Case report

Rhinoscleroma

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Summary

A case of rhinoscleroma occurring in a 35 year old casual labourer from the Passara district, a rural area of Sri Lanka, is reported. This is the first report of this disease from Sri Lanka. The growth of a proteus species from the affected tissues on 3 separate occasions, was a special feature.

Introduction

Rhinoscleroma is a chronic, slowly progressive, infectious and mildly contagious disease caused by the bacterium Klebsiella rhinoscleromatis. The disease is acquired by direct contact or indirect contact with the nasal exudate of an infected person. It is usually localized in the nasal fossae, but may later spread to the adjacent skin, upper respiratory tract and the lacrimal apparatus. The disease produces an infiltrating granuloma with a marked tendency to sclerosis and subsequent obstruction [1].

The disease occurs sporadically almost all over the world and is endemic in certain countries, such as central Europe, Russia, China, India, Indonesia, Africa, Central and South America [2,3]. No previous report to this could be traced from Sri Lanka.

The infection occurs in both sexes at any age but it is more frequent between 20 and 40 years. It is more common in rural areas [1].

Case report

A 35 year old casual labourer from the Passara district was referred to our department, with a nasal granuloma with deformity of the nose of 1½ years duration. The lesion had started as erythematous papules on the throat and nasal septum which gradually enlarged and coalesced to form an erythrematous plaque which later become ulcerated. At first he had nasal stuffiness and obstruction with a burning sensation of the throat. There was also a sero-sanguinous nasal discharge. Later there developed a nasal quality to his voice and a change in the contour of his nose. The symptoms were mild and did not disturb him enough to seek medical treatment previously. There were no other constitutional symptoms.

On presentation he had an erythematous, granulomatous lesion with irregular borders extending outwards from the floor of the nasal vestibule into the upper lip. The anterior part of the nasal septum was destroyed with flattening of the nose and near total obstruction of the nasal passage. The lesion was indurated with a warty surface and had surface ulceration. A similar plaque of irregular outline with well demarcated margins was seen in the oropharynx. There was no significant lymphoadenopathy. There was no other skin or mucosal lesions.

All basic investigations including the ESR, WBC/DC, chest X-ray and mantoux test were within normal limits.

Pathological and bacteriological features

A biopsy from the nasal mucosal lesions revealed a dense infiltrate chiefly of plasma cells, but also having large, round vacuolated histiocytic cells with pale cytoplasm and an eccentric nucleus, the Mikulicz cells, and the non-nucleated elliptical structures with hormogeneous cytoplasm which turns bright red, the Russell body. These changes were pathognomonic of rhinoscelroma.

Bacterial culture from the nasal mucosa, done on 3 separate occasions in 2 different laboratories each yielded a proteus species, further indentified on one occasion as P. mirabilis.

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Management

The patient was managed on a combination of streptomycin 1 g daily and tetracycline 500 mg 6 hourly for 4 months. There was significant regression of the nasal lesions and a considerable relief from the symptoms of nasal obstruction and stiffiness. The voice gradually lost its nasal tone. A deformity of the nose however remained.

Discussion

The clinical manifestations of rhinoscleroma run a very slowly progressive course and the patient seeks medical attention after many years. Our patient had his symptoms for 1½ years before presentation. The symptoms progress through 3 over lapping stages: rhinitic, proliferative (infilterated and noduler) and fibrotic.

At presentation our patient had been through the rhinitic phase with a nasal discharge. The infiltrative phase with nasal obstruction with exhuberant, friable granulation tissue with extensions to the pharynx and larynx was evident in our patient. In the later noduler period the disease has spread forward invading the nasal lobule in our patient, giving rise to the so called "Hebra nose". After treatment clinical improvement occurs as in our patient but the previous proliferative lesions are replaced by fibrosis, leading to persistant nasal distortion. This was also seen in our patient after treatment.

The most frequent symptoms of nasal obstruction 94%, nasal deformity 32%, voice change 12% were all present in our patient. The nose is involved in 95% of cases, and the pharyx in 18% of cases reported by Miller[5].

The causative organism has been identified as Klebsiella rhinoscleromatis and a granulomatous inflammation with a similar histopathology has been reproduced in mice[6]. 3 attempts at isolating the organisms from the nasal mucosa in our patient, resulted in the growth of a proteous species. Though secondary infection is a possible cause, it is interesting to consider the possibility of a proteus being the cause, both organisms being gram negative rods.

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