



# RHINOSCLEROMA AS A DIFFERENTIAL DIAGNOSIS OF MUCORMYCOSIS: A CASE REPORT

## RINOESCLEROMA COMO DIAGNÓSTICO DIFERENCIAL DE MUCORMICOSIS: REPORTE DE CASO

Julio Cesar Layseca-Ortiz <sup>1,a</sup>, Juan Manuel Combe-Fernandez <sup>1,b</sup>, Angela Gabriela Vasquez-Leon <sup>1,c</sup>,  
Jorge Roberto Mosqueira-Sanchez <sup>2,b</sup>

### ABSTRACT

**Introduction:** Rhinoscleroma is a chronic granulomatous disease, rare in our country, but endemic in some areas around the world such as North Africa, Middle East and Central America. It is most caused by *Klebsiella rhinoscleromatis*, mainly affecting the nasal cavity and nasopharynx. The clinical presentation primarily involves the upper respiratory tract. Symptoms such as fetid rhinorrhea, nasal obstruction, nasal deformity, and destruction of nasal cartilage occur months after the infection and depend on the stage of the disease. The diagnosis is based on histopathological features, with the presence of Mikulicz cells. Antibiotics are the cornerstone of treatment, using them for a prolonged period: 2 to 3 months. In cases of airway obstruction, surgical intervention would be required. We present the case of a 60-year-old woman with a diagnosis of type 2 diabetes mellitus and hypertension, who arrives at the hospital with a pathological anatomy report showing thin and septate hyphae, suggesting a mucormycosis infection. A subsequent biopsy performed at the hospital ruled out mucormycosis and confirmed the diagnosis of rhinoscleroma.

**Keywords:** Rhinoscleroma; mucor; mucormycosis; differential diagnosis. (Source: MESH-NLM)

### RESUMEN

**Introducción:** Rinoscleroma es una enfermedad granulomatosa crónica, rara en nuestro país, pero endémica en algunas áreas alrededor del mundo como el norte de África, América central y Oriente Medio. En la mayoría de los casos es causada por *Klebsiella rhinoscleromatis*, afectando principalmente la cavidad nasal y nasofaringe. La presentación clínica comprende comúnmente el tracto respiratorio superior. Síntomas como rinorrea fétida, obstrucción nasal, deformidad nasal, y destrucción del cartílago nasal ocurren meses después de la infección y depende del estadio de la enfermedad. El diagnóstico se basa en las características histopatológicas donde observamos las células de Mikulicz. Los antibióticos son la piedra angular del tratamiento los cuales se usan por tiempo prolongado (2 a 3 meses). En casos de obstrucción de la vía aérea, puede requerirse una intervención quirúrgica. Presentamos el caso de una mujer de 60 años con diagnóstico de diabetes mellitus tipo 2 e hipertensión arterial, que llega al hospital con un informe de anatomía patológica que muestra hifas delgadas y septadas, por lo que se sugiere infección por mucor. La biopsia hospitalaria posterior descartó mucormicosis y confirmó el diagnóstico de rinoescleroma.

**Palabras clave:** Rinoescleroma; Mucor; Mucormicosis; Diagnóstico diferencial. (Fuente: DeCS- BIREME)

<sup>1</sup> Hospital María Auxiliadora. Lima, Peru.

<sup>2</sup> Department of Internal Medicine, Clínica San Pablo. Lima, Peru.

<sup>a</sup> Internal Medicine Resident

<sup>b</sup> Internist

<sup>c</sup> Intern



Cite as: Layseca-Ortiz JC, Combe-Fernandez JM, Vasquez-Leon AG, Mosqueira-Sanchez JR. Rhinoscleroma as a differential diagnosis of mucormycosis: A case report. Rev Fac Med Hum. 2025;25(1):149-153. [doi:10.25176/RFMH.v25i1.6436](https://doi.org/10.25176/RFMH.v25i1.6436)

Journal home page: <http://revistas.urp.edu.pe/index.php/RFMH>

Article published by the Journal of the Faculty of Human Medicine of the Ricardo Palma University. It is an open access article, distributed under the terms of the Creative Commons License: Creative Commons Attribution 4.0 International, CC BY 4.0 (<https://creativecommons.org/licenses/by/4.0/>), which allows non-commercial use, distribution and reproduction in any medium, provided that the original work is duly cited. For commercial use, please contact [revista.medicina@urp.edu.pe](mailto:revista.medicina@urp.edu.pe)





## INTRODUCTION

Rhinoscleroma (RS) is a chronic granulomatous infectious disease of the upper airways caused by *Klebsiella rhinoscleromatis*. This bacterium, frequently, affects the nasopharynx<sup>(1)</sup>, but also the paranasal sinuses, orbit, and larynx, leading to the deformation of the nasal tracheal and bronchial mucosa (nodular type) developing tracheal stenosis<sup>(2)</sup>.

RS is endemic to Central America, Egypt, Central Africa, India, Middle Eastern countries and Indonesia<sup>(1)</sup>. There are several predisposing factors such as low socioeconomic level, malnutrition, poor hygiene, and iron deficiency anemia which facilitate the infection process<sup>(3)</sup>. It is transmitted by direct or indirect contact with nasal exudate from an infected person<sup>(2)</sup>. It is more predisposing in women with a 13:1 ratio and occurs approximately in the middle-aged population<sup>(3)</sup>. The incidence and prevalence of RS is low; however, more than 16 000 cases have been reported worldwide since 1960<sup>(4)</sup>.

The diagnosis of RS is based on clinical, imaging, histopathological and microbial findings<sup>(1)</sup>. Clinically and pathologically, there are 3 phases: The catarrhal/exudative phase causes non-specific early symptoms that can last for weeks to months with fetid and purulent rhinorrhea<sup>(1,3)</sup>. Histologically, squamous metaplasia with polymorphonuclear infiltrate, hyperkeratosis and atrophy are usually observed<sup>(3,5)</sup>. The granulomatous/hypertrophic phase lasts months to years and is characterized by the presence of masses obstructing the nasal passages, epistaxis, deformity and destruction of the nasal septum ("stringy" nose)<sup>(1,5)</sup>. Also, in this phase the biopsy is usually diagnostic, because Mikulicz cells are identified due to the presence of large foamy cells surrounded by plasma cell infiltrates with lymphocytes, neutrophils, and Russel bodies<sup>(1)</sup>. In the third and last sclerotic/cicatrical phase, inflammatory cells, extensive fibrosis formation and adhesions leading to nasal stenosis are observed; few or no Mikulicz cells are seen in this phase<sup>(1,3)</sup>.

Radiological imaging is essential for the early diagnosis of RS, therefore computed tomography (CT) and/or magnetic resonance imaging (MRI) is recommended to evaluate the lesions. In CT, lesions are homogeneous, hyperdense without enhancement<sup>(6)</sup>. In MRI, we can identify in the second phase the Mikulicz cells and Russel bodies as hyperintense areas in T2 and T3 sequences. In the fibrotic phase, hypointense areas can be observed as a heterogeneous pattern suggesting fibrosis in the T2 sequence<sup>(6)</sup>.

RS is effectively treated with antibiotics, debridement and surgical repair<sup>(1)</sup>. The duration of treatment is usually prolonged, approximately 2 to 3 months or until cultures and pathological examination are negative<sup>(1,7)</sup>. Ciprofloxacin is recommended as the first line of treatment, but other drugs such as rifampicin, trimethoprim sulfamethoxazole, ciprofloxacin and doxycycline can also be used as dual therapy depending on the bacterial susceptibility test of *K. rhinoscleromatis* or therapeutic response<sup>(1,5)</sup>.

The present case aims to report to the medical community a rare case of rhinoscleroma, so that it can be considered as a differential diagnosis in some cases of atypical presentation of mucormycosis. We consider that it will be of great importance to have this case as a background, especially for otorhinolaryngologists physicians, head neck and maxillofacial surgeons and infectious disease specialists.

## CLINICAL CASE

A 60-year-old female patient diagnosed with diabetes mellitus type 2 and long-standing hypertension was admitted to the emergency department with left retroocular headache and gaze deviation for 5 months (Fig. 1). She reports that the clinical picture began with constant and persistent rhinorrhea so she goes to the medical center where a brain and turbinates tomography is performed, and the report describes sinusitis for which treatment is prescribed.



The patient reported that her symptoms persisted and worsened, prompting her to visit the Instituto Nacional de Ciencias Neurológicas of Peru, where a new tomography study was requested, which described "expansive lesion of neoformative aspect locally advanced with epicenter in the pterygo maxillary fossa with invasion of the maxillary sinus and lateral extension to the middle and upper fossa towards the orbital cavity and discreetly to the cavernous sinus".

Due to this result, she was referred to the Instituto Nacional de Enfermedades Neoplásicas of Peru where a biopsy of the lesion was performed and the result was "nasal mucosa infected by thin and septate hyphae associated with necrosis, suggestive of infection by mucor". After this result, the patient was referred to the internal medicine service of the Hospital María Auxiliadora, where medical treatment of

mucormycosis was started with amphotericin b deoxycholate at a dose of 1mg/kg/day; however, the patient only received 3 doses of this drug since in coordination with the head and neck surgery service it was suggested that it was not a mucor infection since the patient had been ill for approximately 5 months.

The patient was admitted to the operating room for the removal of the lesion. During the surgical procedure, the macroscopic characteristics of mucor infection were not found, so we had to wait for the result of the microscopic study of the tumor that finally revealed the diagnosis rhinoscleroma. Treatment was started using ciprofloxacin at a dose of 750 mg po every 24 hours. The patient improved and was discharged 7 days after starting antibiotic treatment.

Informed consent was obtained from the patient.



**Figure 1.** Brownish, crusted lesions on the dorsal nasal wall, with slight periorbital erythema and increased volume in the left mid-facial third, as well as deformity in the nasal architecture.

## DISCUSSION

Rhinoscleroma was first described in 1870 by Von Hebra; then, in 1882, Von Frisch saw the possible association with a microbial agent. Finally, Mikulicz demonstrated the foam cell's presence in these tissues (Mikulicz cells)<sup>(8)</sup>. Rhinoscleroma is an endemic disease with high prevalence in Africa, Asia and Latin America, but many cases have been recently reported in other

areas such as Central Europe<sup>(9,10)</sup>. Rhinoscleroma occurs predominantly within middle-aged low-income women with poor nutritional and hygienic conditions<sup>(9,11)</sup>. This entity is associated with *Klebsiella rhinoscleromatis*, an encapsulated, gram-negative bacilli which has great affinity for nasal mucosas and spreads by contact with infected patient's nasal



exudates<sup>(8,12)</sup>. Humans are the only known hosts, and the nasal cavity is the most affected site (95%); then, nasopharyngeal (18-43%), larynx (15-40%), trachea (12%) and bronchi (2-7%)<sup>(9)</sup>. Rhinoscleroma progresses through three stages: catarrhal, granulomatous, and sclerotic<sup>(10,11)</sup>.

The mainstay of the diagnosis is histopathology, where we can find lymphocyte infiltration, plasma cells and the presence of Mikulicz cells which are a pathognomonic clue to rhinoscleroma. All these changes are usually seen at the granulomatous stage. At this point, intracytoplasmic bacilli can be demonstrated by special stains such as Gram, PAS, Giemsa and others. Also, in granulomatous inflammation, bacilli are present in 50% of tissues cultures<sup>(8,12)</sup>.

The recommended imaging techniques are MRI and CT scan where characteristic lesions are homogeneous, hyperdense and non-enhancing masses with well-defined borders<sup>(10,12)</sup>. In context of granulomatous diseases, the main differential diagnosis to be ruled out is tuberculosis, especially in our country. Indeed, others like Actinomycosis, leprosy, fungal infection, mucocutaneous leishmaniasis, or even inflammatory diseases and lymphomas should also be considered<sup>(9,11)</sup>. In the context of fungal infections, it is important to consider mucormycosis as a differential diagnosis because this disease, is currently the third cause of invasive fungal infection after candidiasis and aspergillosis. The classic clinical presentation with rhinosinus involvement is linked to diabetic ketoacidosis. The distribution of clinical forms according to frequency is as follows: sinus 39%, pulmonary 24%, cutaneous 19%. The prerequisites for diagnosis are a high index of suspicion, recognition of risk factors and rapid assessment of clinical symptoms.

A necrotic ulcer with blackish eschar (cutaneous or mucosal) is the characteristic lesion of mucormycosis and should be a red flag in all patients with risk factors,

but the definitive diagnosis is made by biopsy culture<sup>(8,9)</sup>. In our case, we saw a patient with a mucocutaneous lesion with characteristics of soft tissue destruction according to tomography studies; in addition, a necrotic lesion was found in biopsy with characteristics suggestive of mucormycosis; however, the time of clinical presentation did not typically correspond to this fungal infection which in more than 90% of cases are of rapid progression (days)<sup>(10,12)</sup>.

Mucormycosis was ruled out by the final microscopic study of the lesion, and the diagnosis of rhinoscleroma was confirmed<sup>(9,11)</sup>. Treatment strategies for rhinoscleroma include long term combinations of antibiotics often requiring dual or triple therapies. Examples include rifampin and trimethoprim, sulfamethoxazole and ciprofloxacin, ciprofloxacin and cotrimoxazole, and ciprofloxacin and doxycycline with duration of therapy lasting at least 2 months. The surgical treatment is reserved for obstructive lesions<sup>(9,12)</sup>. Because of the high recurrence, early proper diagnosis and treatment are essential for the patient's outcome<sup>(8-12)</sup>.

## CONCLUSION

We believe that clinical evaluation and diagnosis are currently undervalued, with more emphasis placed on complementary examinations, even when clinical features suggest otherwise. As we present in this case, both the imaging studies and the first pathological anatomy indicated a diagnosis of mucor infection; however, the clinical picture was not compatible, so the diagnosis was reconsidered. The new approach established a new entity which was confirmed with the result of the surgical specimen and, from this, the specific treatment was indicated.

Therefore, it is essential to prioritize the importance of the clinical presentation over diagnostic support studies. Indeed, it is imperative to consider a broad of differential diagnoses in immunosuppressed patients with granulomatous lesions. This case will serve to recognize and diagnose similar cases.





**Author contributions:** JRMS: Conceptualization, investigation, formal analysis, and writing – original draft. JMCF: Conceptualization, methodology, formal analysis, and writing – review & editing. AGVL: Investigation, data curation, writing – original draft, and writing – review & editing. JCLO: Methodology, supervision, and writing – review & editing. All authors approved the final version to be published.

**Conflict of interest statement:** The authors declare no conflicts of interest.

**Received:** March 15, 2024.

**Approved:** September 19, 2024.

**Funding:** Self-funded.

**Correspondence:** Jorge Roberto Mosqueira-Sanchez.

**Email:** [jorge.mosqueira@urp.edu.pe](mailto:jorge.mosqueira@urp.edu.pe)

## REFERENCES

1. Umphress B, Raparia K. Rhinoscleroma. Arch Pathol Lab Med. 2018;142(12):1533-1536. doi: [10.5858/arpa.2018-0073-RA](https://doi.org/10.5858/arpa.2018-0073-RA)
2. Malkud S, Mahajan P. Rhinoscleroma: an usual presentation. Indian Dermatol Online J. 2018;9(3):191-193. doi: [10.4103/idoj.IDOJ\\_134\\_17](https://doi.org/10.4103/idoj.IDOJ_134_17)
3. Fanconi M, Greco A, Cattaneo CG, et al. Social geography of rhinoscleroma and qualitatively and quantitatively abnormal cell-mediated immunity. Infect Genet Evol. 2018;62:17-19. doi: [10.1016/j.meegid.2018.03.018](https://doi.org/10.1016/j.meegid.2018.03.018)
4. Peerani R, Shah M, Minnema B, et al. Long-standing laryngeal rhinoscleroma with rare Mikulicz cells. Clin Case Rep. 2023;11(6):e7490. doi: [10.1002/ccr3.7490](https://doi.org/10.1002/ccr3.7490)
5. Mandale MS, Bhavthankar JD, Humbe JG, et al. Rhinoscleroma: report of an erratic palatal swelling. Contemp Clin Dent. 2018;9(Suppl 2):S365-S368. doi: [10.4103/ccd.ccd\\_282\\_18](https://doi.org/10.4103/ccd.ccd_282_18)
6. Ibrahim D, Fayed A. Report of a case of giant rhinoscleroma: CT and MRI. BJR Case Rep. 2018;4(4):20180027. doi: [10.1259/bjrcr.20180027](https://doi.org/10.1259/bjrcr.20180027)
7. Bazzout A, Lachkar A, Benfadil D, et al. Rebellious headache revealing an extensive rhinoscleroma: a case report and review of the literature. Ann Med Surg (Lond). 2021;63:102166. doi: [10.1016/j.amsu.2021.02.012](https://doi.org/10.1016/j.amsu.2021.02.012)
8. Ortega JR, Velasco FJ, Fornes S. Rinoescleroma. Una entidad a considerar en la rutina diaria del patólogo. Rev Esp Patol. 2016;49(1):32-36. doi: [10.1016/j.patol.2015.06.004](https://doi.org/10.1016/j.patol.2015.06.004)
9. Ndiaye M, Diouf MS, Ndiaye C, et al. Rhinoscleroma: report of 2 cases and literature review. Rhinol Online. 2019;2:115-118. doi: [10.4193/RHINOL/19.032](https://doi.org/10.4193/RHINOL/19.032)
10. Bazzout A, Lachkar A, Benfadil D, et al. Rebellious headache revealing an extensive rhinoscleroma: a case report and review of the literature. Ann Med Surg (Lond). 2021;63:102166. doi: [10.1016/j.amsu.2021.02.012](https://doi.org/10.1016/j.amsu.2021.02.012)
11. Ortega JR, Velasco FJ, Fornes S. Rinoescleroma. Una entidad a considerar en la rutina diaria del patólogo. Rev Esp Patol. 2016;49(1):32-36. doi: [10.1016/j.patol.2015.06.004](https://doi.org/10.1016/j.patol.2015.06.004)
12. Mariz BALA, Sanchez-Romero C, Romañach MJ, et al. Respiratory scleroma: a clinicopathologic study of 51 cases from Guatemala. Oral Dis. 2020;26(3):670-676. doi: [10.1111/odi.13264](https://doi.org/10.1111/odi.13264)