Case Report

Rhinocerosoma with Involvement of the Maxillary Sinus

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Abstract

Rhinocerosoma is a chronic granulomatous infection of the upper respiratory tract, particularly affecting the nose. It is a bacterial disease mainly caused by rhinoscleroma subspecies of Klebsiella pneumoniae. We described a case of a 72-year-old Chinese man presented with repeated epistaxis for more than 2 years without any inducing factors accompanied by nasal obstruction, purulent discharge, nasal itching irritation of the left facial skin for 9 months and right ear for 3 months. Computer tomography showed a mucosal thickening in the nasopharynx, a shrinking of the nasal cavity structure, and a maxillary sinus inflammation. MRI showed a superficial thickening of the nasopharyngeal mucosa, a disappearance of the bilateral pharyngeal orifice of the Eustachian tube, and a high signal in the nasopharynx, a shrinking of the nasal cavity structure, and a maxillary sinus inflammation. A histopathology report showed a mixed inflammatory cell infiltrate and a collection of large, foamy Russell bodies and vacuolated Mikulicz cells beneath the nasal squamous epithelium. Rhinoscleroma is a quite rare case in the Otorhinolaryngology department but it is important to consider the etiology, diagnosis, and treatment of the disease. Rhinoscleroma requires a prompt diagnosis and treatment which may be fatal if the treatment is delayed. We managed our patient conservatively using intramuscular streptomycin and hemostatic anti-inflammatory drugs.

Keywords rhinoscleroma; maxillary sinus; Mikulicz cells

1. Introduction

Rhinocerosoma is caused by Klebsiella (K.) rhinoscleromatis, a gram-negative aerobic coccobacillus. Rhinoscleroma is a worldwide disease that occurs under all climatic conditions. Most cases are from tropical and temperate zones. The endemic regions are characterized by common environmental factors, such as low level of hygiene, malnutrition, and overcrowded housing. The inflammatory process usually starts in the nasal cavity and then it spreads on to nostrils, pharynx, and larynx, [1] but extension into the soft and hard palates, upper lip, and maxillary sinuses has also been reported [2]. The diagnosis can be established on the basis of histopathological, bacteriological, and genetic examinations. The initial complaint is usually sore throat, which is associated with nonspecific pharyngitis, progressive nodular infiltration of the soft and hard palates, tonsillar fossa, and oropharynx. There is a wide variation in clinical presentation depending on the site of disease and the duration of the disease. It usually affects the sinonasal cavities but all respiratory passages can be involved [3,4].

The disease manifests as 3 overlapping phases: exudative, proliferative, and fibrotic. The exudative phase is characterized by watery nasal discharge, frontoethmoidal headache, and difficulty in breathing, with nasal obstruction and epistaxis. The nasal mucosa and nasal septum may be hyperemic, edematous, or hypertrophic, with yellowish crusting. Sore throat and hoarseness may be present if the nasopharynx is involved. The proliferative phase includes a progressive infiltration of the nasal mucosa and septum, leading to formation of granulomatous waxy nodules. This leads to nasal obstruction, expansion to the anterior naris, and deformity of the upper lip. Proliferative granulomatous infiltration of nasal cavity leads to destruction of the nasal cartilage, causing “Hebra nose.” The fibrotic or cicatricial phase shows scarring or stenosis of the airway, leading to obstruction, and further involvement of an adjacent vital organ.

2. Case report

A 72-year-old Chinese man with low socioeconomic status, a resident of an urban slum area, was referred to our ENT department. The patient complained of recurrent epistaxis for 2 years without any inducing factors, accompanied by nasal obstruction, purulent discharge, and nasal itching. The amount of blood loss varied each time, a maximum amount of blood loss noted was 100 mL, and bleeding was fresh blood. After pressing the nasal cavity by fingers bleeding stopped. In recent 6 months, the patient suffered 4 episodes of bleeding so he went to local county hospital for treatment. He was treated with nasal packing, antibiotics, vitamins, and hemostatic drugs. A biopsy of nasal mucosa done in the local hospital was reported as nasal mucosa inflammation. After the drugs were stopped, the patient again suffered from nasal bleeding. The irritation of the

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facial skin was treated with steroid ointment, after the use of ointment for 1 week the irritation disappeared. Recent 2 months, the patient suffered from nasal bleeding and irritation of the right ear, and the irritation on the left facial skin reoccurred. For the better diagnosis and treatment, the patient came to our outpatient department of our hospital where he underwent a biopsy of nasal cavity mucosa and right ear skin. Nasal cavity mucosa biopsy was reported as rhinoscleroma, whereas skin of the right ear was reported as inflammatory granulation tissue. The patient was diagnosed as rhinoscleroma and admitted to our department. Since the onset of illness, the patient has no history of coma and loss of consciousness, no fever, no head and face pain, no visual changes, or other symptoms. He has also no history of nasal bone trauma or fracture. The general mental state, sleep, and appetite are normal, and no significant changes in body weight. No other surgical or medical history was revealed by the patient. He denied the use of any Chinese herbal medicine in the nasal cavity by himself to cure the disease.

On physical examination, we found right ear skin redness and swelling, especially in the central of redness which was prone to bleeding. Nasal endoscopy examination revealed the nasal mucosa erosion and it was prone to bleeding; most of the nasal cavity was covered by scab and secretion crusts. Turbinate structure was not clear. Cleaning all the secretions in the nasal cavity showed an atrophic mucosa without polyloid growth. Proptosis of the left facial skin was also present during the examination (Figure 1). On palpation, the left facial skin region was rigid. Computer tomography showed a mucosal thickening in the nasopharynx, a shrinking of the nasal cavity structure, and a maxillary sinus inflammation (Figure 2). MRI showed a superficial thickening of the nasopharyngeal mucosa, a disappearance of the bilateral pharyngeal orifice of

Figure 1: Clinical photograph showing proptosis in the patient’s left facial skin and right ear skin was also present. Published with patient’s permission.

Figure 2: Computer tomography showed a mucosal thickening in the nasopharynx, a shrinking of the nasal cavity structure, a maxillary sinus high-density, and a maxillary sinus inflammation (arrows).
Figure 3: MRI showed the top of the nasopharyngeal mucosa thickening, both of the pharyngeal orifices of the Eustachian tube disappearing, and a flake high signal in the nasal cavity and maxillary sinus (arrows).

Figure 4: Photomicrograph displays foamy cells beneath the nasal squamous epithelium and plasmacytes with visible traits of hyaline degeneration in the form of Cornil cells and Russell bodies. (a), (b) During the 6 months before admittance to the hospital. (c), (d) During the inpatient treatment. (hematoxylin and eosin stain; ×10 magnification).

the Eustachian tube, and a flake high signal in the nasal cavity and maxillary sinus (Figure 3). Hematology results were WBC 13.38 × 10^9/L, RBC 2.93 × 10^{12}/L, and HGB 86.90 g/L. The renal function result revealed that creatinine was 77 umol/L, with endogenous creatinine clearance rate of 56.09 mL/min. Other hematologic and biochemical results were within normal limits.

Considering the clinical course of the disease and the radiology findings, chronic granulomatous diseases such as tuberculosis, actinomycosis, histoplasmosis, malignancy of the maxillary sinus (lymphoma), and nasopharyngeal tumor were considered as the differential diagnosis. Because of the presence of severe trismus, the patient once again performed a nasopharyngeal endoscopic guided nasal cavity biopsy. Subsequent staining with hematoxylin and eosin showed a mixed inflammatory cell infiltrate and a collection of large, foamy Russell bodies and vacuolated Mikulicz cells beneath the nasal squamous epithelium (Figure 4).

The patient was prescribed intramuscular streptomycin injection 0.375 g daily, accompanied with cefotaxime, cephalosporin antibiotics, vitamins, and hemostat drugs. After 1 month of therapy, the frequency and severity
of irritation in the skin of face and ear was decreased, with occasional nasal bleeding. Throughout therapy in 3 months with streptomycin, the patient did not present with nasal bleeding or skin irritation. Adverse gastrointestinal discomfort or any other allergic reaction was not seen. The nasal mucosa recovered to normal condition. No nasal bleeding, purulent discharge or crusts were seen.

3. Discussion
Rhinoscleroma is a progressive granulomatous disease starting in the nose and later extending into the nasopharynx and oropharynx, larynx, and, sometimes, the trachea and bronchi. Scleroma has been reported in many unusual sites in the head and neck, including the orbit, lacrimal system, facial skin, and anterior cranial fossa [5,6].

Clinical differential diagnosis of the second and third stages of rhinoscleroma involves all destructive lesions associated with granulomas and ulcer formation, including sarcoidosis, Wegener’s granulomatosis, leishmaniasis, tuberculosis, syphilis, fungal infections, leprosy, nasopalatine duct cyst, Rosai-Dorfman disease, intranasal maniasis, tuberculosis, syphilis, fungal infections, leprosy, including sarcoidosis, Wegener’s granulomatosis, leishmaniasis, tuberculosis, syphilis, fungal infections, leprosy, nasopalatine duct cyst, Rosai-Dorfman disease, intranasal maniasis, tuberculosis, syphilis, fungal infections, leprosy, including sarcoidosis, Wegener’s granulomatosis, leishmaniasis, tuberculosis, syphilis, fungal infections, leprosy.

In this case, the patient was put on antibiotic therapy for 3 months because the nasal mucosa was involved. K. rhinoscleromatis is not found in normal nasal secretions; therefore, a positive culture is considered diagnostic of scleroma. The presence of gram-negative diplobacillus in Mikulicz cells or intercellular spaces is considered pathognomonic of rhinoscleroma. Immunochemistry adds a great specificity in the diagnosis of scleroma when histology is not diagnostic. An immunoperoxidase technique may also be helpful in the identification of K. rhinoscleromatis when cultures are negative.

Antibiotics constitute a standard treatment in the early stage of the disease. Surgery and laser ablation can be used only after the disease is under control with antibiotics and the cultures are negative. Antibiotics do not reach an effective level in relatively avascular scar tissue; therefore, patients presenting with areas of fibrosis and scarring require surgery as well as antibiotic therapy. Surgical debridement could also be considered if there is a significant airway obstruction or cosmetic deformity. Relapses in rhinoscleroma are common, hence the need for prolonged antibiotic treatment and close follow-up to detect early recurrences. Other rarer complications to keep in mind include stenosis leading to respiratory obstruction, hemorrhage, intracranial invasion, and malignancy transformation.

In the present case, the patient had the characteristic signs and symptoms of nasal tract infection over many years, although he sought medical treatment in local hospital without definitive diagnosis which was not effective. Using streptomyacin and other drugs in our hospital after a definitive diagnosis confirmed by a histopathological study was found to be effective. The patient recovered well after the drug course of 3 months. Diagnosis of the disease is based on clinical, bacteriologic, and histopathologic findings and supported by radiologic examination. Without the benefit of a prior clinical diagnosis, laboratory findings, or suggestive clinical signs and symptoms, granulomatous diseases may be difficult to differentiate radiologically. Although individual granulomatous diseases may have overlapping findings at imaging, certain radiologic findings should prompt the inclusion of granulomatous diseases in the differential diagnosis, thus facilitating appropriate clinical management [9]. We highlight this condition to raise awareness of the disease in order to aid in early diagnosis of patients. Without treatment, this condition can result in significant complications, including involvement of the lower airways. Early diagnosis and appropriate treatment help to reduce morbidity.

References