Rhinoscleroma: A Case Report

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Abstract

Background: Rhinoscleroma is a chronic granulomatous disease caused by gram negative bacteria; Klebsiellarhinoscleromatis. Respiratory mucosa is predominantly affected involving mainly the nasal cavity and nasopharynx and further extending to the respiratory tract. Systemic involvement is rare. The bacilli can be stained for histopathological examination by Hand E, Gram's stain and Giemsa stain. Treatment being antibiotics and anti-inflammatory drugs.

Case Presentation: Hereby presenting a case of 52 year old male with nasal blockage; clinically found to have a mass encasing the bilateral chonchae and nasopharyngeal wall being investigated for malignancy, but proved to be diagnosed with rhinoscleroma on histopathology.

Conclusion: Rhinoscleroma, usually difficult to diagnose, may be confused with granulomatous disease, carcinoma, tuberculosis and leprosy. Seen majorly affecting the respiratory tract, adequate biopsy material sent for histopathology may prove to be useful in the diagnosis; along with a proper clinical history. The presence of inflammatory background containing Mikulicz cells are highly suspicious of rhinoscleroma. The disease often presents with nasal mass therefore clinically mimics with neoplastic mass. Due to its chronic course, it often proves challenging to treat. Early diagnosis thus ensures timely treatment further avoiding relapses and complications.

Keywords: *Rhinoscleroma, Mikulicz cells, Klebsiellarhinoscleromatis, Microscopy.*

Introduction

Rhinoscleroma is a slowly progressive infectious disorder caused by gram negative rod shaped bacilli; Klebsiellarhinoscleromatis.(1) The bacilli mainly involves the upper respiratory passage, with further extension into the lower respiratory passage. Rarely the involvement of other systems have been identified.(2) The disease has been divided into three clinicopathologial stages: Catarrhal-atrophic, granulomatous and sclerotic.(3) Usually the patients present with nasal blockages, foul smelling purulent discharges, crust formations and repeated epistaxis with progression to permanent defects of nasal deformities, anosmia, dysphonia and stridor.(4)

Clinical Case Presentation: A 52 year old married male came to Otorhinolaryngology OPD with the complaints of bilateral nasal blockage since 6 months. On clinical examination, it was found that the patient had bilateral inferior turbinate hypertrophy and congested mucosa; along with a mass encasing the bilateral chonchae and nasopharyngeal wall on both sides. The probable clinical diagnosis were nasopharyngeal carcinoma or hemangiopericytoma of nasopharynx. Radiological investigations were carried out. Computed Tomography (CT) of Paranasal Sinuses findings were suggestive of Nasopharyngeal carcinoma. A biopsy specimen with multiple, whitish brown tissue pieces aggregating 1 x 1 cm were sent for histopathological examination.

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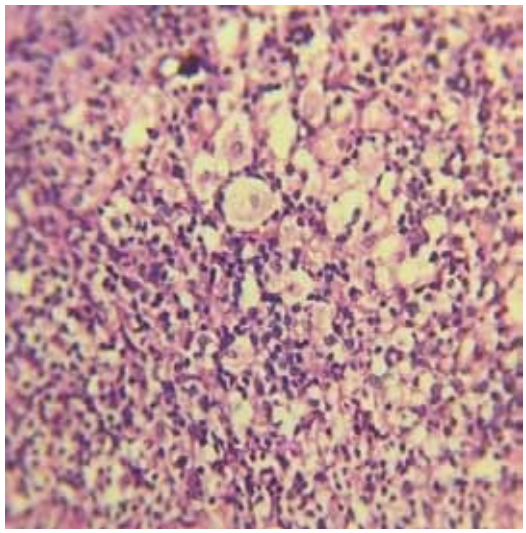
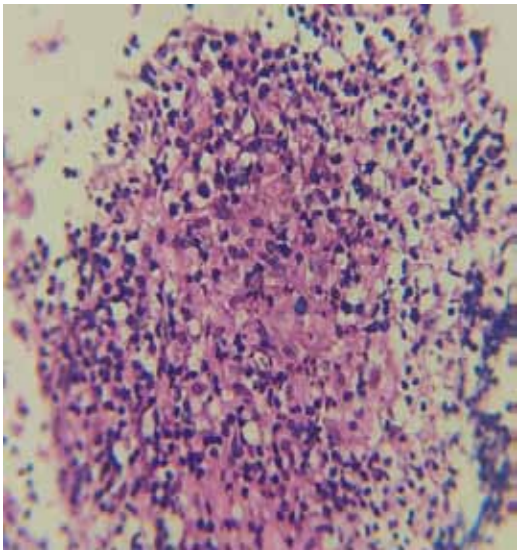


40X microphotograph showing foamy macrophages (Mikulicz cells) in the background of inflammatory infiltrate.



40X microphotograph showing epitheloid granuloma in the background of inflammatory infiltrate.

Discussion

Rhinoscleroma is a chronic granulomatous disease; commonly affecting the respiratory tract. [1] Most common site being the nose (95-100%), followed by nasopharynx, larynx, trachea and bronchi. [1,3] The disease is caused by a immotile, short and encapsulated gram negative organism known as Klebsiellarhinoscleromatis.[1] The only host for this organism are the humans.[1] Inhalation of nasal droplets of the infected person is the mode of transmission.(1) Opportunistic disease in immunocompromised people and must be differentiated from other granulomatous diseases.[2] It is seen to affect majorly the people with low socio-economic status and with poor hygiene. [2]

It is categorized as non-neoplastic mass of the respiratory tract. (5) The progression of the disease is seen in three stages: (A) Catarrhal stage- starting with rhinitis and progressing to rhinorrhoea, crust formation and nasal obstruction; can last uptoweeks or months. (B) Hypertrophic stage- It involves granulation tissue formation with nasal pyramid widening and destruction of nasal septum. Symptoms involve epistaxis, anosmia, reduced sensations of the soft palatefollowed by others. (C) Sclerotic phase- Involves extensive scarring and stenosis of the nasal vestibule.[2]

Histopathology plays a key role in the diagnosis of rhinoscleroma. The bacilli can be demonstrated by H and E, Gram's stain, Geimsa stain, PAS. (1) Diagnostic features seen in rhinoscleroma are the dense inflammatory infiltrate, Mikulicz cells and the granulomas.[6] K. rhinoscleromatis is positive in routine cultures on blood or MacConkey agar in 50%-60% of patients in the granulomatous stage.[7] Detection of Klebsiella antigen, CD68 and altered CD4:CD8 ratio are useful in the diagnosis.[1]

Malkud S, et.al; (2017) documented a case of 34 year old female with complains of swelling of lips and gums since 9 months. On clinical examination, a swelling of the peri-oral area, hypertrophy of gingival and crust formation with fissures was observed over the lips. The nose and throat were found to be uninvolved. The biopsy specimens were sent to histopathology for further examination. The slides stained by H and E, Gram and PAS stains were observed under microscope. Presence of inflammatory infiltrates of plasma cells with Russel bodies and Mikulicz cells were the diagnostic findings that were seen. The patient showed significant improvement with Ciprofloxacin and Doxycyclin and advise of follow-up.[1]

Ahmed A, et.al; (2015) conducted a retrospective study to spot the potential diagnostic features of nasal rhinoscleroma when mikulics cells are absent. Predominance of plasma cells with Russell bodies and neutrophils with squamous metaplasia along with nasal crust formations was found to be diagnostic. This also helped in early diagnosis and treatment of the cases.[4]

In our case, male aged 50 years presented with blockage in the nasal cavity and a nasal mass was found on examination. The biopsy specimens sent for

histopathology examination were stained with H and E, Giemsa and Gram's stains. On H and E stain, Mikulicz cells along with epitheloid granuloma was observed which confirmed the diagnosis of rhinoscleroma. The patient was treated indoor with antibiotics and advised for follow-up.

Conclusion

Rhinoscleroma, usually difficult to diagnose, may be confused with granulomatous disease, carcinoma, tuberculosis and leprosy. Seen majorly affecting the respiratory tract, adequate biopsy material sent for histopathology may prove to be useful in the diagnosis; along with a proper clinical history. The presence of inflammatory background containing Mikulicz cells are highly suspicious of rhinoscleroma. The disease often presents with nasal mass therefore clinically mimics with neoplastic mass. Due to its chronic course, it often proves challenging to treat. Early diagnosis thus ensures timely treatment further avoiding relapses and complications.

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Conflict of Interest: Nil.

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