CASE REPORT

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Rhinoscleroma of the sinuses*

Axelle Bailhache[1], Danièle Dehesdin[1], Arnaud François[2], Jean-Paul Marie[1], Olivier Choussy[1]

[1] Department of Otorhinolaryngology, Rouen University Hospital, Rouen, France
[2] Department of Pathology, Rouen University Hospital, Rouen, France

SUMMARY

Introduction: *Rhinoscleroma is a rare chronic, granulomatous disease of the respiratory tract.*
Objective: *The aim of this paper was to report 2 unusual cases of rhinoscleroma and to review the literature.*
Material: *We present two cases of sinus rhinoscleroma diagnosed and treated at the ENT Department of a French University hospital.*
Results and conclusion: *Rhinoscleroma primarily affects the nasal cavity but the nasopharynx (18%-43%), larynx (15%-40%), trachea (12%) and bronchi (2% to 7%) can also be involved. However, the paranasal sinuses are usually free of disease. Rhinological signs are generally the first reported by patients. CT scan and MRI are useful for diagnosis. Diagnosis of rhinoscleroma was based on histological characteristics and presence of Klebsiella rhinoscleromatis on biopsy cultures. In most cases treatment involves prolonged antibiotic therapy with aesthetic surgical reconstruction when necessary. However, rhinoscleroma is difficult to eradicate and its recurrence rate is high.*

Key words: chronic sinusitis, rhinoscleroma

INTRODUCTION

Rhinoscleroma is a chronic infectious granulomatous disease caused by *Klebsiella rhinoscleromatis*. To our knowledge, this pathology is rare in Western Europe but commonly found in semi-desert types of climates [(1)]. In France the current incidence of rhinoscleroma is largely due to immigrants from endemic areas [(2)].

The nasal cavity is the most common site of involvement and although lesions may arise in any area of the respiratory airway, the sinuses are rarely affected [(1)].

The aim of this paper was to report 2 unusual cases of sinus rhinoscleroma together with a review of the literature.

CASE REPORTS

Case 1

A 72-year old woman, presented with right jugal pain over a period of several months. This patient was born in France with no previous medical history. Twelve years previously she went to Tunisia on holiday, which was the sole risk factor. On clinical investigation, there was no nasal discharge or epitaxis. A previous anterior rhinoscopy was normal. Cranial nerves and neurological testing were also normal. CT scan of sinuses was performed due to the severe and persistent pain (Figure 1). Imaging revealed opacification of the right posterior ethmoid and sphenoid with osteolysis of the sphenoid floor. A magnetic resonance imaging (MRI)(Figure 2) complemented the CT scan findings and confirmed the tumoural appearance of this mass using gadolinium enhancement. However, the lesion was localised to the sinus with no extra-sinus extension. An endoscopic surgical exploration was carried out with dissection of haemorrhagic and adherent soft tissue. Histology revealed granulomatous and necrotizing lesions with vacuolated histiocytes, Mikulicz cells and plasmocytes with Russel bodies. The results of Grocott and Wharthin-Starry staining were negative.

Table 1. Nasal rhinoscleroma, literature cases in non-endemic areas.

Authors	Year of publication	Number of cases	Geographical origin
Ammar [7]	2001	1	USA
Avery [14]	1995	1	USA
Trautmann [17]	1993	1	Germany
Badia [20]	2001	1	UK
Di Carlo [21]	2001	1	Italy
Kim [22]	2003	1	Korea
Fernandez [23]	2004	3	Spain
Verma [24]	2005	1	Canada
Chan [25]	2007	1	USA

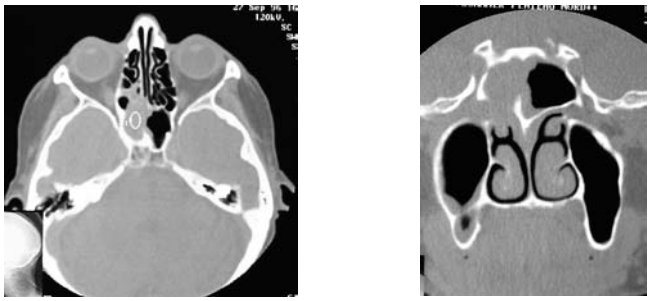

Figure 1. Pretherapeutic Sinus CT scan. Right posterior ethmoidal and sphenoidal opacity with osteolysis of the sphenoidal floor.

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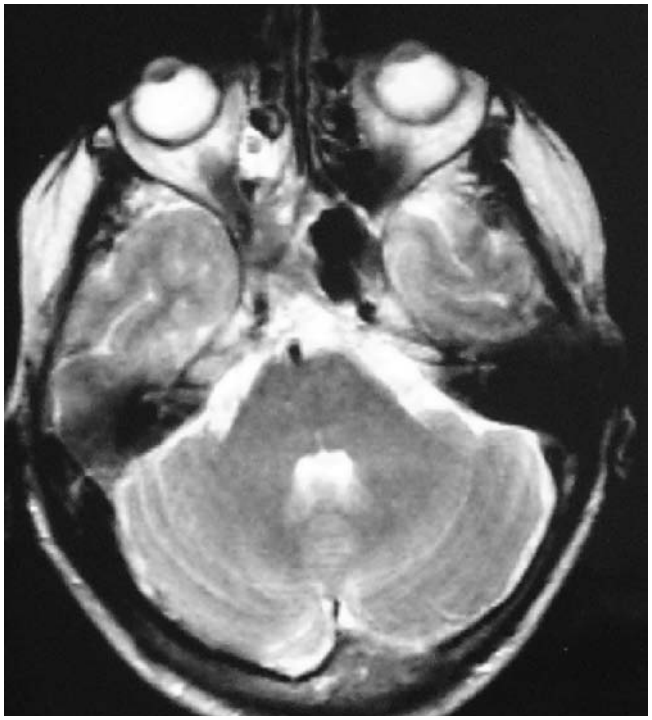

Figure 2. Soft tissue mass with moderate gadolinium enhancing on the initial MRI imaging.

Gram-negative intra-cellular organisms were found with identification of *K. rhinoscleromatis*. These results suggested a diagnosis of rhinoscleroma. Trimethoprime-sulfamethoxazole was prescribed alternating with ciprofloxacin due to poor tolerance for 6 weeks. Clinical examination and MRI constituted the follow-up assessment. Six years later due to loss of visual acuity as a result of an intra orbital lesion, further antibiotic therapy with ciprofloxacin and systemic corticosteroid for 10 days was initiated, which resulted in total recuperation of the visual acuity. Two years later a new deposit was found near the middle meatus. The result of the biopsy suggested rhinoscleroma but no *K. rhinoscleromatis* was discovered. Further antibiotic therapy with ciprofloxacin was started for a 6-week period, and the patient then completely recovered.

Case 2

A 74-year-old woman presented with bilateral nasal obstruction, a long period of anosmia and a diagnosis of nasal polyposis despite 9 months of corticosteroid treatment.

On clinical examination we found a mass in the left superiomedial corner of the orbit with inferior and lateral exophthalmia. The rest of the ophthalmologic examination was normal. Nasal endoscopy revealed a polyp and inflammatory mucosa. Sinus CT scan (Figure 3) revealed left frontal and ethmoidal opacification with bone erosion. A frontal mucocele was diagnosed and surgical treatment was performed. Histological tests revealed Mikulicz cells with intra cellular organisms and rhinoscleroma was suggested. Ciprofloxacin was administered for 6 weeks. At 9 years follow-up, there was no recurrence of the rhinoscleroma. Polyposis remained present and was subsequently treated medically with topical nasal corticosteroid.

DISCUSSION

These 2 cases are unusual due to their geographic origin, the age of the patients and the sinus presentation. Cases reported in the literature are summarized in Table 1.

Epidemiology

Rhinoscleroma is a granulomatous disease of the respiratory tract. Von Hebra and Kohn described the first case in 1870 [3] but Von Frish [4] in 1882 first detected the causal agent, i.e. *K. rhinoscleromatis*. In Western Europe, this pathology is rare and major factors are economic deprivation and poor hygiene and nutrition [1]. Endemic geographical areas include eastern and central Europe, Central and South America, East Africa and the Indian sub-continent [1]. Airborne disease transmission requires prolonged contact [1]. There is no gender prevalence and the young adults in the second or third decades are most commonly affected [5]. HIV positive patient can also be affected [2] but this does not occur frequently [6].

Clinical manifestation

Rhinoscleroma includes 3 clinical stages [2,7]. The catarrhal-atrophic stage is characterized by blood streaked mucus or mucopurulent nasal discharge and fetid crusts. Non-specific symptoms such as epitaxis, nasal obstruction and anosmia characterize the infiltrating or granulomatous stage during which granulomatous nodules may develop. The last stage is a sclerotic stage where nodules are replaced by fibrosis tissue causing nasal stenosis and external deformity. At this stage nasal endoscopy can be useful to reveal abundant crusts.

Nasal cavity pathology is almost always present (95-100%) [7,8]. Minor symptoms can be present (rhinorhea, crusts, anosmia, polyposis) [7] but the involvement can be extensive with nasal deformity (32%) [8] and a Hebra nose (10%) [3]. In the last fibrosis stage, nasal stenosis or nasolacrimal duct stenosis may occur [9].

The other affected areas are larynx (26%), eustachian tube (27%), trachea (20%), maxillary sinus (22%) and pharynx (in 18 to 43%) [10].

Radiological findings

CT scan and MRI are the recommended examinations. On CT scan, the nose is frequently invaded. Characteristic lesions are homogeneous with well-defined borders and without enhancement [11]. Lesions varying from localized mucosal thickening to obliterative soft tissue extending from the septum to the lateral wall, opacification of sinuses and in extreme cases bone and cartilage involvement may also be present [7]. Le Hir et al. reported that the [6] middle and inferior turbinate are usually affected. There are no clear feautures of rhinoscleroma on MRI, according to the literature. Some authors [6] have reported rhinoscleroma as a homogenous lesion on T1-weighted images of isosignal intensity and hyposignal intensity on T2-

weighted images relative to the muscle. Razek et al. [12] reported that, on T1-weighted images, rhinoscleroma showed striking or mild high signal intensity relative to muscle and cerebro- spinal fluid, but less hyper intensity than fat. On T2-weighted images, the nasal masses may show homogeneous high signal intensity or heterogeneous high signal intensity associated with hypo intense foci.

Bacteriology

Cultures from biopsy are in most cases positive and confirm the presence of *K. rhinoscleromatis* [13]. Cultures are carried out on the nasal discharge or crust, are only positive in 50 to 60% of cases [2]. The bacteriological laboratory should be informed regarding the possibility of *K. rhinoscleromatis* due to the specific nature of the culture required.

Pathology

Biopsies must be performed from the most active sites of this pathology or from the septum and inferior turbinate. Mickulicz cells are not pathognomonic but are characteristic. A chronic inflammatory infiltration of monocytes and lymphocytes has also been described [14].

Biological findings

There are no specific or sensitive tests required to perform the diagnosis. Some authors have reported cellular immunity modifications [7,15] but with no benefit as regards the diagnosis.

Treatment

Aggressive therapy must be carried out due to the high recurrence rate of rhinoscleroma [7]. Antibiotics are the basis of the treatment. Tetracycline is an historical choice due to its low-cost and efficacy (87% of *K. rhinoscleromatis* are sensitive [16]). However, this is contraindicated in children or pregnancy and its recommended dosage is four times daily. Cephalosporin is effective *in vitro* but pathological relapse may occur when treatment is stopped [17]. Rifampicin is one of the most effective antibiotics [18]. Ciprofloxacin seems to be an antibiotic with the best treatment compliance and efficacy but with a high cost [14,16]. The optimal cost benefit ratio is obtained with the combination trimethoprim-sulfamethoxazol but with greater toxicity than ciprofloxacin [16]. Antibiotic therapy should be continued for several months (3 to 6) until biopsy and culture results are negative [19].

Surgery can be associated with antibiotics to clear the nasal cavity crusts and in the reconstruction of nasal deformity.

CONCLUSION

Rhinoscleroma is a rare pathology in economically developed countries. The nose is always affected but sinuses are rarely affected. Our case study reports 2 cases of sinuses rhinoscleroma with specific clinical and radiological findings. Diagnosis was made based on pathology and bacteriology. The natural history includes a tendency of this disease to recur, to extend locally causing deformity. Treatment is based on several months of antibiotic therapy, whereas surgery is reserved for diagnosis (biopsies), nose cleaning and reconstruction.

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Dr. Olivier Choussy
Service d'ORL et chirurgie cervico-faciale
CHU de ROUEN - Hôpital Charles Nicolle
1, rue de Germont
76031 Rouen cedex
France

Tel: +33-23-288 6614
Fax: +33-23-288 8359
E-mail: olivier.choussy@chu-rouen.fr