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Case Report

Osteopetrosis Complicated by Maxillary Osteomyelitis: A Case Report

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Abstract

Introduction:
Maxillary osteomyelitis is a rare phenomenon. If it occurs, evaluation for underlying disease especially osteopetrosis must be considered. Osteomyelitis occurs as a complication in 10% of the cases of osteopetrosis.

Case report:
This is a case report of maxillary osteomyelitis presented in a 15-year old boy with osteopetrosis. In this case, the disease represented mainly with facial pain and swelling and also drainage from facial skin fistulas in bilateraral infraorbital area and lower eyelids. Diagnosis was confirmed by clinical and radiological findings. Treatment consisted of surgical debridment and intravenous antibiotic therapy which led to partial response.

Conclusion:
Maxillary osteomyelitis is a rare phenomenon. If it occurs, evaluation for underlying disease especially osteopetrosis must be considered.

Keywords:
Hereditary disorder, Maxillary bone, Osteoclast, Osteomyelitis, Osteopetrosis

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Introduction

Hence osteomyelitis of the maxilla is very rare, probably due to the thin cortical bone and rich collateral blood supply, the possibility of predisposing immunosuppressive conditions or underlying bony pathology should be considered in the presence of osteomyelitis (1).

Osteopetrosis refers to a group of rare genetic disorders characterized by greatly increased bone density caused by severe impairment of osteoclasts function (2,3). The prevalence of the disease is low (about 0.005% of the general population) (4). Four types of osteopetrosis have been defined; two lethal autosomal-recessive and two benign autosomal dominant forms. Autosomal-dominant type II (marble bone disease, Albers-Schönberg disease) is the most frequent type with a normal life expectancy (3).

Clinical presentation is different and depends on the type and severity of the disease. Pathologic fracture, failure to thrive, frequent infections, small stature, dysmorphic features and hepatosplenomegaly are some of the presentations. Neural palsies, especially involvement of the optic, oculomotor and facial nerves may occur (3). The patients may develop severe anemia due to bone marrow obliteration (2).

The diagnosis is made by radiographic manifestations of the great increase in the bone density.

Increased serum levels of osteoclast-derived tartrate-resistent acid phosphatase and the brain isoenzyme of creatine kinase are the only significant lab findings (2).

Treatment is difficult. Bone marrow transplantation, low calcium/high-phosphate diet and methylprednisolone maybe used. Also surgical intervention and orthopedic management are required for some complications (2).

Osteomyelitis complicates osteopetrosis in 10% of the cases due to impaired white cell function and reduced vascular supply (1). The most common site of involvement is the mandible. Osteomyelitis of the maxilla is not very common (4).

Case report

A 15-year old boy referred to our center by his otolaryngologist with chief complaint of facial swelling and pain. Since he was 9 years old, he had a history of multiple dental extractions due to severe dental caries which led to facial pain and swelling. The patient received multiple courses of intravenous antibiotic therapy over the next 3 years, sometimes with impression of maxillary sinusitis, other times with impression of local dental infection. He underwent three surgical interventions on the alveolar ridge. After each of these procedures there was a recurrence of symptoms. During this period halitosis and destruction of alveolar mucosa were added to his previous problems. At the age of 12, bilateral sinus tract formation on the chick was developed and the lower eyelids retraction was occurred. Since 2 weeks before admission the patient received antibiotic therapy due to another episode of facial swelling and pain without significant response to oral antibiotic therapy, so he was referred to our center and was hospitalized. The patient had history of multiple fractures. He was also complaining of hearing and visual problems. Family history for osteopetrosis was negative.

At admission time he was febrile, had diffuse bilateral facial swelling, sinus tract drainage in both infraorbital regions (Fig 1). Physical exam revealed hepatosplenomegaly. Ophthalmoscopy was in favor of bilateral optic atrophy. Audiometry showed low frequency hearing loss. Blood test resulted in microcytic hypochromic anemia with Hb about 5.5. Serum calcium, phosphorus and alkaline phosphate levels were normal. The chest x-ray showed diffuse bone sclerosis and the skull x-ray revealed increased bone density especially in the

skull base. Also mastoids were poorly aerated and maxillary sinuses were absent. Roentgenograms of long bones and spinal column were obtained (Fig 2).

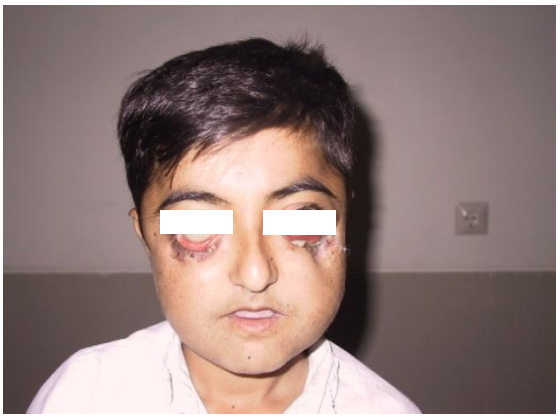

Fig 1: This photo (taken from the patient during hospital course) demonstrating bilateral lower eyelid retraction, orifice formation with discharges in infraorbital area

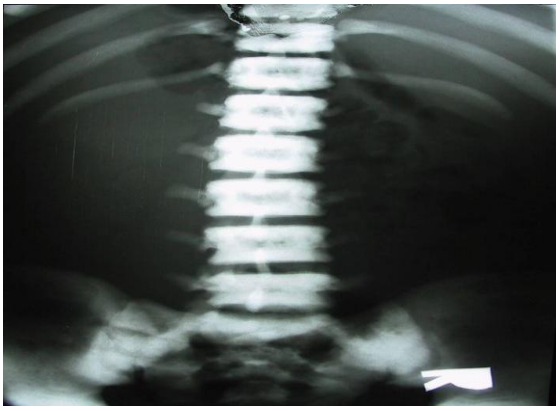

Fig 2: Roentgenogram of spinal column demonstrating diffuse vertebral sclerosis

Generalized sclerosis especially in diaphyseal regions and widening of metaphyseal regions were detectable and modularly spaces were absent. Para nasal sinuses CT-scan showed destruction of both nasal antrum accompanied with sequestration formation and sclerosis of maxilla, zygomatic and ptyregoid plates (Fig 3). A fistulous tract between the maxillary bone and oral cavity was also seen. Biopsy of the exposed bone resulted in chronic osteomyelitis.

The patient received intravenous clindamicin 160 mg/q6h for 2weeks and underwent surgical procedure that included curettage, sequestrectomy and the sinus tracts excision. During the next 9 weeks after operation his condition improved, oral mucosa healed and drainage decreased but did not completely subside.

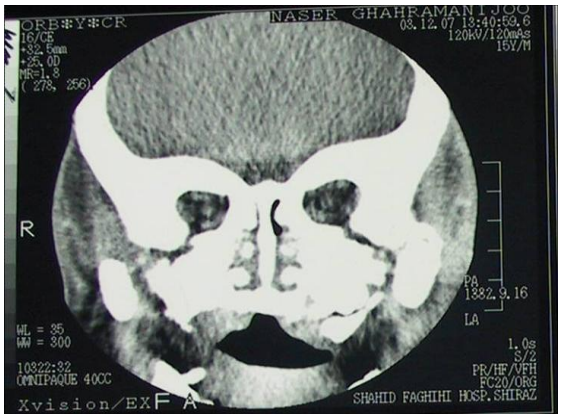

Fig 3: Coronal computed tomography from nasal cavity and facial bone showing destruction of both nasal antrum accompanied with sequestration formation and sclerosis of maxilla, zygomatic and ptyregoid plates

Discussion

Osteopetrosis, originally described by Albers-Schönberg in 1904 (5) is believed to be the consequence of dysfunctional osteoclasts, resulting in defective remodeling of bone and increased bone density. Its diagnosis is based on a history of multiple fractures and radiological findings of osteosclerosis (2).

Osteopetrosis is a hereditary disorder (3), although it was not possible to determine the hereditary pattern in our case since there was no family history of osteopetrosis. Classification of hereditary bone disease is often difficult because of their complex nature and similar characteristics. However, each entity can be distinguished by considering historical, clinical and radiological findings. In our case other forms of human sclerosing disorders were ruled out because of the absence of dwarfism (pycnodysostosis),

mental deficiency (Camurati-Engelmann disease), and specific long-bone osteopetrosis (Pyle disease) (6).

Otoneurological presentation has been reported in 16% of the patients with autosomal-dominant type osteopetrosis (7). The trigeminal and the acoustic nerves involvement are primarily seen in the patients with type I autosomal-dominant osteopetrosis, whereas the facial nerves involvement are more frequent in type II autosomal-dominant osteopetrosis. The most commonly affected nerve is the optic nerve, with the facial nerve being second in the literature (7). In this case, the facial and trigeminal nerves were intact but the optic and acoustic nerves were involved.

Some patients with osteopetrosis may be asymptomatic and many cases are initially diagnosed when the patient presents with osteomyelitis. Osteomyelitis is a known complication of osteopetrosis, which is refractory in most cases because of decreased blood supply, anemia and neutropenia (1). The present case also demonstrates maxillary osteomyelitis, which did not properly respond to the conventional treatment.

Approximately two thirds of patients with autosomal-dominant type osteopetrosis (64%) had stomatological manifestations (7). The risk of osteomyelitis increases in the presence of local infection such as odontogenic infection. Therefore dental treatment for patients with osteopetrosis, especially extraction of the teeth, must be carried out with great caution. In our case, manifestation of the disease was begun with stomatological complaints, including multiple periods of exacerbations and remissions of osteomyelitis after teeth extraction.

Controversies exist in treatment of osteomyelitis secondary to osteopetrosis, although the principles of treatment remain the same as other types of osteomyelitis. These principles include: antibiotic therapy, teeth extraction, incision and drainage, sequestrectomy, saucerization, decortication, bone resection, and hyperbaric oxygen. Resection and hyperbaric oxygen therapy are the only methods mentioned as successful treatments in cases with osteomyelitis which is secondary to osteopetrosis (8). Radical surgical management was not done in this case because of the extended bony destruction and social factors, although some authors suggest that radical approach is preferable for patients with severe complications resulting from osteopetrosis.

The filling of the maxillary defects can be achieved by obturators. Free bone grafting is not recommended due to impaired blood supply. Free vascularized, myo-osseous flap may be more useful but may be precluded due to lack of a suitable donor site in these patients (9).

Reports of successful treatment are rare, and in many cases the osteomyelitis remains unresolved.

Taking history of multiple fractures must be considered in each case which is presented by maxillary or mandibular osteomyelitis. Skull and chest radiographs, and full blood count must be performed. Renal and bone profiles should also be obtained to screen for abnormalities that are seen in osteopetrosis.

If osteopetrosis is confirmed, medical consultation is mandatory, as management of the underlying osteopetrosis may lead to a better outcome.

Some reports have mentioned that pentoxifylline and tocopherol plus antibiotic therapy might be useful in the treatment of patients with osteopetrosis complicated by osteomyelitis and to avoid radical surgery (8).

Conclusion

As mentioned earlier maxillary osteomyelitis is a rare phenomenon. If it

occurs, evaluation for underlying disease especially osteopetrosis must be considered. In spite of an increase in the life expectancy of patients due to progressive medical care, increasing frequency of this condition also leads to a situation which requires more attention.

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