Rhinoscleroma: A case report

Bhagyalakshmi A., Rao C.V., Krishna Kishore T., Kartheek B.V.S.

ABSTRACT

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Case Report: We report a case of rhinoscleroma in a 32-year-old female presenting with extensive swelling of mid-face encroaching onto the lower two-thirds of the nose, upper lip and oral cavity and nasal blockage with a duration of more than five years. A biopsy was taken for histological diagnosis which confirmed the diagnosis of rhinoscleroma followed by excision of the lesion and reconstruction of the nose and upper lip. For five years, there was no evidence of recurrence.

Conclusion: When the disease progresses with proliferation, it may simulate a tumor, as in our patient with typical Hebra nose.

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Keywords: Chronic granulomatous infection, Hebra nose, *Klebsiella rhinoscleromatis*, Mikulicz cells, Rhinoscleroma

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INTRODUCTION

Rhinoscleroma or scleroma is a chronic granulomatous disease caused by Gram negative bacillus called *Klebsiella rhinoscleromatis* or Frisch bacillus. Nasal cavity is the most common predilection site and may extends to nasopharynx, oropharynx, larynx, trachea and bronchi. Females are more frequently affected between 10–30 years of age. Rhinoscleroma is mostly endemic in tropical countries [1]. The disease was first described by the dermatologist Ferdinando Von Hebra in 1870 [2]. We present a case of Rhinoscleroma with typical Hebra nose.

CASE REPORT

A 32-year-old female from tribal area of Visakhapatnam district, Andhra Pradesh (India) presented with extensive swelling of five years duration on mid-face occupying the lower two-thirds of the nose, upper lip and oral cavity (Figure 1). The swelling started as a small nodule over the tip of the nose and gradually increased in size. The patient had applied some native treatment on the swelling. The swelling occupied the entire area described. The surface was red and irregular and was bleeding on touch. There

was crust formation all over producing offensive smell mimicking a malignant lesion. She was able to open the mouth and take food as the tongue; lower lip were not involved. The patient was breathing through the oral cavity. It was painless swelling with no regional lymph nodal involvement. The lesion was biopsied twice and the initial histological diagnosis was an inflammatory lesion. As the lesion was large, distorting the facial configuration, radical excision of the swelling and reconstruction of the nose and upper lip using forehead skin was planned.

Accordingly, the patient was operated upon under general anesthesia and the lesion was excised and reconstruction of the nose and upper lip was done using oblique classical Indian forehead flap in stages. On Gross examination the excised lesion was measured 7x7 cm, grayish white on cut section and firm in consistency (Figures 2 and 3). The histopathological examination with hematoxylin and eosin staining (H&E) revealed hyperplasia of surface squamous epithelium with subepithelial sheets of mixed inflammatory cell infiltrates composed of neutrophils, lymphocytes, plasma cells, Mikulicz cells and homogenous eosinophilic round to ovoid bodies (Russel bodies) and areas of fibrosis. (Figures 4–6). Special staining with Gram's stain and periodic acid (PAS) stain revealed small bacilli in Mikulicz cells (Figures 7 and 8). The diagnosis was confirmed to be rhinoscleroma. Postoperative period is uneventful and the patient was discharged (Figure 9). We could follow the case for five years, there was no evidence of recurrence.

DISCUSSION

Rhinoscleroma is a chronic infectious disease caused by *Klebsiella rhinoscleromatis*, a Gram-negative, facultative intracellular, non-motile encapsulated bacillus identified by Von Frisch in 1882 [3]. The nose, pharynx, larynx, trachea, and occasionally also the skin of the upper lip are distorted and infiltrated with hard, granulomatous masses. The disorder always begins in the nose [4]. The nose is affected in 95–100% of cases, the pharynx in 18–43%, larynx in 15–80%, trachea in 12%, and bronchi in 2–7% [5].

The disease evolves through three stages:

- (1) Catarrhal stage: It is characterized by foul smelling purulent nasal discharge and crusting resembling atrophic rhinitis.
- (2) Granulomatous stage: In this painless and nonulcerative granulomatous, nodules form in the nasal mucosa. There is also subdermal infiltration of external nose and upper lip giving a 'woody' feel.
- (3) Cicatricial stage: This causes stenosis of nares, distortion of upper lip, adhesions in the nose, nasopharynx and oropharynx.

There may be subglottic stenosis with respiratory distress [6]. Rhinoscleroma spreads from person-to-person by air-borne transmission. The pathogenesis

Figure 1: Clinical photograph showing nasal lesion leading to external expansion of nose known as Hebra nose.

Figure 2: Gross photograph of the excised lesion measuring 7x7 cm.

Figure 3: Gross photograph of the cut section of the lesion showing grayish white, firm in consistency with focal yellowish areas.

Figure 4: Photomicrograph showing sheets of inflammatory cell infiltrates and bands of fibrosis (H&E stain, x400).

Figure 7: Photomicrograph showing Small rods in Mikulicz cells (Gram's stain, x1000).

Figure 5: Photomicrograph showing mixed inflammatory cell infiltrates composed of neutrophils, lymphocytes, plasma cells and homogenous, eosinophilic round to ovoid bodies (Russell bodies) (H&E stain, x100).

Figure 8: Photomicrograph showing Mikulicz cell with bacilli (Periodic acid schiff stain, x400).

Figure 6: Photomicrograph showing lymphocytes, plasma cells and Mikulicz cells (H&E stain, x100).

Figure 9: Postoperative photograph of the patient after excision of the lesion and reconstruction of the nose and upper lip using oblique classical Indian forehead flap.

of rhinoscleroma is still unclear. Host susceptibility is thought to be important in development of the disease. Altered immune response with impaired cellular

immunity also plays an important role in the development of the disease apart from the infection. However, an alteration in CD4:CD8 population in blood has been postulated as a cause of chronicity of this disease. The altered proportion of CD4+ and CD8+ lymphocytes in the lesion may produce disabled macrophages, allowing bacterial multiplication inside them and an ineffectual delayed type hypersensitivity response [7].

The patient in our case had presented typically with a hypertrophic stage lesion involving the nose along with hypertrophy of nose ring areas as she used to put nose rings in both nostril areas as seen in the clinical and gross photograph.

Sood et al. [7] studied the cytohistological features of rhinoscleroma and correlated the cytological findings with the histopathological findings along with immunostain CD68. We could not do cytological examination and immunohistochemical study in our case.

Histopathological features are characterized by a chronic granulation tissue with abundant plasma cells and Mikulicz cells. Russell bodies (plasma cells with retained globules of immunoglobulins) are frequently observed. The characteristic cell is the Mikulicz cell, a large histiocyte with a pale, vacuolated cytoplasm containing causative bacilli which are diagnostic of rhinoscleroma. They can be seen faintly in sections stained with hematoxylin and eosin but are better visualized with the Giemsa stain or a Warthin-Starry silver stain. They are also stained magenta color by the PAS technique. They are Gram-negative rods that measure 2–3 µm in length and appear round or ovoid in cross section. Humans are the only identified host of *Klebsiella Rhinoscleromatis* [8].

A positive culture in MacConkey agar is diagnostic of rhinoscleroma, but it is positive in only 50–60% of patients. The diagnosis is confirmed by histology. Differential diagnosis includes other granulomatous conditions, e.g., tuberculosis, leprosy and fungal infections [9].

The mucosal epithelium overlying the cellular infiltrate often exhibits hyperplasia, which may be so pronounced as to give rise to a mistaken diagnosis of squamous cell carcinoma. Special stains and associated clinicopathologic features usually enable to distinguish rhinoscleroma from other conditions^[4]. In our case, marked hyperplasia of surface squamous epithelium but there is no doubt of malignancy.

In initial catarrhal stage, treatment with antibiotics streptomycin and tetracycline eradicates the infection. Combined surgery and antibiotic treatment are required in granulomatous and cicatricial stage. In our case, combined antibiotic treatment and surgery was curative and the case had been followed-up for five years.

CONCLUSION

Scleroma (rhinoscleroma) is a chronic granulomatous condition caused by *Klebsiella rhinoscleromatis* which is

endemic in northern parts of India. This report presents a case of nasal rhinoscleroma from South India presenting in granulomatous stage. When the disease progresses with proliferation, it may simulate a tumor, as in our patient with typical Hebra nose.

Author Contributions

Bhagyalakshmi A. – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Rao C.V. – Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Krishna Kishore T. – Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Kartheek B.V.S. – Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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