

Rhinoscleroma: A diagnostic challenge



Medical Science

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ABSTRACT

Rhinoscleroma is a rare chronic, granulomatous disease of the respiratory tract. Rhinoscleroma primarily affects the nasal cavity but the nasopharynx (18%-43%), larynx (15%-40%), trachea (12%) and bronchi (2% to 7%) can also be involved. However, the paranasal sinuses are usually free of disease. Rhinological signs are generally the first reported by patients. CT scan and MRI are useful for diagnosis. Diagnosis of rhinoscleroma is based on histological characteristics and presence of Klebsiella rhinoscleromatis on biopsy cultures. In most cases treatment involves prolonged antibiotic therapy with anaesthetic surgical reconstruction when necessary. However, rhinoscleroma is difficult to eradicate and its recurrence rate is high.

INTRODUCTION:

Rhinoscleroma ("hard nose") is a chronic infectious granulomatous disease caused by *Klebsiella rhinoscleromatis*, a gram-negative encapsulated bacterium of low infectivity. It is worldwide but has peculiar geographic distribution being commoner in semi-desert types of climates¹. The disease process usually involves the nasal cavity and the nasopharynx, but it can also involve the larynx, trachea, bronchi, middle ear and orbit. It often affects the entire respiratory tracts and this condition is termed as 'respiratory scleroma'. The sinuses are rarely affected².

It is a poorly communicable infection although transmission after prolonged household contact with infected family members has been reported³.

Rhinoscleroma often begins at a young age (first to third decade). Infection requires prolonged exposure to the pathogen.

CASE REPORT:

10 year old girl came with a complaint of mass in nose since 2 years, difficulty to eat & swallow since 1 year, difficulty to breathe since 1 year, decreased sense of smell since 3 months.

On examination of nose-swelling present in lower part of nose 3x2 cm in size involving both nasal cavities from the vestibule involving nasal septum & lateral wall. Mass with smooth surface, pink in colour, do not bleed to touch. Oral cavity shows ulceration on hard & soft palate.

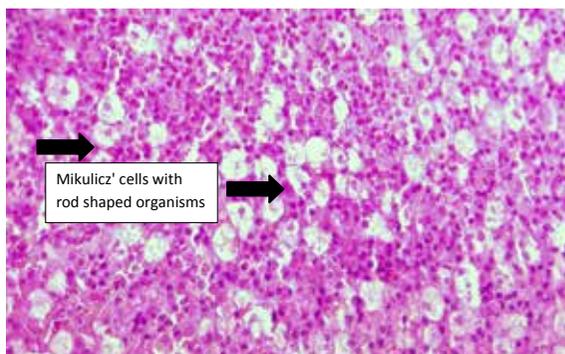
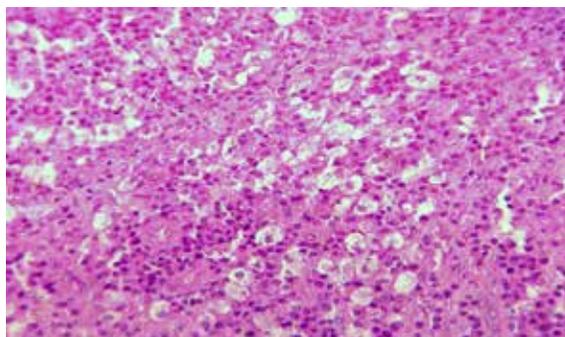
Biopsy from mass from left nasal vestibule was sent.

On gross examination-multiple grey white soft tissue bits all aggregating 1.5 cm.

On microscopic examination- tissue lined by pseudostratified columnar epithelium. Few foci of metaplastic squamous cells also seen. Underlying stroma shows chronic granulation tissue with plasma cells, Russell bodies & foamy histiocytes (Mikulicz' cells). Within the cytoplasm of Mikulicz' cell, bacilli (*Klebsiella rhinoscleromatis*) are seen which are round to ovoid bacilli. Active granulation tissue with proliferating capillaries seen.

Special stains: PAS stain & Giemsa stain: positive

A sprinkling of foamy histiocytes (clear cells) is seen in area of inflammatory cells (blue staining nuclei of lymphocytes and plasma cells)



Discussion:

Rhinoscleroma is a rare chronic infectious granulomatous disease⁴. There is no gender prevalence. The young adults in the second or third decades are most commonly affected⁵. HIV positive patient can also be affected² but this does not occur frequently⁶.

Clinical manifestation-Rhinoscleroma includes 3 clinical stages^(2,7). The catarrhal atrophic stage is characterized by blood streaked mucus or mucopurulent nasal discharge and fetid crusts. Non-specific symptoms such as epistaxis, nasal obstruction and anosmia characterize the infiltrating or florid or granulomatous stage during which granulomatous nodules may develop.

The last stage is a sclerotic stage where nodules are replaced by fibrosis tissue causing nasal stenosis and external deformity. At this stage nasal endoscopy can be useful to reveal abundant crusts. Nasal cavity pathology is almost always present (95-100%)^(7,8). Minor symptoms can be present (rhinorrhoea, crusts, anosmia, polyposis)⁷ but the involvement can be extensive with nasal deformity (32%)⁸ and a Hebra nose (10%).

In the last fibrosis stage, nasal stenosis or nasolacrimal duct stenosis may occur⁽⁹⁾. The diagnosis is usually made during florid phase.

The other affected areas are larynx (26%), Eustachian tube (27%), trachea (20%), maxillary sinus (22%) and pharynx (in 18 to 43%)⁽¹⁰⁾.

Affected patients usually have a hyperplastic respiratory epithelium (although squamous metaplasia might be seen). Diagnostic characteristics are most commonly found in granulomatous stage and it shows chronic inflammation comprising of lymphocytes, plasma cells, neutrophils and large macrophages with foamy cytoplasm. Russell bodies are also seen. The multinucleated macrophages, which are most abundant during the florid phase, are referred to as Mikulicz' cells. True granulomatous inflammation is not seen. The application of a tissue H & E stain, Gram's stain or PAS stain will detect the gram-negative rods in the foamy macrophages, which are usually numerous and easily identified. Organisms are diagnosed by culture.

In some cases, it is necessary to exclude massive lymphadenopathy with sinus histiocytosis (Rosai-Dorman disease) or other chronic granulomatous processes (e.g., leprosy or atypical mycobacterial infections).

Biopsies must be performed from the most active sites of this pathology or from the septum and inferior turbinate. Mikulicz' cells are not pathognomonic but are characteristic. A chronic inflammatory infiltration of monocytes and lymphocytes has also been described⁽¹⁴⁾.

Treatment

Aggressive therapy must be carried out due to the high recurrence rate of rhinoscleroma⁽⁷⁾. Antibiotics are the basis of the treatment. Tetracycline is an historical choice due to its low cost and efficacy (87%) of *K. rhinoscleromatis* are sensitive⁽¹⁶⁾. However, this is contraindicated in children or pregnancy and its recommended dosage is four times daily. Cephalosporin is ineffective *in vitro* but pathological relapse may occur when treatment is stopped⁽¹⁷⁾. Rifampicin is one of the most effective antibiotics⁽¹⁸⁾. Ciprofloxacin seems to be an antibiotic with the best treatment compliance and efficacy but with a high cost^(14, 16). The optimal cost benefit ratio is obtained with the combination trimethoprim-sulfamethoxazole but with greater toxicity than ciprofloxacin⁽¹⁶⁾. Antibiotic therapy should be continued for several months (3 to 6) until biopsy and culture results are negative. Surgery can be associated with antibiotics to clear the nasal cavity crusts and in the reconstruction of nasal deformity.

CONCLUSION

Rhinoscleroma is a rare pathology in economically developed countries. The nose is always affected but sinuses are rarely affected. Diagnosis was made based on pathology and bacteriology. The natural history includes a tendency of this disease to recur, to extend locally causing deformity. The causative organism is resistant to most antibiotics and, being intracellular, is not always exposed to sufficient concentration of the drug. Treatment is based on several months of antibiotic therapy, whereas surgery is reserved for diagnosis (biopsies), nose cleaning and reconstruction.

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