Unusual Case of Rhinoscleroma: A case report

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Abstract: Rhinoscleroma is a progressive chronic granulomatous disease of the respiratory tract. It usually involves the nose but can rarely progress to involve the nasopharynx, oropharynx, larynx, trachea and bronchi. We report an unusual case of a 60 year old female patient who presented with progressive bilateral nasal obstruction and decreased sense of smell. Histopathology played a vital role in the diagnosis. Surgical debulking of the sinonasal mass was done and patient was simultaneously treated with Rifampicin 600 mg daily for 6 weeks. Treatment was effective on the infection.

Keywords: Chronic granulomatous disease, Frisch bacillus, Mikulicz cells, Russell bodies.

INTRODUCTION
Rhinoscleroma is a chronic granulomatous disease of the respiratory tract starting in the nose and can extend to nasopharynx, oropharynx, larynx, trachea and bronchi [1]. Causative organism is Klebsiella Rhinoscleromatis (Frisch Bacillus) which is a Gram negative organism.

CASE REPORT
A 60 year old female presented to Adichunchanagiri institute of research and medical sciences with history of bilateral gradually progressive nasal blockade since 2 years. Left side nasal blockade greater than on the right side. There was history of mucoid nasal discharge and decreased sense of smell since 2 years. However there was no history of foul smelling nasal discharge, bleeding or crusting in the nose. On examination there was a pinkish, smooth, highly vascular mass attached to left inferior turbinate and seen obliterating the left nasal cavity. There was a pinkish vascular mass in the right nasal cavity between the inferior turbinate and the floor of the nasal cavity. Posterior rhinoscopy revealed mass completely filling the posterior choana. Bacteriological culture showed Klebsiella Rhinoscleromatis. Computed tomography scan with contrast revealed hypodense soft tissue mass in both nasal cavities, bilateral maxillary, anterior and posterior ethmoids and left sphenoid sinus and nasopharynx. Patient had concurrent swelling of the right upper eyelid. FNAC of the swelling revealed chronic inflammation. Surgical debulking of the mass was done. Patient was treated with Rifamicin 600mg oral for 6 weeks. Patient had good response to combined surgical and medical management and the mass completely resolved. Patient had no recurrence during the 3 months follow up period.

Fig-1: Endoscopic view showing the smooth vascular mass involving the left inferior turbinate

Fig-2: Endoscopic view left nasal cavity
Fig-3: Endoscopic view showing the mass arising from posterior part of right inferior turbinate

Fig-4: Histopathology picture showing Mickulicz cells and Russell bodies

Histopathological examination showed fibrocollagenous tissue infiltrated by sheets of chronic inflammatory cells comprising predominantly of plasma cells and foamy histiocytes (Mickulicz cells). The plasma cells were seen around the blood vessels. Many Russell bodies were seen. Lymphocytes were also seen.

Fig-5: C T image showing the mass occupying bilateral nasal cavities with no bony erosions

Fig-6: C T image showing the mass extending posteriorly in to the nasopharynx

DISCUSSION
Rhinoscleroma is an airborne disease [2]. It mostly occurs in young adults between second to third decade with no gender prevalence [3]. As in the present case the patient belonged to 6th decade which increased the diagnostic dilemma.

Endemic areas with a high incidence of rhinoscleroma include Africa, Middle East, Central and South America, South, Central and Eastern Europe, India, China and Indonesia[3-6]

There are 3 stages of Rhinoscleroma

Atrophic stage:
Crust formation and foul-smelling nasal discharge with atrophic changes in floor of nose, septum and turbinate.

Granulation or nodular stage:
Non-ulcerative nodules develop which at first are bluish red and rubbery and later become paler and harder.

Cicatrizing stage: Adhesion and stenosis of the respiratory tract
As the patient had presented with smooth, vascular swelling without prior history of nasal crusting and foul-smelling nasal discharge the current case posed a diagnostic dilemma.

Differential diagnosis to needed to be ruled out are tuberculosis, actinomycosis, leprosy, histoplasmosis, blastomycosis, sarcoidosis, wegener'sgranulomatosis,lymphomas.

Treatment to achieve cure and to avoid recurrence is challenging. Treatment must be intense and prolonged to eradicate the disease completely. Most effective antibiotics are streptomycin, rifampicin and tetracycline given in large doses for a minimum of 4-6
weeks and continued until 2 consecutive cultures from biopsy material are proved to be negative. Surgical debridement was done in this case because there was significant airway obstruction. In late cases where disease is eradicated plastic reconstruction can be done.

CONCLUSION
Rhinoscleroma as in this case can be a diagnostic dilemma. A deeper study into the pathophysiology and the treatment modalities is required because of the high rate of recurrence.

REFERENCES