Rhinoscleroma: An Unusual Presentation

Abstract

Rhinoscleroma is a chronic granulomatous disease caused by *Klebsiella rhinoscleromatis*. It commonly affects the nasal cavity and nasopharynx, but it can also involve the larynx, trachea, bronchi, middle ear, and orbit. We are reporting a rare presentation of rhinoscleroma in a middle-aged female patient involving the lips and gums.

Keywords: *Gums, lips, rhinoscleroma*

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Introduction

Rhinoscleroma is a slowly progressive, chronic granulomatous infectious disease caused by *Klebsiella rhinoscleromatis*.[1] It is a Gram-negative rod-shaped bacilli. The disease is endemic in Middle East, Central and South America, and Eastern Europe whereas sporadic cases of rhinoscleroma have been reported throughout the world.[2-6] In 95% to 100% of cases nose is the most affected part.[7] There are reports of rhinoscleroma involving pharynx, nasopharynx, paranasal sinuses, orbit, larynx, trachea, and bronchi in the world literature.[8-12] Involvement of lips and gums are uncommon. Here we are reporting a rare presentation of rhinoscleroma involving lips and gums.

Case Report

A 34-year-old female patient presented with asymptomatic swelling of lips and gums since 9 months. It was insidious in onset and gradually progressive. She was otherwise healthy with no systemic complaints and no history of nasal lesion in the past.

On cutaneous examination, there was diffuse swelling of peri-oral area, gingival hypertrophy and areas of crusting and fissuring over lips. A verrucous lesion was noted over the inner aspect of lower lip [Figures 1a and b]. On palpation the swelling was non-tender, firm in consistency, and did not bleed on touch. The examination of nose and throat was normal.

Based on history and clinical findings granulomatous diseases were suspected such as foreign body reaction, mycobacterial infection, cheilitis glandularis, sarcoidosis, granuloma cheilitis, Crohn's disease, Wegener's granulomatosis, Melkersson-Rosenthal syndrome, and histoplasmosis. On investigating, complete blood count, liver function tests, renal function tests, and chest X-ray were normal. The serological tests were negative for syphilis, hepatitis B, and human immunodeficiency virus (HIV). Two punch biopsies were taken from lips for histopathology and tissue culture. The slides were stained with hematoxylin and eosin (H and E), Gram, and PAS (periodic acid-Schiff) stains. All slides were examined under light microscopy. On H and E stain, diffuse inflammatory infiltrate of plasma cells with Russel bodies and mononuclear cells with large foamy macrophages (Mikulicz cells) were seen [Figures 2a and b]. Gram [Figure 3a] and PAS [Figure 3b] stains demonstrated intracytoplasmic bacilli. We did not find any growth on tissue culture. The diagnosis of rhinoscleroma was made based on classical histopathological findings and demonstration of intracytoplasmic bacilli by special stains. The patient was further investigated for nasal and upper respiratory tract involvement. The diagnostic endoscopy of nose and oropharynx did not reveal any lesion. X-ray of nose and PNS (paranasal sinus) was normal. The patient was started on ciprofloxacin 500 mg and doxycycline 100 mg twice daily for 4 months. The patient showed good

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response to treatment [Figures 4a and b] with no clinical relapse after 1 year of regular follow-up.

Discussion

Rhinoscleroma was first described by Hebra in 1870. It is a chronic granulomatous disease of the upper respiratory tract caused by *K. rhinoscleromatosis*.[7] Which is a short, encapsulated, and immotile Gram-negative bacillus. The organism has an affinity for nasal mucosa. The disease is transmitted by direct or indirect contact with nasal exudate of infected person.[13] Nasal cavity is the most commonly involved site (95–100%) followed by nasopharynx (18–43%), larynx (15–40%), trachea (12%), and bronchi (2–7%).[8]

The disease predominantly occurs in middle-aged woman, living in poor hygienic and nutritional condition. The exact pathogenesis of the disease is not known. Altered immune response with impaired cellular immunity is thought to play a role in the pathogenesis of the disease. However, an alteration in CD4:CD8 population in blood has been postulated as a cause of chronicity of the disease.[14]

Humans are the only host for K. rhinoscleromatosis. The route of transmission is mainly by inhalation of nasal droplets or contaminated material facilitated by poor hygiene and malnutrition.[15] Rhinoscleroma usually occurs at the epithelial transition zones, especially at the junction where stratified squamous epithelium of the vestibule meets ciliary epithelium of the nose.[16]

Three stages are described in rhinoscleroma: catarrhal-(rhinitic), granulomatous (proliferative or nodular), and sclerotic (cicatricial or fibrotic).[7] Patients with initial catarrhal phase present with nonspecific rhinitis that evolves into chronic foul smelling purulent discharge, epistaxis, crusting, and nasal obstruction.[7] On histopathology, it shows a nonspecific subepithelial infiltrate of polymorphonuclear leucocytes with granulation tissue.[2] The clinical features are more prominent in granulomatous phase. In this phase, patient develops granulomatous nodular mass which leads to nasal obstruction. The dense infiltration of lymphocytes, plasma cells, Russell bodies, and pathognomonic large Mikulicz cells are the diagnostic histopathological changes seen in granulomatous stage.[2] If left untreated patient goes into sclerotic phase. Permanent complications are the features of sclerotic phase which includes nasal deformities, anosmia, oral anesthesia, dysphonia, dysphasia, and stridor.[7] In sclerotic phase, extensive fibrosis and less inflammatory cell infiltration are the main histopathological findings.[2,7]

Histopathology is the mainstay in the diagnosis of rhinoscleroma. Intracytoplasmic bacilli can be demonstrated by special stains using Gram, PAS, Geimsa, Warthin-Starry, and Grocott's silver methenamine stains. The detection of Type III *Klebsiella* antigen, CD 68 and altered CD4:CD8 ratio are helpful in the diagnosis.[17,18] In granulomatous stage, tissue culture can demonstrate the organism in about 50% of cases.[17]

As the diagnosis of rhinoscleroma was confirmed by classical histopathology, and positive findings of Gram and PAS stains, investigations for other granulomatous disorders were not done. It was further supported by therapeutic response with ciprofloxacin and doxycycline.

Lips and gums involvement is rare in rhinoscleroma. The upper lip involvement in rhinoscleroma is seen in only 12% of the patients. Ten percent of rhinoscleroma patients present with upper lip swelling as initial presentation.[19] In our case, patient initially manifested with swelling of both lips without any preceding nasal lesion. To the best of our knowledge primary involvement of both the lips in

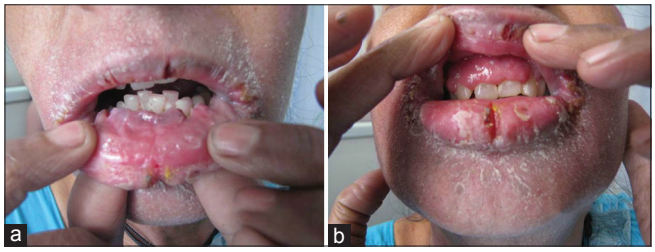

Figure 1: (a and b) Hypertrophy of gums with verrucous lesion over lower lip

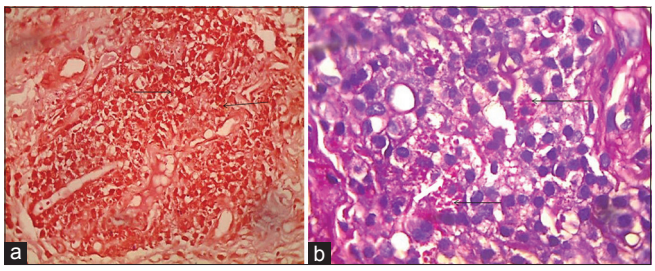

Figure 3: Section showing (a) Gram negative (×40) and (b) PAS positive intracytoplasmic bacilli (×100)

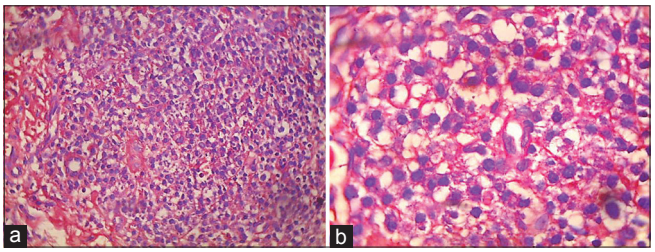

Figure 2: Histopathological section (H and E stain) showing abundant plasma cells, Russell bodies and Mikulicz cells (a) ×40, (b) ×100

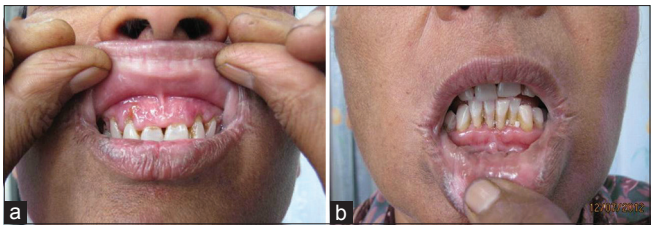

Figure 4: (a and b) Improvement of lesions after 4 months of treatment

rhinoscleroma has not been reported in literature and hence this case is being reported.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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