Rhinoscleroma is a chronic, infectious, granulomatous disease that can affect the lining mucosa of the entire respiratory tract. There are three stages of the disease: catarrhal/atrophic, hypertrophic/granulomatous, and the final sclerotic phase. The diagnosis is possible by the histological findings of Mikulicz cells and plasma cells degenerated into Russell bodies and by the finding of Klebsiella rhinoscleromatis in the tissue biopsy. Treatment can be performed by specific antibiotics and by surgery in some situations. Imaging findings are not-specific, but are helpful in staging and for treatment follow-up. We present a case report with Mikulicz cells, plasma cells degenerated into Russell bodies, and endoplasmic bacilli present in the biopsy material.

Keywords: rhinoscleroma, radiology, computed tomography.
maxillary sinus mucosal thickening and a cystic lesion related to one of the roots of the tooth number 12, consistent with a periradicular cyst (Figure 2). The finding of such cystic lesion in the present case could represent a radiological indication of low level of oral hygiene, since these lesions are associated with chronic dental inflammatory processes leading to pulp necrosis. CT of the neck, performed from the base of the skull to the level of the carina, did not show additional similar-looking lesions at any other regions of the respiratory tract or enlarged cervical lymph nodes.

The lesion biopsy revealed vacuolated and distended histiocytes diffusely infiltrating the submucosa, with round and eccentrically located nuclei (Mikulicz cells), lymphocytic inflammatory infiltrate, some contained Russell bodies and mild fibrosis (Figure 3). The Mikulicz cells were positive for CD68 and negative for pan cytokeratin AE1/AE3, demonstrating the histiocytic nature of the injury. The Giemsa, Periodic Acid Schiff, and Warthin-Starry stains demonstrated the presence of endoplasmic bacilli.

After the diagnosis of rhinoscleroma was established, the patient was treated with antibiotics with complete remission, resulting in minimal residual nasal stenosis requiring no surgery due to significant clinical improvement.
DISCUSSION

Rhinoscleroma is caused by Klebsiella rhinoscleromatis, (Enterobacteriaceae group), an intracellular, gram-negative, non-motile, encapsulated, facultative and glucose-fermenting diplobacillus which causes diseases in humans.3,4

It is a chronic specific granulomatous infection, of indolent and progressive course, that predominantly affects the upper airway tract, affecting the nose in 95% to 100% of the cases. It afflicts young individuals (2nd to 3rd decades of life), without predilection for race, with a slight predominance in females and people of low socioeconomic status and of poor hygiene.5,6

Ferdinand von Hebra, an Austrian Dermatologist, first described the disease in 1870, initially described as a sarcoma.7 In 1876, Jan Mikulicz-Radecka described the histological features and the vacuolated cell, which was named after him.8 In 1882, Anton von Frisch discovered the causative agent.9 The first case published in Brazil dates from 1890 by Adolpho Lutz.10

The disease is clinically and pathologically divided into three stages. First stage, catarrhal/atrophic in which the patient has nonspecific symptoms of a common cold or rhinitis, with persistent rhinorrhea, the lining mucosa may be atrophic. There is evidence of squamous metaplasia, subepithelial infiltration of neutrophils and some granulation tissue on histology. In the second phase (hypertrophic/granulomatous), the patient may present with epistaxis, nasal deformity from enlargement of the nasal pyramid and erosion of the septal cartilage and/or the nasal bone. Histologically, there may be atrophy or hyperplasia, the last being more common, known as epitheliomatous pseudo-hyperplasia, with infiltrates of chronic inflammatory cells, monocytes, lymphocytes, and histiocytes.5,11 The Mikulicz cells are histiocytes with numerous large vacuoles containing bacteria that may or may not be viable and can be found below the basal layer.5,15 The Russell bodies are eosinophilic structures within the cytoplasm of plasma cells.2,4,5 The final or sclerotic phase is characterized by progression of stenosis and deformity of the nasal vestibule, with granulomatous areas surrounded by fibrotic tissue. At this stage, the Mikulicz cells and Russell bodies are hardly seen.12

The differential diagnosis of scleroma is extensive and may include other granulomatous lesions, fungal infections, and neoplasms, including tuberculosis, histoplasmosis, blastomycosis, Wegener’s granulomatosis, lethal mid-line granuloma, actinomycosis, paracoccidioidomycosis, tertiary syphilis, leishmaniasis, sarcoidosis, lymphoma, basal cell carcinoma, and verrucous squamous cell carcinoma.3,4,11,14

By our knowledge there are a few large series of reports establishing an imaging pattern based on CT scans. On the other hand, reports of single cases are more numerous, in which the CT scan shows mass unilateral or bilateral, with homogeneous density, specific limits, with standard variable-enhanced and destruction of bone and nasal septum.

Razek described the signal behavior of the scleroma on magnetic resonance imaging through an analysis of 15 patients. By his series, in T1 weighted images, the lesions were slightly hyperintense related to the muscles and to the cerebrospinal fluid, but less hyperintense than fat. In T2 weighted images, the lesions were homogeneously hyperintense varying to heterogeneous hyperintense with hypointense foci.6

Diagnosing rhinoscleroma is challenging and it is not always the first diagnostic hypothesis by the time of clinical examination or by imaging studies. In the clinical setting of a young patient presenting with a long standing infiltrating lesion in nasal pyramid, involving and obstructing one or both nasal vestibules, with a personal positive history of poor hygiene and coming from an endemic area, would lead to a suspicious candidate for the disease, requiring for this reason the proper work out.

The computed tomography pattern observed in the present case was in agreement with current literature. These imaging findings, although non-specific, are useful in determining the extent of the disease and in evaluating the therapeutic response. From the clinician standpoint, facing the suspicion of rhinoscleroma with signs, symptoms, and positive epidemiological data, the accurate diagnosis can be certainly made by finding the bacteria itself (which is not always possible), or by finding the specific histiocytes through specimen biopsy. The majority of the cases will exhibit a favorable response after a full course of specific antibiotic treatment.

ACKNOWLEDGEMENTS

Márcia Cabral A. T. de Cabral
Pushikin Pires Leal
Tânia Mara C. Rossi.

REFERENCES