Rhinoscleroma: case report
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1. Introduction
Rhinoscleroma is a chronic granulomatous, slowly progressive infection that affects the nose and other respiratory tract structures. It was first described in 1870 by dermatologist Ferdinando Von Hebra and posteriorly named respiratory scleroma, which emphasizes involvement of upper and lower airways [1]. It occurs frequently in the nasal fossae, eventually extending itself to the larynx, the rhinopharynx, the mouth and the paranasal sinuses; the lips, trachea, and bronchi may also be affected to a lesser degree. Extraspiratory involvement has rarely been described. Scleroma is endemic in some Central American countries, Indonesia, India, Poland, Hungary, Russia, and some African countries. In China the frequency of this disease is low. The regions of endemicity is Shandong province. The infection is due to a capsulate gram-negative bacterium, Klebsiella rhinoscleromatis, which was first described by Von Frisch in 1882 [2].

2. Case report
In December 2007, a 73-year-old married male patient who was a native of Beijing was brought to otolaryngology ward at the Beijing Tongren Hospital by his son and presented a mass extruding through the left nasal vestibule (Fig. 1), with progressive bilateral nasal obstruction for ten years. He also had a long history of baryodmia, no nasal pruritus or yellowish rhinorrhea, no daily intense frontal headaches. The specimen was referred to pathology for examination in other hospital and suspected “rhinoscleroma”. On examination with the laryngoscopic mirror, left nasal fossae were not pervious. A microlaryngoscopic exploration was performed and showed an enlarged nose due to a wax yellow-colored mass with small ulcers, a hardened consistency, asperate surface, and irregular borders, which extruded from the left nare. There was a similar

Fig. 1. Preoperative view: Mass extruding through the left nasal.
mass in the back of nasal septum in the right nasal cavity. Computed tomography of the paranasal sinuses showed soft tissue attenuation material occupies the nasal fossa, pachymucosa of bilateral frontal sinus, and submucous cyst of the left maxillary sinus. The cartilaginous septum is deviated to the right. There is no bone destruction. The specimen was referred to pathology for examination. Histopathologic features revealed nasal mucosa containing a diffuse lymphplasmacytic inflammatory process with large vacuolated macrophages typical of Mikulicz cells and many plasmocytes transformed into Russel bodies. The patient was treated with antibiotic therapy. Cefradine was used (500 mg, 12/12 hours for 1 month) based on literature data.

Three months later, the patient again consulted for progressive bilateral nasal obstruction, cough and thick phlegm. A microlaryngoscopic examination revealed the scar of the front rhinoscleroma and the spreading inflammation of the nasal mucosa were persistent. A nodulous mass through bilateral nasal vestibule. Treatment included surgery followed by antibiotics. Surgical removal of the tumor from the nasal vestibule. The specimen was referred to pathology for examination. The second histopathological examination showed an inflammatory reorganization, including plasmocytes, lymphocytes, and giant histiocytes. The patient accepted surgical therapy and the tumor was resected with a safety margin. In order to provide cover of the wound, a free skin flap of abdominal part was designed.

The flap measured 3 cm (in width) by 5 cm (in length) and was raised beginning at its lateral end. Depending on degrees of skin redundancy and elasticity, the flap was extended longitudinally as necessary to optimize the length-to-width ratio that would allow for primary closure without undue tension. The full-thickness skin graft was harvested with a number 10 blade and defatted using sharp scissors. The skin graft was sutured peripherally to the nasal vestibule defect. Postoperatively no complication was noted and the flap survived. The patient returned to the otorhinolaryngology unit last week was in good physical condition and no signs of recurrence in the nasal mucosa (Figs. 2 and 3).

3. Discussion

Rhinoscleroma is a long-standing disease that begins in the nasal mucosa and extends to other respiratory tract organs, producing airway obstruction and sometimes threatens the patient life [3]. Rhinoscleroma is found predominantly in rural areas and is commoner where socioeconomic conditions are poor. Acquisition of disease ID facilitated by crowding, poor hygiene and poor nutrition [4]. Early diagnosis and treatment are important to eradicate Klebsiella rhinoscleromatis and limit the fibrotic scarring sequelae.

The disease progresses in three stages [5,6]. (1) The catarrhal stage: patients have non-specific rhinitis symptoms that progress to fetid rhinorrhea, crusting, and nasal obstruction. The histopathology of this stage was squamous metaplasia, subepithelial infiltrate of neutrophils, and some granulation tissue. (2) The hypertrophic stage: this stage includes granulation tissue with deformity by widening of the nasal pyramid and nasal septum cartilage destruction. Epistaxis, anosmia, and anesthesia of the soft palate, among other symptoms may occur. Histopathological shows an infiltrate of chronic inflammatory cells, “Mikulicz cells” and “Russell bodies” were visualized as hallmark for diagnosis of rhinoscleroma. (3) The sclerotic stage: this stage is characterized by extensive scarring and nasal vestibular...
stenosis in severe cases. Histopathological includes large amount of fibrous and cicatricial tissues and few or no Mikulicz cells or Russell bodies.

Rhinocleroma treatment involves prolonged antibiotic therapy, in the attempt to eradicate *K. rhinoscleromatis*, because the relapse rate is high. Antibiotics with demonstrated efficacy are streptomycin, doxycycline, tetracycline, rifampicin, second- and third-generation cephalosporin, sulfonamides, ciprofloxacin and ofloxacin.

The patient in our case had a tumor that extruded from the right nare. It was typically a hypertrophic stage lesion. Cure is obtained with antibiotics, but the therapeutic scheme is controversial [7]. Until now, there has been no report of resistance to extended-spectrum cephalosporins in *K. pneumoniae*, which explains why we chose this antibiotic for the present case. The patient accepted cephalosporin by mouth for one month. Because of the drug concentration in nasal secretions and macrophages requires a prolonged course of treatment, our patient was not relieved by antibiotic therapy. So we took surgical therapy when he consulted again and showed to be of value. There was no relapse one year after surgery.

From this case, we thought drug treatment combined with surgery in cases with granulomatous lesions or scarring stenosis may be a good choice for curing rhinoscleroma. Granulomatous tissue, crusting and fibrotic scarring are lesions that produce several degrees of airway obstruction and cosmetic deformity, so that surgical therapy is indicated in some cases. Surgical procedures must be postponed until no residual disease activity is evident in the tissues to be debrided; otherwise, there is high risk of relapse or iatrogenic dissemination [8].

### 4. Conclusion

Rhinocleroma is a chronic disfiguring and debilitating disease. We have much to learn about its epidemiology and pathogenesis. We still have a long way from providing optimal therapy. Finally, relapses occur and prolonged follow-up is needed to spot early relapse.

### References


