Case Report

Rhinoscleroma: A Report of Three Cases with Unusual Headache Presentation and Literature Review

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Abstract

Rhinoscleroma is a rare endemic disease in low socioeconomic settings characterized by nasal growth, nasal blockage, and epistaxis. The causative organism is not an usual nasal commensal, and diagnosis is often delayed. Characteristic Mikulicz cells should be visualized microscopically for definitive diagnosis. We present our experience of rhinoscleroma with unusual presentation of severe headache and orbital mass in three patients over an 11-year period in the pathology laboratory of a tertiary hospital.

Keywords: Epistaxis, orbital mass, rhinoscleroma, severe headache

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Introduction

Rhinoscleroma is a rare endemic disease of the nose and upper airways caused by *Klebsiella rhinoscleromatis*, an immotile short encapsulated Gram-negative bacillus.[1,2] The disease is characterized by chronic granulomatous inflammatory response of numerous foamy histiocytes known as Mikulicz cells, lymphocytes, plasma cells, and Russell bodies.[3] Endemicity of the disease is associated with poor hygiene and low socioeconomic indices, and cases have been previously reported in Central and South America, Africa, the Middle East, the Philippines, and India.[3-5] The disease affects all age groups though more frequently recognized in young adults with no preferential sex predilection, while symptom duration is variable and may exceed 10 years.[2,6-8] We report three patients with an incidental diagnosis of rhinoscleroma with an unusual clinical presentation of severe headache over 11 years (2010–2021) in a tropical pathology laboratory.

Case Reports

Case 1

A 19-year-old female undergraduate of a tertiary institution presented to the general outpatient clinic of our hospital with a 7-month history of throbbing right sided, sometimes global headache with right nasal growth. Nasal growth was not associated with bleeding, difficulty with breathing, nasal discharge, pain, or trauma. The growth occasionally reduced in size but never disappeared completely. She experienced no loss of smell or nasal sensation. She is not asthmatic and had no contributory significant medical history. Clinical examination revealed a pea-sized 5 mm × 5 mm firm-to-hard mass at the floor of the right nasal cavity, which was clinically diagnosed as a nasal cyst. Paranasal sinus computed tomography (CT) scan revealed a rounded nonenhancing hypodense lesion which was located on the right anterolateral nasal floor. The lesion measured 10 mm × 11 mm in coronal section. There was also bilateral mild engorgement of the nasal turbinates. The paranasal sinuses, orbital cavities, and their contents were all within the normal limits. Diagnosis from CT scan was chronic rhinitis with vestibulitis. Based on the clinical and CT scan findings, she had surgical excision of a 5 mm × 5 mm mass at the floor of the right nasal cavity (vestibular) under general anesthesia, which was sent for histopathological analysis. A 10 mm × 5 mm × 5 mm firm regular gray tissue fixed in

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10% formalin was received in the pathology laboratory. The formalin-fixed tissue was processed into paraffin-embedded blocks. Sections of 4 μm thickness were mounted on glass slides and deparaffinized with xylene. The slides were then dried with graded alcohol and stained with hematoxylin and eosin (H and E) and Giemsa stains. Microscopy showed a circumscribed lesion composed predominantly of foamy macrophages (Mikulicz cells) surrounded by a thick fibrous tissue [Figures 1 and 2] and diagnosed as rhinoscleroma. Part of an ulcerated squamous epithelium overlying inflamed granulation tissue was also seen. She was subsequently placed on Augmentin for 4 weeks based on the diagnosis of rhinoscleroma. She did well postoperative and was discharged for clinic follow-up. Her 1-month postoperative follow-up at the clinic was uneventful.

Case 2

A 38-year-old female presented to the ophthalmology clinic of our hospital with a 2-year history of throbbing headache associated with left orbital mass, visual impairment, and proptosis. There was no complaint of nasal discharge, epistaxis, swelling, pain, or history of trauma. There was also the absence of other constitutional symptoms or masses anywhere else in the body. A clinical assessment of a left orbital tumor of unknown type was made by the attending physician. Based on this clinical diagnosis, she had an excision biopsy of a superior medial left orbital cystic mass which was sent for histopathological analysis. A surgically opened 4-g cystic mass measuring 25 mm × 18 mm × 8 mm was received in the pathology laboratory. The cystic mass was fixed in 10% formalin and processed into paraffin-embedded blocks which were sectioned at 4 μm thickness. The sections were mounted on glass slides, deparaffinized with xylene, dried with graded alcohol, and stained with H and E and Giemsa stains. Microscopy showed abundant plasma cells and lymphocytes admixed with marked histiocytic infiltration and numerous Mikulicz cells (large vacuolated nonlipid-containing histiocytes having hyperchromatic small, uniform, round nucleus located peripherally) and occasional multinucleated giant cells containing cellular debris [Figure 3]. Other areas showed fibrocollagenized cystic wall and denuded squamous epithelial lining. However, this patient did not turn up for follow-up postsurgical excision.

Case 3

A 40-year-old female patient was referred to the ear, nose, and throat clinic of the Ahmadu Bello University Teaching Hospital, Zaria, from a general hospital with complaints of severe headache and epistaxis of 10 years duration. She also complained of 2-year history of gradual hearing loss and 5-year history of difficulty with breathing and nasal obstruction. There were no associated complaints of nasal discharge, dysphonia, dysphasia, or palpable masses. Two years before her presentation at our facility, she had surgery at a private hospital for nasal blockage; however, the surgical biopsy was not sent for histopathological analysis. Two weeks post surgery, the nasal blockage recurred until this current presentation at

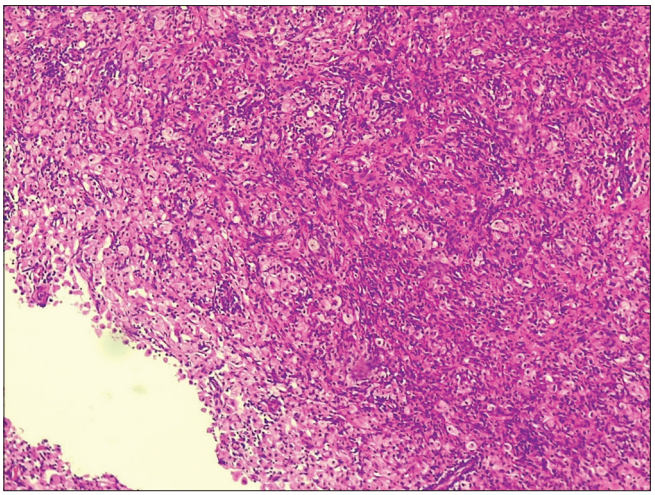

Figure 1: Diffuse infiltration by numerous histiocytes (Mikulicz cells) admixed with lymphocytes and plasma cells (H and E, ×40)

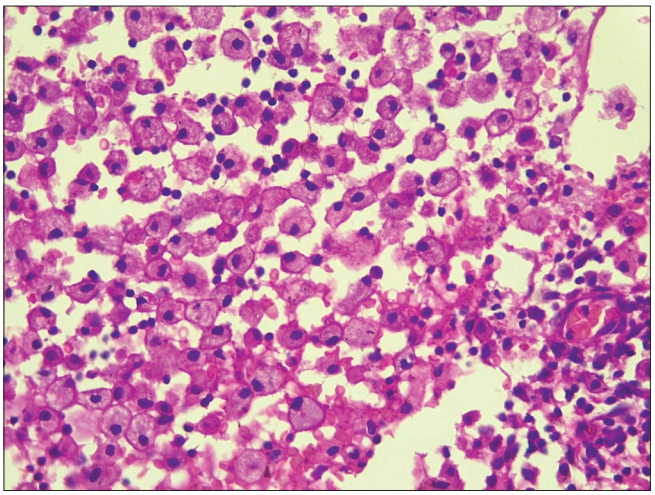

Figure 2: Numerous Mikulicz cells (arrowed) having abundant cytoplasm with hyperchromatic nucleus admixed with lymphocytes and plasma cells (H and E, ×100)

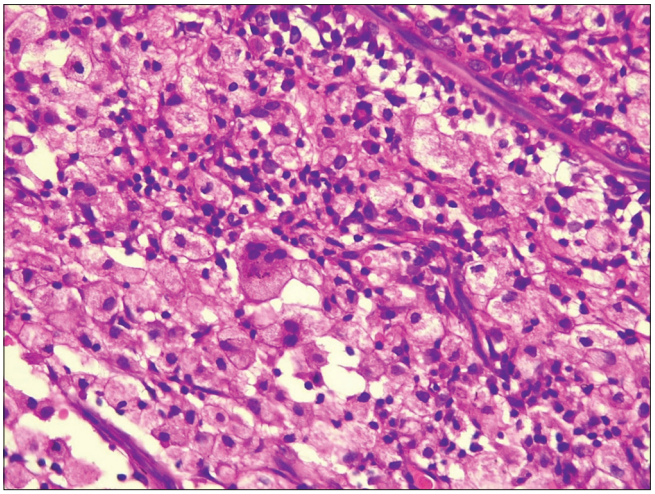

Figure 3: Multinucleated giant cell (arrowed) containing cellular debris admixed with lymphocytes and plasma cells (H and E, ×200)

our facility. She was not a known hypertensive or diabetic and had no other complaints or significant medical history of any other ailment. Clinical examination revealed a healed surgical scar over the right alae of the nose, bilateral nasal obstruction by a firm nonbleeding mass that was attached to the lateral walls of the nose with bilateral nasal bone expansion. The mass was separate from the floor, root, and medial wall of the nose. The palates and floor of the mouth were uninvolved. She had a radiographic (X-ray) examination which showed erosion of the nasal septum with cloudy maxillary antrum and sphenoid sinus. Clinical diagnosis was bilateral nasal polyps with a differential diagnosis of nasopharyngeal tumor. The patient had a bilateral rhinotomy with nasal clearance under general anesthesia. Intraoperative finding was a friable mass that extended from the nasal cavity to the nasopharynx posteriorly. It bled with contact and had a gritty feel. There was also bilateral maxillary antral pus collection.

The tissue biopsy received in our laboratory was gray white and fragmented; it weighed 68 g and measured 9 cm × 7 cm × 3 cm. Histology showed a polypoid lesion lined by focally ulcerated respiratory epithelium overlying sheets of foamy histiocytes having abundant clear cytoplasm and uniform small hyperchromatic nuclei (Mikulicz cells) admixed with lymphocytes, plasma cells, and Russell bodies. Other areas showed fibrosis, mucous-secreting glands, and bony trabeculae. She was placed on 5-month antibiotics therapy which comprised ciprofloxacin, tetracycline, and metronidazole and only complained of occasional rhinorrhea at 10-month follow-up visit in the clinic.

Discussion

Rhinoscleroma, is also known as lupus scrofulous of the nostrils, and more recently as scleroma though it was first described by von Hebra in 1870.[8] The causative organism, *K. rhinoscleromatis,* has transmitted through nasal exudates of infected persons and is not a normal nasal commensal. The disease symptoms vary from those of common cold to nasal bleeding, nasal obstruction, aphonia, loss of smell, and palatal thickening. However, the clinical manifestation is dependent on the stage of the disease. The common symptom of all three patients in this report was severe headache, an unusual presentation. A similar unusual rebellious headache was reported by Bazzout *et al*. in a 70-year-old known diabetic and hypertensive female with extensive rhinoscleroma.[9] However, epistaxis and respiratory distress, as seen in one of the patients, were also reported as the presenting signs of rhinoscleroma by Simao *et al*. in Portugal and Bonacina *et al*. equally reported epistaxis in an Egyptian.[10,11] Orbital mass with proptosis and visual impairment as presenting signs of rhinoscleroma is uncommon, and only four cases have been reported in the literature.[12] Lubin *et al*., in their report of rhinoscleroma with exophthalmos, postulated that the causative organism causes pressure atrophy on the optic nerve by infiltrating through the paranasal sinuses into the lacrimal apparatus, base of the skull, and into the orbit for this clinical manifestation to occur.[12] Adekanye *et al*. reported a case of rhinoscleroma coexisting with an undifferentiated malignancy and documented bony destruction.[4] It is noteworthy that the causative organism of rhinoscleroma is not a known carcinogen and has not been implicated in the malignant transformation of any tumor.

Respiratory difficulty and nasal obstruction associated with extensive disease may also involve the cervical lymph nodes and extend to the orbit, as seen in the patient with orbital mass though there were no palpable cervical lymph nodes. The first reported rhinoscleroma patient in Finland had respiratory symptoms with cervical lymph node involvement.[13] Lassikri *et al*. also documented cervical lymph node involvement in their case report.[14] The disease severity may also cause soft-tissue distortion and bony destruction, which may lead to an erroneous clinical diagnosis of a neoplastic lesion rather than an infection, as seen in our patients. The disease extent and concomitant soft tissue and bony destruction can be identified with radiological examination such as X-ray and CT scan, though definitive diagnosis is based on histopathology of the tissue biopsy.

Several reports have documented preferential female disposition in the second to third decades of life, while some reports established no sex predisposition and affectation of all age groups.[2,5,6,15,16] The youngest documented patient is a 5-year-old boy in Portugal.[10] The patients in this report are females in the second and fourth decades of life.

The clinical manifestations of rhinoscleroma as identified by endoscopic and microscopic parameters comprise the catarrhal, nodular/granulomatous, and sclerotic stages.[2,16,17] The catarrhal stage is associated with recurrent sinusitis and rhinorrhea, and the nodular stage is characterized by mass formation with attendant respiratory obstructive symptoms, while the third stage of sclerosis is marked by fibrosis and extensive scarring. All three patients in this report were in the late phase of the nodular stage of the disease at the time of presentation. This nodular stage is associated with a delayed or missed diagnosis with neoplastic lesions because of the characteristic mass formation.

The differential diagnoses of rhinoscleroma include other causes of chronic granulomatous infections such as leishmaniasis, leprosy, rhinosporidiosis, syphilis, paracoccidioidomycosis, and Wegener's granulomatosis, an ulcerative destructive disease characterized by necrotizing inflammation, arteritis, and reactive giant cells. The common microscopic feature in all the aforementioned differential diagnoses is the presence of granuloma formation and mononuclear cellular infiltrates, whereas the definitive diagnosis of rhinoscleroma is the presence of Mikulicz cells which are most numerous during the nodular/granuloma stage.[3,17] Mikulicz cells are also identifiable with stains such as H and E, Giemsa, Warthin–Starry, and Gomori methenamine silver (GMS). The Lepra (Virchow) cell seen in leprosy may mimic Mikulicz cell; however, the Fite-Faraco stain is useful in identifying the acid-fast bacilli of leprosy. Giemsa stain is also useful

in identifying the leishmania bodies in leishmaniasis, while the GMS stain will identify the spores of rhinosporidiosis and paracoccidioidomycosis. Microbiological culture of the representative tissue biopsy using MacConkey agar will also identify the Gram-negative bacillus, *K. rhinoscleromatis*.

The treatment of choice is a combination of antibiotics, and the trio of augmentin, ciprofloxacin, and tetracycline has been used successfully in the treatment of rhinoscleroma.[2,6,7,18,19] However, antibiotic therapy is recommended for durations not <6 months for effective disease clearance because rhinoscleroma is associated with high recurrence rates.[3,6,15,18] Many studies have used different combinations of antibiotics such as streptomycin, doxycycline, tetracycline, rifampicin, sulfonamides, and cephalosporins to achieve complete resolution of the infection.[3,7,15] Surgical intervention such as debridement is indicated in relieving pressure symptoms of nasal blockage, as well as cosmetic repair of destroyed bony and soft tissues in severe and advanced disease stage. The use of corticosteroids in antibiotic-resistant rhinoscleroma cases has not been widely documented, while untreated cases have been associated with soft tissue and bony destruction with resultant facial deformities and fatalities.[7,12]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conclusion

A high index of suspicion should be entertained for rhinoscleroma in patients presenting with headache, orbital and nasal masses in our setting. Treatment and disease resolution is achievable with at least 6-month antibiotic therapy, while surgical intervention is indicated for relief of obstructive symptoms and cosmetics.

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Conflicts of interest

There are no conflicts of interest.

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