

Dacryoscleroma - A Pandora's Box

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Abstract

Rhinoscleroma is a chronic granulomatous disease with an ability to cause extensive disfiguration. A patient presented with extensive stenosis of the nares and sudden onset lacrimal abscess. In this case, we see Rhinoscleroma has spread involving the lacrimal sac system as well, thus the term Dacryoscleroma. The patient underwent dacryocystorhinostomy, multiple staged endonasal surgical release of adhesions with external drainage of the lacrimal abscess and Silastic sheet stenting of bilateral nasal cavity. This case demonstrates that a high index of suspicion is needed in suspecting this rare cause for lacrimal abscess and also emphasizes on the need for staged management in relieving the various sequelae caused by the disease process to attain optimized disease clearance and adequate relief of symptoms with a chronically disfiguring disease such as rhinoscleroma.

Keywords: *Dacryoscleroma; Rhinoscleroma; Lacrimal abscess; Nasal endoscopy*

1. Introduction

Scleroma is a chronic granulomatous disease of the upper respiratory tract caused by *Klebsiella rhinoscleromatis* [1]. It presents with mass lesions in the respiratory tract which can be found anywhere from the nose up to the trachea. It affects primarily the nose and less commonly the pharynx, larynx, trachea, bronchi and atypically lacrimal apparatus, cervical lymph nodes and cranial cavity [2-4]. Classically it affects the transitional zones at the entry of the nose and hence the name Rhinoscleroma. The nose is involved in 95%-100% of cases [5]. It is endemic to Egypt, Central, and South America, Africa, South East Asia and areas of Europe in descending order of prevalence [2-6]. It commonly affects in 2nd or 3rd decade of life.

However, there is paucity of data regarding the incidence and prevalence around the world [2-6]. Except for the series of cases documented by Badrawy in 1962, there have been no such case reports documented in the recent decade involving the lacrimal sac [2].

2. Case Report

A 34-year-old male presented to the ENT clinic with bilateral progressive nasal obstruction for 8 years. The patient also noticed a sudden onset swelling since 4 days over the inner aspect of the left eye, which was gradually progressive to the size of an almond and was associated with a throbbing type of pain. He had undergone septal surgery elsewhere, eight years ago, for nasal obstruction. He is a known diabetic on treatment since the past ten years with well-controlled sugar levels. On examination a solitary swelling of about 20 mm -30 mm was present near the medial canthus of the left eye occupying the lacrimal area, the borders were well defined and skin over the swelling was shiny with areas of erythema. On palpation, tenderness was present with a local rise of temperature and periorbital edema around the left eye. The right eye examination revealed mucoid discharge when pressure was applied over the medial canthus (FIG. 1A). The rest of the eye examination was unremarkable. Examination of the external nose revealed depressed and broadened dorsum with flared out ala and weak tip recoil. The quality of the skin was thick. Bilateral nasal valve area stenosis was seen on tip elevation which obviated the need for anterior rhinoscopy. However, posterior rhinoscopy revealed marked concentric narrowing of the choana (FIG. 1). Ear, throat, and neck examinations were clinically normal. Routine blood investigations were normal except for elevated Erythrocyte Sedimentation Rate. Computed Tomography (CT) of the paranasal sinuses revealed, thick well defined peripherally enhancing collection in the medial aspect of the left orbit, communicating with the nasolacrimal duct approximately measuring 16 mm to 11 mm. There was a similar thick well defined peripherally enhancing collection in the medial aspect of the right orbit communicating with the nasolacrimal duct measuring 7.5 mm to 4.5 mm with no underlying bony erosion. Minimal preseptal and premaxillary soft tissue swelling with adjacent fat stranding were noted on the left side, suggestive of edema. Additionally, mucosal thickening of bilateral maxillary sinus and ethmoidal air cells was present suggestive of sinusitis (FIG. 2). Hence a diagnosis of bilateral infective dacrocystocele and sinusitis was made.

The patient was taken up for surgery under anesthesia. Owing to the marked stenosis in the valvular region, a 2.7 mm zero-degree endoscope was used to visualize the nasal cavity. Marked synechiae were noted between the turbinate's and the septum in both the nasal cavities and were released (FIG. 1). On the left side, Endonasal Dacryocystorhinostomy was done and a marked fibrotic lacrimal sac was noted. However, on incising the sac wall, no pus was expressed. Hence a 0.7 mm linear incision was taken over the prominent part of the abscess externally and drainage was done. Incidentally, it was an extra-saccular collection of pus. Before the insertion of silastic sheets in the nasal cavity, multiple biopsies were taken from bilateral nasal cavities and the left lacrimal sac and sent for histopathology reporting, both of which were reported as chronic inflammatory infiltrate comprising of lymphocytes and plasma cells admixed with Mikulicz cells and Russell Bodies suggestive of rhinoscleroma of both the nose and the lacrimal sac (FIG. 3). The patient was treated with antibiotics comprising of Ciprofloxacin, Rifampicin and Metronidazole along with topical saline nose drops, antibiotic eye drops and multivitamins. Local application of a 2% Acriflavine solution mixture was done over the nasal mucosa. The patient underwent serial diagnostic nasal endoscopies 1 week apart and all the synechiae were released from non-corresponding raw areas. The silastic sheet stents were removed after 6 weeks and was followed up to 10 months, during which the patient showed significant improvement symptomatically and clinically (FIG. 1).

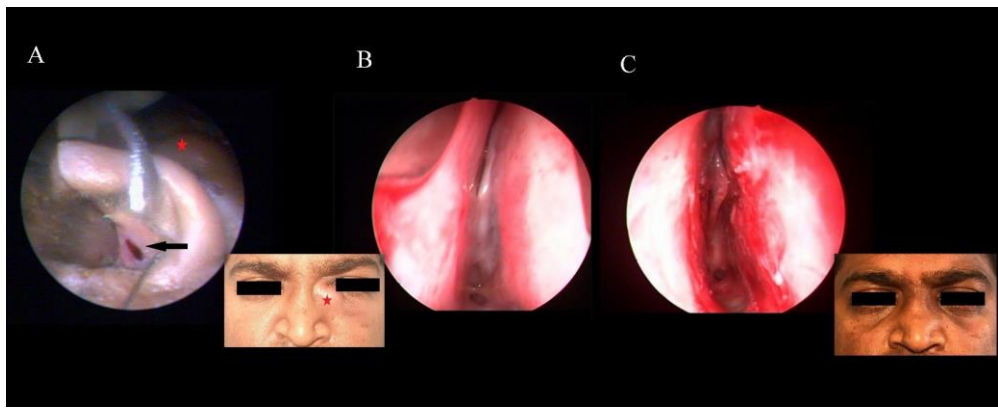


FIG. 1. A: Preoperative presentation, (black arrow) showing marked valvular stenosis and (red star) showing the left lacrimal abscess. B: Nasal Endoscopic image showing stenosis of the left nasal cavity with adhesions and stenosis of the choana. C: Follow up endoscopic image showing widening of the nasal cavity and improvement in the external deformity with no swelling in the left medial canthus.



FIG. 2. Showing left lacrimal sac abscess in contrast CT PNS (red arrows).

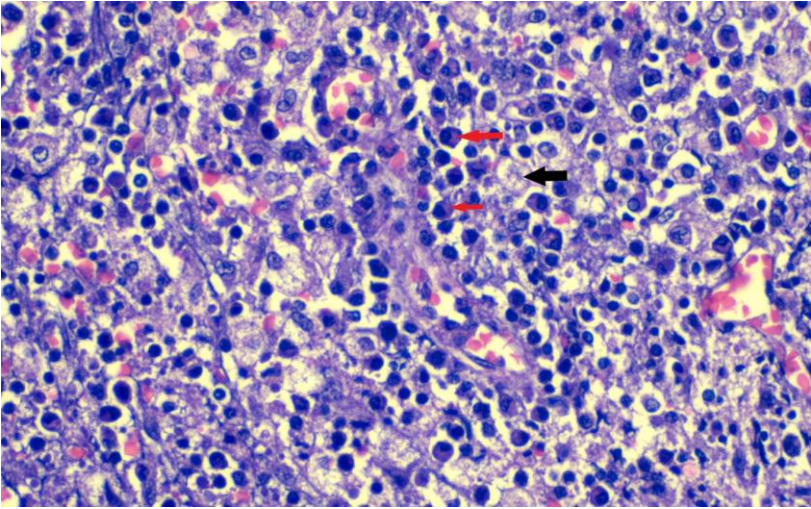


FIG. 3. H&E stained histopathology slide showing Mikulicz cells (black arrows) and Plasma cells (red arrows).

3. Discussion

In the year 1870 Von Hebra was the first to describe the term rhinoscleroma that he assumed to be a form of sarcoma while describing a nasal disorder. Later in 1877 histological criteria for this disease was given by Mikulicz-Radecki, emphasizing its non-neoplastic inflammatory nature [5]. In 2011, Von Frisch identified the causative organism as a Gram-negative coccobacillus, more recently known as *Klebsiella rhinoscleromatis* [4,5]. The disease progresses in three different overlapping stages as described by Canalis et al. [1,4]. In this case report, the patient presented with overlapping stages of granulomatous with cicatricial stage involving the nose and lacrimal sac causing marked stenosis of the nasal cavity and fibrosis of the lacrimal sac and thus interestingly mimicking a simile spectrum of a Pandora's box. Badrawy (1962) was the first to describe the lacrimal extension of the disease and termed it dacryoscleroma. The junction of the stratified squamous epithelium of the nasal vestibule extending into the inferior meatus and ciliated columnar epithelium of the nasolacrimal duct as classically occurred in our case reiterates that the disease affects the transitional zones. He also described the possible pathological pathway to the formation of lacrimal abscess owing to intraluminal plug action of the granuloma or the narrowing of the lumen by granuloma by extramural pressure and extension [3]. Thus, possibly explaining the basis behind the extra-saccular collection of pus that occurred in our case. A study conducted by Elwany et al, on 52 patients showed that the nasal involvement was predominant followed by nasopharynx; and one case presented with the additional involvement of both lacrimal sac along with the nose. Serial biopsies taken from different sites confirmed the diagnosis [2]. The patient was unwilling to undergo biopsy from the right lacrimal sac as it was not significantly symptomatic.

According to a retrospective study done by Molumi et al, on 134 patients with nasal and nasopharyngeal stenosis, fibrotic tissue was debrided followed by recanalization of nasal cavity and nasopharynx using Celestine tube placed such that one end was in the nasal cavity and the other end was in the nasopharynx [6]. Emphasis on CT scan plain and contrast is mainly to assess the extent of the abscess and sinus mucosal thickening. Additionally, the presence and extent of synechiae can also be gauged thus aiding in effective preoperative evaluation and planning. A study done by AbdelRazek et al, concluded that rhinoscleroma of the nose in the hypertrophic stage appeared as a well-defined mass on CT, and involvement of the inferior and middle turbinate were more common. On Magnetic Resonance Imaging granulomas can be identified in the T1 weighted images owing to the high protein content within Mikulicz cells and Russell bodies [10]. However, histopathological confirmation remains the gold standard wherein the presence of Mikulicz cells, Russell bodies, plasma cells, and leukocytic infiltration conclusively prove the presence of the disease [1,4].

Similarly the patient's diagnosis was confirmed only after histopathological reporting of the biopsy specimens. In an attempt to eradicate *K. rhinoscleromatis*, which has a high relapse rate, the treatment of rhinoscleroma requires prolonged antibiotic therapy [6]. Antibiotics with demonstrated efficacy are streptomycin, doxycycline, tetracycline, rifampicin, second- and third-generation cephalosporins, sulfonamides, ciprofloxacin and ofloxacin [7-9]. 2% Acriflavine for local application is known for its strong antibacterial activity against both gram-positive and gram-negative organisms [11]. Indications for surgery include those for relief of airway obstruction and reconstruction of cicatricial defects [8]. Even though the drug of choice being ciprofloxacin, other drugs have aided in the treatment of this patient. Also, to relieve the morbidity associated with the marked valvular stenosis, surgical release of the non-corresponding synechiae in a stage-wise manner was done to supplement easy and early healing. Nasolacrimal duct stenting was not done in this scenario as it was an external abscess however it would be advisable otherwise. The patient had a follow-up period of 10 months.

With very few reports mentioning the involvement of the lacrimal apparatus as a complication of rhinoscleroma, there is no standard management protocol advocating the exact methodology of treatment. Hence, based on the experience of treating cases of rhinoscleroma along with a good knowledge base about its possible routes and sites of involvement and its associated complications has led us to craft this method of management in a stage-wise manner to achieve total clearance of the disease as well as the sequelae.

4. Conclusion

In the modern antibiotic era, being in the endemic region, a high degree of suspicion is needed to diagnose Rhinoscleroma of the nose as well as other areas. Early histopathological confirmation of the disease with a proper complete medical line of treatment along with planned nasal endoscopic surgical procedures to treat sequelae such as nasal stenosis can yield befitting results and prevent disfiguration. It can also lead to the prevention of dangerous complications associated with the disease. However, long term follow-ups are necessary to prevent recurrence associated with relapse.

5. Limitations

Due to paucity of literature in the management of these rare conditions, it should be emphasized to report such cases and the treatment protocol adopted in order to formulate standard management algorithms to aid the clinician in the future.

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There are no financial disclosures to be made.

7. Conflict of Interest

There are no conflicts of interests among authors.

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