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Case Report

Recurrent osteomyelitis in patient of osteopetrosis: A rare case report and management

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ABSTRACT

Introduction: Osteopetrosis is a rare heritable disease. It sometimes presents as incidental finding with no symptoms to even life threatening complications. It may present as multiple fractures, compressive neuropathies, hypocalcaemia, and life-threatening pancytopaenia.
Case Report: In this article, A 15 years old female reported for evaluation and treatment of pain and pus discharge in the left side of mandible since 1 year after the extraction multiple deciduous tooth. pus discharge in the left side of mandible with draining fistula. Intraoral examination revealed exposed necrotic bone in mentioned region.
Discussion: osteopetrosis of mandible may develop osteomyelitis. The osteomyelitis is usually due to tooth extraction or pulpal necrosis. However, its treatment is controversial.
Conclusion: Treatment regimens include high-dose systemic antibiotics along with thorough debridement of necrotic bone and primary closure of soft tissues & may require Hyperbaric oxygen.

1. Introduction

Osteopetrosis is variability referred to a "Marble bone disease" aur Albers Schonberg disease, after the German radiologist describe it firstly in 1904.[1] The Greek word 'osteo' means 'bone 'and 'petros' means 'stone'. In this disease cortical bone density increased at the expense of medullary bone. Term osteopetrosis given by Karshner in 1926.[1–4] It is a rare hereditary bone disorder which was different range of clinical features and is characterized by an increase in bone density and reduction of bone marrow spaces. That results a defective osteoclast functioning and accordingly decrease in bone remodelling.it has been shown that the normal count of osteoclast are present but have defective functioning in this disease osteoclastogenesis and remodeling of the bone is affected.[1,4]

Osteopetrosis has two varieties as malignant osteopetrosis that is transmitted as mendelian recessive trait X-linked manner which is seen immediately at birth or during early childhood. The patients suffering from malignant osteopetrosis have a bone marrow defects and increased bone density.[2,5] The growth of the bones is impaired resulting in short stature. Frontal bossing develop within the first year resulting in typical facial appearance.[5,6] The skull changes may lead to choanal stenosis and hydrocephalus the expanding bone can have narrow nerve foramina that results in blindness, deafness, facial palsy.[7] Hearing loss, Tooth erruption defects and severe dental carries.[8] Children develop risk of hypocalcemia with attendant tetanic seizures and secondary hyperparathyroidism. Most severe complication is bone marrow suppression.[1,3] Patient suffers from hepatosplenomegaly, anemia, thrombocytopenia, and

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neurological manifestation like optic atrophy. Due to anemia or secondary infections which is around 1 year duration the life expectancy is very less in such patients. The treatment of choice available for such patient is allogenic hemopoietic stem cell transplantation. It should be offered at earliest possible time to decrease the further complication.[2,4]

The second variety of osteopetrosis is benign osteopetrosis. It is transmitted as a mendelian dominant trait. It is diagnosed at the age of 30 or 40 years by radiographic findings. Accordingly to clinical and radiological signs there are two varieties of this disease. That are autosomal dominant osteopetrosis type 1 (ADO I) and autosomal dominant osteopetrosis type II (ADO II).[2,3]

ADO type 1 is characterized by pronounced and symmetrical osteosclerosis of the skull and enlarged thickness of cranial vault. The fracture rate of this type is negligible.[1]

In ADO type 2 there is less sclerosis of the skull and is more prominent or pronounced in the base. Clinically ADO type 2 are dominated by long bone fractures. It can be occur with or without trauma. In 78% of the patient with ADO type 2 includes hip osteoarthritis, facial nerve Palsy and osteomyelitis of mandible.[1]

There is one more type of osteopetrosis, described by Beighton et al. That is intermediate type of osteopetrosis. It is more frequent in practice. Tips and Lynch reported no racial or sexual predisposition. Osteoporosis is caused by failure of osteoclast differentiation or functions or mutations in at least 10 genes have been identified as causative in humans.

Osteoclasts are highly specialised cells, which degrade bone minerals and organic bone matrix. This processes are crucial for the bone remodelling and the maintenance of the bone mechanical stability and mineral homeostasis. Due to defective osteoclastic function relative bone deposition rate is more than bone resorption results in excessive formation of immature bone, cortical bone thickening and narrowing or obliteration of medullary cavities. For bone resorption into the extracellular space osteoclast release the necessary lysosomal enzymes.[1]

Mutation and defect in different genes leads to phenotype with osteopetrosis. This defect includes mutations in gene encoding carbonic anhydrase 2, the proton pump gene and chloride channel gene. Now a days the immune system responsible in the pathogenesis of various metabolic bone diseases including osteopetrosis.

2. Case Report

A 15 year old female patient came to our institute with the chief complaint of pain and swelling over left side of face since last 4 years. On examination patient had a diffuse swelling over left side of face. Patient also had draining sinus tract over the site of swelling. Frontal bossing and a flat nose were observed. The patient's height was 150 cm, and her body weight was 33 kg. Because of the short stature of the patient, her head appeared disproportionately large. On intraoral examination sequestrum was seen on left side of mandible and also had multiple unerupted teeth.

Medical history revealed that patient was diagnosed with Osteomyelitis 5 years back and sequestrectomy was done on the left side of mandible. There was no relevant family history. Radiographs of chest, skull, long bone and mandible indicated an increased density throughout the ribs, calvarium and facial bones. A panoramic radiograph revealed dense sclerotic bone in the whole mandible, with areas of lytic destruction typical of chronic Osteomyelitis. The maxilla was characterized by increased radiodensity. There were un-erupted molars in the left and right posterior areas. Therapeutic management of the chronic.

Osteomyelitis was considered and the following treatment plan was selected-

1. Antibiotic therapy guided by the findings of bacterial culture and sensitivity tests.
2. Conservative surgical debridement or sequestrectomy.
3. Removal of extraoral and intraoral sinus tract followed by primary closure.
4. A series of hyperbaric oxygen therapy.

The potential benefits and risks of hyperbaric oxygen therapy were explained to the patient and consent for the same was obtained.

32 hours sessions of hyperbaric oxygen (2 ATM pressure) were administered over a 30 day period in conjugation with cephalexin, 1 gm three times a day. Clinical examination following this period revealed moderate improvement, with drainage ceasing both extraorally and intraorally. However there was continued presence of exposed bone intraorally on left side of the mandible. The bone appeared avascular but was not demarcating or forming sequestrum. Bone biopsy was avoided in this patient coz of poor healing capacity of bone.

3. Discussion

Patients with osteopetrosis frequently visit the dentist for a variety of consequences, the most prevalent of which is dental caries. Other complications include enamel hypoplasia, malformed roots and crowns, and thickening of the lamella dura. Bone necrosis and dental caries are caused by the narrowing of the canals that house the neurovascular bundles that feed the teeth and jaws, as well as by the destruction of the marrow cavities and dental pulp chambers.[3] In 10% of instances, osteomyelitis will eventually result from these conditions. Because anaemia and neutropenia are present, osteomyelitis may be severe and have a long course.[1]

Fractures may occur even in minor incidents due to the shift in bone structure's pronounced inclination towards

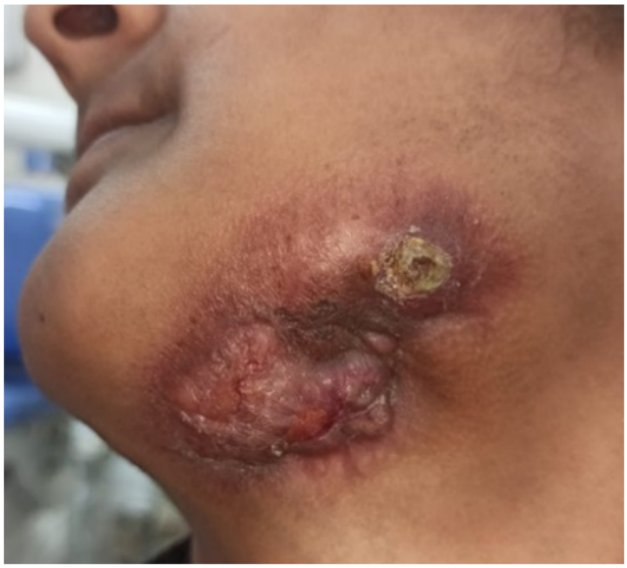

Fig. 1: Extraoral draining sinus

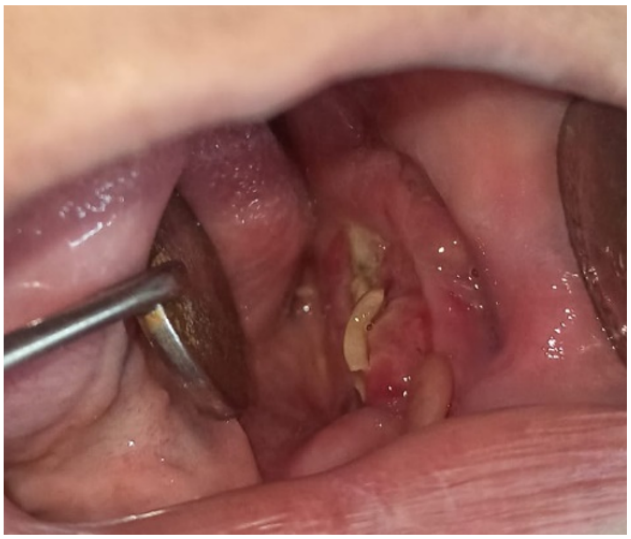

Fig. 2: Intraoral exposed sequestrum

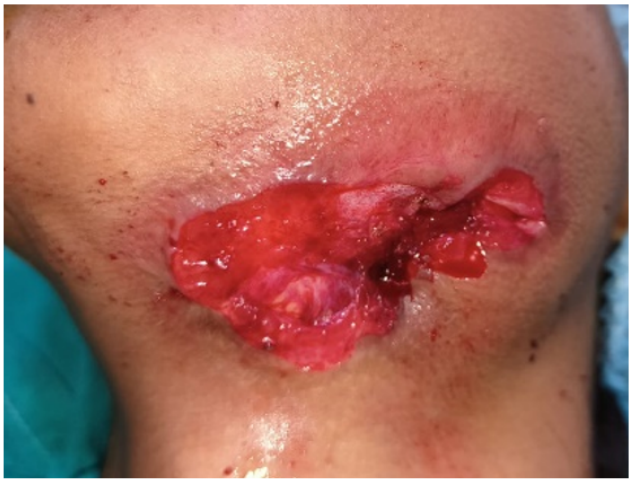

Fig. 3: After sinus tract removal

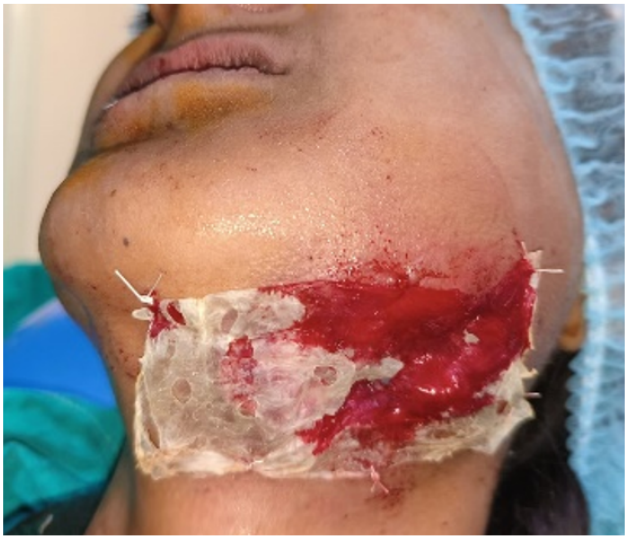

Fig. 4: Secondary closure using collagen sheet

fragility.[3,5] Patients with osteopetrosis may experience pathologic fractures as a result of structural weakness brought on by poorly organised bone and persistent deposits of immature bone and calcified cartilage. The phrase "marble bone" refers to the sclerotic and opaque radiographic changes that occur as the disease progresses in the bones.[4,5]

10% of people with osteopetrosis go on to develop osteomyelitis, which typically affects the jaw. Osteomyelitis is typically brought on by pulpal necrosis or tooth extraction. Lack of appropriate bone vasculature is assumed to be the main factor contributing to the higher likelihood of infection. Large volumes of poorly vascularized bone and progressive marrow space obliteration in the afflicted areas make infection treatment challenging. Bony sequestrum and multiple draining fistulae are frequent clinical findings.[8] As a result of the maxilla's thin cortical bone and abundant collateral blood flow, osteomyelitis of the maxilla is extremely uncommon. Treatment for osteomyelitis brought on by osteopetrosis is debatable High-dose systemic antibiotics are often combined with extensive debridement of necrotic bone and, when practical, primary closure of soft tissues in treatment plans.[2–4] Chronic osteomyelitis has been treated with hyperbaric oxygen (HBO).[3,4] Enhanced leucocytic killing, osteoclastic resorption of the dead osteomyelitic tissue, fibroblast division, collagen production, neovascularization, and enhanced permeation of specific antibiotics (aminoglycosides) across bacterial cell walls within the necrotic tissue are some of the mechanisms

by which HBO works to treat osteomyelitis.[1]

Its activity is heavily dependent on oxygen since osteoclasts are 100 times more metabolically active than osteocytes.[3] There are few examples showing that treatment is effective; osteomyelitis frequently persists for a very long time.[2,8] In a review of 57 cases of osteomyelitis brought on by osteopetrosis, it was discovered that the majority of cases were chronic and treatment-resistant.[2] Unfortunately, there doesn't appear to be a permanent cure for maxillary or mandibular osteopetrosis short of total bone removal. Patients with osteopetrosis should be urged to maintain good dental hygiene and oral health in order to reduce such issues, as there is a chance that surgical operations could promote osteomyelitis.[6] Management of osteopetrosis has to be individualised because of the wide spectrum of clinical symptoms and complications. Medical management of osteopetrosis revolves around modulation of the osteoclasts, either to stimulate the remaining host osteoclasts or to provide an alternative source of the same. Restriction of calcium intake, high-dose calcitriol therapy, steroids, parathyroid hormone and recombinant human interferon gamma-1b, have all been attempted to stimulate the host osteoclasts with variable success rate.

Although the adoption of a microvascularized osseous free flap may be advantageous, it may not be possible in these patients due to the lack of a suitable donor location.[1]

The best option seems to be palliative care, which includes nerve decompression and debridement. Preventive management combined with regular dental care appears to be the best course of action. Try endodontic therapy first before extracting teeth since periosteal bone stripping raises the possibility of asymptomatic bone becoming necrotic and sequestering. Only extremely necrotic bone should be removed through sparing flap dissections during any debridement, which should be as minimal as possible. Due to the systemic nature of the disease, there are typically no visible differences between affected and healthy bones.[9,10]

4. Conclusion

It is an uncommon condition in which there is inadequate bone remodelling.

5. Source of Funding

None.

6. Conflict of Interest

None.

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