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Rhinoscleroma Presenting As Nasal Mass – A Case Report

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Abstract

Rhinoscleroma is a human specific chronic granulomatous disease caused by Gram negative bacilli, Klebsiella rhinoscleromatis (1). The specific histologic feature is accumulation of foamy macrophages called Mikulicz cells. We report a case of 35 years old female who presented to the ENT department with firm, non-tender swelling over the dorsum of the nose. Excision was done and specimen was sent for histopathlogical examination which confirmed the diagnosis of rhinoscleroma.

Key words: *Rhinoscleroma; Klebsiella; Mikulicz cells.*

Introduction

Rhinoscleroma is a chronic granulomatous infection affecting the upper respiratory tract. It is a progressive infectious disease which may lead to severe consequences or death [(2)]. Rhinoscleroma is caused by bacterium Klebsiella pneumonia subspecies rhinoscleromatis [(3)]. It is a gram-negative , non-motile, encapsulated bacillus [(3)]. It is endemic in developing countries with poor hygiene infrastructures and is found in Eastern Europe, Middle East, tropical Africa, South and Central America and South East Asia [(1)]. Though

K.rhinoscleromatis can infect any structure in upper respiratory tract, it has an affinity for nasal mucosa (4). In this case report, we present a case with swelling on dorsum of nose which on histopathological examination revealed the features of Rhinoscleroma.

Case Report

A female patient, aged 35years presented to the ENT department with swelling on the right side over the dorsum of the nose since 3 months. The swelling started as small papule and gradually increased in size. There was no history of pain, trauma, nasal obstruction, epistaxis, nasal dryness or other nasal symptoms. No other respiratory symptoms were present. On examination, swelling over the upper 1/3rd of the right side of the dorsum of the nose measuring 1X1cm was noted. The swelling is firm in consistency with well defined margins. Skin over the swelling is pinchable and swelling is adherent to underlying structures. Clinically it was suspected to be dermoid cyst and surgical excision was done under local anesthesia. Curvilinear incision was given and swelling which was adherent to bone was dissected. Excised specimen was sent for histopathological examination.

Grossly we received globular gray-white soft tissue mass measuring 1X1cm. Histopathological examination revealed fibrocollagenous tissue with sheets of foamy macrophages admixed with lymphocytes and plasma cells (Figure 1 & 2). These foamy macrophages showed PAS positive material in the cytoplasm (Figure 3). Foamy macrophages with disrupted membrane forming microcysts are also noted (Figure 4). The above features are consistent with diagnosis of Rhinoscleroma.

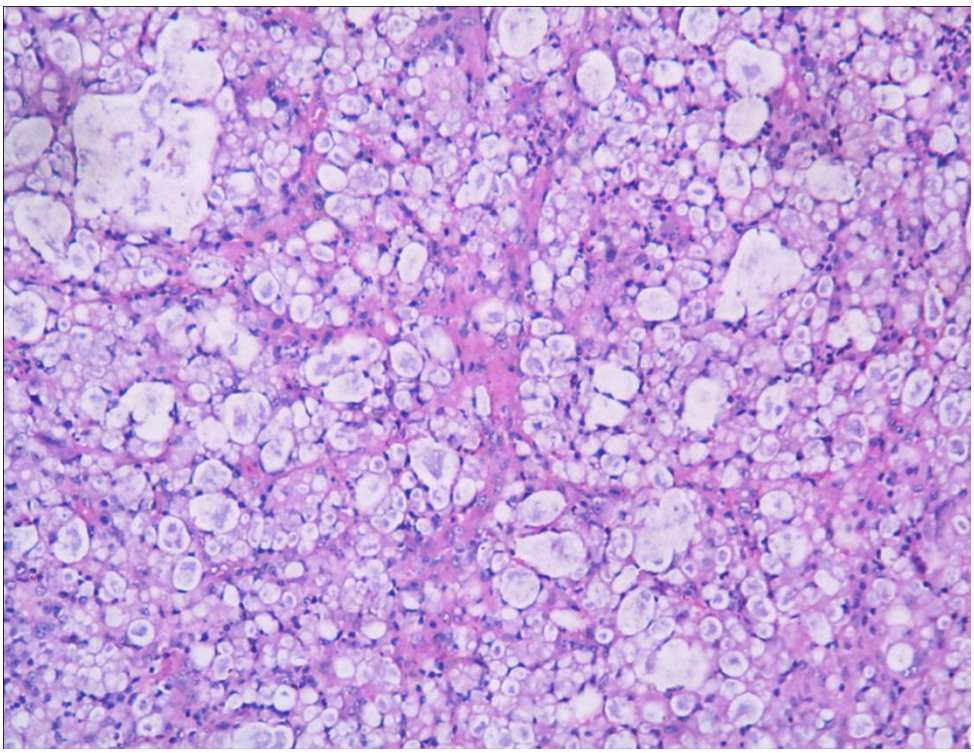

Figure 1: Sheets of foamy macrophages admixed with lymphocytes and plasma cells (H&E X100)

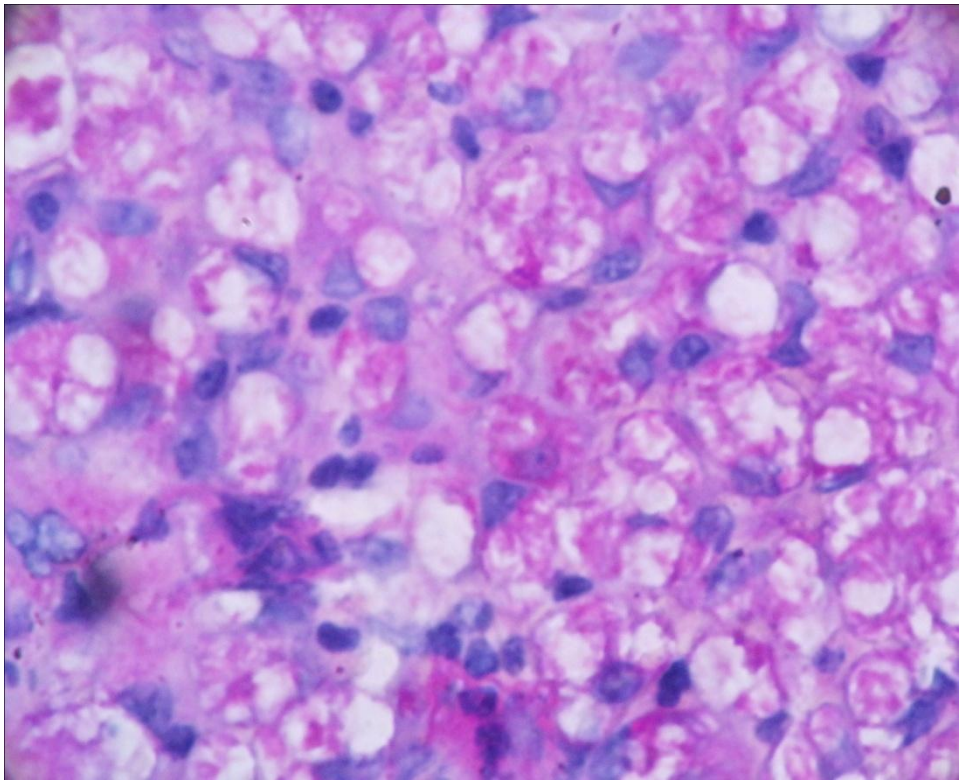

Figure 2: Sheets of foamy macrophages admixed with lymphocytes and plasma cells (H&E X400)

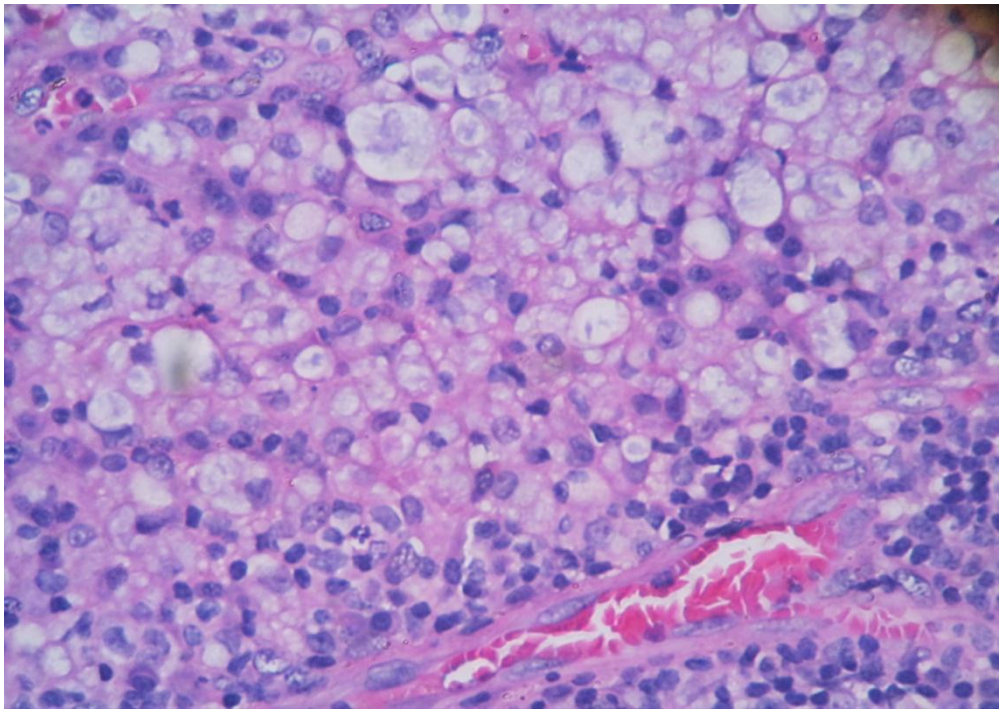

Figure 3: Non-lipid containing foamy macrophages containing PAS positive material in the cytoplasm (PAS X400)

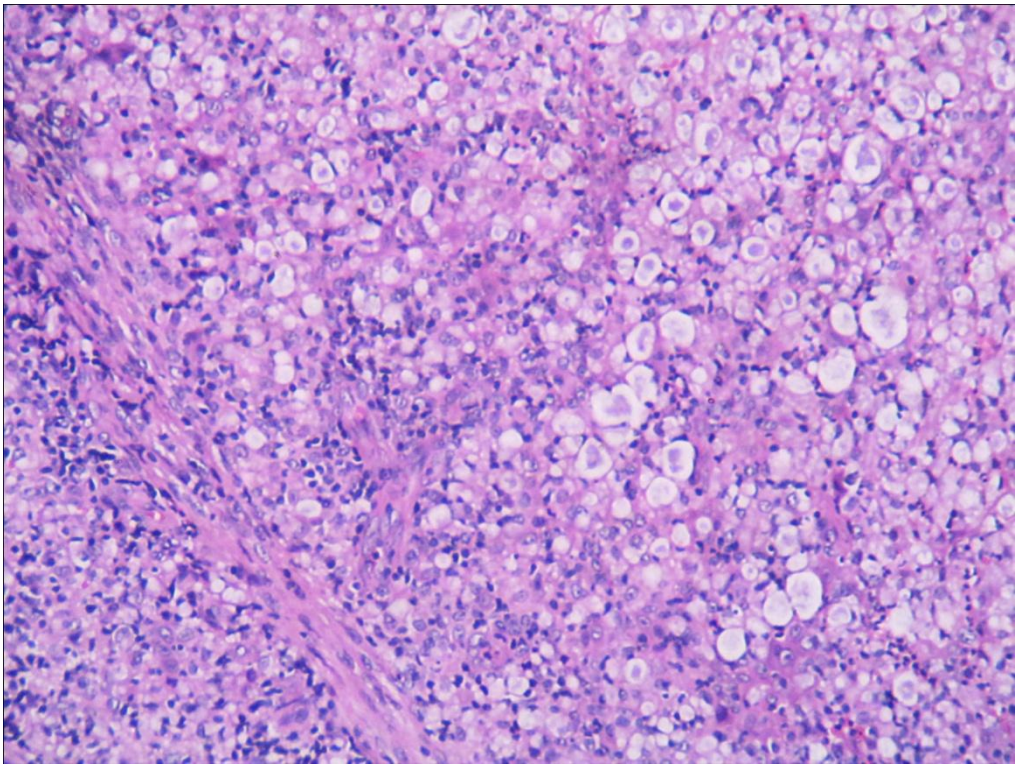

Figure 4: Foamy macrophages with disrupted membrane forming microcysts (H&E X100)

Discussion

Rhinoscleroma is a human specific chronic granulomatous disease caused by enterobacteria, klebsiella rhinoscleromatis which is closely related to pneumoniae [(5)]. It is a gram-negative, encapsulated non-motile diplobacillus which can involve any structure in upper airways with specific affinity towards nasal-mucosa, leading to the term "Rhinoscleroma".

The histological features of Rhinoscleroma was first described in 1877 by Johann Von Mikulich, a polish surgeon in Wroclaw. The etiologic organism was identified in 1882 by Von Frisch. In 1932, the term 'Scleroma respiratorium' was proposed by Belinor as the disease process not only involves the upper airways but also the lower airways. It was demonstrated that K.rhinoscleromatis was an etiologic factor in the inflammatory changes typical of scleroma by Steffen and Smithn in 1961. Genetic factors also are considered in infection caused by K.rhinoscleromatis, due to the occurrence of familial disease in endemic areas [(6)].

This condition is more common in the second or third decades of life and shows female preponderane with female to male ratio of 13:1. It is endemic to areas of Africa, South East Asia, Mexico, central and south America and central and eastern Europe. Sporadic cases occur usually due to immigrants from endemic regions [(7)]. Low socioeconomic status, immunosuppression, poor hygienic conditions, and contact with infected persons are considered to be some of the predisposing factors [(8)].

Klebsiella rhinoscleromatis cannot produce the disease in the healthy individuals but produces the

infection in the individuals with impaired cellular immunity. The lesion shows an altered CD4/CD8 cell ratio with reduction in the levels of CD4 lymphocytes. This possibly leads to diminished T cell response. Macrophages are not fully activated. In these patients there is ineffective phagocytosis of the organism by the macrophages which is contributed by the mucopolysacharides in the capsule of bacilli [9].

Rhinoscleroma is spread from person-to-person by direct inhalation of droplets or contaminated material. The disease process initially starts at the areas of transition such as the vestibule of the nose, area between the nasopharynx and oropharynx and the subglottic areas of the larynx [10]. The most common site for the Rhinoscleroma is the nasal cavity but lesions may also be seen in the larynx, paranasal sinuses, nasopharynx, oral cavity, trachea, bronchi and lip [10].

Rhinoscleroma is a progressive disease, clinically occurring in three stages. The first stage is rhinitic or atrophic stage, which is also described as catarrhal or exudative phase. During this phase histopathological findings are non-specific. There are abundant neutrophilic infiltrate and cellular debris. Very careful examination may reveal few Mikulicz cells and plasma cells. This is followed by second stage which is infiltrative or granulomatous phase. This is followed by second stage which is infiltrative or granulomatous phase. This stage is characterized by the presence abundant Mikulicz cells, plasma cell infiltrate, lymphocytes and few neutrophils. Granulation tissue rich in blood vessels is noted in this stage. Granulomas are also seen in this phase. The second stage evolves into the cicatrical phase which is the third stage of disease. This stage is also described as fibrous or scarring stage due to the presence of abundant connective tissue. Mikulicz cells, plasma cells are rare in this stage.

The Mikulicz cell found in the Rhinoscleroma is a large (10-100µ) mononuclear cell having vacuolated cytoplasm. Cytoplasm shows single, irregular vesicle with granular and fibrillar debris. The vacuolated cells coalesce to form microcysts. PAS staining showed positive material within the majority of vesicles and also in the periphery of these structures. Warthin and Starry silver stain demonstrates bacteria in some of the vacuoles and microcysts. Giemsa and Gram stain can also be used to demonstrate bacteria [11]. Bacteria are present in abundance in the areas of acute inflammation and are rarely found in the fibrous areas.

In Rhinoscleroma, frequent fragmentation of the limiting membrane of the phagosomes is noted which can be because of increased osmotic pressure within the vacuole due to the mucopolysacharide of the bacillus. Vacuolization in the macrophage is produced by the monosaccharides of molecular weight more than 200,000. In Rhinoscleroma, the slime layer of K.rhinoscleromatis is a polysaccharide which has molecular weight above 300,000. It has been proposed that the slime layer or/ bacilli enters the macrophage by endocytosis and resides in phagosome which has membrane permeable to water but impermeable to mucopolysacharides.

Due to this semi permeability of the phagosomal membrane, water enters the phagosome, causing increased intravacuolar pressure which later leads to fragmentation of membrane [12].

Electron microscopy shows phagosomes filled with bacilli and fibrillar or finely granular material surrounding them, arranged in radial pattern. The granular material represents accumulated antibodies on the surface of bacteria (Type A granules) and also around the aggregates of bacterial mucopolysacharides (Type B granules).

Rhinoscleroma should be suspected in patients presenting with nasal obstruction or unremitting, persistent rhinitis. Other clinical conditions presenting with similar symptoms and which are considered in the differential diagnosis of Rhinoscleroma are Tuberculosis, Actinomycosis, Syphilis, Sarcoidosis, Leprosy, Leishmaniasis, Wegeners granulomatosis, Verrucous carcinoma, Basal cell carcinoma and lymphoma. The diagnosis of Rhinoscleroma can be confirmed by the histopathological examination which reveals large vacuolated non lipid containing macrophages (Mikulicz cells) and granulomatous inflammation [13]. Culture on MacConkey agar can be used in conjunction with histopathological findings to confirm the diagnosis. However only 50%-60% of patients show positive results on culture [9].

Treatment of Rhinoscleroma disease includes long term antibiotic therapy combined with surgical intervention in patients with symptoms of nasal or pharyngeal obstruction.

Conclusion

K.rhinoscleromatis is an intracellular facultative bacilli that proliferates in macrophages and is resistant to the digestion of macrophages. Possibility of Rhinoscleroma should be considered in cases of chronic rhinitis. Major cause of tissue injury in Rhinoscleroma is due to the granulomas and fibrosis occurring in the cicatrical stage.

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