Case Report

Seemingly neoplastic destructive lesion of maxillary sinus, eroding the bone and extending into skull base-Atypical Rhinoscleromaor Antroscleroma

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Abstract

Rhinoscleroma (RS) is a chronic granulomatous infectious disease predominantly affecting the nose and upper respiratory tract caused by *Klebsiella rhinoscleromatis* (KR), a gram negative diplobacillus. In India, it is prevalent in Northern and central parts. Belinov, in 1932 proposed the term "scleromarespiratorium" as the disease process may involve not only the upper, but also lower respiratory pathways. A 35 year old male from South India, presented with mass in the right maxillary and ethmoid sinuses eroding the bone, extending into infratemporal fossa and skull base, clinically mimicking malignancy. Histopathological examination confirmed rhinoscleroma. Nasal cavity free, isolated ipsilateral involvement of sinusesmakes this case interesting and one among the few case reports reported in the past. Rhinoscleroma should be differentiated from other granulomatous conditions and neoplastic diseases like lymphoma & Sinonasal carcinoma. Correct diagnosis is very crucial as sinus-positive cases are linked with antibiotic resistance and early recurrence after medical treatment.

Keywords: Rhinoscleroma, Klebsiella rhinoscleromatis, sinus involvement, early recurrence, antibiotic resistance.

Introduction

RS, Scrofulous lupus is a chronic slowly progressive granulomatous obstructive infectious disease that frequently affects the respiratory mucosa, especially nose and nasopharynx. In 1870, Ferdinand Von Hebra, Viennese dermatologist coined the term RS. It was believed to be a form of sarcoma till 1877, when Johann von Mikulicz described the histological features of the disease. Von Frisch in 1882, identified *Klebsiellar hinoscleromatis* [1,2]. Another causative agent of RS is Klebsiella ozaenae, very few cases of which are evident in the literature. Identification of subspecies is important from the treatment aspect as K. ozaenae is susceptible to ampicillin, unlike KR which is most commonly resistant.

147 years have passed since the discovery of RS, unfortunately little is known about its epidemiology and pathogenesis. It is endemic in countries of Middle East, tropical Africa, India, Indonesia, South East Asia, Central & South America. In India, it is prevalent in northern and central parts. In Karnataka, most cases are from Southern highlands province, a belt north of Vindhya Mountains [3]. We report a case of RS in a 38 year old man from non-endemic zone of Karnataka.

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Host susceptibility plays an important role in the development of disease. Defective cell mediated immune response, altered CD4+/CD8+ lymphocyte ratio with a decrease in CD4+ lymphocytes and increase in CD8+ lymphocytes has been documented in affected individuals. However, humoral immunity remains intact; this explains predominance of plasma cells in the tissue biopsy [3,4]. The disease manifests as three overlapping clinicopathological phases, catarrhal/ atrophic, proliferative/ granulomatous and sclerotic/ cicatrical phases. Most patients are diagnosed in granulomatous phase because of clinical manifestations.

Treatment of RS involves antibiotics coupled with surgical debridement, especially in cases of airway obstruction or cosmetic deformity. Dual antibiotic therapy with ciprofloxacin and cotrimoxazole or

combination therapy with ciprofloxacin and doxycycline for a period ranging from 2 months to 1year gives good results. Our patient was treated with doxycycline and ciprofloxacin for 3 months. After 3 months of treatment he presented with residual growth in right maxillary sinus.

Case History

A 36-year old man, from Karnataka (Southern part of India) presented with headache, nasal blockade, right sided facial pain and epiphora, presentfor 8 months. He denied history of travel / contact with affected individuals. His haematological and biochemical parameters were normal. Anterior / posterior rhinoscopyand neurological examinations were normal. CT scan of paranasal sinuses showed a destructive mass in the right maxillary sinus, eroding the bone and extending into ethmoid and infratemporal fossa, mimicking malignancy.

Frozen section ruled out malignancy but provisional diagnosis of plasma cell neoplasm/ IgG4 gammopathy was given. However, Immunohistochemistry disproved both. Maxillary antrectomy and debulking of the entire tumor was done. Histology showed necrotizing and granulomatous lesions with vacuolated histiocytes, Mikulicz cells and plasma cells with Russell bodies (Fig 1).

Geimsa (Fig 2) and Gram's stainshowed Gram negative intracellular coccobacilli. Stains for acid fast bacilli and fungal organisms were negative. Patient was treated with combination of ciprofloxacin (500mg, BD) and doxycycline (100mg, BD) for 3months. Follow-up CT (Fig 3 & 4) scan showed residual disease in maxillary sinus with soft tissue extension into infratemporal fossa, pterygomaxillary fissure and pterygopalatine fossa and intra-orbital extension through the inferior orbital fissure.

Endoscopic exploration and clearance was done. Histology showed disease in sclerotic phase and culture did not yield growth. Previous medicationalong with rifampicin and intralesional acriflavine was given. Patient responded to treatment and is doing good.

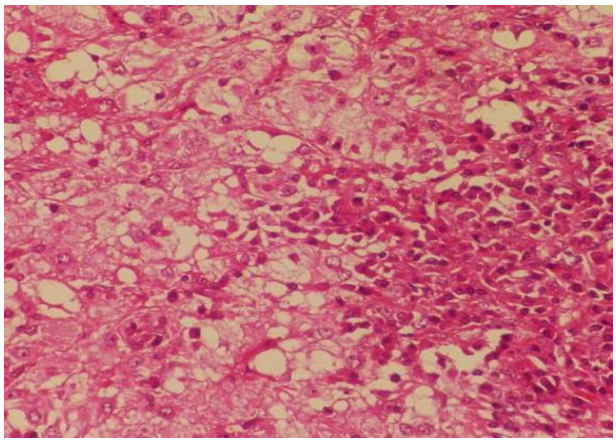

Fig 1

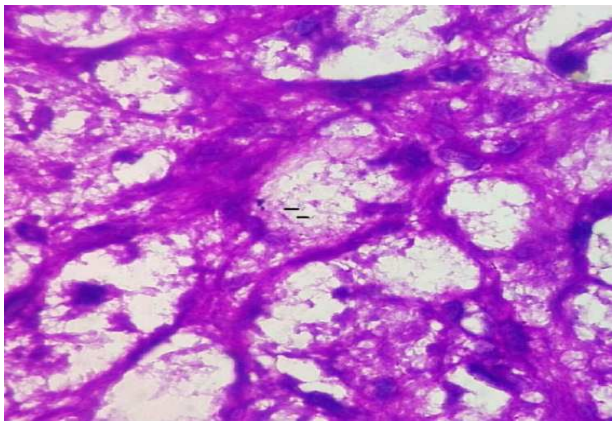

Fig 2

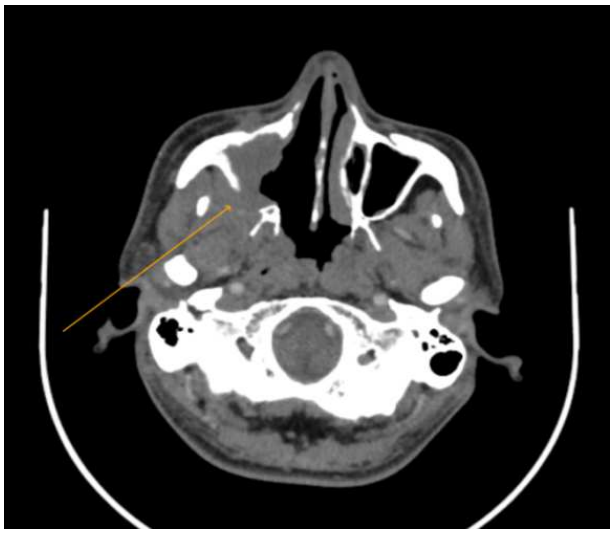

Fig 3

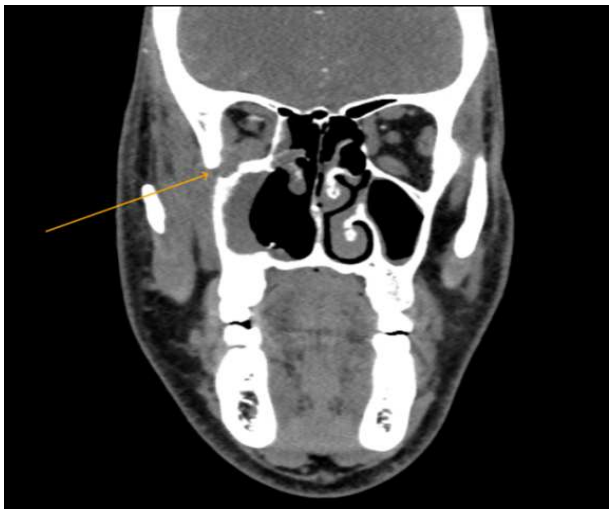

Fig 4

Legends for Figures

Fig 1: Histomorphological picture showing Mikulicz cells and plasma cells with Russell bodies (H & E, 40x).

Fig 2: Picture showing bacilli within the foamy cell, marked with the arrow (Geimsa stain, 40x).

Fig 3: Axial CT- Soft tissue within right maxillary sinus with retroantral extension through a large defect in the posterior wall.

Fig 4: Coronal CT- Intra-orbital extension of soft tissue through the inferior orbital fissure with loss of fat plane within posterior aspect of inferior rectus.

Discussion

RS is an ancient disease. Since its discovery in 1870 many cases involving both upper and lower respiratory tracts with typical and atypical presentations have been published. Hence the term proposed by Belinov, scleromarespiratorium appears more appropriate [5]. RS affecting predominantly sinuses with or without extra sinusextension is aptly termed antroscleroma/ethmoid scleroma based on site of involvement [6,7].

In a study conducted by Ahmed Atef et al, it was observed that more than 1/3rd (9 out of 23) of the cases were associated with histologically proved ethmoid

sinus lesions. Among 9 cases, only 6 showed mucosal granules, rest had normal ethmoid mucosa, this clearly suggests, sinus involvement is underrepresented clinically [8].

Maxillary and ethmoid sinuses are most commonly involved among sinuses. Although some data have emerged about the possibility of sinus involvement, isolated sinus involvement is a rare occurrence posing diagnostic and treatment challenges.

Ours is one such case with isolated ipsilateral maxillary and ethmoid sinus involvement, clinically mimicking malignancy and histologically masquerading as plasma cell neoplasm on frozen section. Rarity and unfamiliarity of atypical presentations lead to delay in diagnosis, posing treatment challenges.

Literature on CT & MRI findings in RS is scant. In a study done by Ahmed Abdel Khalek et al, it was observed that hypertrophic stage had mild to marked high signal intensity on both T1- and T2- weighted MR images, a characteristic finding that differentiates RS from neoplastic pathology, which gives low to intermediate signal intensity [9].

Histopathological examination is the only reliable time saving confirmatory test available at present scenario, especially when cultures are not available and also when done may yield growth only in 50-60% of cases.

However, scarcity or absence of pathognomonic Mikulicz cells during catarrhal or sclerotic phase pose diagnostic difficulties.

Ahmed RH Ahmed et all, has defined histologic diagnostic features in the absence of Mikulicz cells into major criteria including dominance of plasma cells and absence of eosinophils and minor criteria including young age, female gender, bilateral nasal involvement, nasal crustation, squamous metaplasia, Russell bodies and neutrophils [10].

Another challenge is chronicity and early relapses due to antibiotic resistance. Defective phagocytic function of histiocytes whichfail to transform into epithelioid cells andsinus involvement which act as reservoir of infection lead to chronicity and resistance to medical treatment [8].

Conclusion: Isolated ipsilateral sinus involvement isa rare presentation in rhinoscleroma, but when present can pose diagnostic and therapeutic challenges. Histopathological examination remains the mainstay for diagnosis with atypical presentation. Where as, imaging techniques and culture aid as supplementary diagnostic modalities.

Unfamiliarity with respect to histomorphological findings can result is erroneous diagnosis of plasma cell neoplasm / IgG 4 related gammopathy as in this case.

Hence, strong clinical suspicion and familiarity of the histomorphological findings is necessary for early diagnosis as prolonged antibiotic treatment is the basis for successful treatment of rhinoscleroma.

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