

Rhinoscleroma

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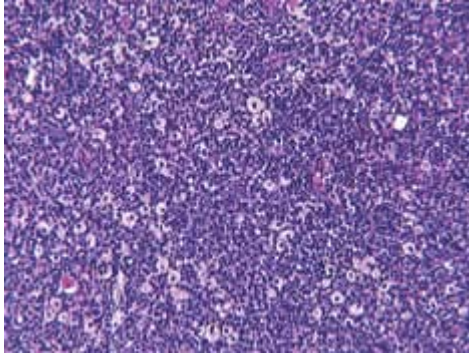


Figure 1. A sprinkling of foamy histiocytes (clear cells) is seen in a sea of inflammatory cells (blue staining nuclei of lymphocytes and plasma cells).

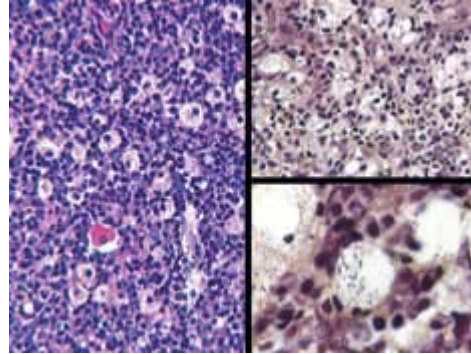


Figure 2. A: The foamy histiocytes stand out against the background plasma cells and lymphocytes. B: An intermediate-power view of Mikulicz' cells stained with Warthin-Starry silver demonstrates a few organisms. C: A higher-power view demonstrates the rod-shaped organisms in the cytoplasm of the histiocytes.

Rhinoscleroma ("hard nose") is caused by *Klebsiella rhinoscleromatis*, a gram-negative encapsulated bacterium of low infectivity. The disease is uncommon in the United States; most cases are found in the Middle East (especially Egypt), in parts of Latin America, and in Eastern Europe. The disease process usually involves the nasal cavity and the nasopharynx, but it can also involve the larynx, trachea, bronchi, middle ear, and orbit.

Rhinoscleroma often begins at a young age (first to third decade). Infection requires prolonged exposure to the pathogen. Clinically, rhinoscleroma progresses in three phases: rhinitic (characterized by mucopurulent discharge); florid or granulomatous (characterized by a hyperplastic mucosal reaction that can lead to nasal obstruction); and fibrotic (which represents sclerotic disease resolution). The diagnosis is usually made during the florid phase.

Affected patients usually have a hyperplastic respiratory epithelium (although squamous metaplasia might be seen). This epithelium overlays a submucosa that has been infiltrated by mixed inflammation, which is made up of lymphocytes, plasma cells, neutrophils, and large macrophages with foamy cytoplasm (figure 1). The multinucleated macrophages, which are most abundant during the florid phase, are referred to as Mikulicz' cells (figure 2). True granulomatous inflammation is not seen. The application of a tissue Gram's stain or a Warthin-Starry silver stain will detect the gram-negative rods in the foamy macrophages, which are usually numerous and easily identified.

The most important aspect of the differential diagnosis is a failure to consider rhinoscleroma. In some cases, it is necessary to exclude massive lymphadenopathy with sinus histiocytosis (Rosai-Dorfman disease) or other chronic granulomatous processes (e.g., leprosy or atypical mycobacterial infections). Tissue cultures yield the causative agent in only about 50% of cases. The treatment of choice is long-term therapy with tetracycline.

Suggested reading

Brandwein M. Rhinoscleroma. In: Barnes L, ed. *Surgical Pathology of the Head and Neck*. 2nd ed. New York: Marcel Dekker, 2001:2032-5.

Quevedo J. Scleroma in Guatemala, with a study of the disease based on the experience of 108 cases. *Ann Otol Rhinol Laryngol* 1945;58: 613-45.