Rhinoscleroma: Report of an Erratic Palatal Swelling

Abstract

Clinicians come across cases of palatal swellings that present with ambiguous features. They may vary in their etiology from numerous infectious and noninfectious causes to a wide array of neoplasms. Accurate diagnosis of such lesions is vital for their prompt and precise management. Rhinoscleroma (RS), as its name suggests, is a persistent, specific, granulomatous disease that results in sclerosis of the affected organ – most frequently the nose. Although its occurrence in the adjacent sites has been reported, the clinical findings did not offer much deviation from the expected. Reported here is a case of RS involving the palate which not only detoured from its usual site and course of spread but also gave off a confusing façade in terms of its clinical presentation.

Keywords: *Chronic granulomatous inflammation, palatal swellings, rhinoscleroma*

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Introduction

Rhinoscleroma (RS) or scleroma is an uncommon, chronic granulomatous disease with a widespread distribution and is endemic in certain developing countries in Africa and Southeast Asia. It is a localized, slowly progressive, infectious disease caused by *Klebsiella rhinoscleromatis* – a Gram-negative bacilli.[1] Although the nasal cavity is the most commonly affected site, its extension into the pharynx, oral cavity, larynx, trachea, and bronchi is not unusual.[2,3]

The following report is one such account of a palatal swelling which turned out to be manifestation of an infrequent bacterial infection.

Case Report

A 33-year-old male patient reported to us with the chief complaint of a painless swelling involving right half of the palate for 5 years from the date of reporting. The swelling initially appeared as a small mass in the central palatal region and then grew over time to involve the entire right half of the hard palate [Figure 1]. The patient gave a history of being treated for RS which had manifested as a nasal swelling associated with chronic rhinitis 3 years back and reported a similar lesion at the same site 6 months back which was suggestive of recurrence.

General examination did not reveal any deviation from the routine findings. Upon extraoral examination, slight facial asymmetry was noticed owing to the nasal swelling on the right side which resulted in obliteration of nasolacrimal fold.

Intraorally, the lesion presented as an ovoid swelling involving the right side extending from the palatal area corresponding to right canine up to approximately 1 cm beyond the third molar and transversely from the gingival margins of the teeth till midline of the palate. The swelling was grossly 4 cm × 2 cm in size with regular, well-demarcated margins, and the overlying surface was smooth and of the same color as the adjacent palatal mucosa. On palpation, the swelling was nontender and soft to firm in consistency.

Based on the history and clinical findings, a provisional diagnosis of palatal extension of RS was made.

Minor salivary gland tumor, radicular cyst, and periapical abscess were considered in differential diagnosis. Subsequently, radicular cyst and periapical abscess were ruled out the due absence of any dental involvement.

Investigations

Pulp vitality test – The teeth were tested vital during the electric pulp testing.

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Radiographic investigations – The patient was subjected to radiographic investigations, wherein the panoramic radiograph was deemed insufficient to comment on the lesion and the patient was further advised cone-beam computed tomography (CBCT). CBCT revealed a palatal swelling appeared as an isodense mass on the right side of the palate measuring 37.4 mm × 20.6 mm in size. In the coronal section, radiopaque structures suggestive of the stents placed to correct the nasal obstruction were present bilaterally, around which soft-tissue masses appeared to grow. On sagittal view, areas of bone erosion were apparent in the posterior palate and floor of the nose indicating continuity in nasal and palatal lesion [Figure 2].

On hematological examination, the patient was found to have hypochromic microcytic anemia. Preanesthetic evaluation was conducted to rule out other systemic malfunctions such as diabetes mellitus and immunosuppressive diseases.

Cytopathology – Cytological evaluation of the aspirate revealed clumps of red blood cells and few inflammatory cells.

Microbial colony culture – No microbial colonies were observed after 48 h of incubation on MacConkey's agar.

Histopathological examination

Incisional biopsy of the lesion gave the impression of a chronic granulomatous inflammation. Haematoxylin and eosin stained section exhibited fibrocellular connective tissue with severe degree of chronic inflammatory cell infiltrate with lymphoplasmacytic predominance and moderate degree of vascularity. Scattered at places were several large macrophages (foamy histiocytes) having a central nucleus and vacuolated cytoplasm with rod-shaped bacilli at places resembling the Miculicz cells (MCs). Also evident were hyalinized plasma cells which were seen either discretely (Russell bodies) or in clusters (Mott cells) [Figure 3].

Special staining

Histopathology was consistent with the clinical diagnosis and further confirmation was done using special staining techniques to demonstrate the microorganisms. Gram staining showed the presence of Gram-negative rod-shaped bacilli. Periodic acid-Schiff (PAS) and Giemsa stain were also employed for demonstration of the *Klebsiella* spp. The organisms were stained magenta with PAS and light blue with Giemsa stain [Figure 4].

Based on the histopathological findings, the diagnosis was given in favor of RS.

Treatment and prognosis

The patient was referred to the department of Ear–Nose–Throat surgery for further treatment. The lesion was surgically excised and antibiotic therapy with tetracycline one gram twice a day along with Ciprofloxacin 500 mg/day for 1 month was started and continued as deemed fit by the physician. No recurrence has been observed up to the 6-month follow-up. The patient is kept under regular observation to monitor any incidence of recurrence.

Discussion

RS or scleroma is a rare, chronic, indolent, granulomatous, and debilitating bacterial disease primarily involving the nose and is designated as respiratory scleroma when it involves subglottis, trachea, and bronchi.[4] Its notoriety in involving other neighboring sites such as nasopharynx, eustachian tube, paranasal sinuses, soft tissues of lips and nose, and rarely orbit is well established. The current case is one such example of extranasal RS.

Anton Von Frisch in the year 1882 is credited with the identification of the causative organism *K. rhinoscleromatis*, a Gram-negative aerobic bacteria belonging to the *Klebsiella pneumoniae* family.[5,6] RS has female predilection (13:1) and usually occurs in the middle-aged population. Low socioeconomic status, malnutrition, and iron deficiency anemia are considered as the predisposing factors that lower cellular immunity while preserving humoral immunity.[2-4] The presence of HLA-DQA103011-DQB0301 haplotype has been found to be a strong risk factor.[7]

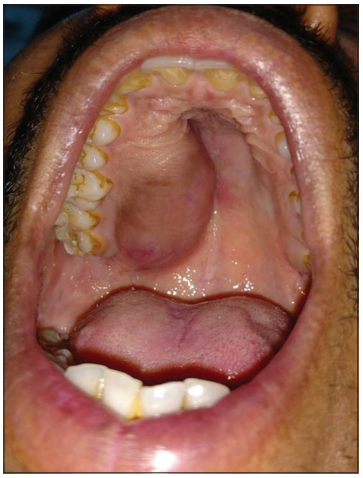

Figure 1: Ovoid swelling involving right half of the palate

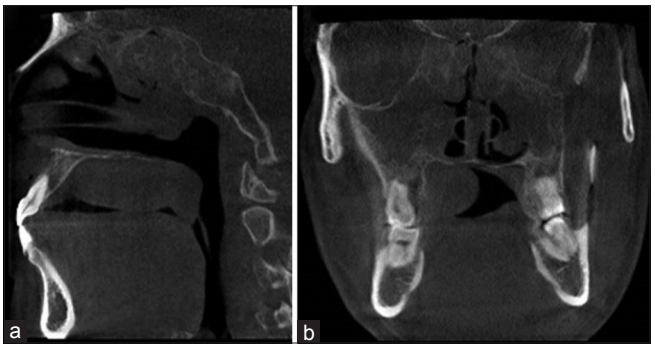

Figure 2: (a) Sagittal view showing isodense mass extending from the incisal alveolar process till the soft palate along with thinning of the palatal process. (b) Bilateral isodense mass involving the nasal cavity and right side of palate and erosion of palatal bone

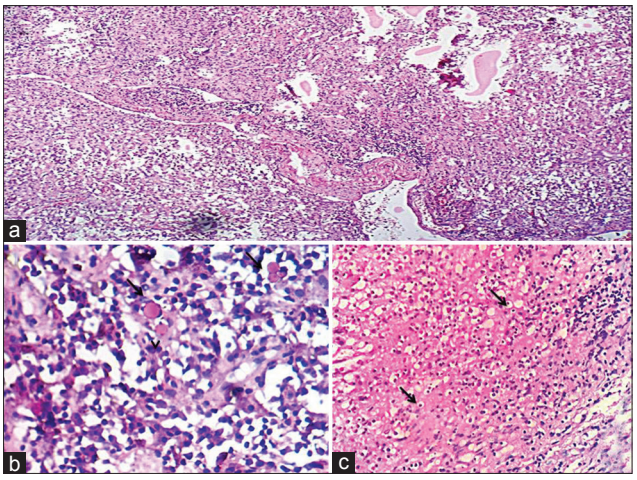

Figure 3: (a) Scanner view shows dense fibrocellular connective tissue. (b) Russel bodies – Pale, homogenous, eosinophilic bodies representing excess immunoglobulin produced by the plasma cells (c) Chronic inflammatory cell infiltrate composed of plasma cells, lymphocytes, and foamy macrophages with centrally placed nucleus – Miculikz cells.

Clinically and pathologically, RS is characterized by three overlapping phases in terms of progression: rhinitic, florid, and fibrotic. The rhinitic/catarrhal stage causes earliest symptoms of nonspecific rhinitis which are known to last for weeks or months and often evolves into purulent and fetid rhinorrhoea with crusting. Histologically, squamous metaplasia along with subepithelial infiltrate of polymorphonuclear cells is usually observed. Bacteria are incompletely digested in the subepithelial layer and further released into tissues. The second florid/granulomatous stage is characterized by the development of bluish-red nasal mucosa and intranasal rubbery nodules or polyps and manifests with epistaxis and nasal deformity, destruction of the nasal cartilage, and bone destruction. In advanced cases, the destruction of the nasal cartilage with the formation of nodules causes a severe deformity referred to as Hebra nose. Histology shows the appearance of Mikulicz cells that are pathognomic of RS.[8,9]

Interleukin-10, an anti-inflammatory cytokine, has been demonstrated as being crucial for the establishment of a proper environment leading to the phenotypic maturation of Mikulicz cells. It controls the metabolic reprogramming of macrophages through inhibition of mechanistic target of rapamycin (mTOR) signaling pathway. Deregulation of mTOR signaling may lead to metabolic changes such as hyperproliferation of macrophages and granuloma formation which in turn will contribute to disease progression in human granulomatous inflammations.[9,10]

The last phase is the sclerotic/fibrotic stage that is characterized by formation of adhesions, extensive fibrosis leading to scarring, and possible nasal stenosis which will ultimately result in distortion of anatomy.[8,9]

In our case, the patient reported with a palatal swelling and involvement of the posterior part of the oral cavity which differed strikingly from its usual manifestation like nodular enlargement observed in the lower nasal and upper labial region.

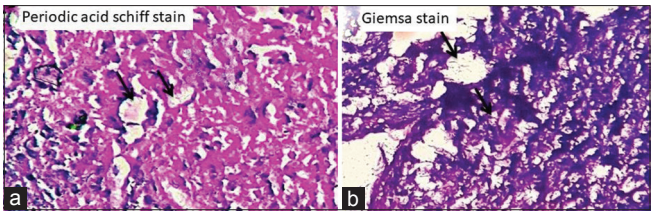

Figure 4: (a) Periodic acid–Schiff staining confirming the presence of pale pink rod-like bacilli. (b) Pale blue-stained microbes on Giemsa staining

In 1969, Mikulicz and Woyke *et al.* described the microscopic picture of the disease along with the first description of the ultrastructure of granulation tissue in RS.[3] Microscopic deduction shows an initial infiltration of neutrophils, plasma cells, and macrophages which is followed by granulomatous inflammation, characterized by numerous plasma cells along with Russell bodies and MCs. Russell bodies, which represent excess immunoglobulins produced by the plasma cells, are round to ovoid, homogeneous, eosinophilic, and up to 40 um in diameter. MCs are large phagocytes with a single shrunken nucleus, displaced to the cell periphery because of the presence of multibacillary cytoplasmic vacuoles, 10–100 um in diameter.[7,11,12] In adjunction to the routine histopathology, various special stains have also been employed in detection of the *K. rhinoscleromatis*, these include PAS stain that stains the bacilli pink, Giemsa stain which gives pale blue color to the organism and Warthin–Starry silver stain which imparts a distinct black color to the bacilli making it more prominent as compared to the previously discussed stains. Recent advances in the diagnosis, in conjunction with the existing methods, include serotyping and immunohistochemistry. De Pontual *et al*. in their study noticed seropositivity in culture for O2:K3 type of Klebsiella.[6]

Among the wide spectrum of nasal and palatal lesions, the differential diagnosis of RS may include:

- Fungal infections such as histoplasmosis, blastomycosis, and rhinosporidiosis
- Bacterial infections – tuberculosis, leprosy, and syphilis
- Noninfectious conditions – Wegener's granulomatosis, natural killer cell lymphoma, lethal midline granuloma etc.[13]

The fungal etiology may be ruled out by the negative culture of the causative organisms as well as by applying the special stains such as PAS, Gomori Silver Methenamine to demonstrate the fungal hyphae. Presence of acid–fast bacilli is the diagnostic feature of other granulomatous infectious diseases such as Tuberculosis and leprosy. Efared *et al*. in 2017 applied immunochemical histiocyte staining for CD68 to rule out the diagnosis of certain lymphomas or malignancies which may mimic the morphology of RS.[5]

Routinely followed treatment for management of RS includes surgical correction and antimicrobial treatment with ciprofloxacin, streptomycin, rifampicin, sulfonamides, clofazimine, or fluoroquinolones.[7] Treatment must be continued for months, which frequently leads to poor patient compliance. Recurrence is quite common in cases of RS and its chronicity also plays a pivotal role in unsatisfactory prognosis of the disease.

Conclusion

RS usually presents with very deceptive clinical features which often may lead to its misdiagnosis. This report presents one such incidence of this disease with widespread involvement including oral cavity. In this scenario, histopathology has proven to be an excellent aid in clearing the dilemma surrounding the diagnosis of the condition. Currently, newer advances in the diagnosis by serotyping and immunohistochemistry are being explored which will lead to more prompt diagnosis thus resulting in better prognosis.

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Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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