

Rhinoscleroma: Pros and Cons in Diagnosis and Prognosis.

¹Dr. Vaishali Sangole, ² Dr. Rachana Tiwari
^{1, 2}(Department of ENT, MGM Medical College, Kamothe, India)

ABSTRACT: *Rhinoscleroma, also known as Mikulicz disease, is a chronic disfiguring and debilitating disease. This case in a 10 year old girl presented with unusual features and mimicked many other diseases. The characteristic MR features are described. The disease is uncommon in Western Maharashtra. Its scenario in Indian subcontinent is discussed.*

Keywords: *Rhinoscleroma*

I. Introduction

Rhinoscleroma is a chronic progressive specific granulomatous infectious disease affecting the upper respiratory tract. It has affected man for at least 1500 years, yet we know very little of its epidemiology and pathogenesis. Rhinoscleroma is a world wide disease that occurs under all climatic conditions. Most cases are from tropical and temperate zones. The disease has been widely reported from countries in the Middle east, India, and Southeast Asia. Polish surgeon Johann von Mikulich in Wroclaw described its histological features in 1877. Von Frisch identified the organism in 1882. In 1932, Belinov proposed the use of the term scleromarespiratorium because the pathological process in rhinosclerosis may involve not only the upper but also the lower airways. In 1961, Steffen and Smith demonstrated that *K rhinoscleromatis* conformed to Koch's postulates and that it was an aetiological factor in the inflammatory changes typical of scleroma. We present a case of rhinoscleroma in a 10 year old female which posed difficulty in diagnosing due to clinical resemblance with chronic granulomatous conditions like Wegener's granuloma, Rhinosporidiosis, Actinomycosis, Tuberculosis, Histoplasmosis, Sarcoidosis and neoplastic diseases like lymphoma, nasopharyngeal tumour.

II. CASE REPORT

A 10 year old female patient, resident of Nanded, Western Maharashtra came to the ENT services of our hospital, Mahatma Gandhi Medical College, Kamothe, Navi Mumbai with the chief complaint of nasal mass in bilateral nostrils, progressive nasal obstruction and impaired sense of smell since the past 2 years. She also noticed nodular lesions over the palate causing difficulty in eating and swallowing since the past one year. The mass noticed in the left nostril initially was as small as of a peanut, gradually progressed to fill not only bilateral nostrils but also extended to involve the vestibule crossing the midline over the columella and adjacent part of the upper lip. The lesion was painful on touch with occasional episodes of unprovoked bleeding. There was progressive bilateral nasal obstruction causing the patient to mouth breath. There was gradually progressive impairment in the sense of her smell. An year later she noticed ulcerative-nodular lesions covered with slough over her hard palate which enabled her to eat hot and spicy edibles and caused difficulty in swallowing. There was no history of rhinorrhea, hoarseness, difficulty in breathing, ear related complaints, drug allergies, taking long term medications, past surgery. She was non diabetic with no history of past tuberculosis or contact with tuberculosis. She had a congenitally underdeveloped right eye causing total blindness on the same side.

On examination, a pinkish nasal mass, filling both the nostrils, extending beyond the midline over the columella and adjacent part of the philtrum. It was tender, did not bleed on touch and was firm in consistency. The nasal lesions could not be probed, nor evaluated endoscopically due to extreme tenderness. A small hard nodular lesion was present over the tip and supratip region. Multiple indurated painless ulcers of the size 1cm x 1.5cm in size, covered with slough was observed on the hard palate. Posterior rhinoscopy was unremarkable. Ophthalmological examination on the right eye revealed microphthalmia and microcornea with no perception of light. Her systemic examination was normal.

Haematological and biochemical investigations were within normal limits. Erythrocyte sedimentation rate (ESR) was 25 mm in first hour. Her P- ANCA done by Elisa was negative (3.40U/ml). Her 2D ECHO test was normal. Stains for Acid fast bacilli (AFB, Gomori silver methenamine) were negative for the nasal swabs. Nasal swab was also sent for culture sensitivity which showed growth of staphylococcus aureus sensitive to all the antibiotics.

A plain MRI of the face was performed using T1, T2 and STIR sequences which showed iso to hyperintense nodular infiltrative lesions on T1, nodular T2 and hyperintense lesion on STIR involving the nose & extending along both anterior nasal cavities. The lesion also extended in the oropharynx along the inferior aspect of hard palate, posteriorly involving the soft palate. In the posterior aspect of hard palate there was

evidence of a focal defect through which the lesion was seen to be extending superiorly occupying the nasopharynx and posterior nasal cavity predominantly on the right side. Moderate narrowing of nasopharyngeal airway was noted due to the lesions. Mucosal thickening was seen involving bilateral maxillary and ethmoid sinuses. Right orbit was underdeveloped (small size with contour abnormalities) with evidence of dislocated lens. Hyperintensities were seen on bilateral mastoid air cells suggestive of mastoiditis. Findings corresponded to granulomatous lesions with a differential diagnosis of Wegener's granulomatosis, Rhinosporidiosis radiologically.

The first superficial biopsy tissue measuring 0.5cm x 0.3 cm was documented as Vasculitis with Granulation tissue. In view of vasculitis as per the first histopathology report with provisional diagnosis of Wegener's granulomatous, Prednisolone in the dose of 2 mg/kg/day was started. But after showing initial response of reduction to minimal inflammation further response of the lesion stopped showing any regressive changes.

In view of the above modality of treatment giving optimal response to the patient a deeper tissue biopsy was then planned in accordance with the histopathologist. H& E stained sections studied showed tissue lined by pseudo stratified ciliated columnar epithelium. Few foci of metaplastic squamous cells in underlying epithelium was also seen. Underlying stroma showed chronic granulation tissue with abundant plasma cells and foamy histiocyte (Mikulicz cells). Within the cytoplasm of Mikulicz cells, bacilli (Klebsiella rhinoscleromatis) were seen which are round to ovoid bacilli. Active granulation tissue with proliferating capillaries were seen. On the basis of the clinical and histopathological findings, the patient was diagnosed as a case of rhinoscleroma in the granulomatous stage. She was then started with Doxycycline (100mg BD) and Septran (Sulfamethoxazole 100mg + Trimethoprim 20mg).

III. DISCUSSION

Rhinoscleroma is a disabling disease because of its persistent, specific, granulomatous inflammatory nature resulting in fibrosis and sclerosis. Rhinoscleroma almost always affects the respiratory tract. Nose is the most common site (95-100%) as seen in our case.¹ It usually starts in the subepithelium of the nasal mucosa, spreading thereafter to other areas, such as the subepithelium of the pharynx, which is involved in 50% of cases. Extension to the oral cavity or oropharynx is seen in 18% to 43% patients. Other affected sites include the eustachian tube, maxillary antrum, oral cavity, larynx, orbit, trachea, and bronchi.²

Wahi and Misra (1964) reviewed the problem of rhinoscleroma and pointed out that the disease is not uncommon in our country and is prevalent in the Northern and Central parts of India above the line joining Bombay to Vishakapatnam.³ Wahi et al (1959) have shown that the disease is common in Lucknow, Jaipur and Indore. From India cases have been reported by Gupta et al (1961), Somani et al (1964), Kakar et al (1964-65), Misra et al (1965), Popli et al (1966), Kakar et al (1967), and Misra (1970). The largest series of cases are reported by Kakar et al (1965), of 20 cases from Delhi. This entity is seen mostly in North India as reported by some workers.^{4,5} Zafar U et al⁵ found a lower incidence of rhinoscleroma (4.83%) at Aligarh, Uttar Pradesh than that observed by Tondon et al⁴. A case of rhinoscleroma mimicking malignancy was reported by Abrol D et al⁶ from Srinagar. Neelam S et al⁷ reported a case after 20 years from Delhi in 2011. Bhowate R et al recently reported a case from Wardha, Western Maharashtra.⁸ We report this case from rural part of Nanded, Western Maharashtra.

Acquisition of disease is facilitated by crowding, poor hygiene and poor nutrition. There is a familial predisposition, probably owing to infection by intimate contact. There is also a suggestion that iron deficiency may predispose to the disease acquisition.⁹ Iron deficiency can lead to squamous metaplasia and this might be an explanation for the association with poor nutrition and higher incidence in women. The disease most frequently affects persons in the 20-40 year age range. Cases have been reported in paediatric age group in as low as 1 year old child¹⁰ and also in elderly patients. Female are more frequently affected than males (ratio 13:1).

The disease manifests as 3 overlapping phases: exudative, proliferative, and fibrotic (cicatrical). The exudative phase is characterized by watery nasal discharge and crusting (catarrhal stage). Gradually nodulo-ulcerative lesions develop involving the nose, upper lip, palate and adjacent regions of the upper respiratory tract (granulomatous stage). The lesion is dusky red, stony hard, insensitive to touch and minimally tender. Intra-nasal rubbery, polypoid lesions are common. In the advanced stage, there is destruction of the nasal cartilages with resultant deformity (Hebra nose). This is followed by a marked tendency to fibrosis and subsequent obstruction (sclerotic stage). In the granulomatous stage it can be mistaken for mucocutaneous leishmaniasis, rhinosporidiosis, lupus vulgaris, sarcoidosis, Wegener's granulomatosis or a carcinoma.

To our knowledge, only a limited number of studies have described the MR imaging features of rhinoscleroma.¹¹ The high signal intensity of rhinoscleroma on T1-weighted images may be explained by the high protein content within the Mikulicz cells and Russell bodies. The slightly high signal intensity on T2-weighted images may be attributed to the high cellular component of rhinoscleroma, mainly Russell bodies and Mikulicz cells. Hypointense foci within these lesions may be attributed to areas of fibrosis.

The histopathological picture in the granulomatous stage is highly characteristic. Dense plasma cell infiltration with Mikulicz cells and Russell bodies are seen.

Antibiotics constitute standard treatment in the early stage of the disease. Surgery and laser ablation can be used only after the disease is under control. Antibiotics used in the treatment of rhinoscleroma include streptomycin, tetracyclin, ciprofloxacin, trimethoprim-sulfamethoxazole, rifampicin, clofazimine, cephalosporins, and sulfamethoxazole. Tetracyclin has the facility of oral administration, but it requires prolonged therapy in terms of months or years with poor patient compliance. Ciprofloxacin, a fluoroquinolone, is an antibiotic with excellent tissue penetration and a broad antibacterial spectrum of action. Adverse effects are comparatively few and include gastrointestinal symptoms in three to six percent of patients. Keeping in view it is not recommended in patients under 12 years of age because of the risk of arthropathy and Tetracycline to be avoided in the paediatric age group because of teeth staining, a combination of Doxycycline (100mg BD) and Septran (Trimethoprim 20mg + Sulfamethoxazole 100mg) especially for and its effectivity is low cost in third world nations was prescribed in our patient for 6 weeks. Dramatic response to the treatment was found without relapse after 12 months of follow up.

FIGURES



Fig 1: A pinkish nasal mass, filling both the nostrils, extending beyond the midline over the columella and adjacent part of the philtrum. It was tender, did not bleed on touch and was firm in consistency.



Fig 2: A pinkish nasal mass, filling both the nostrils, extending beyond the midline over the columella and adjacent part of the philtrum. Nodulo-ulcerative lesions over the hard palate, soft palate and uvula..

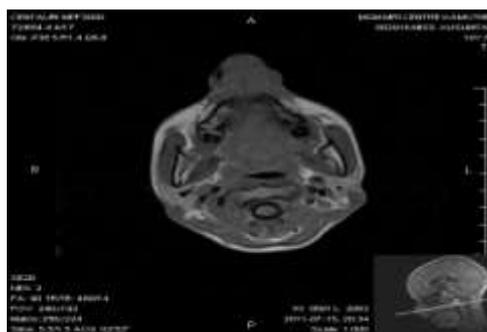


Fig 3: T1 W images axial section shows iso to hyper intense nodular infiltrative lesions involving nose and hard palate.



Fig:4. Axial T2 at the level of nose shows ill defined iso to hypointense nodular lesion involving nose & nasal cavity s/o granuloma.

References

- [1] Hart CA, Rao SK. Rhinoscleroma. *J Med Microbiol* 2000;49:395-6.
- [2] Alfaro-Monge JM, Fernandez-Espinosa J, Soda-Merhy A. Scleroma of the lower respiratory tract: A case report and review of literature. *J Laryngol Otol* 1994;108:161-3.
- [3] Wahi AL, Misra RN. A note on the geographical distribution of Scleroma. *J of Laryngology* 1964;78:573-577.
- [4] Tandon PL, Gulati J, Mehta N. Histological study of polypoid lesions in the nasal cavity. *Indian Journal Otolaryngol head neck Surg* 1971;13:3-11.
- [5] Zafar U, Khan N, Afroz N, Hasan SA. Clinicopathological study of non-neoplastic lesions of nasal cavity and paranasal sinuses. *Indian J Pathol Microbiol* 2008;51:26-9.
- [6] Abrol D, Maqbool M, Afroz F, Ashraf M, Khan NA. Rhinoscleroma mimicking malignancy: A case report. *JK Science* 2008;10:32-3.
- [7] Neelam S, Sanjeev S, Sunaina A, Deepika. Cytological features of rhinoscleroma. *Indian J of Pathology and Microbiology* 2011;54(4):806-808.
- [8] Bhowate R, Degwekar S, Rawlani S, Dangore S. Rhinoscleroma with involvement of the maxillary sinus, orbital floor, and temporomandibular joint: A case report. *J Oral Maxillofacial Surg* 2012;70:135-140.
- [9] Akhounk S, Saad EF. Iron deficiency in atrophic rhinitis and scleroma. *Indian J Med Res* 1987;85:576-579.
- [10] Kohli GS. Rhinoscleroma in a child. *Indian Journal of Otolaryngology* 1978;30(1):36-37.
- [11] Le Hir P, Marsot-Dupuch K, Bigel P, et al. Rhinoscleroma with orbital extension: CT and MRI. *Neuroradiology* 1996;38:175-178.