

Caso clínico

Rhinoscleroma

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Abstract

The case of a 19-year-old male patient from a rural area is presented. He had a 3-year history of nasal obstruction, episodes of facial inflammation, epistaxis, fetid rhinorrhea, hearing loss in the right ear, bilateral axillary and multiple facial bilateral lymphadenopathies. He was referred for consultation to the Mexico Hospital due to an obstructive granulomatous lesion of the nasal septum. The first biopsy revealed the presence of chronic inflammatory infiltrate, with predominance of macrophages with foamy appearance.

A second biopsy was positive for *Klebsiellapneumoniae* subsp. *rhinoscleromatis*, described histologically as a pseudoepitheliomatous hyperplasia with dense inflammatory infiltrate and the analysis of lamina propria showed dense inflammatory infiltrate with lymphocytes, plasmatic cells, Russell bodies and macrophages with vacuolated cytoplasm, with microorganisms and debris. The patient was treated with oral ciprofloxacin therapy for seven months, after which the patient was considered cured from the etiological point of view.

Keywords: *Klebsiella*, chronic granulomatose disease, biopsy.

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Since Friedländer in 1883 demonstrated for the first time the presence of encapsulated bacilli in the lungs of some patients who died of Pneumonia, *Klebsiella* became known like a causal agent of "Friedländer Pneumonia"; this genus of Enterobacteriaceae¹ is characterized by producing of large amounts of capsular polysaccharides in vivo and in

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solid culture media, in addition to other virulence factors such as: fimbriae, siderophores and O antigen. Efforts have been realized to understand the mechanisms of pathogenicity of *Klebsiella*, whose determinants are mainly found in capsular antigens.²

K. pneumoniae and *K. oxytoca* are the two mainly species of this genus that cause disease in humans, have also been isolated from clinical samples: *K. pneumoniae* subsp. *ozeanae*, *K. pneumoniae* subsp. *rhinoscleromatis*, *K. terrigena*, *K. planticola*, *K. ornithinolytica* and *Enterobacter aerogenes*, which is renamed like *K. mobilis*.

Masculine patient, 19 years old, from Guanacaste, referred to the Otorhinolaryngology Department (ORL) of Mexico Hospital, by a severe granulomatous obstructive lesion of the nasal septum, apparent inflammation of crystalloids bodies, *Leishmania* sp and fungi smears were negative; with a biopsy report realized in February of 2008, which stated as clinical history of a nasal granulomatous lesion reminiscent *Leishmania* sp. That report indicates the presence of a chronic inflammatory infiltrative tissue, predominantly foamy macrophages and many particles of crystalloid aspect; without evidence of multinucleated giant cells, or compatible structures of *Leishmania* sp.

The non-pathologic patient's record indicates that works as fencing operator and smokes 5 cigarettes per day average, occasional elitism, no medical allergies; the pathologic record referred a nasal pain since 3 years ago, facial inflammation episodes, epistaxis, fetid rhinorrhea and right low hearing.

In March, 2009 it was detected multiple lymphadenopathies in the face and in both armpits, as also granulomatous injury with both nostrils occlusion (to the lobe of the right ear). It was declared under observation of a lymphoproliferative disease, with a biopsy programming; three weeks later, in ORL session, a study established for possible deep mycosis versus *Klebsiellapneumoniae* subsp. *Rhinoscleromatis* infection. Finally, they took a biopsy of the nasal lesions, which was processed by Pathology and Microbiology division in Mexico Hospital.

The laboratory received three fragments of brown fibrous tissue, with a volume average of 1 cm³, which were macerated in aseptic conditions and were sown in Columbia base agar, with sheep blood at 5%, chocolate agar, McConkey agar and tioglicolate broth. The media were incubated at

35°C for 24 hours (the first two in CO₂-enriched atmosphere). The macerated Gram stain did not show microorganisms.

The post incubation showed cream colonies growing, round, with entire border and mucoid aspect in solid media, except in the McConkey Agar (where only grew three lactose fermenting colonies after 24 hours incubation); Gram Taint was realized with Gram negative bacilli, negative oxidase, the identification and antibiotic sensitivity test was scheduled according to Vitek 2 methodology (System Version 04.01); 24 hours later was identified with a probability of 99% of microorganism like *Klebsiellapneumoniae* subsp. *rhinoscleromatis*, all antibiotic sensitive.

The bacteriological diagnosis was confirmed a month later, by pathologic anatomy, whose biopsy report described pseudoepitheliomatous hyperplasia tissue, and dense lymphocytic inflammatory infiltration in lamina propria, plasmatic cells, Russel Bodies and vacuolated cytoplasm macrophages, with microorganisms and detritus presence. These histopathological changes are characteristic of Rhinoscleroma, with Gram stain was observed Gram negative bacilli with foamy macrophages; with special stain, and there was no evidence of mycobacteria or fungi.

Seven weeks after the biopsy remained initial physical signs and a CT scan showed the absence of maxillary sinuses lesions, that's why Ciprofloxacin 500mg two times a day oral via per three weeks was initiated. Then in that time, ORL documented rhinoscleroma diagnosis based on the bacteriological test. The patient confirmed feeling better, with left nasal nostril patency (LNN), right nasal nostril (RNN) clogged and with crusted and the dosage of the initial treatment lasted two more months.

After this period the patient reported an overall improvement in LNN, but rhinorrhea of the RNN, remained blocked. Treatment continued for four more months, with total rhinorrhea resolution and permanent obstruction of RNN.

In January, 2009, the patient claimed to have any trouble, so that schedule was judged to scar tissue surgical removal present in RNN. In September 2009, as an etiological standpoint, Infectious Diseases Department discharged him.

Discussion

Among the specific pathogen associated *Klebsiella* infections (rare), are the rhinoscleroma and ozena; both diseases presents specific anato pathologic changes. Rhinoscleroma is a chronic granulomatous infection located in high respiratory tract, primarily in the nasopharynx. This

infection is caused by *K pneumoniae* subsp. *Rhinoscleromatis* with is cosmopolitan, but endemic in some Oriental Europe zones, Latin America, Central Africa and South Asia and affects predominantly women. The rhinoscleroma is acquired by direct or indirect contact with nasal discharged from a infected person and may affect bronchi, causing hoarseness, dyspnea, stridor, and productive cough with laryngeal tracheal engagement. The principally warning effect is the obstruction of air flow, which may need endoscopic treatment, thus progresses through three stages: a catarrhal phase (with non-specific inflammation), a proliferative (with one granulomatous aspect reaction) and a phase with the formation of scar tissue in reparation. (See Table 1 for an overview). Histologically, it is characterized by the vacuolated foamy macrophages presence, with bacilli known as Mikulicz cells (see Figure 1); the determination factors that lead their formation are unknown. The auto phagocytosis can contribute to the fagosoma distention and the consequent membrane vacuole destruction, which does not allow adequate bacilli elimination and cause liberation into the interstitium and implicates an immunologic macrophages deprotection.^{8,11}

Ozena is a atrophic chronic rhinitis illness, characterized by nasal mucosal necrosis and, mucus purulent dischargers, however, the *K. pneumoniae* subsp. *ozenae* isolation in this study, indicates the disease, it appears to be endemic in subtropical and temperate regions like South Asia, Africa, Oriental Europe and the Mediterranean, and has been associated to patients that lives in poorly conditions and deficient hygiene.¹²

Because *K. pneumoniae* subsp. *Rhinoscleromatis* is not normally found in nasal secretions, the culture of this type of sample is diagnostic. It has been routinely observed in routine practice that using McConkey Agar media, this microorganism is recoverable only in a 50-60% of the cases.¹¹

The de *K. pneumoniae* subsp. *Rhinoscleromatis* pathogenicity has been attributed to the composition of capsular polysaccharides, serotype K3 specific, which protects the microorganism from phagocytosis. The contact of illness patients with healthy persons do not transmit de disease in a direct way, which suggest the host susceptibility plays an important role in the development of the disease. In the injury sites has been documented, a CD4+/CD8+ lymphocytic relation altered, with a decrease of CD4+ and a CD8+ increased, which possibility induces to alter or diminish Lymphocytes T response.

The Rhinoscleroma Treatment suggests Tetracyclines and Quinolones use for a six month period or until the nasal biopsy culture were negative;⁸ the Ciprofloxacin and rifampicin combination is interesting for the concentrations achieved within macrophages and nasal secretions.^{8,11}

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