

Rhinoscleroma: Mimicking As Acute Invasive Fungal Pan-Sinusitis

S Gulati, J Gulia

Citation

S Gulati, J Gulia. *Rhinoscleroma: Mimicking As Acute Invasive Fungal Pan-Sinusitis*. The Internet Journal of Tropical Medicine. 2008 Volume 5 Number 2.

Abstract

Rhinoscleroma is a chronic indolent granulomatous infection of the nose and the upper respiratory tract. The average time of presentation between the onset and diagnosis is about a decade. An unusual case of Rhinoscleroma in a young immune-competent male with a two month history mimicking as invasive fungal sinusitis is being presented. While Radiological appearance is not pathognomonic of rhinoscleroma, nasal endoscopy is both diagnostic and therapeutic. Histopathology report is confirmatory for rhinoscleroma.

INTRODUCTION

Rhinoscleroma (RS) is caused by *Klebsiella rhinoscleromatis*, which is a gram negative encapsulated diplobacillus. It is a chronic, specific granulomatous disease that manifests an affinity for the mucosa of the upper respiratory tract. Poor hygiene, crowded living environment and mal-nutrition appear necessary for infection and transmission^[1]. It is most frequently recognized in young adults, who may have symptoms for over 10 years prior to diagnosis.^[2]

We present an unusual case of rhinoscleroma in a young HIV negative male, who presented with a short history of two months and was treated as acute sinusitis. Computed tomography was suggestive of invasive fungal pan-sinusitis with breach of the medial wall of the left orbit. Biopsy confirmed the diagnosis of Rhinoscleroma.

CASE REPORT

A 25-year-old male patient presented with a history of nasal discharge and nasal blockage more on the left side associated with headache and low grade fever for two months. He was diagnosed and treated as a case of acute sinusitis with some relief. Patient had a recurrence of symptoms after he discontinued the antibiotics and consulted us on developing pain in the left eye.

General physical examination was normal. Otorhinolaryngological examination revealed DNS to left and thick nasal discharge on both sides. No nasal mass was seen. A Computer Tomography (CT) scan PNS revealed

hyper-dense soft tissue masses with surrounding hypodensities in bilateral maxillary, ethmoid, sphenoid and frontal sinuses with destruction of ethmoid septa, thinning of turbinates and destruction of medial wall of the left orbit. (Fig1, Fig 2). A diagnosis of invasive fungal sinusitis with orbital extension was made and patient was taken for surgery.

Figure 1

Figure 1: Computer Tomography (CT) scan PNS showing hyper-dense soft tissue masses with surrounding hypodensities involving ethmoid, maxillary and frontal sinuses with destruction of ethmoid septa, thinning of turbinates and destruction of medial wall of the left orbit.

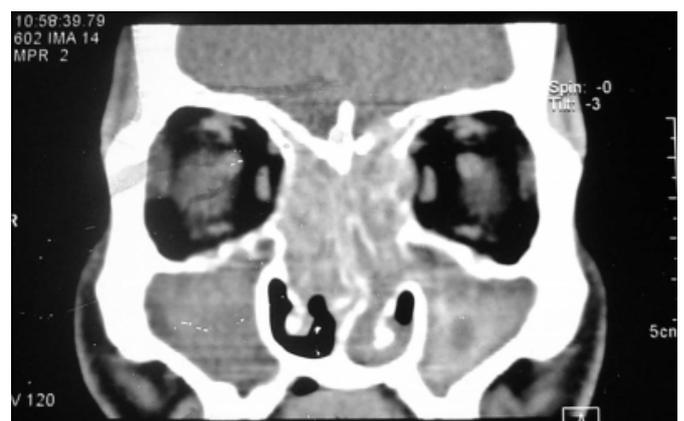
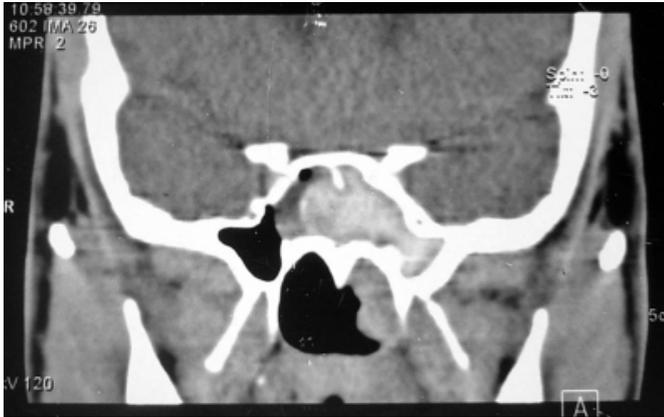


Figure 2

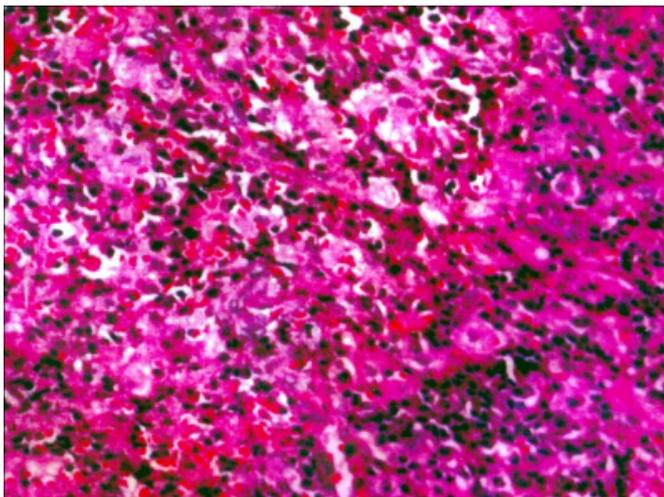
Figure 2: Computer Tomography (CT) scan PNS showing non-homogenous opacity in the sphenoid sinus and mass in posterior part of nasal cavity.



Endoscopic sinus surgery was performed under local anesthesia. Concha bullosa was present on both sides and were full of thick secretions. The area around the uncinate process and maxillary ostium was hyperemic with small polypoid masses, which were sent for histopathology examination. Uncinate was friable. The maxillary, ethmoid and sphenoid sinuses on both sides were full of thick tenacious yellow to brownish secretion, which was suctioned out. Bilateral nasal packs were put in for 24 hours.

Figure 3

Figure 3: Histological section showing numerous foamy macrophages (Mikulicz Cells) with hematoxylin and eosin stain.



Histopathology examination revealed numerous foamy macrophages (Mikulicz Cells) (Fig3). No fungal hyphae were seen. A diagnosis of Rhinoscleroma was made. Patient was put on tablet ciprofloxacin 500mg twice daily and capsule rifampicin 450mg once daily for six weeks. After six months

of follow up patient was relieved of nasal obstruction and headache.

DISCUSSION

Rhinoscleroma usually presents with nasal involvement but it can involve all parts of the respiratory tract in a progressive and often destructive fashion. The disease manifests through three overlapping stages: exudative, proliferative and cicatricial.^[12]

Exudative stage is characterized by watery nasal discharge, frontal-ethmoid headaches and difficulty in breathing with nasal obstruction and epistaxis. The nasal mucosa may be hyperemic, edematous or hypertrophic, yellowish crusts may be noted. Sore throat and hoarseness may be present if the naso-pharynx and larynx are involved.^[3]

The proliferative stage is characterized by the disappearance of the coryza, progressive infiltration, and coalescence of the nodules to form large granular masses. These waxy, hard nodules arise from the floor of the nose and nasal septum. The tissues become indurated and have a cartilaginous consistency. There is slow spread of the nodules to involve both the medial and lateral walls of the nose, leading to nasal obstruction, expansion of the anterior nares and deformity of the upper lip.^[23]

In the cicatricial stage adhesions of the soft structures and contraction of the involved tissue may lead to stenosis and virtually occlude the naso-pharynx, larynx and air passages, causing death from severe airway obstruction ^[13].

Rhinoscleroma is most frequently recognized in young adults who may have symptoms for over 10 years prior to diagnosis^[2]. Our patient had a very short history of two months.

Almost all patients of Rhinoscleroma have nasal involvement with nasal obstruction (94%), nasal deformity (32%), and epistaxis (11%) being the presenting feature. The disease involves oral cavity in 18%, pharynx in 18-43%, eustachian tube in 27%, larynx in 26%, trachea and bronchi in 10%, cases. The nose is involved in almost all cases, yet the reported rate of sinus involvement is low with maxillary sinus being involved in about 22% cases. Sinus involvement may also be due to secretion retention, secondary to ostium blockage in the nasal cavity.^[145] After reaching the maxillary sinus the disease can invade orbit after breaching the lamina papyracea.^[6] Rhinoscleroma can invade the anterior cranial fossa simulating a meningioma.^[7]

Radiologic findings of rhinoscleroma include a diffuse soft-tissue shadow in the nasal cavity usually obstructing the nasal cavity, maxillary and ethmoid sinus involvement is seen mainly as concentric mucosal thickening and cloudiness.[8]

On CT scan PNS, lesions may vary from localized mucosal thickening to soft tissue densities extending from the septum to the lateral nasal wall with bony nasal expansion. Lesions characteristically are homogenous, do not enhance, and have well-defined borders. There can be complete erosion of the turbinates and medial maxillary wall and resorption of the septal cartilage due to pressure necrosis.[5] In the present case the CT scan the lesions were non-homogenous suggestive of fungal sinusitis. Rhinoscleroma on both T-1 and T-2 weighted MRI images shows mild to marked high signal intensity. Further the retained sections can be clearly differentiated from lesions of rhinoscleroma.[9]

A positive culture of *K. rhinoscleromatis* is diagnostic of rhinoscleroma. Cultures from biopsy are positive in 98% of all clinically diagnosed cases, while pus swabs are positive in only 60% cases of biopsy proven scleroma cases. A biopsy is taken from areas of active disease, with the nasal septum and the anterior aspect of the inferior turbinate being the easily accessible site. The organisms are most adequate and the histopathology changes are most characteristic in proliferative stage.[23]

The typical histopathology of rhinoscleroma consists of plasma cells, lymphocytes, Russell bodies, and gram positive bacilli and Mikulicz cells. The later is foamy vacuolated histiocyte containing bacteria and undergoing hydropic degeneration at different stages. These cells are well stained with haematoxylin-eosin[210]. They are characteristic but not pathognomonic feature of rhinoscleroma[11].

Klebsiella rhinoscleromatis has a slime coating, which is a non-digestible muco-polysaccharide material and it is important in the pathogenesis of disease[10]. It increases the osmotic pressure and forms multiple hydropic vacuoles that

rupture the Mikulicz cells. Release of the muco-polysaccharide would initiate a cycle that would prolong the disease in the absence of bacteria[10]. Further, the pre-mature rupture of the Mikulicz cells allows liberation of some morphologically viable bacilli. This ineffective destruction allows the infiltration process to continue and is responsible for the chronicity of the disease.[12]

Ciprofloxacin and Rifampicin are proven drugs for the treatment of rhinoscleroma[13]. Further a combination of two drugs produces speedier eradication of the infection if they both are bactericidal.[3]

CORRESPONDENCE TO

Dr Joginder Singh Gulia House no.20/9J Medical Campus, Pt. B.D. Sharma PGIMS Rohtak. 124001(Haryana) India
Phone: 09416287404 E-mail: jsgulia@hotmail.com

References

1. Shum TK, Whitaker CW, Meyer PR. Clinical update on rhinoscleroma. *Laryngoscope* 1982; 92:1149-53.
2. Holinger PH, Gelman HK, Wolfe CK. Rhinoscleroma of the lower respiratory tract. *Laryngoscope* 1977; 87:1-9.
3. Ssali CLK. The management of Rhinoscleroma. *J Laryngol Otol* 1975; 99:89-91.
4. Gamea AM. Role of endoscopy in diagnosing scleroma in uncommon sites. *J Laryngol Otol* 1990; 104: 619-21.
5. Ammar ME, Rosen A. Rhinoscleroma mimicking nasal polyposis. *Ann Otol Rhinol Laryngol* 2001; 110: 290-92.
6. Lubin JR, Jallow SE, Wilson WR, Grove AS, Albert DM. Rhinoscleroma with exophthalmos: a case report. *Br J Ophthalmol* 1981; 65: 14-17.
7. Hara HJ. Scleroma. Observations on seven cases in one family for 10 years and another case which developed intracranial complications after five years. *Laryngoscope* 1957; 67: 695-708.
8. Shehata MA, El-Sallab, Mandour MA. Radiographic study of scleroma. *J Laryngol Otol* 1975; 89: 1243-61.
9. Razek AAKA, Elasmfour AA. MR appearance of rhinoscleroma. *Am J Neuroradiol* 1999; 20: 575- 8.
10. Hoffmann EO, Lopse LD, Harkin JC. The Mikulicz Cell in rhinoscleroma. *Am J Pathol* 1973; 73: 47-58.
11. Caldarelli DD, Friedberg SA, Harris AA. Medical and surgical aspects of the granulomatous diseases of the larynx. *Otolaryngol Clin North Am* 1979; 12: 767-81.
12. Canalis RF, Zamboni L. An interpretation of the structural changes responsible for the chronicity of rhinoscleroma. *Laryngoscope* 2001; 111: 1020-26.
13. Avery RK, Salman SD, Baker AS. Rhinoscleroma treated with Ciprofloxacin: A case report. *Laryngoscope* 1995;105: 854-5.

Author Information

S.P. Gulati

Senior Professor, Department of Otolaryngology, Pt. B.D Sharma University of Health Sciences

Joginder Singh Gulia

Associate Professor, Department of Otolaryngology, Pt. B.D Sharma University of Health Sciences