Rhinoscleroma-A Rare Granulomatous Disease in a Child

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Abstract

Rhinoscleroma (RS) is a chronic granulomatous disease caused by *Klebsiella rhinoscleromatis*. In all the patients, the nose is affected but sometimes extends to any parts of the airways. Diagnosis of this disease is done on the basis of the histopathological examination or direct evidence of the bacteria in nasal exudates. The differential diagnoses of the RS are syphilis, malignancy and midline granuloma. Streptomycin is drug of choice, although fluoroquinolones and cephalosporin also provide good results. Surgery is opted in case of obstruction or deformity. Here, we reported a case of 11-year girl diagnosed as RS on the basis of the histopathological examination. This case was treated by endoscopic removal of the mass in the nasal cavity followed by a course of ciprofloxacin. She was found asymptomatic at the last visit of 1 year after treatment and there was no evidence of recurrence.

Keywords: *Rhinoscleroma, nasal cavity, pediatric patient,Mikulicz cells, Russel bodies.*

Introduction

Rhinoscleroma (RS) is an uncommon granulomatous disease often found in mucosal lining of the respiratory tract and occurred by the causative agent of the *Klebsiella rhinoscleromatis*, rod shaped Gram-negative bacteria.[1] There are several cases of RS reports reported from several parts of the world like South East Asia, tropical Africa,Middle East, India,South and Centralparts of America but in recent years, few cases are also detected in non-endemic zones as of high migration of population from different part of the world.[2] After the nasal cavity, other common sites affected by RS are nasopharynx(18-43%), paranasal sinuses(22%) and the larynx(15-40%).[3] RS is common in rural areas where socio-economic status is very low. This disease is more in crowding area, low quality nutrition and low quality hygiene. It is common in females than males (13:1) and found in the 2nd and 3rd decades of life.[4] It is thought that decreased iron in the body may leads to this disease. [4] Here, we are reporting this case of RS in a 11-year-old child.

Case Report: An 11-year-old girl attended outpatient department of Otorhinolaryngology with complaints of rhinorrhea,nasal bleeding and nasal block in the right nostril since 3 months. Anterior rhinoscopy showed a friable mass in the right nostril. Examinations of throat and ear were within normal limits. Diagnostic nasal endoscopy demonstrated the same appearance of the nasal mass with presence of granulomatous lesions inside the nasal cavity attaching to the floor of the nose (Fig.1). Routine blood tests and serological tests for

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HIV were within normal limit. Chest X-ray was normal. Computed tomography(CT) scan revealed opacity/ mass in the right nostril (Fig.2). The re was no evidence of bony erosion in the nasal wall. Biopsy was taken from right nasal cavity and sent for histopathological examination (HPE). HPE revealed extensive inflammatory infiltrates along with lymphoplasmacytic and histiocytic predominance. The plasma cells showed Russel bodies (Fig.3). Intrahistiocytic bacilli correspond to *K.rhinoscleromatis* which was also recognized. After confirmation of the diagnosis, the treatment was done with ciprofloxacin (250 mg twice daily) for 3months with satisfactory follow up. The endoscopic examination revealed no evidence of recurrence after 1 year of treatment.

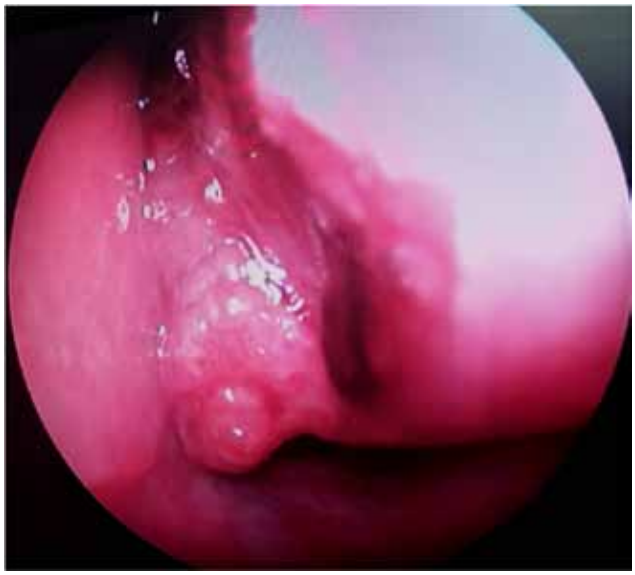

Fig. 1: Diagnostic nasal endoscopy revealed granulomatous mass in the right nasal cavity.

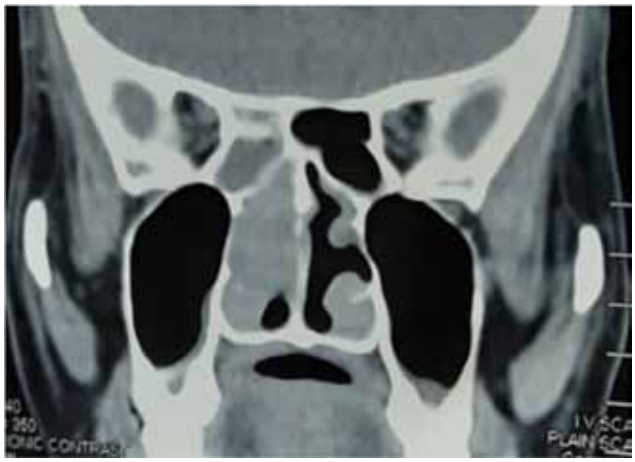

Fig. 2: CT scan of the paranasal sinus showing radio-opaque soft tissue mass in the right side nasal cavity.

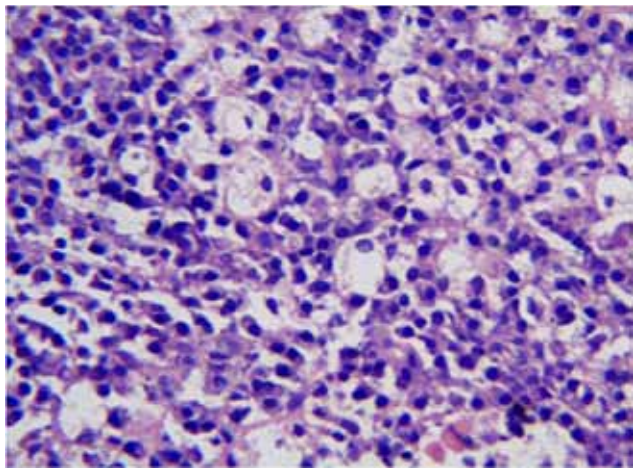

Fig. 3: Histopathological picture shows Mikulicz cells which are large vaculated foamy cells with small nuclei and Russel bodies which are bright red degenerated plasma cells with background of several plasma cells (HEX400).

Discussion

RS is a chronic granulomatous disease due to infection of *Klebsiella rhinoscleromatis*, gram-negative encapsulated bacilluswhich belongs to Enterobacteriaceae family. In anaerobic condition, it forms spores in anaerobic condition which can produce exotoxin and leads to scleroma formation without presence of bacteria.[5]It is often associated with poor socioeconomic condition,malnutrition,poor hygiene, HIV infection and overcrowding. Rhinoscleroma is common among females and often affects the middle age group people.Bilateral nasal cavities are usually affected. In our case, the lesion is confined to one side nasal cavity. Sometimes, it affects the nasopharynx and larynx. Involvement of the larynx may lead to severe stridor. Pediatric patients are rarely affected, however very few cases are reported in the medical literature. The cellular immunity may be affected in this patient;however, this diseased person was immunocompetent.

This disease has three stages: Catarrhal stage, granulomatous stage and fibrotic stage. The catarrhal stage is nasal variety where findings and histopathology may not be specific. In case of granulomatousstage of RS, granulomatous lesions are seen this can block the nostrils. The biopsy is diagnostic in granulomatous stage.[6] This stage may be associated with extra-nasal extension involvement of larynx is the commonest (60-80%) and may be complicated to stridor.[7] In the third stage or stage of fibrosis, there is extensive fibrosis which leads to nasal block rather than proliferative masses.

The fibrotic variety is usually difficult to diagnose as the causative bacteria is usually not seen.

Clinical symptoms of the patients with RS are nonspecific and often presents with mucopurulent nasal discharge,cough,breathing difficulty due to nasal block, dyspnea, nasal bleeding and headache.[8]This disease slowly progress with characteristic of remission and relapse. Patients often require medical advice only when the obstructive granulomatous stage leading to obstruction at the nasal cavity.

The investigations of RS include diagnostic nasal endoscopy, imaging and histopathological examination. Nasal endoscopic examination helps to find the lesion in nasal cavity and its extension to other parts of the upper airway. Computed tomography (CT) scan of the nasal cavity andsinuses reveal lesions in the sinonasal area. Histopathological examination confirms diagnosis. One study documented RS in children where the lesions appear as amyloid like protein in histopathological examination which may be due to an autoimmune reaction.[9] One study revealed RS in three siblings staining at the non-endemic area and thought to be due to neutropenia acting as a predisposing factor.[5] The differential diagnosis of the RS includes tuberculosis, actinomycosis, leprosy syphilis, histoplasmosis, paracoccidioidomycosis, sporotrichosis and parasitic infections such as mucocutaneous leishmaniasis.[10] The RS should also be differentiated from Wegner's granulomatosis, carcinoma and lymphomas. The specific diagnosis of the RS is based on the histopathological examination and the bacilli appear rod-shaped. This bacillusis positive to Warthin-Starry stain and periodic Acid-Schiff (PAS).Staining of immunoperoxidase with anti-capsular antiserum is also helpful for identification of the causative bacteria.Bacterial culture with MacConky agar or blood is also helpful for identification of the bacilli in approximately 50% of the cases.[11] The histological picture of the RS is characterized by the presence of Mikulicz cells, inflammatory cells consisting of multiple plasma cells along with few eosinophils and Russel bodies. These Russel bodies are originated from disintegration of plasma cells. The Mikulicz cells are vaculated cells with clear cytoplasm possessing the bacteria. The cause of transformation of the histiocytes into Mikulicz's cells is unknown.[12] Untreated patients of RS often progress to involve other areas of the respiratory tract. If laryngotracheal part is involved, this may lead to the obstruction of the airway which threatens to life.[13] The sclerotic stage of the RS shows extensive fibrosis, which may cause stenosis and disfigurement. Biopsy from fibrotic stage is often not specific. In this patient, the stage of the disease was with granulomatous type as multiple numbers of Mikulicz cells were found. Immunoperoxidase monolayer assay (IPMA) method for antigen of the capsule of *Klebsiella rhinoscleromatis* is very reliable for the confirmation of the RS. The differential diagnosis is usually other granulomatous diseases of the noses, nasal polyps and sarcoidosis.Presence of extra-nasal sites should be carefully evaluated by diagnostic nasal endoscopy or imaging tests in order to disease process which often need prolonged treatment.[3]

The choice of drug is streptomycin although cephalosporins, fluoroquinolones and ciprofloxacin have had some good outcome. Ciprofloxacin has good antibacterial spectrum and excellent tissue penetration with fewer side effects. Surgical treatment is useful inpatients with severe nasal obstruction or nasal deformity. The treatment period is around three to six months which targets intracellular bacilli. Currently criteria comprise the antibiotics fluoroquinolones therapy with demonstrable efficacy but the clinical and endoscopic examination must be necessary until negative biopsies for eradication of the disease.[13] Surgery is often reserved for the disease particularly orolaryngotracheal forms where stenosis is detected.[13] These may require endoscopic laser or debridement of the stenotic part of the airway or may need open surgery preceded by tracheostomy.[14]Mortality due to RS is extremely rare,but may happen due to upper airway obstruction in case of undiagnosed disease or due to complication of the surgical procedure.

Conclusion

Rhinoscleroma (RS) is a rare clinical entity and should be ruled out in case of nasal mass of a pediatric patient. Surgical removal cannoteliminate the lesion and may lead to recurrence in the stage of fibrosis. The clinical presentations are usually non-specific and granulomatous stage of this disease often evokes the possibility of the RS. Histopathological examination confirms the diagnosis where Mikulicz cells are pathognomonic for *Klebsiella rhinoscleromatis*. The diagnosis of the RS is often difficult and delayed because of its clinical polymorphism. However, early diagnosis and prolonged treatment are important criteria for avoiding recurrence and late squeal.

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Ethical Permission: Approved

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