CASE REPORT

Rhinoscleroma Mimicking Malignancy

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Abstract

Rhinoscleroma is an uncommon chronic granulomatous disease of the upper airways affecting nasal cavity, nasopharynx, larynx, trachea, and bronchi. The oral cavity, para nasal sinuses, and soft tissues of the lips and nose can be affected. In rare cases, rhinoscleroma spreads to the orbit. We present a case that was being evaluated for bilateral neck nodal mass, was confused with malignancy and subsequently, on thorough clinicopathological evaluation, was diagnosed as rhinoscleroma. This case underlines the importance of thorough clinical evaluation and diagnostic workup before instituting any sort of treatment in oncology.

Key Words

Rhinoscleroma, Malignancy, Chronic Granuloma

Introduction

Rhinoscleroma is a chronic progressive granulomatous disease of upper airways mainly targeting nasal passages (1).The first use of name rhinoscleroma dates back to 1870 when Von Hebra and Kaposi described a lesion in the nose (2).Histological features and non-neoplastic inflammatory nature were established by Mikulicz in1877 (3). In 1882, a Gram-negative coccobacillus was identified by Von Frisch and established as the causative agent of this lesion. Same coccobacillus is now known as Klebsiella rhinoscleromatis (4). Clinically and pathologically rhinoscleroma is divided into three stages. Early institution of treatment is necessary to avoid the spread of disease. Rhinoscleroma is widely distributed in the world with an endemic area around eastern Europe (5). A case of rhinoscleroma with nasopharyngeal mass and bilateral neck nodes has rarely been reported from this region which prompted us to report this case.

Case

23 year old female, a resident of Kupwara, presented to the hospital with a four month history of left cervical swelling. Two months prior to coming to hospital she also noticed swelling on right side of neck. There was associated history of high grade fever, dysphagia and pain in head and shoulders. On examination, there was bilateral cervical lymphadenopathy which was tender on touch and nodes were matted .There was bilateral tonsillar hypertrophy, uvula was centrally placed and mucosa congested.Sinonasal endoscopy done by an E.N.T. specialist revealed smooth, glistening, lobulated mass which was friable and filling whole of nasopharynx. Before coming to our hospital she had gone to local hospital where FNAC of cervical lymph node showed metastatic deposits of malignant epithelial cell tumor. Histopathology of excised lymphnode mass showed features compatible with necrotizing lymphadenitis. She was reffered to state hospital where biopsy from nasopharyngeal mass sent to two private laboratories were reported as poorly differentiated carcinoma from one and rhinoscleroma from other. Patient was then reffered to our department for further management and on first look it appeared to be carcinoma and patient was admitted in ward for institution of treatment. On detailed clinical evaluation the diagnosis was doubted and biopsy got reviewed. Review showed features in favor of Rhinoscleroma and patient

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was put on doxycycline and sent for E.N.T. consultation. Keeping in view strong possibility of malignancy, she was put on close follow up and was subjected to various investigations. Elisa was negative for tuberculosis, and serology was negative for EBV. A fresh biopsy specimen was sent to Lal path lab which showed chronic inflammatory granulation tissue. No evidence of malignancy was seen in that biopsy. After one month of treatment patient showed response to doxycycline and was sent for E.N.T. consultation. She again came to our department after two weeks with increased size of lymph nodes because she had stopped taking medicine of her own . Keeping in view recommendations for long treatment for rhinoscleroma,patient was again put on same treatment with addition of anti-inflammatory agents and asked to follow our as well as E.N.T. department regularly. On her last visit on 22-02-08, patient was perfectly alright.

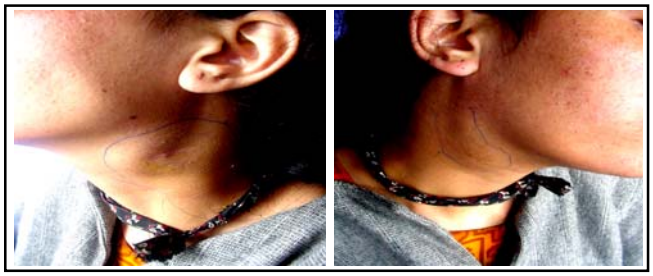

Fig 1: Before treatment

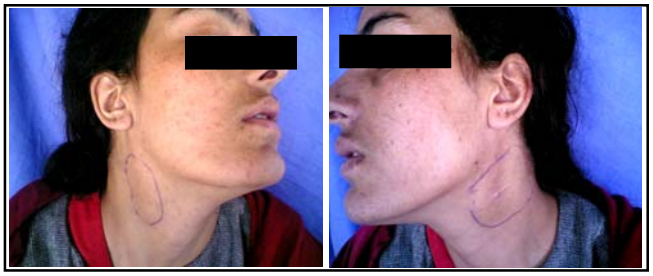

Fig 2: After treatment

Discussion

Rhinoscleroma mostly affects young and females outnumber males (1) Poor hygiene, crowded living conditions and malnutrition are predisposing factors and it may also be associated with diseases lowering immunity. In our case also possibility of both carcinoma and rhinoscleroma coexisting was not ruled out but the way the patient behaved to treatment certainly excludes carcinoma. Rhinoscleroma is a chronic progressive granulomatous disease mostly affecting nose. In a clinicopathological study done in gulf region six out of 25 patients were found to have nasopharyngeal involvement. The low incidence of nasopharyngeal involvement in that series was attributed to difficulty of direct visualization. Rhinoscleroma is usually divided into three stages- the catarrhal, the proliferative, and the sclerotic. Histological findings are more characteristic and diagnostic in proliferative stage and it is difficult for a pathologist to diagnose in catarrhal stage (1). Clinically it should be differentiated from infectious granulomatous processes, sarcoidosis, vasculitis, neoplastic diseases like lymphoma and extra nodal Rosai-Dorfman disease. The histological differential diagnoses included leprosy, malakoplakia and metastatic renal cell carcinoma (1). Treatment should include a long-term antimicrobial therapy and surgical intervention in cases with symptomatic obstruction (5). Tetracycline group remains drug of choice. Mayo clinic carried out a review on rhinoscleroma in 1993 and found ciprofloxacin to be a potential alternative.Badia L reported a case in 2001 who was successfully treated with ciprofloxacin (6). Ciprofloxacin can be a better option for long term use because of few side effects. Surgical intervention is needed only in case of obstruction. Because of high rate of relapses close observation is the solution to the long-term follow-up care of the patient. Being aware of clinical presentations, early diagnosis of this condition is prerequisite to reduce the morbidity caused by this disease.

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