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Association of Actinomycotic Osteomyelitis in the Mandible with Osteopetrosis/Albers-Schoenberg Disease:A Rare Case Report

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ABSTRACT
Osteopetrosis accommodates a clinically and genetically heterogeneous group of conditions that share the hallmark of increased bone density on radiographs. This increase in bone density results from osteoclastic dysfunction. A 42-year-old male reported osteopetrosis complicated by actinomycotic osteomyelitis of the right side mandible. Long-term antibiotic therapy is needed in such patients along with surgical necrotic resection. This case report highlights the association of actinomycotic osteomyelitis with osteopetrosis, which has never been documented so far in the literature. The management and diagnosis of two very rare clinical entities coexisting together is a challenging case for diagnosis and, a very prompt treatment which is for complete healing. Therefore, we are discussing the diagnosis and management of a supportive type of actinomycotic osteomyelitis lesion that has already been diagnosed with osteopetrosis.
Keywords: Osteopetrosis,actinomycotic osteomyelitis, long-term antibiotic therapy

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INTRODUCTION

"Marble bone disease,Osteosclerosis fragilis generalist [1], and Albers-Schoenberg Disease" are the other names of Osteopetrosis which was first described by the Germanradiologist Albers Schoenberg in 1904 [2]. It is characterized by an increase in bone density as reabsorption decreases by osteoclasts,which leads to various defective architecture (brittle bone) such as frequent fractures [1].Many complications occur in Osteopetrosis patients and the most common is Osteomyelitis of the mandible. Actinomycosis is gram-positive bacteria mimicking fungal infections which affect the mandible more commonly and its diagnosis is challenging as it resembles granulomatous disease and neoplasm clinically and radiographically. Hence the title of"Great masquerader of head and neck"region is given to this clinical entity [3].Further to our knowledge based on previous literature, the association of actinomycosis osteomyelitis is rarely seen with osteopetrosis disease. The early diagnosis of two such clinical entities which rarely coexist to gether is a real challenge for diagnosticians as a delay in diagnosis will further determine the condition of the patient, leading to a very poor prognosis. Hence, this article puts great emphasis onthe need for careful clinical, radiographical and histopathological examination to achievean accurate diagnosis and best management modality for such cases to have a better prognosis and a, very less chance of relapse of such infection in osteopetrosis cases with thorough evaluation along with broad-spectrum antibiotics with weeklycurettage, irrigation and drainage of pus from the affected site leaving any foci of infection can lead to a further complication during and post-treatment.

CASE REPORT

A 42-year male patient reported with the chief complaint of pain and swelling in the lower left back tooth region for 5-7 days with a history of uncontrolled diabetes mellitus type II. 9years back he had a fracture in both femur bones while walking and was admitted to the hospital for the same where he was diagnosed with a very rare disease of bone marble bone disease (osteopetrosis)/Albers-Schoenberg Disease. While

taking history he mentioned that his sister had also multiple fractures and suffered from a rare similar condition. There was a history of tuberculosis running in the family which strongly affected his grand parents and parents. Atthetimeofextra-oral examination, diffuse swelling with intact overlying skin was seen on the right side of the face of size 3x4 cm approx. and it involves the inferior border of the mandible which elongates from the corner of the mouth to the angle of the mandible anterior-posteriorly. (Fig 1) Multiple right side submandibular lymph nodes were palpable, tender, and had a matted feel on palpation On Intra oral examination root stump wrt 46 was appreciated with active pus drainage and vestibular obliteration and tenderness. Expansion of the buccal cortical plate was also noticed (Fig 2). After a thorough intra-oral examination and extra oral examination following investigations were advised which include, Panoramic radiograph, CBC, KFT, and LFT, HbA1c was high, Vitamin-D was decreased, and Serum Acid phosphatase level, and Creatinine Kinase level. (All other blood investigations were within normal range).

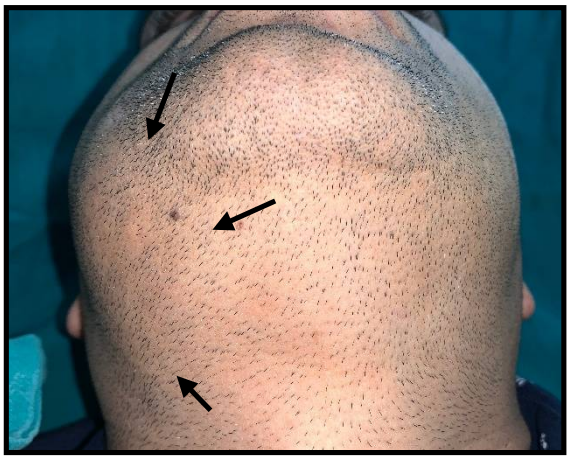

Fig 1. Shows diffuse swelling with intact overlying skin on the right side of face of size 3x4 cm approx.

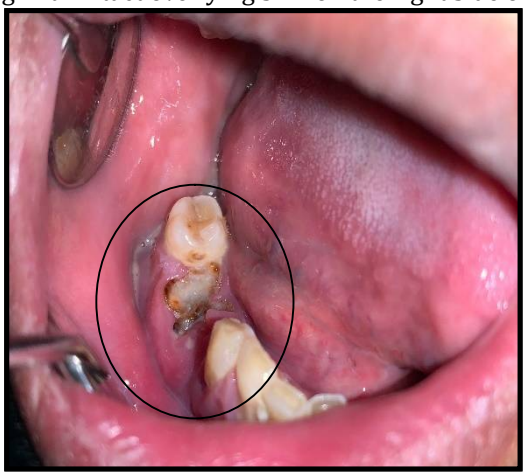

Fig 2. Shows root stump wrt 46 was appreciated with pus discharge and vestibular obliteration.

A panoramic radiograph revealed a well-defined scooped-out radiolucency in the body of the mandible associated with root pieces 46 and 47. The radiolucency was extending from the edentulous area of 45 to the distal of 47. The internal structures have a typical moth-eaten appearance with an area of ill-defined sclerotic bone associated with apical 2/3rd of 47. There was the involvement of the mandibular canal which can not be appreciated. (Fig3)

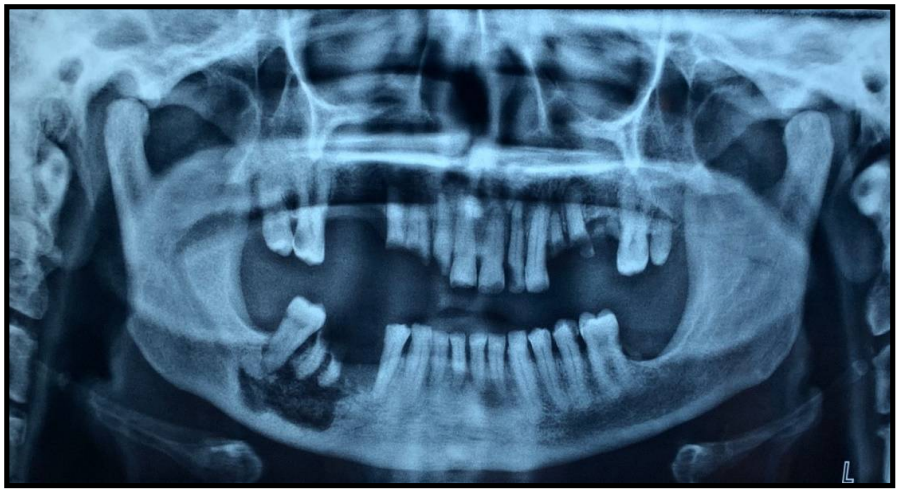

Fig 3 Shows a well-defined scooped-out radiolucency in the body of the mandible associated with root pieces 46 and 47

Advanced imaging CBCT was taken post-operatively after extraction of the involved infected teeth 46,47 and also irrigation and saucerization of dead bone. A well-defined mixed hypodense and hyperdense lesion with HU ranging from 89 to 450 were appreciated. The size of the lesion was 23.9mm anterio-posteriorly, 11.9mm buccolingually, and 16.9mm superior-inferiorly.(Fig 4 a,b) The change in a trabecular pattern depicting"cotton-wool appearance"was appreciated.It has involved superior and inferior border of the mandibular canal which cannot be appreciated on the radiograph.(Fig 4c).

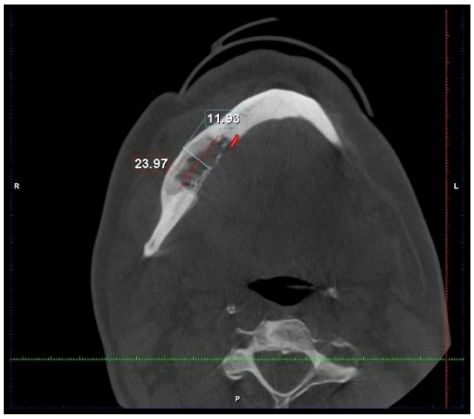

Fig4(a): Axial section shows lesion measuring 23.9 mm anterio-posteriorly and11.9 mm buccolingually.

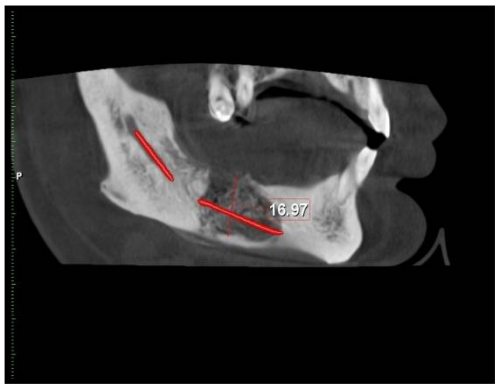

Fig 4(b) showing lesion measuring 16.9 mm superior-inferiorly

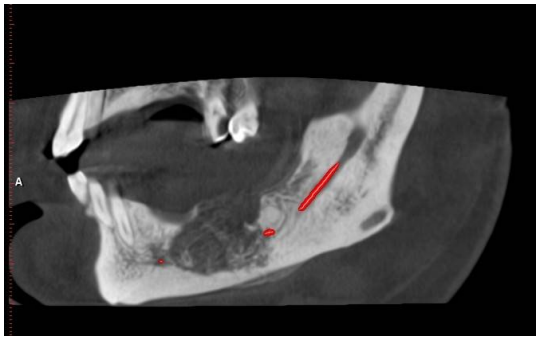

Fig 4(c): Shows a well-defined mixed hypo dense and hyperdense lesion with change in trabeculae pattern depicting "cotton-wool appearance "involving superior and inferior border of the mandibular canal.

After all thorough conventional and higher imaging investigations diagnosis of actinomycoticosteomyelitis was given.An incisional biops yollowed by extraction wrt46 and 47 was done. The histopathological section which show bony bits and soft tissue sections shows granulation tissue, bony spicules, and inflammatory cellslong with clones of actinomycosis suggestive of actinomycotic osteomyelitis of thejaw.(Fig 5)

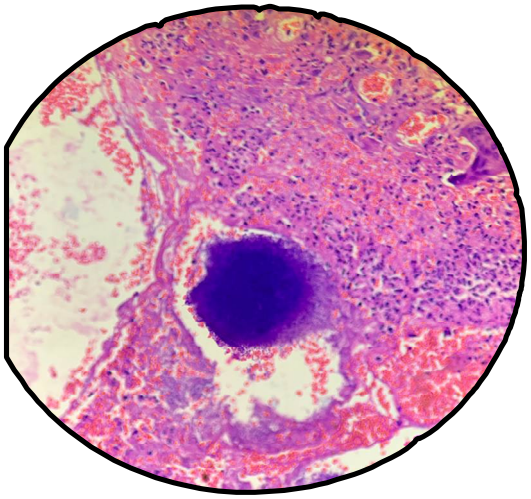

Fig 5. showing granulation tissue, bony spicules, and inflammatory cells along with clones of actinomycosis suggestive of actinomycotic osteomyelitis of the jaw.

The patient has advised along-term broad spectrum antibiotic i.e.Augmentin 625mg TDS along with weekly curettage and irrigation. Follow-up was done weekly for 2-3 months regularly.

DISCUSSION

Osteopetrosis is a set of uncommon bone problems in the circle of relatives of sclerosis bone dysplasia, characterized by using reduced osteoclastic bone resorption that affects excessive bone mass. It incorporates a scientific extent in the range from very mild to excessive disorder phenotype that is fatal with in the 1styear of life [4]. It is subdivided into three categories based on inheritance osteopetrosis with autosomal dominant, osteopetrosis autosomal recessive, and X-linked recessive [1, 4]. Most common in these types is autosomal dominant which shows mild symptoms and consider in "benign" form with an incidence of 1:20,000, autosomal recessive considered in"malignant"infantile form as the patient has a short life expectancy with an incidence of 1:200,000 [1, 4]. Extremely rare form is X-linked recessive [1].In the literature,only the association of osteomyelitis of the jaw with osteopetrosisis documented. It is most often seen in the mandible, which is mostly associated with odontogenic infections and oral tactics.It might also go away because the necrotic bone is exposed and hard to remove due to reduced blood supply and often accompanying anemia and neutropenia [4]. The extraction must be approached judiciously because the carious tooth can be a precursor to osteomyelitis [4]. Actinomycosis is an agreed word which comprises of "Aktino"whichmeanstheradiatinglookof sulfur granules and "mykos"which labels the condition as mycotic sickness. Bollinger defined the organism actinomycosis bovidandits potential to motive"lumpy jaw"infarm animals. In 1848 it was first described by Von Langenback in humans. It was

declared gram +ve in 1960 by Waksman. It usually influences among 3rd and 6th decade of life with particular male predilection (4:1) [6].

Actinomycotic osteomyelitis is uncommon which results in about 12%of total osteomyelitis [6].Cape in 1938 cautioned that the infection can be categorized anatomically as cervicofacial (i.e lumpy jaw),thoracic,or stomach.Cervicofacialis the most not unusual among all and debts for greater than half of the mentionedinstances [7].Unidentified mass, facial swelling, or persistent contamination particularly after endodontics therapy or teeth extraction, regardless of its non-traumatichistory is suggestive of actinomycosis.

Actinomycotic osteomyelitis has manifestations inclusive of paresthesia and after root canal treatments and extraction, it canlead to pathologic fracture of bones, and chronic infection without any history of trauma. In the present case, the affected person had signs and symptoms such as pus associated with root piece[4, 6] and swelling with nohistory of extraction and trauma.

The diagnosis of actinomycotic osteomyelitis of ten depends on medical,radiographic, and microscopic findings. On clinical examination, the disease may be painless orpainful and mimic one-of-a-kind benign or malignant situations. This organism is technically hard in culturing because it calls for an anaerobic environment. Biopsy and histopathological examination are strongly advocated for analysis, which were used for the final prognosis of our patient

Radiologic capabilities depend upon the type and level of the disease,It varies from diffuse lytic adjustments with fuzzy and instinct bony trabecular to scattered patchy, sclerosis of bone often which is expressed as a"cotton-wool" appearance [7]. In our present case panoramic view reveals the classic"cotton wool" appearance in the right mandibular region.

Differential diagnosis with differentiating features include fibro-osseous lesion (ray fungal colonies), bone tumors (active osteoblasts are absent and ray fungal colonies present), chronic granulomatous lesion(ray fungal colonies present, absence of long hand's giant cell, steroid bodies, lepra cells, histiocytes, and Schaumann bodies, Chronic osteomyelitis(ray fungal colonies presence) and Nocardia (granuloma present) [5].

Well-known radiographs for the diagnosis of osteopetrosis is pathognomonic and show parallel bands of dense bone can provide the advent of "bone-within-bone" or "endobones. prominent in long bones.4 Inside the case provided, a 42-year-old man suffered from mandibular actinomycotic osteomyelitis which was associated with tooth extraction, bone trimming, and antibiotic treatment which helps in stabilizing the infection. But, pus drainage and the risk of recurrence of infection are excessive because of the ischemia continuity and infection of the bone. The chance is to maintain normal affected person comply with-up in our department.

Regardless of the context, patient who is suffering from osteopetrosis must be controlled comprehensively,with a multi-disciplinary approach. Sufferers have immune fragility with, in particular, anemia, hypocalcemia, and thrombocytopenia. First off, theseneed to be identified and corrected to enhance sufferance and efficacy of destiny medical care. The drug of choice in antibiotic therapy is amoxicillin with clavulanicacid followed by clindamycin. Other than this local healing can be promoted byhyperbaric oxygen therapy and might be used earlier than or after surgical remedyCombining curettage, sequestrectomy, and minimum bone trimming is theconservative surgical management related to overlaying flaps. To reconstruct bone defects.vancomycin with calciumsulphatecan beused. Actinomycotic osteomyelitis is an extremely rare disease with an association in osteopetrosis withn on specific signs and symptoms and making the prognosis challenging.

CONCLUSION

Early diagnosis can deliver a set-off remedy and as a result higher out comes and a better prognosis of the disease. Any suspected medical lesion has to without a doubt go for incisional biopsy as histopathological prognosisis the"benchmark" for the disorder and the false poor result of microbiological cultures may also obscure the best and most conclusive analysis. A proper follow up which includes regular debridement of the infected site and antibiotic coverage is the key to the healing of such lesions.Also, ensuring to come to a proper diagnosis or thorough clinical evaluation with complete laboratory /blood examination follow by conventional radiography to advance imaging modalities can help in having the proper extent of the lesion. This further aids in lesser post-operative care and complications.

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