Case Report

Rhinocerebral mucormycosis associated with actinomycosis in a diabetic patient: A rare presentation

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Abstract

Mucormycosis is an opportunistic fulminant fungal infection which mainly affects the immunocompromised individuals. It begins in the nose and paranasal sinuses due to the inhalation of fungal spores. The common predisposing factors include diabetes mellitus and immunosuppression. Actinomycosis is a bacterial infection caused by nonspore-forming, anaerobic or microaerophilic bacterial species of the genus *Actinomyces*. It is a suppurative and chronic granulomatous disease characterized by abscess formation, tissue fibrosis and draining sinuses rarely diagnosed in humans. A case of rhinocerebral mucormycosis associated with actinomycosis of the maxilla involving the palate in an uncontrolled diabetic patient is reported.

Keywords: Actinomycosis, diabetes mellitus, maxilla, mucormycosis

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Received: 28.03.2018, **Accepted:** 29.12.2018

INTRODUCTION

Mucormycosis is an opportunistic fungal infection caused by fungi of the order Mucorales.[1] The first case of mucormycosis was reported by Paultauf in 1885.[2] These infections are usually noticed in individuals with immunocompromised status and they generally have uncontrolled diabetes mellitus, hematological malignant disease like leukemia or are on immunosuppressive therapy.[1]

Mucormycosis represents the third most common angioinvasive fungal infection.[3] This fungus occurs in soil, manure, vegetable, fruits and as bread mold.[4] The infection begins in the nose and paranasal sinuses (PNSs) due to the inhalation of fungal spores. The infection can spread to orbital and intracranial structures either by direct invasion or through the blood vessels.[5] The fungus invades arteries leading to thrombosis that subsequently causes necrosis of hard and soft tissues.[6]

Actinomycosis in man was first described by Von Langebeck, in 1845 which was attributed to a fungus. Later in the 1960s, Waksman showed that *Actinomyces* was a Gram-positive bacteria.[7] Actinomycosis is a suppurative and chronic granulomatous disease characterized by abscess formation, tissue fibrosis and draining sinuses rarely diagnosed in humans.[7,8] It is caused by nonspore-forming, anaerobic or microaerophilic bacterial species of the genus *Actinomyces*.[8] It is caused by traumas of the orofacial region, extraction of a tooth or through an infected pulp cavity.[9]

A rare case with combination of rhinocerebral mucormycosis associated with actinomycosis of the

Access this article online

Quick Response Code:

Website: www.jomfp.in

DOI: 10.4103/jomfp.JOMFP_77_18

How to cite this article: Rani SU, Sivaranjani Y, Kumar MP, Rao GV. Rhinocerebral mucormycosis associated with actinomycosis in a diabetic patient: A rare presentation. J Oral Maxillofac Pathol 2019;23:S122-5.

maxilla in a 63-year-old male patient with uncontrolled diabetes is reported.

CASE REPORT

A 63-year-old male reported to the outpatient department with a complaint of bad breath and pain in the upper right back teeth region for 3 months. Initially, the patient was asymptomatic and then gradually noticed radiating pain to the upper jaw, nose region and the forehead. The patient is a known diabetic and hypertensive for 2 years and is under medication. Dental history revealed that he underwent extraction 15 years back. No relevant family history was reported.

Extraoral examination revealed a diffused swelling with ill-defined borders [Figure 1]. Bilateral submandibular lymph nodes were palpable, which were firm and tender on palpation.

Intraoral examination revealed bone erosion of the maxillary arch with pseudomembranous slough. Necrotic bone was seen in the region of the palate and in the maxilla region extending from 18 to 26 region, and exposure of palate in the midline was also noticed. Missing teeth seen in relation to 11, 12, 13, 14, 15, 17, 18, 21, 22, 23, 25 and 28 [Figure 2].

Investigations were performed. The patient's blood glucose levels were tested and the fasting blood glucose level was found to be 230 mg/dl, whereas the postprandial blood glucose level was 380 mg/dl. Computerized tomography scan revealed erosion of anterior maxilla and midpalatal region [Figure 3]. PNS view revealed moth-eaten appearance involving the upper anterior alveolar ridge [Figure 4].

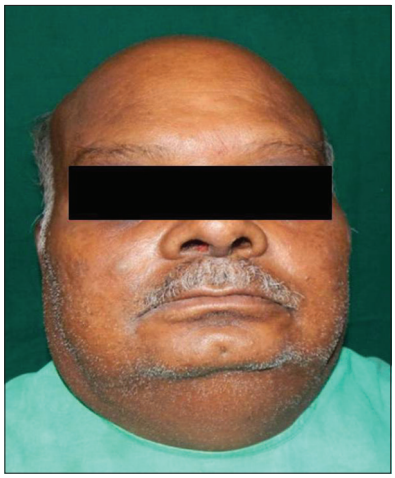

Figure 1: Extraoral picture revealing diffused swelling with ill-defined borders

An incisional biopsy was performed. Histopathological examination revealed numerous thick-walled, irregularly branching nonseptate hyphae in the background of necrotic tissue at the periphery of the bony trabeculae [Figure 5a] and was seen invading into the small blood vessels, which is suggestive of mucormycosis, and the tissue showed a peripheral band of fibrosis encasing a zone of chronically inflamed granulation tissue surrounding large collections of polymorphonuclear leukocytes and colonies of microorganisms. These colonies consist of club-shaped filaments that form a radiating rosette pattern suggesting actinomycosis [Figure 5b]. Surgical excision of necrotic bone and adjacent soft tissue was done and sent for histopathological examination [Figure 6]. Histopathologic findings were in accordance with the incisional biopsy, and a final diagnosis of mucormycosis associated with actinomycosis was given.

DISCUSSION

Mucormycosis is the third invasive mycosis caused by fungi of the class zygomycetes.[3] Based on the clinical presentation and anatomic site, mucormycosis can be divided into at least six clinical categories: (1) rhino-orbito-cerebral (44%–49%), followed by (2) cutaneous (10%–19%), (3) pulmonary (10%–11%), (4) disseminated (6%–11%), (5) gastrointestinal (2%–11%) and (6) miscellaneous.[5,10] Mucormycosis is caused by Rhizopus arrhizus species. Mucormycosis in diabetic patients has the ability to produce enzyme ketoreductase which later utilizes patient's ketone bodies for their nutrition.[6]

Rhinocerebral mucormycosis usually present with malaise, headache, facial pain, swelling and low-grade fever. The disease usually begins in the nasal mucosa or palate and extends to the PNSs spreading through the surrounding vessels.[11] If it invades the mouth, a black, necrotic eschar is found on the palate, and ischemic, necrotic turbines may be found in the nose.[12] In addition, mucormycosis can involve the retro-orbital region by direct extension. As

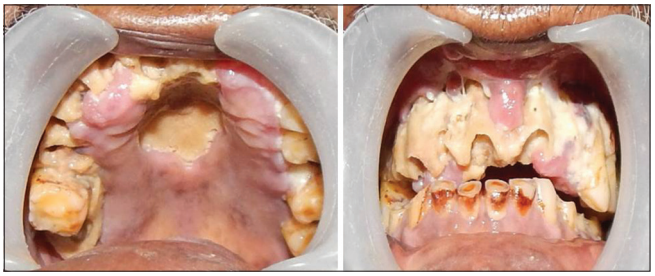

Figure 2: Intraorally necrotic bone was seen in the region of the palate with bony erosion of the maxillary arch

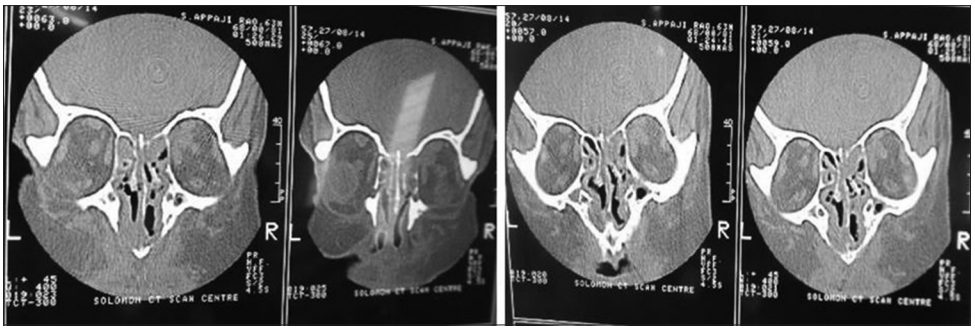

Figure 3: Computerized tomography scan revealed erosion of the anterior maxilla and midpalatal regions

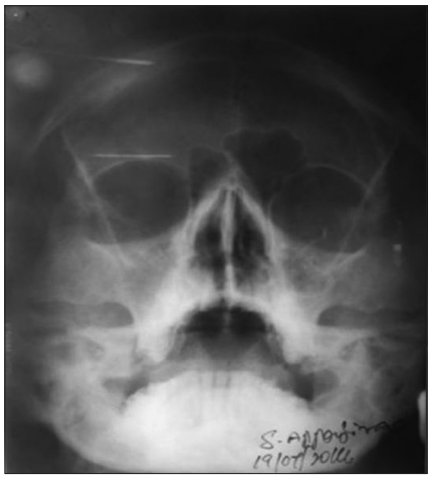

Figure 4: Paranasal sinus view revealed moth-eaten appearance involving the upper anterior alveolar ridge

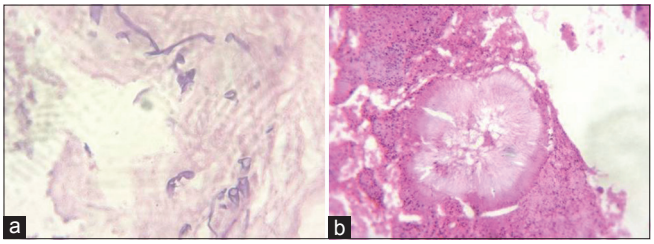

Figure 5: (a) Histopathology revealing numerous thick-walled, irregularly branching nonseptate hyphae in the background of necrotic tissue. (b) Club-shaped filaments forming a radiating rosette pattern

mucormycosis develops, the fungal hyphae start to invade local tissue.[11]

Mucormycosis of the oral cavity can originate from disseminated infection where the portal of entry is by inhalation (usually through the nose) or can also result from direct wound contamination with dissemination to other viscera as a common complication. When arising from the nose and PNSs, the infection may cause palatal ulceration progressing to necrosis.[13]

Histologically, mucormycosis is characterized by extensive tissue necrosis and the presence of numerous, large (5–30 μm), thin-walled fungal hyphae, which are nonseptate, branched at right angles and have a ribbon-like appearance.[12] There is a close histopathological resemblance between the fungal hyphae of mucormycosis and aspergillosis. The differentiating feature is that the hyphae of mucormycosis are nonseptate and branch at right angles, whereas the hyphae of aspergillus species are septate, smaller in width and branch at more acute angles.[3]

The *Actinomyces* species are Gram-positive, pleomorhic and diphtheroidal, or more commonly, delicately filamentous. The cervicofacial, thoracic and abdominopelvic regions and central nervous system are most commonly involved in actinomycosis. Actinomycosis usually occurs in immunocompetent persons but may occur in persons with diminished host defenses.[8]

The common initial symptoms of infection are sudden onset of cervicofacial pain, swelling, erythema, edema and suppuration. Rarely regional lymphadenopathy is seen. However, lymphadenopathy and abscess formation are seen due to secondary infection.[9] Cervicofacial actinomycosis is the most common form of actinomycosis. The most commonly involved sites are submandibular space, cheek, parotid gland, teeth, tongue, nasal cavity, gingiva, etc.[7] In general, the disease has a peak incidence in the fourth to sixth decades of life with a slight male predominance. The mandible is more commonly involved than maxilla (4:1).[14] Similarly, this patient was of middle age. Although this case was immunocompetent, actinomycosis has a predilection for causing infection in immunocompromised hosts such as malignancy, immunosuppressive drugs and diabetes.

Sulfur granules, biopsy regimens and pathologic investigation are the most significant factors in the diagnosis of actinomycosis.[7,9] In the present case, we could not demonstrate sulfur granules probably because the infection was just transitory between the acute and chronic forms, and the mineralization might not have taken place. Further, the periphery of the central core of

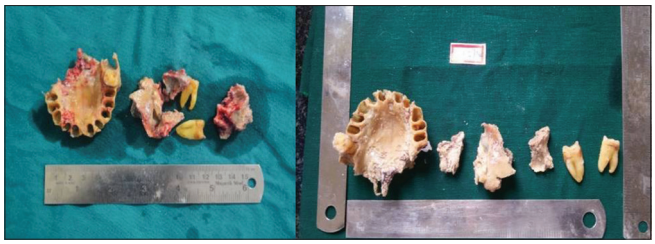

Figure 6: Surgical excision of necrotic bone and adjacent soft tissue

the rosette stained basophilic, whereas the peripheral clubs stained eosinophilic red. This established finding from other studies helped us to arrive at a definitive diagnosis.

CONCLUSION

In summary, we present a complicated association of two uncommon diseases. Mucormycosis is a rare condition which may pose a diagnostic challenge for dental surgeons who may not be familiar with its clinical presentations. Actinomycotic infections of the cervicofacial region are uncommon, but they are important in dental practice because they may mimic more common oral disease, primarily dental-related infection mimicking a malignant disease. Hence, a proper understanding of such infections is needed as the underlying systemic conditions at many times may be quiescent and dentists may identify the debilitated status of the patient.

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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