare chronic granulomatous disease of the upper airways affecting nasal cavity, nasopharynx, and paranasal sinuses. *Klebsiella rhinoscleromatis* is the causative agent of this infection and Mikulicz cells are specific to this lesion. RS is commonly seen in poorer regions such as Central Africa, South America, Middle East, India and Indonesia. It is predominantly found in rural areas and people with poor socio-economic conditions. Most patients present with chronic rhinitis, sneezing, headache and deviated nasal septum similar to current case. An association with oral cavity has not been reported previously, as per authors’ knowledge. This report describes a rare case of RS of nasal cavity extending into the oral cavity.


INTRODUCTION
Rhinoscleroma is an uncommon chronic, slowly progressive, localised infectious, granulomatous disease caused by the bacillus *Klebsiella rhinoscleromatis*.1,2 The name rhinoscleroma was first used in 1870 by Von Hebra and Kaposi.2 The disease has worldwide distribution and endemic in developing countries.1 Microscopic picture of the disease was described by Johann von Mikulicz and Woyke et al. in 1969 who gave first description of ultrastructure of granulation tissue in rhinoscleroma. In 1882, Anton von Frisch discovered *Klebsiella rhinoscleromatis* as the single possible etiological factor of the disease.3 RS is commonly seen in nasal cavity, nasopharynx, larynx, trachea, bronchi, oral cavity and paranasal air sinuses.2 Females are more frequently affected than males (ratio 13:1) and the 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 There is a familial predisposition, probably owing to infection by intimate contact.5-7

We report an unusual case of rhinoscleroma of nose in a 50 years old female patient with an extension into the labial vestibule whose clinical and histologic features are unique.

CASE REPORT
A 50-year-old female patient presented with a growth in the inner aspect of the upper lip since one year. Growth was gradual in onset, slowly progressive and extended into the nasal floor. Patient had a previous history of mucopurulent nasal discharge, difficulty in breathing, nasal pruritis and frontal headache infrequently since 6 months. Clinical examination revealed a well defined, firm and sessile pink coloured growth measuring about 1.5 x 1 cm in the labial vestibule (Figure 1). Clinical differential diagnosis of fibroma was made. Computed tomography showed homogeneous hyperdense area involving entire right maxillary sinus with deviation of the nasal septum. Blood investigation revealed reduced haemoglobin level along with microscopic picture of iron deficiency anaemia. Complete excision of the mass was carried out under general anaesthesia.

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Tissue was fixed under 10% formalin, processed and stained with Haematoxylin and Eosin [H and E] and Gram's staining. H and E stain revealed lymphoplasmocytic inflammatory reaction with large vacuolated macrophages (foam cells) typical of Mikulicz cells and numerous plasma cells transformed into Russel bodies, Mott cells were also evident throughout (Figures 2a and 2b). Diagnosis of rhinoscleroma was made. Gram's staining showed gram negative pink coloured bacilli favouring diagnosis of rhinoscleroma (Figure 2c). The patient was treated by surgical excision of the nasal mass followed by ciprofloxacin 500 mg orally twice a day for 2 weeks. Postoperative period was uneventful. The patient was on routine follow-up and presently is doing well.

DISCUSSION

Rhinoscleroma is a chronic progressive granulomatous disease, affects upper airways mainly targeting nasal passages. Histological features and non-neoplastic inflammatory nature were described by Mikulicz in 1877. In 1882, a Gram-negative coccobacillus was identified by Von Frisch and was established as the causative agent of this lesion. Same coccobacillus was known as Klebsiella rhinoscleromatis. Iron deficiency anaemia may act as a predisposing factor in the initiation of the disease. Present case showed microscopic features of iron deficiency anaemia. Klebsiella rhinoscleromatis predominantly resides intracellularly and can be difficult to isolate in the laboratory. The histological features include granulomatous tissue infiltrating into the submucosa, characterized by the presence of plasma cells, lymphocytes and eosinophils. Besides, there are characteristic scattered large macrophages (foam cells) typical of Mikulicz cells, which have a central nucleus and a vacuolated cytoplasm containing bacilli and Russel bodies. The Mikulicz cells are transformed macrophages which have ingested the bacillus, but the bacillus being resistant to digestion by the macrophage persists intracellularly. The Russell bodies resemble plasma cells with an eccentric nucleus and deep eosin staining cytoplasm. Ultrastructure studies have supported the theory of an intracellular formation of Russel bodies. Histochemical studies have indicated a high content of mucopolysaccharides around the walls of the bacillus and it is hypothesized that this may be responsible for the protection afforded to the organism from antibiotics and the hosts antibodies.

Histological differential diagnosis of donovanosis, leprosy, chronic staphylococcal abscess, mycoses, leishmaniasis, granulomatous processes, sarcoidosis, vasculitis, neoplastic diseases like lymphoma and extra nodal Rosai-Dorfman disease were considered. It is important to consider RS in cases of chronic nasal obstruction. As culture is not always positive (50 - 60%), histopathological examination may be crucial to the diagnosis. The diagnosis of RS requires a high index of suspicion and clinicopathological correlation. Histologically three overlapping stages were described in RS as catarrhal, granulomatous and sclerotic. Catarrhal stage shows atropic, squamous metaplasia and nonspecific subepithelial infiltrate of polymorphonuclear leukocytes with granulation tissue. Granulomatous (proliferative) stage reveals subepithelial Mikulicz cells, which are histologic hallmark for diagnosis of RS. Similar to the present case, Sclerotic (cicatricial/fibrosis) stage displays extensive fibrosis, which may lead to stenosis and disfiguration.

Diagnosis in the atrophic stage is extremely difficult and the disease is usually recognized only in the granulomatous or cicatrising stage. Magnetic resonance imaging in the hypertrophic stage shows a soft tissue mass with mild to marked high signal intensity in both T1- and T2-weighted images. The diagnosis may be further aided by a complement fixation test or by microbiological demonstration of the organism, but it is usually established by a biopsy demonstrating the characteristic histological features. Klebsiella rhinoscleromatis can be demonstrated in H and E stained sections and by other appropriate stains such as Periodic Acid-Schiff (PAS), Giemsa, Gram's and Warthin-Starry stain. Warthin-Starry is the most helpful type of stain as it stains the organisms black, leading to easier detection. Specific diagnosis of RS is made with the bacterial isolation by culture on blood or MacConkey
agar. In the present case section was stained with Gram's staining which revealed pink coloured, rod shaped Gram negative bacilli.

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 RS. 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. The present case was treated by surgical excision of the nasal mass followed by a course of ciprofloxacin. Patient was followed-up for 6 months, prognosis after surgical removal and antibiotic treatment was usually good which was confirmed in this case.

REFERENCES