Rhinoscleroma: A Retrospective Study
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ABSTRACT
Introduction: Rhinoscleroma is an infectious granulomatous disease of the upper respiratory tract that affects mainly the nasal mucosa. The disease was first described by Von Hebra in 1870.1 The causative organism is Klebsiella rhinoscleromatis, a gram negative encapsulated bacillus subsp. of Klebsiella pneumoniae.2,3 Scleroma is endemic in Egypt, Eastern Europe, Southeast Asia, Central and South America, and Mexico, in other countries is related to migration.2,4,5

The involvement sites are nasal mucosa (95%-100%), nasopharynx (18%-43%), larynx (15%-40%), Eustachian tube (30%), less frequently, sinosuses (26%), mouth, orbit, trachea (12%) and bronchi (2%-7%).6

There are 3 stages of the disease:
1. Catarrhal or Atrophic: may last weeks to months, characterized by purulent rhinorrhea, nasal congestion and crusts. Histologically, epithelial squamous metaplasia and granulation tissue are seen.3,5
2. Granulomatous or Proliferative: may last from months to years, with painless granulomas in upper respiratory tract including orbit. Patients present with epistaxis, sequestration, centrofacial headache, anosmia, palate anesthesis, uvala enlargement, hoarseness and airway obstruction. Histologically, Mikulicz cells (large vacuolated histiocytes containing Klebsiella rhinoscleromatis) and Russell bodies (eosinophilic round structures within plasmatic cells) are seen.5,6,15
3. Sclerotic or Fibrotic: last for years, with stenosis of nasal cavity, nasopharynx or larynx. Histologically, few Mikulicz cells and Russell bodies are found.4,16

Specific diagnosis is made with biopsy samples by different methods. Histopathology studies with Warthin Starry stain helps in the identification of the bacteria however immunoperoxidase staining for some capsular antigens may increase accuracy.2,17 Bacteriological studies on blood or MacConkey agar are recommended, but has low sensitivity (50-60%).9 Recently, PCR tests for specific capsular serotypes are been used at a high cost.

The Objective was to evaluate the evolution, during 19 years, of patients diagnosed with Rhinoscleroma in the State of Mexico.

METHODS AND MATERIALS
We performed a retrospective study consisting of 8 cases diagnosed with scleroma during the period of January 1994 to December 2013. We reviewed these cases and analyzed their medical records. Results: The study included five women and three men; their ages ranged from 32 to 50 years, with a mean age of 43 years. Hundred percent of the patients presented bilateral affection of the nose but one had laryngeal disease as well. Biopsy was performed in all patients, but only 5 had a positive culture result (Table 1). Two patients were at catarrhal stage, five granulomatous and one sclerotic.

The most common clinical features were crusts, purulent rhinorrhea, post nasal drip and frontal headache (Figure 1).

Five out of the eight cases were women and three men, their ages ranged from 32 to 50 years, with a mean age of 43 years. Hundred percent of the patients presented bilateral affection of the nose but one had laryngeal disease as well. Biopsy was performed in all patients, but only 5 had a positive culture result (Table 1). Two patients were at catarrhal stage, five granulomatous and one sclerotic.

RESULTS

Klebsiella subsp. can be found in soil, water, fresh vegetables, wood and paper mills, in fesces of carriers however, humans are the only identified host for K. rhinoscleromatis.7 Transmission occurs via direct inhalation of respiratory droplets. Predominantly affects women in a variable ratio from 1.3:1 in European series and 13:1 in Latin-American reports, commonly in the third and fourth decades of life.1,5 However, we found a mean age of 43 years in our patients not only attributed to the delay in looking for medical attention, but also to the risk factors. Rhinoscleroma has historically been associated with low socioeconomic status, poor hygiene, and malnutrition with immunity deficiency.18 Immunocompromised patients should be treated with 404/CDR ratio, a specific phagocytosis deficit6 and DOA1*03011-DQB1*0301 haplotype7 are considered as risk factors. Immunosuppression diseases can also predispose to this entity.9 In our study, 25% of the patients suffered from diabetes mellitus. However, given the low n of the study it is not possible to establish a risk factor between these two diseases.

The clinical presentation with non-specific symptoms indicates that the differential diagnosis should include bacterial sinustisus, atrophic rhinitis, tuberculosis, syphils, carcinomas, lymphomas and rheumatologic diseases.4,14 Imaging diagnoses; using CT scan does not offer pathognomonic findings for scleroma. However, our patients presented mucosal thickening and involvement of the maxillary and ethmoid sinus as reported previously by Racek AA and Gasaf HA, et al.8,19 Treatment depends on the patient’s stage, extent of disease and individual characteristics, so it should be individualized with antibiotics, surgery or both. When the disease is localized to the nose and obstructs breathing, such as in three patients of our study, removal of granulation tissue, fibrosis or adhesions can be performed.1 Klebsiella rhinoscleromatis has a polysaccharide capsule which makes it resistant to some antibiotics.1 Treatment options are streptomycin, tetracycline trimethoprim/sulfamethoxazole, rifampin, third generation cephalosporins and fluoroquinolones. Mexican reports confirm ciprofloxacin 500mg given twice daily for 4 to 12 weeks had shown efficacy.13,20 De Pontual L, et al. and Chan TV, et al. reported ciprofloxacin reaches adequate intracellular efficacy, therefore adequate concentration within macrophages.2,3,20,21 Topical therapy offers high oral drug concentration so it may be useful to reduce the amount of secretions and microorganisms therein. Gupta A, et al. reported a good cure rate with 5% ointment acriflavin.22 Gamez A, described a decrease in the total of Mikulicz cells with rifampin instillations.21 Aminoglycoside nasal irrigation had been described for treatment of different pathologies, although guidelines don’t have enough evidence to recommend it routinely.

CONCLUSIONS
Although rhinoscleroma is endemic in our country, it represents a challenge because there are no specific manifestations, so it should be included in the differential diagnosis for patients with fetid rhinorrhea and nasal congestion. Once diagnosed, treatment should be individualized. Fluoroquinolones and third-generation cephalosporins offer a good treatment option. Local treatment with amikacin can help to control some nasal symptoms.

REFERENCES