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Rhinoscleroma presenting as a nasal-palatal mass with airway obstruction

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CASE REPORT

Rhinoscleroma presenting as a nasal-palatal mass with airway obstruction [v1; ref status: indexed, http://f1000r.es/zi]

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Abstract

We report a case of a 45-year-old male with severe rhinoscleroma. The patient presented to the emergency room with dyspnea from a long-standing nasal-palatal mass. A tracheostomy was required for airway control. While dyspnea in the presence of an upper airway mass is typical of malignancy, consideration of non-oncological etiologies is important. We review the epidemiology, pathology, diagnosis, and treatment of rhinoscleroma.

Article Status Summary

Referee Responses

Referees	1	2	3
v1 published 09 May 2013	? report	✓ report	? report

1 **Rakesh K Chandra**, Northwestern University Feinberg School of Medicine USA

2 **Allen M Seiden**, University of Cincinnati USA

3 **Alkis Psaltis**, Stanford University Medical Center USA

Latest Comments

No Comments Yet

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Introduction

Rhinoscleroma is a chronic bacterial infection caused by *Klebsiella rhinoscleromatis*, a Gram-negative, non-motile, encapsulated bacillus. Due to the low infectivity of the bacteria, chronic exposure is required in order to establish infection. Rhinoscleroma is more frequent in the developing world, and is likely a secondary complication as a result of underdeveloped hygiene infrastructures, poor access to antibiotics, and overcrowded living conditions. Most cases are found in Central America, Africa and the Middle East[1]. The prevalence of sporadic cases outside of endemic areas is usually attributed to immigration[2]. Though rhinoscleroma can involve any structure of the upper respiratory tract, *Klebsiella rhinoscleromatis* has an affinity for nasal mucosa and thus is present in the nasal cavity in 95–100% of cases[3]. It can also be found in the nasopharynx (18–43%), larynx (15–40%), trachea (12%), and bronchi (2–7%)[4]. Here, we present a case with both nasal and palatal involvement resulting in airway obstruction.

Case report

A 45-year-old Central American male presented with a 13-year palatal mass, and new onset stridor in the background of chronic dyspnea. He denied weight loss and night sweats. He worked as a day laborer, drank socially, but never smoked. He had been unable to breathe out of his nose for at least thirty years.

Nasal endoscopy showed obstructed choana bilaterally. Inspection of the oral cavity showed a hard, plaque like growth involving the hard and soft palates, pharynx, and marked foreshortening of the palatoglossal folds (Figure 1). Dentition was poor. Endoscopic visualization of the larynx could only be performed transorally. The patient's airway was tight at the level of the palatoglossal folds and base of the tongue. The vocal cords and epiglottis were uninvolved.

A computed tomography (CT) scan confirmed a palatal mass, and obstructed choana. Thickening of the uvula, and hard and soft palate mucosa was noted. No palatal bony obstruction or lymphadenopathy was seen (Figure 2).

A local awake tracheostomy was performed to provide a secure airway. A palatal biopsy was sent for analysis and demonstrated squamous mucosa with a dense, mixed inflammatory infiltrate containing abundant plasma cells and scattered vacuolated macrophages (Mikulicz cells) (Figure 3). A Warthin-Starry stain revealed rod-shaped bacilli within the vacuolated macrophages. The bacilli were morphologically consistent with *Klebsiella* (Figure 4).

The patient was treated with ciprofloxacin 500 mg BID for 12 weeks. His airway symptoms improved and he was later decannulated without sequelae. He declined surgical nasal airway debridement.

Discussion

Rhinoscleroma generally progresses in three stages. The initial stage is the catarrhal or exudative phase. This is followed by the

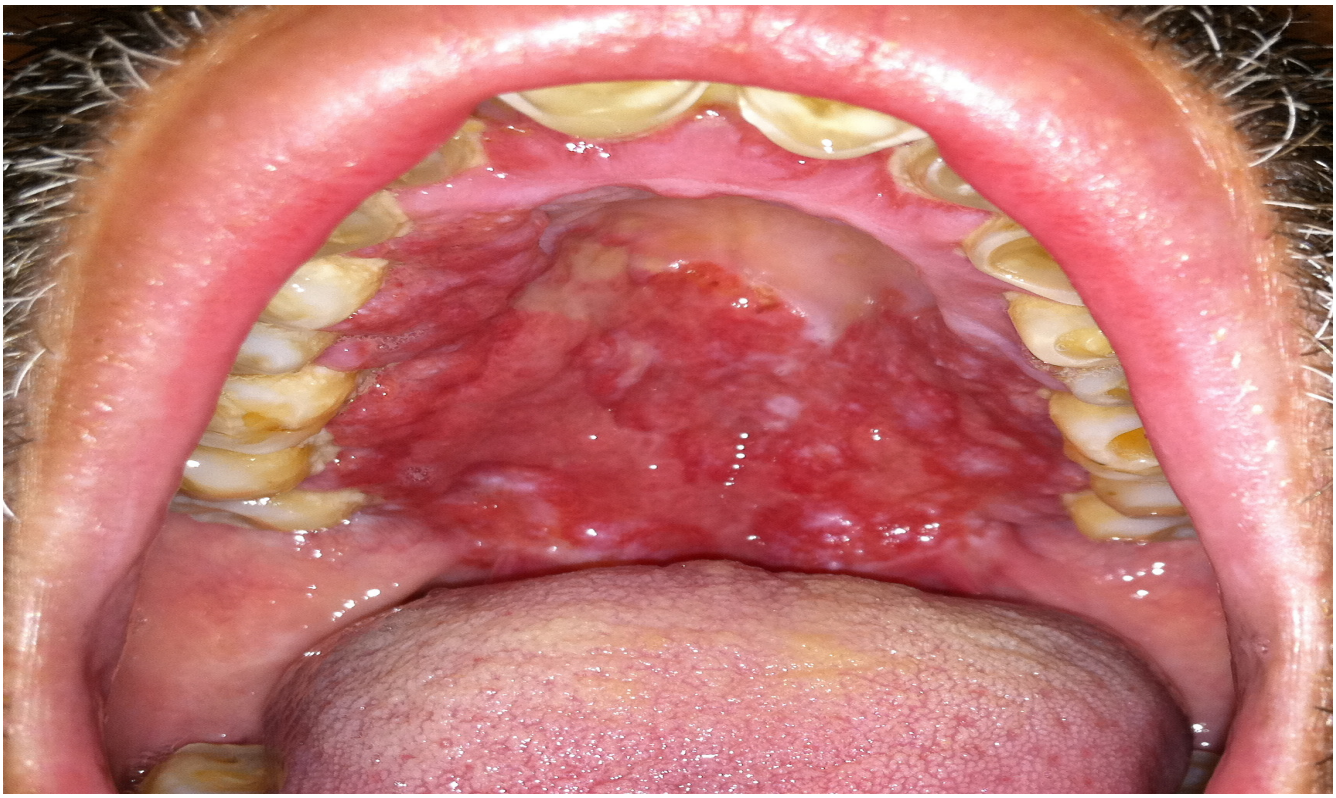

Figure 1. Oral cavity showing a plaque-like erythematous mass involving the gingiva, hard and soft palates.

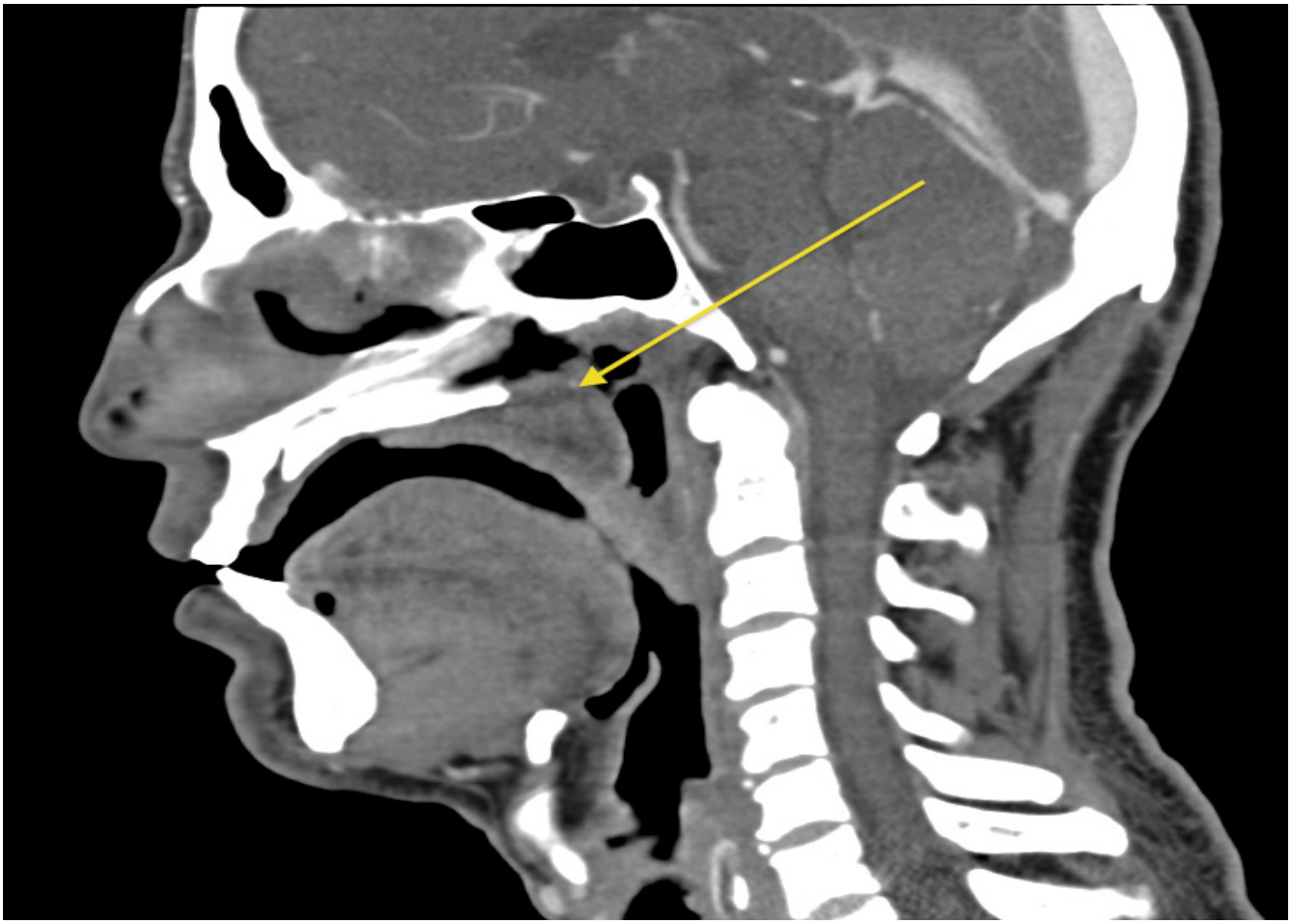

Figure 2. CT neck with contrast in sagittal plane. Heterogeneous soft tissue is present in the nasopharynx.

proliferative or granulomatous phase, which finally evolves into the cicatricial phase[2]. During the catarrhal stage, patients may have persistent rhinitis and mucopurulent discharge. In the second stage, inflamed mucosa coalesces to form granulomas. These granulomas may infiltrate other portions of the airway and then scar, giving rise to the third or cicatricial stage[2]. These stages usually do not exist independently. In many cases of rhinoscleroma, the presence of all three stages can be found at the time of diagnosis.

Rhinoscleroma is spread by person-to-person transmission. However due to the low infectivity of the pathogen, transmission requires a chronic exposure. It has also been proposed that an altered immune response along with an alteration in the CD4+ and CD8+ proportion leads to ineffective macrophage production that are susceptible to bacterial replication[5].

A high degree of suspicion is warranted when patients present with persistent, unremitting rhinitis or nasal obstruction unexplained by other causes. The differential diagnosis of such symptoms should include rhinoscleroma, as well as tuberculosis, syphilis, Wegener's granulomatosis, lymphomas as well as more common carcinomas. Histopathologic evidence of rhinoscleroma includes granulomatous inflammation with large vacuolated histiocytes known as Mikulicz cells[6]. Canalis *et al*. proposed that these Mikulicz cells arise from histiocytes that migrate to areas where neutrophils have failed to contain the *Klebsiella* infection[7]. The histiocytes, however, are unable to lyse their phagocytosed *Klebsiella* cells, leading to the dilation of their vacuoles[7]. Positive culture of rhinoscleroma on MacConkey agar is diagnostic, though culture is only positive in 50–60% of patients. Thus, it is key to have high clinical suspicion in conjunction with positive histopathologic evidence to confirm the diagnosis.

Historically, treatment of rhinoscleroma was with tetracyclines and aminoglycosides such as streptomycin. However, a prospective study done in the Mayo Clinic, USA, by Andraca *et al*. in 1993 demonstrated the efficacy of fluoroquinolones[8]. Treatment with fluoroquinolones also confers the benefit of a lower side-effect profile. Dosing of the antibiotic is variable between different studies, but most agree that long-term therapy for months and sometimes years is necessary to adequately treat the infection[3,7,8]. Despite treatment, recurrence has been reported in up to 25% of cases at 10 years[2,4]. Consideration should be made when addressing whether a patient requires surgical de-bulking of the

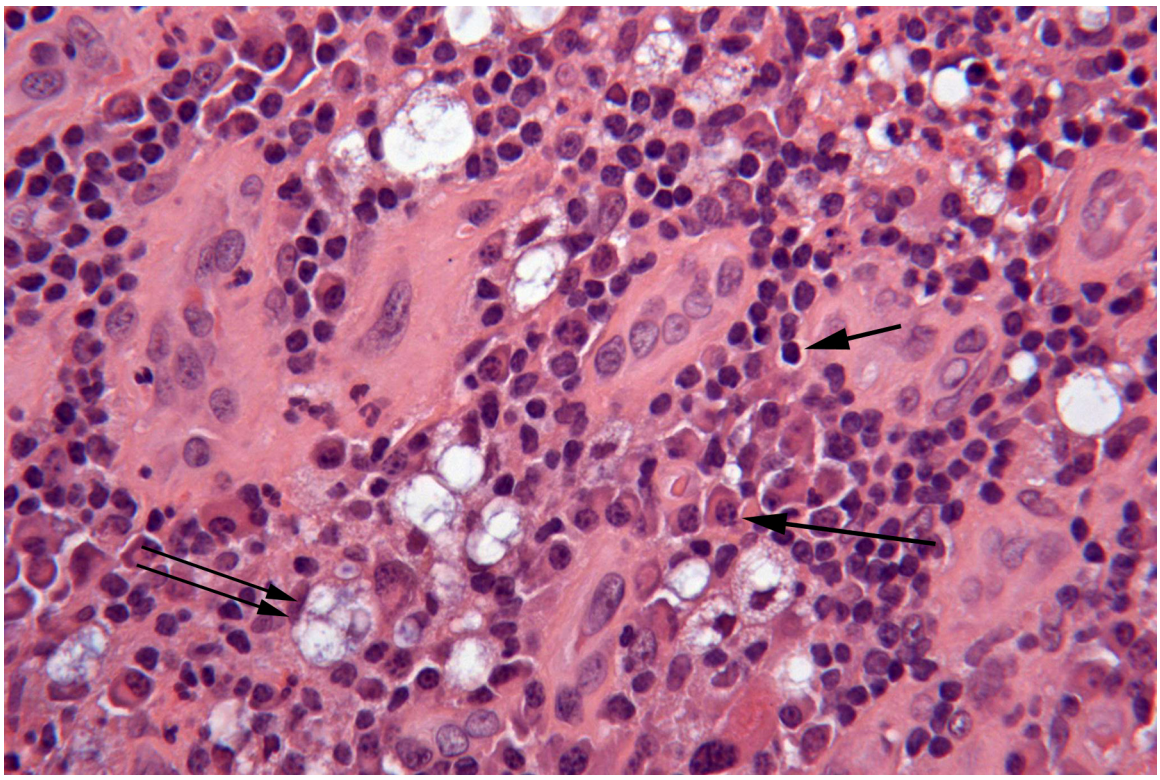

Figure 3. **H&E stain (400×) demonstrated a mixture of plasma cells (arrow), lymphocytes (short arrow) and vacuolated macrophages (Mikulicz cells) (double arrow).**

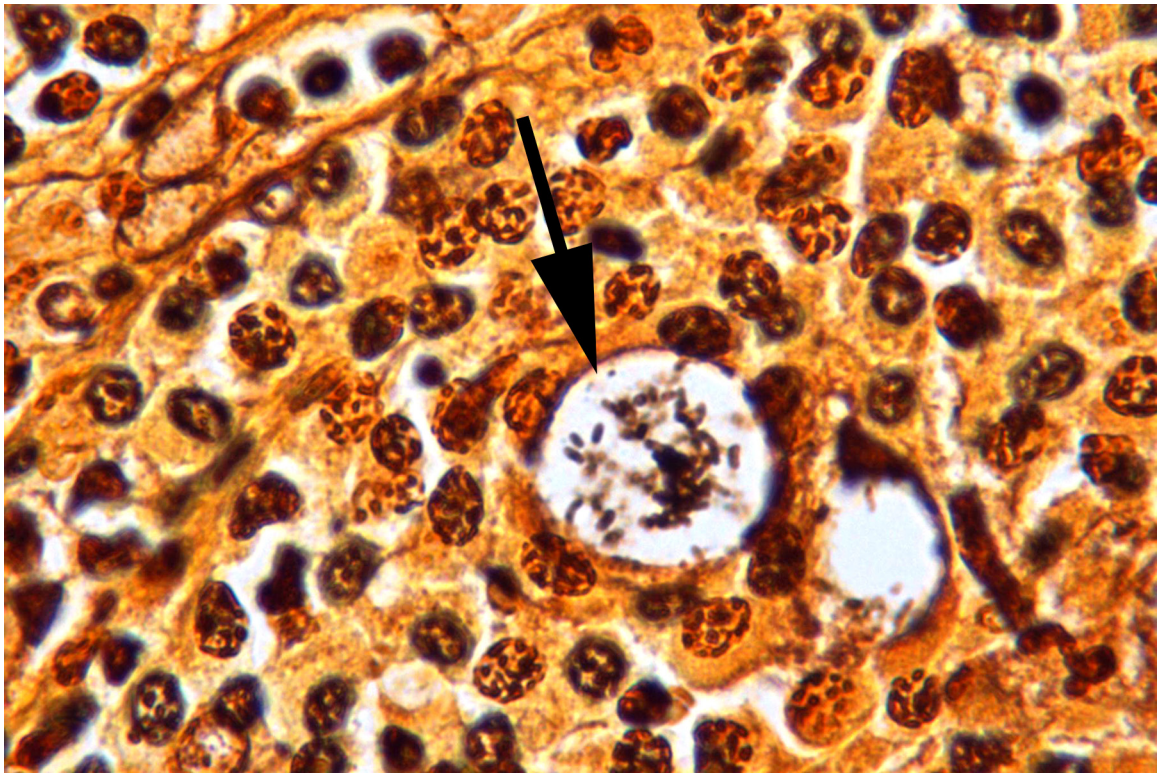

Figure 4. **Steiner stain, (1000×) with rod-shaped bacilli within a vacuolated macrophage (Mikulicz cell) (arrow).**

scar in rhinoscleroma formed during the cicatricial stage. Indications for surgical de-bulking include airway patency, treatment of bulky disease, and cosmesis.

Conclusion

Rhinoscleroma is due to chronic and indolent *Klebsiella* infection. Symptoms may include chronic, unremitting rhinitis or nasal obstruction that is present for years. The presenting symptom can also be more dramatic, such as airway compromise, as seen in this case. A diagnosis of rhinoscleroma is made via pathological specimens. Communication between the clinician and the pathologist as to the possibility of non-oncological processes can aid in determining the diagnosis. A Warthin-Starry stain demonstrating rod-shaped bacilli within vacuolated macrophages (Mikulicz cells) is classic for rhinoscleroma. Mainstay of treatment is long-term fluoroquinolones. Evaluation of airway patency is critical and surgical intervention may be required.

Consent

Written informed consent for publication of clinical details and clinical images was obtained from the patient.

Author contributions

MD treated the patient. DK prepared the pathological slides, which made the diagnosis. AR prepared the first draft of the manuscript. All authors were involved in the revision of the draft manuscript and have agreed to the final content.

Competing interests

No relevant competing interests were disclosed.

Grant information

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PubMed Abstract | Publisher Full Text

Current Referee Status:

Referee Responses for Version 1

Alkis Psaltis
Stanford University Medical Center, Stanford, CA, USA

Approved with reservations: 31 May 2013

Referee Report: 31 May 2013
doi:10.5256/f1000research.1278.r979

The authors present an interesting case report of a condition not commonly seen in developed countries. Its unusual presentation reaffirms the need for otolaryngologists to consider infectious processes in the work up of sinonasal and nasopharyngeal masses. The paper is concise, well written and easy to follow. It provides a nice summary of rhinoscleroma particularly of the histopathophysiological features essential for diagnosis.

Additional photographs demonstrating the encroachment of the mass on the pharyngeal airway (i.e. with the use of a tongue depressor), a nasal endoscopic view and a post treatment view may have been helpful in providing more visual information for the reader.

I agree with the previous reviewer's concern regarding the need for an awake tracheotomy for the management of the airway. Given the superior appearance of the mass on the sagittal CT scan and the fact that the authors state that an adequate visualization of the uninvolved vocal cords and epiglottis was obtained with transoral endoscopy, why did the authors not consider awake fiber-optic endoscopic intubation or at least mention this as an alternative for managing the airway.

The discussion of the response of the mass to treatment is a little brief. I think additional information would be useful to the reader such as: Percentage decrease in the size of the mass with treatment, how the authors deemed that 12 weeks of treatment was appropriate, progress post treatment and also any scheduled future follow up.

I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Competing Interests: No competing interests were disclosed.

Allen M Seiden
Department of Otolaryngology, College of Medicine, University of Cincinnati, Cincinnati, OH, USA

Approved: 16 May 2013

Referee Report: 16 May 2013
doi:10.5256/f1000research.1278.r954

This is an interesting case report describing an unusual presentation for rhinoscleroma, an infectious problem that we see rarely in the USA. It is well written, and well-organized. It would have been interesting to include patient photos post treatment. It can be a difficult diagnosis to make, so a more thorough literature review discussing potential physical findings on presentation would also have been helpful.

I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard.

Competing Interests: No competing interests were disclosed.

Rakesh K Chandra
Department of Otolaryngology - Head and Neck Surgery, Northwestern University Feinberg School of Medicine, Chicago, IL, USA

Approved with reservations: 15 May 2013

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The manuscript is an excellent review of the topic and is well illustrated with a clinical photo, radiology, and histopathology. My only area of concern is that I wonder whether it was indeed necessary to perform a tracheotomy on this patient. If the glottis was visible transorally, and given the findings on the sagittal radiologic image shown, it looks like the patient could have been intubated. Also the lesion is too superior to cause true stridor. It could be argued that tracheotomy was advised because the patient would have had difficulty managing secretions and there was concern the lesion would swell or bleed upon biopsy, but it is difficult to imagine true stridor and that this patient couldn't have been intubated. The report would be improved if the authors justify their decision to perform the tracheotomy in the light of these comments Title and Abstract are appropriate and the conclusions are otherwise balanced and justified.

I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Competing Interests: No competing interests were disclosed.