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Case Report

Ear, Nose and Throat Section

Non Indigenous Rhinoscleroma

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ABSTRACT

Rhinoscleroma (RS) is a granulomatous illness of the upper respiratory tract caused by *Klebsiella rhinoscleromatis*. It is a rare, chronic, granulomatous disease. RS develops in the nasal mucosa subepithelium and can spread to the eustachian tube, maxillary antrum, oral cavity, larynx, orbit, trachea, and bronchi. The present case is of a 29-year-old female who came to the Ear, Nose and Throat (ENT) Outpatient Department (OPD) with chief complaints of bilateral nose block for six months. Examination by anterior rhinoscopy revealed pink nasal mucosa and a swelling arising from both sides of nasal septum, that was soft, non tender, did not bleed on touch. Diagnostic nasal endoscopy showed a midline septal mass which was present just posterior to mucocutaneous junction and a second septal mass was present posterior to first mass at level of posterior part of inferior turbinate. The patient underwent submucosal resection of septal mass and the same was sent for histopathological examination and it was diagnosed as features suggestive of RS. The patient was treated with a course of tablet ciprofloxacin, analgesics, antihistamines. There has been no recurrence during the one year follow-up period. Patient improved symptomatically and had no further complaints.

Keywords: Granulomatous disease, Nasal mucosa, Nasal septal lesion, Rhinoscopy

CASE REPORT

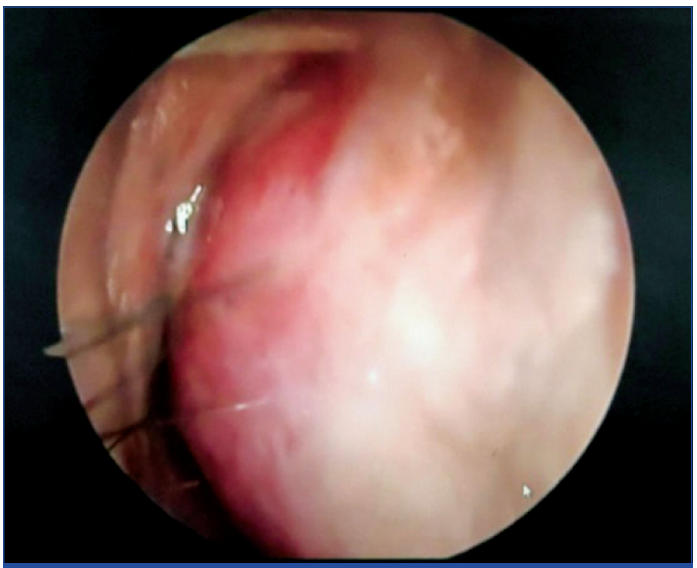
A 29-year-old female visited the ENT OPD with chief complaints of bilateral nose block for six months. It was more during night, relieved on taking nasal drops. On and off, she complained of a hyponasal voice and mouth breathing.There was no history of epistaxis, nasal discharge, recurrent Upper Respiratory Tract Infection (URTI), anosmia, hyposmia, facial heaviness and snoring. On examination of nose, external framework appeared to be normal. On raising the tip of the nose, the vestibule area was normal but a smooth swelling was visualised on both the sides of the nasal cavity. Anterior rhinoscopy revealed pink nasal mucosa and a swelling arising from both sides of the nasal septum, that was soft, non tender, did not bleed on touch and the lateral wall of the nose was partially visualised. No paranasal sinus tenderness was present. Postnasal space by posterior rhinoscopy was found to be clear.

Nasal endoscopy revealed a midline septal mass which was present just posterior to mucocutaneous junction [Table/Fig-1] and a second septal mass was present posterior to first mass at level of posterior part of inferior turbinate. Bilateral inferior and middle meatus were found to be free. Nasopharynx was free.

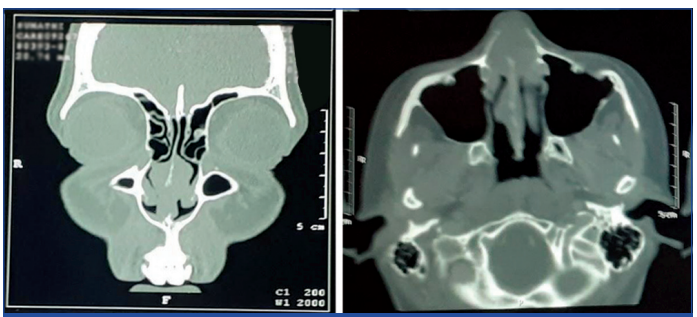
Hence, the patient was provisionally diagnosed with a nasal septal mass with differentials including tuberculosis, leprosy, lupus vulgaris, systemic lupus erythematosus, Wegener's granulomatosis, sarcoidosis.

Computed Tomography (CT) scan of paranasal sinuses revealed nasal septum was deviated to the right with impingement on the right inferior turbinate. Soft tissue density 25×30 mm was noted in cartilaginous section of nasal septum. Mild bony remodelling of septum. Another focal mucosal thickening of size 17×11 mm was noted in bony septum. There was focal ground glass matrix within the bone [Table/Fig-2,3].

Contrast-Enhanced Computed Tomography (CECT) of paranasal sinuses revealed nasal septum was deviated to the right with impingement on the right inferior turbinate. Soft tissue density 25×30 mm was noted in cartilaginous section of nasal septum with mild bony remodelling Another focal mucosal thickening of size 17×11 mm was noted in bony septum with focal ground glass matrix within the bone. Minimal enhancement noted within the lesion



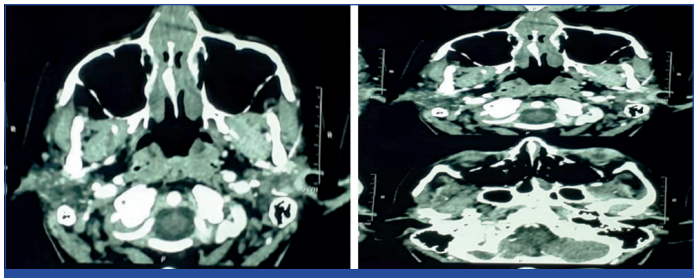
[Table/Fig-1]: Preoperative image showing midline septal mass was present just posterior to mucocutaneous junction.



[Table/Fig-2,3]: Computed tomography paranasal sinuses (plain) show nasal septum deviated to the right with impingement on the right inferior turbinate and focal mucosal thickening. (Images from left to right)

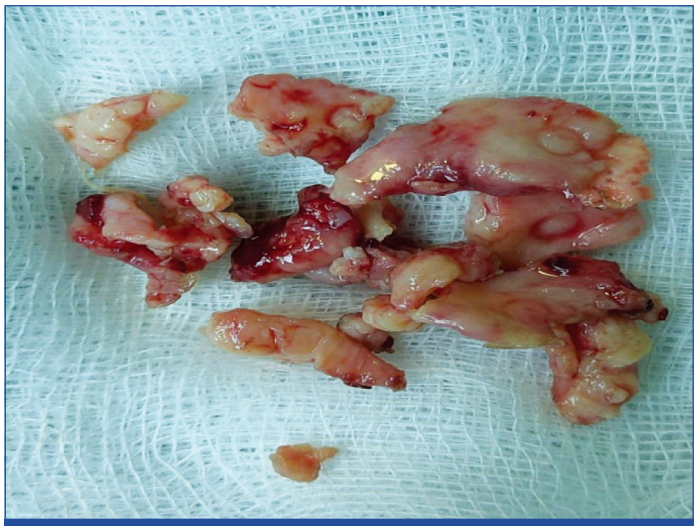
[Table/Fig-4,5]. The differential diagnosis included pseudotumour/ lymphoma.

The patient was diagnosed to have a mass in the nasal septum for which she was planned for a submucosal resection with nasal mass excision biopsy under general anaesthesia. The same



[Table/Fig-4,5]: Contrast-enhanced computed tomography scan showing a soft tissue density in cartilaginous section of nasal septum with mild bony remodelling and minimal enhancement. (Images from left to right)

was performed seven days from the day of first consultation of the patient, after doing the necessary haematological and radiological investigations and getting anaesthetist fitness.Patient was placed in supine position with orotracheal intubation under sterile aseptic precautions with general anaesthesia. Following local infiltration, under endoscopic visualisation, an anterior mucocutaneous junction incision was made up to the roof, and a mucoperichondrial flap adhering to the septal mass was lifted. The dissection plane was found and followed posteriorly. The septal mass was separated from its attachment and was found to be smooth, lobular, and rubbery in texture and was removed [Table/Fig-6]. Excision was carried superiorly upto the roof sparing strip of cartilage and was sent for histopathological examination. Nasal packing done. Patient withstood the procedure and was extubated.

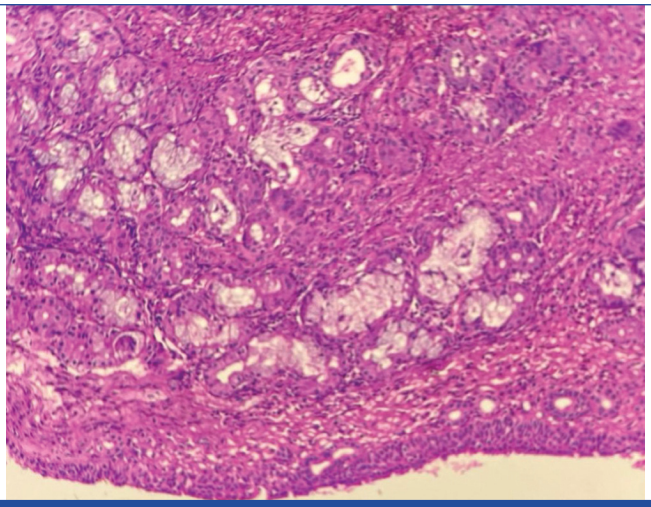


[Table/Fig-6]: Intra-operative specimen.

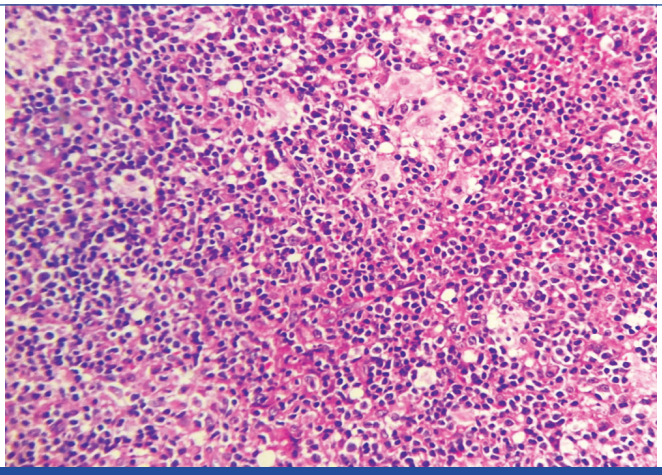
Postoperative period was uneventful. The histopathological report showed mucous secreting glands [Table/Fig-7], lymphoplasmacytic infiltrate [Table/Fig-8], bony spicules [Table/Fig-9], lymphoid follicles with germinal center, [Table/Fig-10] all features suggestive of RS. The patient was treated with a course of tablet ciprofloxacin 500 mg twice daily for a week, tablet pantoprazole 40 mg before food daily, tablet montelukast-levocetirizine 5 mg at night daily for a week. Regular postoperative follow-up for one year has been done. There was no recurrence of the lesion. Patient improved symptomatically and had no further complaints.

DISCUSSION

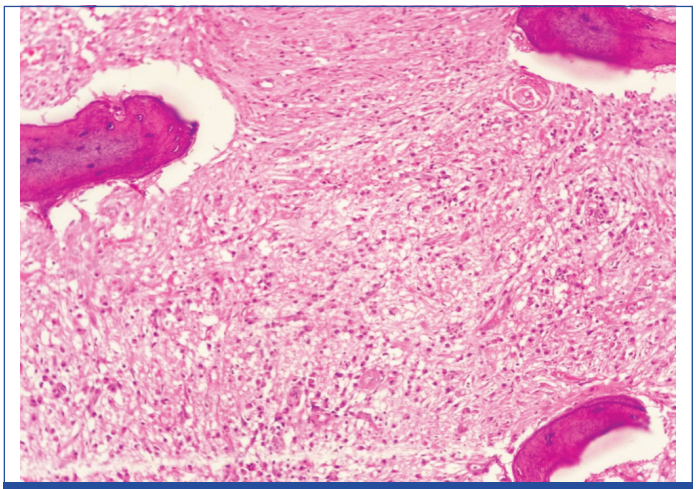
The RS is a granulomatous illness of the upper respiratory tract caused by *Klebsiella rhinoscleromatis*. It is a rare, chronic, granulomatous disease. Von Hebra was the first to invent the word "rhinoscleroma" in 1870 [1]. He described it as a nasal lesion that resembled a variety of sarcoma. This disease is found in Central and Eastern Europe, South and Central America, Eastern Africa, and the Indian subcontinent. This disease chiefly affects young adults in the second or third decades and has no



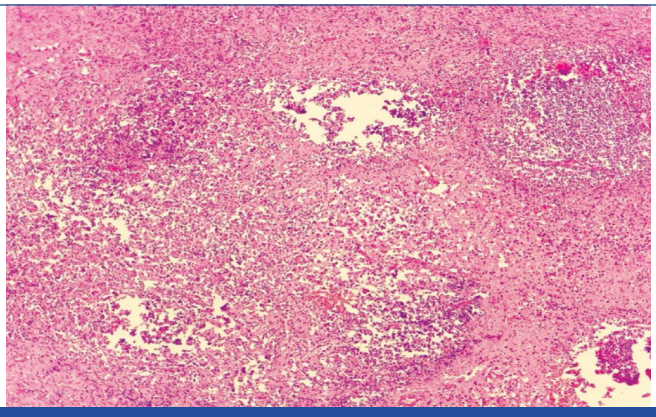
[Table/Fig-7]: Mucous secreting glands seen on histopathology (H&E,20X).



[Table/Fig-8]: Shows lymphoplasmacytic infiltrate (H&E,20X).



[Table/Fig-9]: Shows bony spicules (H&E,20X).



[Table/Fig-10]: Shows lymphoid follicles with prominent germinal center (H&E,10X).

gender prevalence [2]. Poor socio-economic status can also be a contributing factor for the emergence of this disease [3]. Patients with RS may have a variety of immune system disorders. The study done by Fusconi M et al., state that in RS there is a qualitative reduction of the lymphocytes which are hyporeactive to the bacilli [3].

The RS develops in the nasal mucosa's subepithelium and spreads to the eustachian tube, maxillary antrum, oral cavity, larynx, orbit, trachea, and bronchi. The pathogenesis of RS is divided into three stages. The initial stage, known as the catarrhal stage, is characterised by rhinorrhea and recurrent sinusitis. It lasts for weeks to months. Histologically, it is characterised by squamous metaplasia with a neutrophilic infiltration. The second stage known as granulomatous stage is characterised by nasal mass development and tissue damage. This stage lasts for months to years and is characterised by nasal enlargement, deformity, and destruction of cartilage. Histologically, this stage is characterised by Mikulicz cells, which are large, foamy mononuclear cells containing numerous gram negative bacilli and may be surrounded by a plasma cell infiltration with substantial Russell bodies, lymphocytes, and neutrophil foci. The third stage, the sclerotic stage, is characterised by extensive scarring, fibrosis, and chronic inflammatory cells, with few to Mikulicz cells. Deformity and tissue destruction may be marked in the final stage [4]. In index case, patient was in second stage i.e. granulomatous stage.

Transmission usually occurs through direct inhalation or inoculation by respiratory droplets following prolonged contact. Histopathologically, *Klebsiella rhinoscleromatis* invades the subepithelium, multiplies, and promotes capillary growth. In reaction, polymorphonuclear cells enter the subepithelium, phagocytose the bacterium, but disintegrate quickly before the organism is completely digested. Finally, macrophages arrive and phagocytose the *Klebsiella* bacilli. These phagosomes undergo significant enlargement and transform into Mikulicz cells. The cycle continues, when these macrophages rupture and release both active and inactive organisms into the interstitium [5]. The virulence factors that mediate in the pathogenesis of *Klebsiella rhinoscleromatis* likely define the severity and chronicity of the disease. The Capsule Polysaccharide (CPS) prevents complement components from being deposited on the bacteria, preventing adherence and phagocytosis of the bacterium by macrophages and epithelial cells [6]. The altered proportion of CD4+ and CD8+ lymphocytes affects phagocytosis by macrophage leading to all delayed type hypersensitivity response [7].

Clinical presentation varies greatly depending on the organ affected and the duration of the disease. It mainly affects the sinonasal cavities, but the entire respiratory tract may be affected [8]. The RS appears on radiographs as a homogeneous, non enhancing tumour with clear edges with some bony or cartilaginous erosion. In the hypertrophic stage, Magnetic Resonance Imaging (MRI), may reveal masses obstructing the osteomeatal complexes, as well as increased signal intensity on T1- and T2-weighted images. Biopsies of the specimens are taken and examined histologically and bacteriologically to determine the final diagnosis. According to literature, only 60% of the time, cultures establish the presence of *Klebsiella rhinoscleromatis*, although negative cultures do not rule out the diagnosis [9].

Differential diagnosis include sarcoidosis, Wegener's granulomatosis [10], leprosy, tuberculosis, rhinosporidiosis. These can be differentiated from RS with presence of other systemic symptoms and signs specific for each of those diseases. Diseases like, tuberculosis, Wegener's granulomatosis, sarcoidosis may have lung and skin involvement, and need to be evaluated further.

The patient in index case came during the granulomatous stage, her symptoms were predominantly nose block and recurrent URTI. The patient was treated with surgical removal of the mass. After that, the patient was given a three-month course of tablet ciprofloxacin 500 mg twice daily for a week and was followed-up regularly. There was no evidence of any recurrence. Relapse and clinical remission are common findings. Remissions rarely occur. The course of treatment is determined by the stage of the disease at the time of diagnosis [11]. A case study show pressure atrophy of nasal bones caused by tissue fibrosis. This fibrous tissue deposition causes blockage of the lymphatics [12].

The nasal mucosa is found to be a major site of infection as a result of droplet transmission. Without prompt treatment, the condition could spread throughout the respiratory tract over a period of years leading to extensive scarring and adhesions of the nose, palate, and larynx during the healing process. Subglottic stenosis is a life-threatening late stage symptom that requires rapid surgical intervention [13]. A study done by Yigla M et al., on laryngo-tracheo bronchial involvement in a patient with non endemic RS reported that, when the disease spreads to trachea and bronchi causing airway constriction and thick secretions resulted in abrupt lethal airway blockage and death of the patient [14]. If there is a severe airway blockage or cosmetic deformity, surgical debridement is necessary. When debridement is required, laser therapy can be utilised to reduce the amount of cicatricial tissue, reduce disease spread and postoperative oedema [15]. Thus, there is need for long-term antibiotic treatment and regular monitoring to detect early recurrences and spread of disease.

CONCLUSION(S)

To summarise, a case of nasal mass must always be evaluated to rule out granulomatous conditions, infections and also malignancies. The RS is not common among Indian population, yet it should be considered as a differential diagnosis in cases of nasal septal mass. RS has the tendency to spread rapidly leading to airway impairment, leading to tissue loss and scarring. Hence, prompt diagnosis and therapy is necessary. Patients with RS are prone to recurrence, so, they should be monitored closely and regular follow-up should be done.

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