Rhinoscleroma is a chronic granulomatous disease caused by Gram-negative bacillus called Klebsiella rhinoscleromatis or Frisch bacillus. We are presenting a case of Rhinoscleroma who has had repeated surgeries for same but had a poor follow up which has resulted in severe disfigurement and loss of vision.

ABSTRACT

Rhinoscleroma is a chronic granulomatous disease caused by Gram-negative bacillus called Klebsiella rhinoscleromatis or Frisch bacillus.1 Diagnosis is important and it is essential to differentiate this condition from other granulomatous diseases and neoplastic conditions because management is different. Although imaging findings can be suggestive of rhinoscleroma, definitive diagnosis needs histopathological examination. Long-term therapy for months and sometimes years is necessary to adequately treat the infection. Despite treatment, recurrence has been reported in up to 25% of cases at 10 years. Nasal cytology is an easy non-invasive investigation, which is simple, reliable and time saving for follow up. Close observation is the key to long term follow up care. We are presenting a case of Rhinoscleroma who has had repeated surgeries for same but had a poor follow up which has resulted in severe disfigurement and loss of vision.

Case Report

A 43 year old male patient with complain of frontal headache was admitted in the Department of Otorhinolaryngology, Medical College Kolkata and a CT scan of nose and PNS was done which showed features suggestive of granulomatous disease. Biopsy from the growth in the nasal cavity was done which on microscopic examination showed features compatible with rhinoscleroma. He was operated on April 2010 and approximately 1 year after the surgery, he attended ENT out patient department (OPD) with complain of right orbital swelling and again found to have recurrent rhinoscleroma for which he was again operated in November 2011. After the procedure, the swelling decreased in size and the visual acuity in the right eye was 6/60 at that time. At present, he has again presented with proptosis and loss of vision in right eye for the last 2 years and difficulty in nasal breathing for the last 6 months associated with nasal intonation of voice. (Fig. 1) On examination, proptosis of right eye with conjunctival congestion and restriction of ocular movements is noted. Hematological investigations were within normal limits. Ultrasonography of right orbit demonstrated a hypoechoic lesion in the retroocular space with destruction of medial wall of orbit. Few hyperechoic foci were also seen within the lesion, suggestive of calcifications. The post-op Contrast Enhanced CT (CECT) scan showed an ill-defined enhancing soft
tissue density lesion in nasal cavities, right orbit and anterior cranial fossa with proptosis and septal perforation (Fig. 2A, 2B). A contrast enhanced MRI (CEMR) revealed CEMRI scan of the brain and paranasal sinuses reveals a large retroocular soft tissue enhancing lesion in the right orbit with posterior extension in the basifrontal regions bilaterally. Bony destruction is seen involving the ethmoidal air cells, and right maxillary antrum with extension of the lesion into the nasopharynx (Fig. 3).
Discussion

Rhinoscleroma is a chronic granulomatous disease caused by Gram-negative bacillus called Klebsiella rhinoscleromatis or Frisch bacillus. Though rhinoscleroma can involve any structure of the upper respiratory tract, Klebsiella rhinoscleromatis has an affinity for nasal mucosa and thus is present in the nasal cavity in 95-100% of cases. It can also be found in the nasopharynx (18-43%), larynx (15-40%), trachea (12%), and bronchi (2-7%). Immunological status revealed general immunological competence except in effective phagocytosis of the organism by the Mikulicz cells - that is humoral immunity is preserved, cellular immunity is impaired. It has also been proposed that an altered immune response along with an alteration in the CD4+ and CD8+ proportion (an inversion of the CD4+/CD8+ ratio) leads to ineffective macrophage production that are susceptible to bacterial replication. There was an absolute increase of the CD56+ cells and cytotoxic cells that coexpress CD8+CD56+ antigens. There was a relative reduction of the CD3+ cells, and the CD19+ cells tended to show an ambiguous beha-vioural pattern.

Clinical features: The disease runs through the following stages:

(a) Atrophic stage. It resembles atrophic rhinitis and is characterised by foul smelling purulent nasal discharge and crusting.

(b) Granulomatous stage. Granulomatous nodules form in nasal mucosa. There is also subdermal infiltration of lower part of external nose and upper lip giving a 'woody' feel. Nodules are painless and non-ulcerative.

(c) Cicatrical stage. This causes stenosis of nares, distortion of upper lip, adhesions in the nose, nasopharynx and oropharynx. There may be sub-glottic stenosis with respiratory distress.

In our case nasal endoscopy showed partially obstructed choana while inspection of the oral cavity showed a firm growth involving the soft palate, and nasopharynx. Bronchoscopic examination revealed no obvious abnormality. However, in some advanced cases, it may not be possible to perform endoscopic visualization of the larynx transorally due to marked foreshortening of the palatoglossal folds and base of the tongue. The vocal cords and epiglottis were uninvolv ed. Histopathological examination shows infiltration of submucosa with plasma cells, lymphocytes, eosinophils, Mikulicz cells and Russell bodies. The latter two are diagnostic features of the disease. A Warthin-Starry or Steiner stain demonstrating rod-shaped bacilli within vacuolated macrophages (Mikulicz cells) is classic for rhinoscleroma. Positive culture of rhinoscleroma on MacConkey agar is diagnostic, though culture is only positive in 50-60% of patients. Klebsiella rhinoscleromatis can be differentiated from other subspecies on the basis of biochemical characteristics (test results positive for methyl red, negative for urease, and negative for citrate reductase) and has the characteristic somatic (O antigen) - capsular (K antigen) antigenic combination O2:K3.

Differential diagnosis: Rhinoscleroma and Rosai-Dorfman disease can rarely coexist; both have a predilection for the respiratory tract and cervical lymph nodes, a protracted course in younger individuals. Distinction is important since therapy is different. Empiric treatment and S-100-positive histiocytes confirm the diagnosis of Rosai-Dorfman disease. Infectious granulomatous processes may include those caused by bacteria (tuberculosis, actinomycosis, syphilis, leprosy), fungi (histoplasmosis, blastomycosis, paracoccidioidomycosis, sporotrichosis), and parasites (mucocutaneous leishmaniasis). Being atypical infection, possibility of underlying AIDS should be investigated and ruled out.

Treatment: Both streptomycin (1 g/day) and tetracycline (2 g/day) are given together for a minimum period of 4-6 weeks and repeated, if necessary. Other antibiotic drugs such as 3rd generation cephalosporins, clindamycin, ciprofloxacin, minocycline, or rifampicin are also used to treat the bacterial infection. Treatment is stopped only when two consecutive cultures from the biopsy material are negative. Steroids can be combined to reduce fibrosis and decrease inflammation. An endoscope may be
used to widen the passageway (dilation), tracheostomy may be required to establish the airway and the growth can be removed by laser excision or surgery (rhinoplasty). Because the mass is not highly vascular, there is no role of endovascular embolisation in the management of these cases.

**Conclusion**

The main goals of managing a patient with rhinoscleroma are eradication of infection, reduction of morbidity, and prevention of complications. Accurate diagnosis and long term therapy for months and sometimes years is necessary to adequately treat the infection. An inadequate follow up usually results in significant disfigurement and functional loss.

**References**