CASE REPORT

Rhinoscleroma: an unusual form of presentation and evolution. Case report

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ABSTRACT

Rhinoscleroma is a chronic infectious condition found mainly in patients living in underdeveloped regions. It affects the respiratory tract (in 95% of the cases the nasal cavity is the first affected), but it can also involve the orbit, the middle ear, the lip, the gums and the cutaneous tissue. It is linked to infection with Klebsiella rhinoscleromatis. Females are far more frequently affected than males. Symptoms typically appear in the second and third decades of life. Diagnosis is confirmed based on the pathology result of the biopsy and also by microbiology exam, but it has a low-rate specificity. Treatment consists of prolonged antibiotic therapy and surgical intervention. Follow-up of the patient is necessary due to the high relapse rate of the disease.

We present a rare case of rhinoscleroma presented initially with cutaneous manifestations, which progressed rapidly causing invasion of the right frontal sinus by lysis of its anterior wall.

KEYWORDS: rhinoscleroma, cutaneous manifestation, Klebsiella rhinoscleromatis.

INTRODUCTION

Rhinoscleroma is a granulomatous condition with a slow evolution caused by the infection with Klebsiella rhinoscleromatis, an encapsulated gram-negative bacillus. It is a rare disease; thus, an accurate incidence is not yet known. Rhinoscleroma is usually found in rural areas, affecting predominantly the population from the lower socio-economic class[1]. Endemic regions are represented by underdeveloped countries – Central and South America, Africa, Central and Eastern Europe, Southeast Asia, but sporadic cases have been reported all around the world[2]. Rhinoscleroma mainly affects parts of the respiratory system: the nasal cavity (95-98%), the larynx (15-40%), the nasopharynx (18-43%), the trachea (12%) and the bronchi (2-7%), but other uncommon localizations have also been described, in addition to those mentioned above. The diagnosis is confirmed by microbiologic (50-60% positive specificity) or histopathologic exams, currently the gold standard method of diagnosis. Treatment consists of antibiotic therapy and, in some cases, surgical intervention[1].

We present a rare case of rhinoscleroma with cutaneous manifestations and rapid, aggressive evolution, which in only 6 months caused lysis of the anterior wall of the frontal sinus.

CASE REPORT

A 49-year-old man presented with a tumefaction located on the upper right lateral part of the nose bridge, which extended to the medial angle of the eye. The swelling appeared six months before presentation and it had a gradual evolution, becoming painful about one month before presentation. No nasal symptomatology was associated. We would like to mention that the patient is from a high economic class, and he has well-controlled diabetes. Before his presentation in our clinic, he had been consulted by multiple doctors, had undergone various investigations, including a craniofacial computed tomography (CT) scanning, performed 6 months prior, that showed

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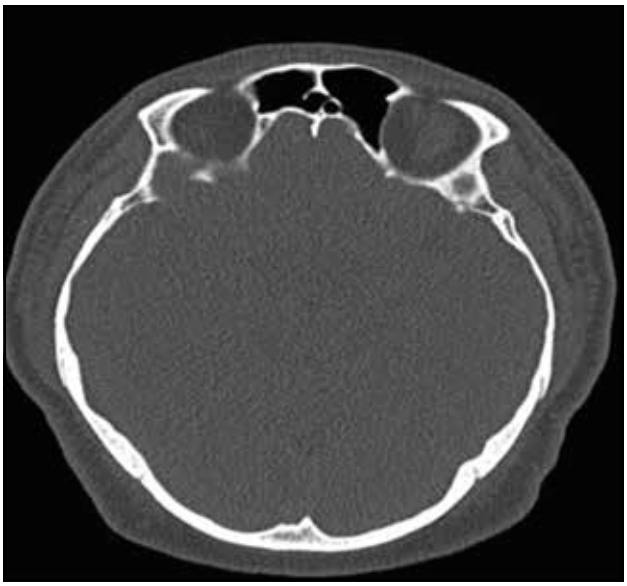

Figure 1. Cranio-facial CT (axial view), showing no lysis of the anterior wall of the frontal sinus (performed 6 months prior to the initial evaluation in our clinic).

no evidence of any bone lysis (Figure 1). As treatment, the patient used various topical medication, but without any improvement.

ENT examination at the admission in the hospital revealed a solid, ovoid mass of approximately 3-4 cm in size, situated on the upper-right part of the nose bridge, immobile against the underlying bone surface, that was painful at palpation, with no other cardinal signs of inflammation; mild swelling around the tumor and towards the medial angle of the eye was noticed (Figure 2 A).

No associated regional adenopathies were discernible on palpation and no abnormalities were detected on endoscopic naso-pharyngo-laryngoscopy exam. Chest X-ray was normal. Complete blood count, renal function tests and liver function tests were all within normal limits. Also, serological tests for hepatitis B and C, for HIV and for syphilis were negative. The nasal and pharyngeal swab cultures were negative for bacteria and fungi.

The cranio-facial native computed tomography scanning (CT) (Figure 2 B, C) completed by magnetic resonance imaging (MRI) (Figure 2 D, E, F) showed a

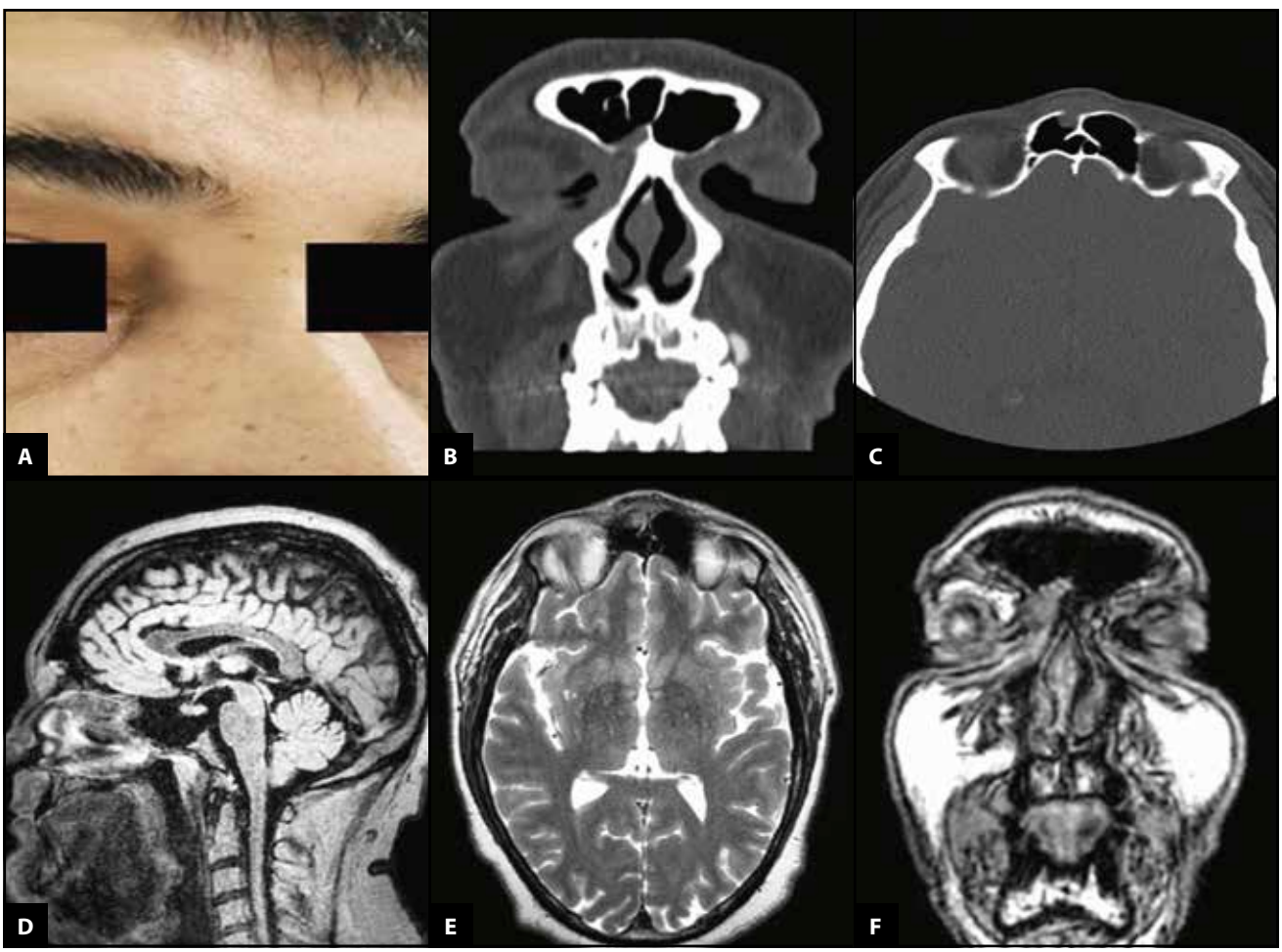

Figure 2. Macroscopic aspect of the lesion with integrity of the cutaneous tissue and no signs of inflammation **(A)**. Cranio-facial CT scan, coronal **(B)** and axial view **(C)**, and cranio-facial MRI sagittal **(D)**, axial **(E)** and coronal view **(F)**, revealing the lysis of the anterior wall of the right frontal sinus and the extension of the mass into the frontal sinus.

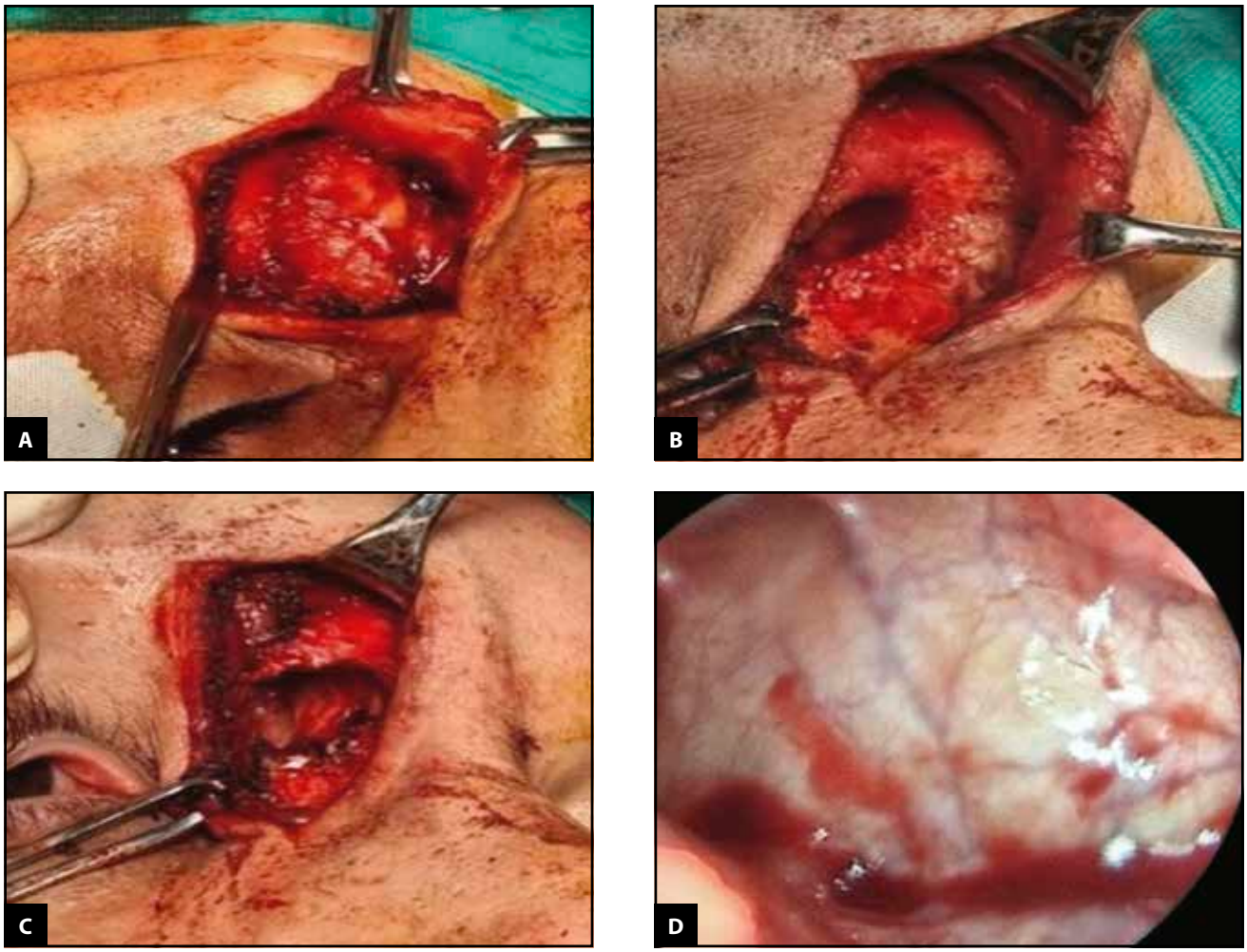

Figure 3. Intra-operative aspect of the mass **(A), (B), (C)**. Intra-operative endoscopic view of the frontal sinus after the careful removal of the mass and surrounding mucosa **(D)**.

hypodense nodular lesion with a diameter of approximately 10 mm, developed "en plaque" with a large base of implantation, well defined margins, located in the soft tissue, situated near the medial angle of the right eye, in intimate contact with the right ethmoidal bone. The imaging examination also revealed erosion of the anterior wall of the right frontal sinus and extension of the tumor into the sinus.

Due to the rapid evolution of the mass – approximately six months, as well as the invasive imagistic aspect, surgery was the treatment of choice. Intraoperatively, a tumoral mass of solid consistency was revealed, adherent to the bone. The tumor incorporated the procerus muscle and extended superiorly in the right frontal sinus, through a bone discontinuity orifice of approximately 1.5 cm across, situated on the anterior wall of the frontal sinus (Figure 3). Excision of the tumor, of the surrounding frontal sinus mucosa and of the anterior wall of the frontal sinus was performed, in order to establish an accurate diagnosis and to avoid possible complications. The tumoral mass along with the frontal sinus mucosa and bone fragment of the anterior wall of the right frontal sinus were sent for histopathology exam.

The histological exam of the excised specimen revealed granulomatous inflammation with large foamy macrophages, multiple rod-shaped strictures were visible within their cytoplasm using Warthin-Starry staining (Figure 4), Mikulicz cells and Russell bodies were also found. The immunochemistry study confirmed these findings, supporting the diagnosis of rhinoscleroma.

DISCUSSIONS

Rhinoscleroma is a chronic granulomatous infectious condition which is mainly present in underdeveloped countries. It is caused by Klebsiella rhinoscleromatis, and it was described in 1870 by Herba[3]. The presence of familial disease showed that, in endemic locations, genetic control of the host response to Klebsiella rhinoscleromatis may be an important factor[4]. Females are significantly more likely than males to be affected, and the condition

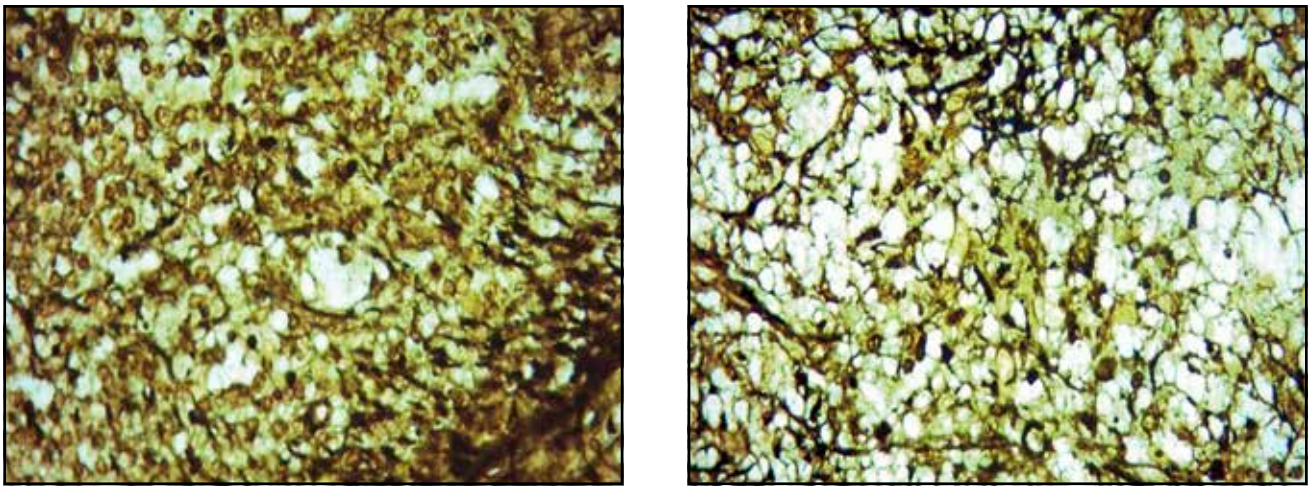

Figure 4. IHistopathological section (Warthin-Starry stain x 60) showing foamy macrophages with multiple rod-shaped strictures within their cytoplasm.

usually manifests in the second to third decades of life[5].

Most of the cases reported in literature had the respiratory tract as a starting point: the nose (95-98%), the larynx (15-40%), the nasopharynx (18-43%), the trachea (12%) and the bronchi (2-7%)[1]. Less common and rare localizations such as the orbit, the skin, the middle ear, the lip and gums have also been described[6–8]. In this case, the patient initially presented only with cutaneous manifestations. So, despite its name, rhinoscleroma is not a disease limited to the nose and nasopharynx. Even though it has an affinity for the nasal mucosa, it can have other localizations. Even with the rapid evolution and invasion of the frontal sinus, no nasal symptoms were observed, which is extremely rare.

Rhinoscleroma has the capacity to erode the surrounding structure by subepithelial invasion of Klebsiella rhinoscleromatis, histiocytes and neutrophils which extend into the interstitium and the associated cartilage and bone structures[9]. In the presented case, the tumor extended inside the frontal sinus from its primary cutaneous location through an area of bone discontinuity of approximately 1.5 cm in the anterior sinusal wall. The rhinoscleroma evolution was aggressive and fast. In only six months it produced the lysis of the anterior wall of the right frontal sinus, causing pain in the projection area of the frontal sinus. Even so, the patient did not accuse any nasal manifestations.

Humans are the only hosts for Klebsiella rhinoscleromatis. The bacteria is spread in the community by inhaling the nasal droplets from an infected person[10]. High rates of infection are found in endemic areas, especially in crowded areas[11].

Rhinoscleroma is classified into three clinical and pathological stages – the catarrhal stage, the granulomatous stage and the fibrotic stage[12]. The histological exam represents the mainstay method for the diagnosis of rhinoscleroma and the findings are characteristic especially in the granulomatous (proliferative) stage[13]. The histopathological stages are represented by: catarrhal / atrophic stage (characterized by squamous metaplasia, nonspecific neutrophil infiltrate with granulation tissue); granulomatous / proliferative stage (pseudoepitheliomatous hyperplasia, chronic inflammatory cells: Mikulicz cells and Russell bodies); sclerotic stage (extensive fibrosis, less inflammatory cell infiltration)[13-14].

The characteristic findings in the histopathology exam are the Mikulicz cells – huge histocytes with multiple vacuoles carrying nonviable bacteria[15], Russell bodies – plasma cells with eosinophilic structures[16] and positive Warthin-Starry stains, but in early stage these characteristics may be noncontributory[13]. In our patient, the histopathological exam confirmed the disease to be in the granulomatous stage.

50-60% of the patients that are in the granulomatous stage test positive for Klebsiella rhinoscleromatis on MacConkey agar culture or on routine blood cultures[7].

Radiological exams, such as computer tomography (CT) scan and magnetic resonance imaging (MRI), are recommended and allow an accurate evaluation of the extension of the lesions[1].

The differential diagnosis depends on the site of infection and can be made with other granulomatous infections caused by bacteria – syphilis, actinomycosis, tuberculosis, as well as fungi – sporotrichosis, histoplasmosis and by parasites – leishmaniosis. Rhinoscleroma can also be difficult to differentiate from non-infectious inflammatory processes and neoplasms such as Wegener granulomatosis, sarcoidosis, basal cell carcinoma, verrucous carcinoma, lymphomas, etc[7].

The treatment for rhinoscleroma is challenging, including long-term antibiotic therapy – at least 4 weeks, but it may be required for up to 1 year, depending on the disease stage. Several antibiotics such as streptomycin, tetracycline, ciprofloxacin, second and third generation

cephalosporins, doxycycline, rifampicin, fluroquinolone, are used. Corticosteroids therapy and surgical intervention have also been proven helpful in the treatment of rhinoscleroma. Continuous follow-up of the patient is required until the disease is completed eradicated, due to the high relapse rate of the disease[1].

The complications of the rhinoscleroma are destructive. The disease has the capacity to erode the surrounding structures, leading to permanent local deformation. Other complications such as anosmia, dysphonia, oral anaesthesia, stridor, tracheal stenosis, vestibular stenosis and dysphagia could happen.

Remarkable for this case are the patient's history, his socio-economic status, the fact that complete blood count, renal and liver function tests were all normal, along with negative nasal and pharyngeal swab for bacteria and fungi, all of which are uncharacteristic for this type of infection. Given the rapid extension (six months) of the mass with bone lysis and intrasinusal extension, it was necessary the exclusion of any neoplastic etiology such as lymphoma or basal cell carcinoma. As such, the first therapeutic gesture was the surgical excision of the mass along with frontal sinus mucosa and a fragment bone of the anterior wall of the right frontal sinus, in order to ensure safety margins for a potential neoplastic etiology. Only after the pathological result was received, antibiotherapy with ciprofloxacin was instituted for a period of 1 month to eradicate the disease and prevent its recurrence. No relapse was observed at the 6-month routine follow-up.

To avoid the progression of the disease and its destructive complications, a prompt diagnosis and treatment of rhinoscleroma is necessary[17]. In this case, since the patient addressed quite early to the doctor, the progression of the rhinoscleroma along with its destructive complications were minimal, without provoking any major damage to surrounding regions.

CONCLUSIONS

Due to various considerations such as its rarity, difficult differential diagnosis, low sensitivity of diagnostic tools and varying forms of presentation, diagnosing rhinoscleroma can be particularly difficult and the histopathological exam remains most trustworthy and accurate diagnosis method.

Rhinoscleroma is a rare, intriguing disease, that requires more research for a better understanding of its pathophysiology and to enhance treatment regimens for the prevention of relapses and their consequences. For the moment, it represents a diagnostic and a therapeutic challenge both for the clinician and for the pathologist.

We reported this case to highlight the fact that, despite a complete clinical and paraclinical examination, histopathological examination is the key to arrive at the correct diagnosis.

Conflict of interest: The authors have no conflict of interest to disclose.

Contribution of authors: All the authors have equally contributed to this work.

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