Osteopetrosis-An Uncommon Entity With Recurrent Osteomyelitis of Mandible

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Abstract: Osteopetrosis is an uncommon genetic failure arising from congenital defect in the development or function of the osteoclasts, as a result, fragile bones which are sensitive to fracture and infection occur. As a complication of osteopetrosis, mandibular osteomyelitis is important. This case report presents a case of osteopetrosis with recurrent osteomyelitis of the mandible in a 42-year-old male.

Key Words: Osteopetrosis; Osteomyelitis; Osteosclerosis; Mandible

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I. Introduction

Osteopetrosis is an uncommon hereditary and familial bone disease characterized by dense and fragile bones which are capable of fracture and infection. In this disease, the lack of resorption of normal primitive osteochondrous tissue by the osteoclast is the primary pathology. The diminution of this primitive tissue hinders the formation of normal mature adult bone. This bone disease contains dense and fragile bones, these abnormal bones are tenderin fracture and infection because of the fault of bone remodeling. (1) Firstly, Albers-Schönberg in 1904 mentioned this bone disease as postponed physical development allied with bone fragility. According to most studies, osteopetrosis is related to some situations such as hepatosplenomegaly, increased possibility of infection, anaemia, respiratory tract infection, cardiac failures and multiple fractures. (2)

Osteopetrosis is an uncommon disease that has an incidence of 1 in 250 000 births. In ten per cent of osteopetrosis cases, osteomyelitis generally develops in the mandible. In maxilla, development of osteomyelitis infrequently occurs, likely due to the thin cortical bone and rich collateral blood supply. In osteopetrosis cases, there is lack of bone marrow cavity in abnormal bones, so that there is fault to withstand normal physical loads, and bones are capable of fractures because of the lack of remodelling. (3)

The osteomyelitis is frequently caused by tooth extraction or pulpal necrosis. The high point reason of the increased rate of infection is thought to be a lack of adequate bone vasculature. (4) Here we present a case of osteopetrosis associated with recurrent osteomyelitis of the mandible in 42-year-old man.

II. Case Report

A 42-year-old male was referred to the Department of Dentomaxillofacial Radiology in Mustafa Kemal University with chief complaints of chronic abscess in the left side of the face. It was learned that osteopetrosis was diagnosed when the patient was about 10 years old. Additionally, it was learned that the patient had aplastic anemia in his anamnesis. The patient explained that the swelling started 3 years ago after the extraction a molar of the mandible and it was treated in Department of Oral and Maxillofacial Surgery. Corticosteroid, antifungal agent, mouthwash, folic acid, protein support, sodium fusidate and ciprofloxacin had been prescribed. Patient was advised for Panoramic radiography and Computed Tomography for advanced evaluation. On general examination, patient was short statures.

Intraoral examination revealed extensively exposed necrotic bone with sequestrum in the left mandibular molar area (Figure 1). Past medical history revealed the diagnosis of osteopetrosis, with many bone fractures. X-ray AP view of pelvis showed multipl dense regions with femoral fracture and the patient was bearer of a hip joint prosthesis (Figure 2a, b).

Panoramic radiograph showed diffuse osteosclerosis of the maxilla and with indistinct trabeculae and a ‘moth eaten’ appearance of the left ramus and angle of the mandible. Generalized alveolar bone loss, multiple carious teeth, impacted tooth and residual root were also seen (Figure 3).
In CT examination, there was an increase in bone density, with obliteration of the medullary spaces and loss of distinction between the cortex and medulla. A hypodense image showing destruction was seen in the posterior region of the left side of the mandible which was suggestive of recurrent osteomyelitis (Figure 4a,b,c).

The pathological result was reported as inflammatory necrotic bone tissue. After clinical, radiological and pathological examination, the diagnosis was determined as osteopetrosis-associated osteomyelitis.

Infection was suppressed by intravenous antibiotics and mouth rinses. Surgery was planned. Mobile segment of bone was removed and sent for histopathological examination. Bone surface was debrided until healthy bleeding bone was reached (Figure 5a,b). After sequestrectomy and curettage, visits of the patient was scheduled at 1 month intervals for 6 months. At each visit, exposed site was evaluated and saucerization was done when necessary to create a uniform bed. Soft tissues were conserved and no local anesthesia was administrated. Patient was motivated to irrigate the site with povidone-iodine mixed with saline for infection control. The site was covered by mucosa spontaneously at the end of 6th month.

### III. Discussion

Osteopetrosis is an uncommon metabolic bone disease characterized by a generalized increase in skeletal mass because of the congenital defects in the development or function of osteoclasts. There are 3 types of osteopetrosis:

1. Juvenile malignant type: This type is fatal within first few years of life.
2. Intermediate type: It occurs during the first decade of life but it has not malignancy potential.
3. Autosomal dominant type I and type II: There were many orthopedic problems.

In osteopetrosis, the most important factor for healing is connected with the vascularity of the bone. As a result, in patients with osteopetrosis, the healing occurs slowly, the result is frequently unsatisfied, and the bone is predisposed to infection. (7)

Pathological fracture which heals with normal callus is the most observed complication of osteopetrosis but after injury, the onset of callus formation is changeable. (2) In our patient, there was pathological fracture and hip joint prosthesis.

Oral characteristics of osteopetrosis are points to consider, since this disease is frequently diagnosed under favour of oral changes. In patients with osteopetrosis, because of the decreased bone vascularity and damage of white cell function can be the reason of osteomyelitis after dental extractions. The most determined factors are narrowing of canals housing the neurovascular bundles that supply the teeth and jaws. In the 10% of osteopetrosis patients, osteomyelitis occurs and usually happens in the mandible. (8) Osteomyelitis of the maxilla is a very uncommon situation, likely due to the thin cortical bone and rich collateral blood supply. (9) There is reduced blood circulation to bone resulting in a potentially severe infection that runs a protracted course, due to the accompanying severe anemia and neutropenia. (10)

There was also dental pathological changes in osteopetrosis patients. In these patients, incidence of dental caries increase. Delayed eruption and early loss of teeth, enamel hypoplasia, malformed roots and crowns, and thickening of the lamina dura can be the other dental changes. (11)

According to Bakeman et al. (12), osteopetrosis radiographically seems as an increased density of the entire skeleton (e.g., ribs, pelvis, clavicle, femur, skull base, and jaws). In the long bones, increased cortical thickening and decreased marrow space are observed, and there is clubshaped formation. The pelvis and the scapula may have endobone phenomenon. (12) According to only Smith’s study, the entity of hypercementosis that contains different teeth was also reported. (7)

Pyknodysostosis, metaphyseal dysplasia, diaphyseal sclerosis, melorheostosis, osteopetrosis striata, osteopoikilosis, Engelmann’s disease and infantile cortical sclerosis should be considered as important differential diagnoses. (13)

Treatment of the patients with osteopetrosis involves an extensive approach to clinical problems such as hematological and metabolic abnormalities, fractures, skeletal deformity, back pain, bone pain, osteomyelitis, and neurological sequelae. Medical treatment of osteopetrosis is based on efforts to the host mesenchymal cells to differentiate into normal osteoclasts. For stimulation of host osteoclast, calcium restriction, calcitriol, steroids, parathyroid hormone, and interferon have performed. (14) Bone marrow transplantation is the only permanent treatment for osteopetrosis. (15)

Hyperbaric oxygen is useful for the treatment of mandibular osteomyelitis. (14) In osteomyelitis cases, mechanisms of hyperbaric oxygen involve enhanced leukocytic killing, osteoclastic resorption of the dead osteomyelitic tissue, fibroblastic division, collagen production, neovascularisation, and enhanced permeation of certain antibiotics (aminoglycosides) across bacterial cell walls within the necrotic tissue. Osteoclasts are 100 times more metabolically active than osteocytes and its function is highly oxygen dependent. (13)

There are few reports mentioned the success of treatment; in many cases, the osteomyelitis can not certainly be solved. (9, 16) In a review of 57 cases of osteomyelitis depending on osteopetrosis, most cases were determined as chronic and resistant to treatment. (16) Similarly, in our patient, we were faced with recurrent

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osteomyelitis that was treated before. Unfortunately, there isn’t any definitive treatment for osteopetrosis of the maxilla or mandible without complete removal of the affected bone. For reducing the problems, patients with osteopetrosis should be encouraged to maintain good dental care and oral hygiene, because in surgical procedures, these patients have potential risk for development of osteomyelitis.(17)

Use of a microvascularized osseous free flap may be useful but may be impossible due to absence of a suitable donor site in these patients.(9)

Palliative treatment that involves nerve decompression and debridement, seems to be the best way among the other treatment procedures. The best treatment is preventive management by the way of providing routine dental care. Teeth should be endodontically treated, if possible, tooth extraction isn’t supposed, because periosteal stripping of bone may predispose asymptomatic bone to become necrotic and to sequester. Any debridement should be as conservative as possible, only grossly necrotic bone through limited flap dissections should be removed. Because of the systemic disease process, there is frequently no clear delineation between affected and unaffected bone.(8)

As a result, because of the infection risk of the patients, it must be considerablycarefulduringdentalprocedures, especiallythetoothextraction.

IV. Conclusion

Based on clinical and radiological examination, our patient was diagnosed recurrent osteomyelitis of mandible related to osteopetrosis. The patient was referred to Department of Maxillofacial Surgery for further management.

References
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FIGURES

Figure 1: Clinical photograph showing exposed necrotic bone with sequestrum in the left mandibular molar area

Figure 2a. X-ray AP view of pelvis with multiple dense regions and broken screw
b. Femoral 1/3 distal diaphysis fracture in the patient who was treated by the way of osteosynthesis with plate-screw
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Figure 3: Panoramic radiograph showing diffuse osteosclerosis of the maxilla and with indistinct trabeculae and a ‘moth eaten’ appearance of the left ramus and angle of the mandible.

Figure 4: CT showed a generalized increase in bone density and in the posterior region of the left side of the mandible was seen a hypodense image suggestive of recurrent osteomyelitis. 

a. Axial scan b. Coronal scan c. Sagittal scan
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Figure 5:a. Bony sequestrum after curettage
b. Photograph during surgical operation in left mandibular molar area