

## Severe Bilateral Nasal Obstruction Revealing a Rhinoscleroma

Niema Benkhraba<sup>1,3\*</sup>, Razika Bencheikh<sup>2,3</sup>, Hanaa Rahim<sup>1,3</sup>, Ahmed Ould Mohamed<sup>1,3</sup>, Mohamed Anas Benbouzid<sup>2,3</sup>, Leila Essakalli Houssyni<sup>2,3</sup><sup>1</sup>Resident Physician in Otorhinolaryngology, Department of Otorhinolaryngology, Head and Neck Surgery, Ibn Sina University Hospital, Rabat, Morocco<sup>2</sup>Professor of Otorhinolaryngology, Department of Otorhinolaryngology, Head and Neck Surgery, Ibn Sina University Hospital, Rabat, Morocco<sup>3</sup>Faculty of Medicine and Pharmacy of Rabat, Mohammed V University, Rabat, MoroccoDOI: [10.36347/sjams.2021.v09i05.006](https://doi.org/10.36347/sjams.2021.v09i05.006)

| Received: 11.03.2021 | Accepted: 18.04.2021 | Published: 11.05.2021

\*Corresponding author: Niema Benkhraba

### Abstract

### Case Report

Rhinoscleroma is a chronic granulomatous disease of the upper airways. Several predisposing factors such as living under conditions of poor hygiene and nutritional deficiencies have been described and are apparently necessary for transmission of the disease. The diagnosis is histological and the treatment is based on surgical debridement and long-term antibiotic therapy. We report the case of a 33-year-old female patient who presented a unilateral nasal obstruction, with chronic rhinorrhea, intermittent epistaxis, and anosmia. The clinical exam found a deformation of the nasal pyramid, a septal deviation, and an anterior nasal mass. The CT-scan of the paranasal sinuses shows a lesional process centered on the left nasal cavity with lysis of the nasal septum and turbinates. An endonasal endoscopic complete exeresis was realized. The histological study returned in favor of a granulomatous inflammatory process of the nasal cavity responding to a rhinoscleroma.

**Keywords:** Rhinoscleroma, Nasal cavity, Surgery, Antibiotherapy.

**Copyright © 2021 The Author(s):** This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CC BY-NC 4.0) which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use provided the original author and source are credited.

## INTRODUCTION

Rhinoscleroma is a chronic granulomatous disease of the upper airways. It rarely occurs nowadays in Europe, where it was first described in 1870 [1]. However, it remains endemic in tropical and subtropical areas (North and Central Africa, Southeast Asia and Central and South America) [1]. Several predisposing factors such as living under conditions of poor hygiene and nutritional deficiencies have been described and are apparently necessary for transmission of the disease [2]. With no gender preference, it more frequently affects young adults [3]. *Klebsiella rhinoscleromatis* is the etiologic agent of rhinoscleroma [1]. Three clinical and histological stages have been described: the catarrhal-atrophic stage, the granulomatous stage and the sclerotic stage [2]. We present a case report of a young patient diagnosed with rhinoscleroma, and we review the existing literature.

## CASE REPORT

This is a 33-year-old female patient with no notable pathological history, who suffered for 2 years from unilateral nasal obstruction, with chronic rhinorrhea, intermittent epistaxis, and anosmia. The evolution was marked by the appearance of a

tumefaction of the homolateral cheek. On examination, we found a deformation of the nasal pyramid, a septal deviation, and an anterior nasal mass (occupying the entire left nasal cavity). There was a mass effect on the right nasal cavity, the nasolabial fold, and an elevation of the nasal wing. The nasal flow is abolished on the left side and decreased on the nasal cavity (Figure-1).

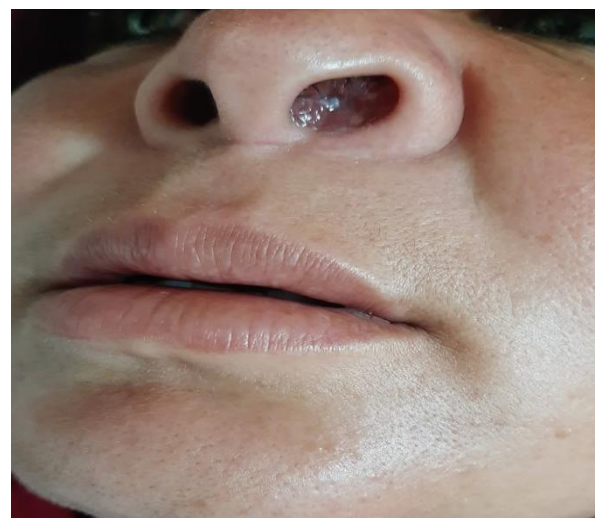


Fig-1

A computed tomographic scan of the paranasal sinuses was performed and showed a lesional process of tissue density, measuring approximately 41 \* 37 \* 83mm, centered on the left nasal cavity, with regular

contours. There was heterogeneous enhancement with intravenous contrast. Additional, a lysis of the septum and turbinates was observed. Also, a frontal- maxillary- sphenoid sinusitis, especially in the left.



**Fig-2: CT-scan of the paranasal sinuses shows a lesional process centered on the left nasal cavity with lysis of the nasal septum and turbinates (A: coronal sections, B: axial section)**

An endonasal endoscopic complete exeresis of the tumor combined with a Caldwell-Luc approach to control the left maxillary sinus was realized. The histological study returned in favor of a granulomatous inflammatory process of the nasal cavity responding to a rhinoscleroma with characteristic intracellular of *Klebsiella rhinoscleromatis*. Therefore the patient received ciprofloxacin 500mg twice a day for 4 weeks. She was seen for endoscopic control and presented a very favorable evolution.

## DISCUSSION

Rhinoscleroma is a chronic, infectious, granulomatous disease that may present with mass lesions in the respiratory tract anywhere from the nose to the trachea [1]. The nasal cavity is most often affected (95–100 per cent) but lesions may also involve the: larynx (15–40 per cent); nasopharynx (18–43 per cent); oral cavity; paranasal sinuses (26 per cent); soft tissues of the lips and nose; trachea (12 per cent); bronchi (2–7 per cent); and, rarely, the orbit or middle ear [1]. The condition is often indolent, progressive, prone to recurrence and extremely difficult to cure. The infectious agent, *Klebsiella rhinoscleromatis* (of the Enterobacteriaceae family), was first described by von Frisch in 1882 [2]. This Gram-negative, non-motile, encapsulated, facultative, glucose-fermenting, intracellular diplobacillus is hosted by humans alone. No racial predilection exists, but females are slightly more affected than males (about 1.3 to one) and patients are commonly affected in the second and third decades of life [3].

Although the infectious agent has been well characterized, the mechanism of infection and the pathophysiology of disease progression are poorly understood. A mean of contamination could be via direct inhalation or inoculation by respiratory droplets, but only after prolonged contact. Rhinoscleroma is

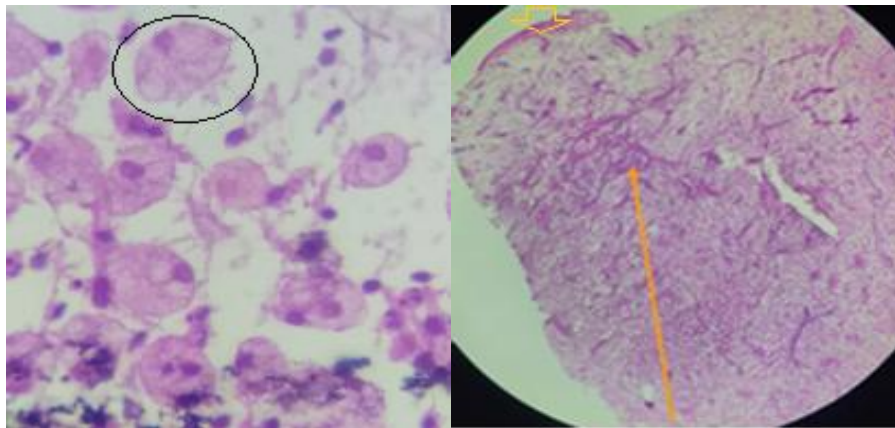
more common in developing countries and rural areas and is endemic in Africa, South-East Asia, Mexico, Central and South America, and Central and Eastern Europe [1, 3]. There appears to be an association between poor hygiene, poor nutrition, crowded living conditions and the development of rhinoscleroma.

Infection and subsequent mass lesions occur in areas of transition between squamous and ciliated respiratory epithelium, such as the nasal vestibule and the subglottic area. Iron deficiency has been proposed as a risk factor for rhinoscleroma and may contribute to this site predilection by altering epithelial regeneration and causing squamous metaplasia [4]. This may explain the association of rhinoscleroma with poor nutrition, and also why menstruating and pregnant women have been anecdotally observed to have a more severe course than other rhinoscleroma patients [4].

Classically, there are three clinical and histological stages of rhinoscleroma: catarrhal-atrophic, granulomatous and sclerotic. The catarrhal-atrophic stage may last weeks to months and may begin with a non-specific rhinorrhea. This may evolve to a foul-smelling, purulent rhinorrhea with crusting and nasal obstruction. Patients with this presentation are often treated for recurrent sinusitis. Histologically, squamous metaplasia of the epithelium is seen, with an underlying infiltrate of polymorphonuclear cells and some granulation tissue.

The granulomatous or hypertrophic stage is usually the point at which the clinical and histological findings are most easily recognized. Patients present with bluish-red or polypoid, anterior nasal tumor which are non-tender, rubbery and may be prone to bleed. This tumor most often affect the antero-inferior septum, but sometimes the maxillary antrum may be involved and act as a reservoir for infection, that was the case of our patient and the reason why we associated a Caldwell-

Luc approach during surgery. The destructive process of the disease may leave the patient with anosmia, anaesthesia of the soft palate, enlargement of the uvula, dysphonia or various degrees of airway obstruction. The soft palate may appear thickened in its junction with the hard palate. Histologically, there is pseudoepitheliomatous hyperplasia, Mikulicz cells and evidence of chronic inflammation with many monocytes, lymphocytes and macrophages [5]. Russell bodies are eosinophilic structures within the cytoplasm of plasma cells and are found characteristically during the granulomatous stage (Figure-3). In addition, sometimes the inflammatory infiltrate is found to be angiocentric and causes a vasculitis-like picture, with hyalinization of the vessels [5].



**Fig-3: Histological appearance showing Mikulicz cells (circle) and tumor-like formations with inflammation and hypervascularization. (Short arrow: Epithelial coating; Long arrow: Inflammatory mass)**

Other granulomatous, neoplastic and infectious lesions must also be considered in the differential diagnosis. These include sarcoidosis, Wegener's granulomatosis, vasculitis, lymphoma, actinomycosis, leishmaniasis, leprosy, tuberculosis, sporotrichosis, syphilis, rhinosporidiosis, nasopalatine duct cyst and Rosai–Dorfman disease.

Establishing the diagnosis is often challenging. When rhinoscleroma is suspected, a brush biopsy specimen of the nasal or respiratory tract mass or an incisional biopsy of an easily accessible lesion should be sent for cytology and culture. A positive culture of *Klebsiella rhinoscleromatis* on blood or on a specific medium confirms the diagnosis of rhinoscleroma, but only in 50–60% [1]. The histology changes according to the stage of the disease but characteristically is marked by subepithelial Mikulicz cells and transformed plasma cells with Russell bodies [5]. These pathologic findings, along with pseudoepitheliomatous hyperplasia, are usually present in the hypertrophic or granulomatous stage. An immunoperoxidase technique for the *Klebsiella rhinoscleromatis* capsular antigen has been shown to increase the specificity of histological findings in culture-negative cases [1, 5].

Imaging studies are somewhat helpful in determining the extent of disease but should not be

Finally, in the sclerotic stage, former masses are replaced by extensive scarring, deformity and stenosis. Few Mikulicz cells or Russell bodies are found during this stage.

Patients may present at any of these three stages and with a host of non-specific complaints, including: nasal obstruction, rhinorrhea, epistaxis, dysphagia, nasal deformity, anaesthesia of the soft palate, difficulty breathing or stridor, dysphonia, and anosmia. On examination, the diagnosis of rhinoscleroma should be considered if the patient comes from an endemic area and has nasal lesions involving the nasal septum with relative sparing of the sinuses.

relied upon for diagnosis. On computed tomographic scans, rhinoscleroma appears as a homogenous, non-enhancing mass with distinct margins. Occasional bony or cartilaginous erosion may be seen, but adjacent fascial planes are not usually invaded. Magnetic resonance imaging may show masses obstructing the osteomeatal complexes and may also show high signal intensity on T1- and T2-weighted images in the hypertrophic stage.

Rhinoscleroma remains a difficult entity to cure, and relapses are common [6]. Currently, the recommended treatment of rhinoscleroma consists of a combination of surgical debridement and long-term antibiotic therapy. Treatment is also stage-dependent. Streptomycin was initially widely accepted as the drug of choice for treatment of rhinoscleroma. Tetracycline soon became the preferred antibiotic, with its potential for oral administration and avoidance of the vestibulotoxic side effects of streptomycin. However, the need for a prolonged course of therapy resulted in poor compliance. The use of tetracycline was further limited by its contraindication in children and pregnant women. Ciprofloxacin, trimethoprim–sulphamethoxazole, topical and systemic rifampin, and topical acriflavine have also been used with some success. A shorter course (four to 12 weeks) of

ciprofloxacin 500 mg twice daily, along with nasal lavage twice daily, has been stated in a number of case reports to be an effective and affordable option [6, 7]. None of these case reports offer a suggested duration of treatment, but they all continued treatment for at least a month after clinical resolution had been documented. Despite its high overall cost, ciprofloxacin for four weeks was found to be more cost-effective and clinically efficacious than the standard six-month combination of rifampin and co-trimoxazole normally used at one institution in an endemic area [6]. Increased compliance, due to shorter duration of treatment, twice-daily dosing and a low side-effect profile, makes ciprofloxacin an appealing drug of choice. Moreover, ciprofloxacin achieves superior tissue penetration and is concentrated within macrophages [8]. In one *in vitro* study, ciprofloxacin was found to have the greatest bacteriocidal activity against *K rhinoscleromatis* of any agent, when compared with streptomycin, tetracycline, rifampin, trimethoprim-sulfamethoxazole and broad spectrum cephalosporins [9]. Trimethoprim sulphamethoxazole remains a good, inexpensive alternative in developing countries.

There are several indications for surgery in rhinoscleroma, including relief of airway obstruction and reconstruction of cicatricial defects. Bronchoscopy can be used for small, early lesions in the lower airway. Tracheostomy should be considered for laryngeal obstruction in either the granulomatous or sclerotic stages. Reconstructive surgery is needed for nasal or lower airway stenosis from scarring or imperforation. For tracheal lesions, the laryngofissure approach is recommended for patients without evidence of subglottic stenosis. Carbon dioxide laser is currently the most effective surgical method for eradication of clinical and histological disease. In one study, patients at the sclerotic stage fared better with CO<sub>2</sub> laser resection than did those with granulomatous disease, as their tissue involvement was often more limited and easier to resect completely [10]. However, both subsets showed a favorable initial clinical response, and only at long-term follow up (18 months) did about 50 per cent of the granulomatous patients show histological recurrence. Uvulopalatopharyngoplasty is recommended to address palatal scarring and thickening.

Recurrence is common in patients with rhinoscleroma, and they should be followed regularly with nasal endoscopy and nasal cytology. In addition, a

prolonged course of antibiotic therapy, lasting weeks to months, may stave off recurrences.

## CONCLUSION

Rhinoscleroma remains seldom in general. It is common in areas with a dry, semi-desert climate and in disadvantaged social classes. It is systematically sought before any centropacial granuloma. Early diagnosis, appropriate treatment and rigorous and prolonged surveillance are necessary to eradicate this infectious disease and to avoid recurrences and sequelae.

## REFERENCES

1. Malowany MS, Chester B, Allerhand J. Isolation and microbiologic differentiation of *Klebsiella rhinoscleromatis* and *Klebsiella ozaenae* in cases of chronic rhinitis. *Am J Clin Pathol.* 1972; 58:550—3.
2. Evrard I, Gruyer X, Desse P, Francois A, Marie JP, Dehesdin D, et al. Spheno-ethmoidal rhinoscleroma. Report of a case and review of the literature. *Ann Otolaryngol Chir Cervicofac.* 1998; 115:85—8.
3. Shum TK, Whitaker CW, Meyer PR. Clinical update on rhinoscleroma. *Laryngoscope.* 1982; 92:1149—53.
4. Akhnoukh S, Saad EF. Iron-deficiency in atrophic rhinitis and scleroma. *Indian J Med Res.* 1987; 85:576—9.
5. Meyer PR, Shum TK, Becker TS, Taylor CR. Scleroma (rhinoscleroma): a histologic immunohistochemical study with bacteriologic correlates. *Arch Pathol Lab Med.* 1983; 107:377—83.
6. Borestein J. Ciprofloxacin for rhinoscleroma and ozena. *Lancet.* 1993; 342:122.
7. Badia L, Lund V. A case of rhinoscleroma treated with ciprofloxacin. *J Laryngol Otol.* 2001; 115:220—2.
8. van den Broek PJ. Antimicrobial drugs, microorganisms and phagocytes. *Rev Infec Dis.* 1989; 11:213—45.
9. Perkins BA, Hamill RJ, Musher DM, O'Hara C. *In vitro* activities of streptomycin and oral antimicrobial agents against clinical isolates of *Klebsiella rhinoscleromatis*. *Antimicrob Agents Chemother.* 1992; 36:1785—7.
10. Maher AI, el-Kashlan HK, Soliman Y, Galal R. Rhinoscleroma: management by carbon dioxide surgical laser. *Laryngoscope.* 1990; 100:783—8.