

Osteomyelitis In Mandible in A Case of Infantile Osteopetrosis: A Case Report And Review of Literature

*Dr.Rajarshi Bandyopadhyay¹, Prof.(Dr.)Amit Ray², Dr. Anjan Maity³.

^{1,3} 2nd Year PGT, Department Of Oral & Maxillofacial Surgery , Guru Nanak Institute Of Dental Science Panihati, Kolkata

² Professor & Head Of The Department Of Oral & Maxillofacial Surgery, Guru Nanak Institute Of Dental Science & Research, Panihati, Kolkata

Corresponding author: *Dr.Rajarshi Bandyopadhyay

Abstract: Osteopetrosis is a rare genetic disorder that causes generalized sclerosis of the bone due to defect in bone resorption and remodelling. Infantile osteopetrosis is a rare form of osteopetrosis. Osteomyelitis is a well-documented complication of osteopetrosis. Any associated dental abnormality may be attributed to the pathological changes in bone remodelling. This case report discusses a case of osteopetrosis with osteomyelitis as a complication in a 9-year-old girl.

Keywords: Osteopetrosis, Osteomyelitis.

I. Introduction

Considered a rare inherited skeletal disease, osteopetrosis(OP) is characterized by a considerable increase in bone density resulting in defective remodelling, caused by failure in the normal function of osteoclasts, ranging in severity.¹ According to its severity it can be asymptomatic to fatal² and it is often diagnosed by radiographic exams³, not being essential a bone biopsy.⁴ Globally the estimated prevalence of OP is 1 in 100000 to 500000. It is usually subdivided into three types: benign autosomal dominant OP, subdivided into type I and II. Type II is also known as 'Albergs-Schonberg disease' is the most common form of OP; intermediate autosomal recessive OP; and malignant autosomal recessive infantile osteopetrosis (MIOP), considered the most serious type. This is associated with a decreased life expectancy, with most children dying in the second decade of life with complications of bone marrow suppression.¹

MIOP is a rare recessive disorder characterized by dense, sclerotic, fragile, radio-opaque bones; neurological abnormalities; anaemia and thrombocytopenia with subsequent extramedullary haematopoiesis and impaired vision and hearing caused by encroachment of the foramina and nerve canals.⁶ They usually die during the first years of age because of anaemia or secondary infection. At the moment, allogenic haemopoietic stem cell transplantation is the only curative treatment of autosomal recessive OP and should be offered as early as possible⁷. Infants are diagnosed with this form of OP immediately or shortly after birth. These patients often have pathological fractures, osteomyelitis of long bones and repeated rate of infections.⁸ Some differential diagnosis to consider include fluorosis; beryllium, lead and bismuth poisoning; myelofibrosis; Paget's disease (sclerosing form); and malignancies (lymphoma, osteoblastic cancer metastases).⁹

The development of dentition is severely disturbed in children with OP. Dental findings include delayed tooth eruption and impaction, aplasia, unerupted malformed tooth, enamel hypoplasia and early tooth loss.^{6,9} Regarding patients with OP, osteomyelitis is the most common and well documented maxillofacial complication and can be severe and difficult to treat. We describe a case of chronic osteomyelitis in the mandible of a patient with Malignant Infantile Osteopetrosis (MIOP).

II. Case report

A young 9-years-old girl (weight:20 kg, height:109cm) had reported to the Department of Oral and Maxillofacial Surgery, of G.N.I.D.S.R. with the chief complaint of swelling, pain and discharging sinus on the right side of mandible since 6 months in January, 2017.



(A)



(B)

Fig1: Clinical photograph showing swelling in the mandibular right posterior region(A) and genu valgum in legs(B).

It was a diagnosed case of OP when she was 2 and half years old required regular transfusion; she also had an history of jaundice and her elder sibling-boy died at the age of 5 years (cause of death unknown). On general examination, she was of slight build and small stature, pale, and moderately nourished with dolicocephaly, prominent eyes and genu valgum. The chest was of a pigeon-breast shape. The spleen and liver were enlarged and palpable. The cervical, axillary, and inguinal lymph nodes were not palpable and sensitive but the right submandibular lymph nodes and submental nodes are palpable. The head was enlarged, with prominent frontal bosses. Vision and fundi were normal. There was no hearing defect.



Fig 2: Showing constricted hypopharynx

On extraoral examination, discharging sinuses were seen on the right submandibular region with whitish to yellowish pus discharging and tender right body of the mandible. On palpation, the submandibular regional lymph nodes were found to be enlarged and mild tender. Intraoral examination revealed exposed bone with right mandibular primary 1st molar with exposed mesial and distal roots with carious involvement. Many teeth were yet to erupt and missing and poor oral hygiene. OPG shows diffuse increased bone density over the right lower border of the mandible with loss of trabecular pattern throughout the mandible. Spine AP & Lateral was suggestive of generalised increase density of bones. Right wrist AP & Lateral was suggestive of sclerotic bone formation. The chest radiograph (AP view) showed increased bone density with bilateral clear lung fields.

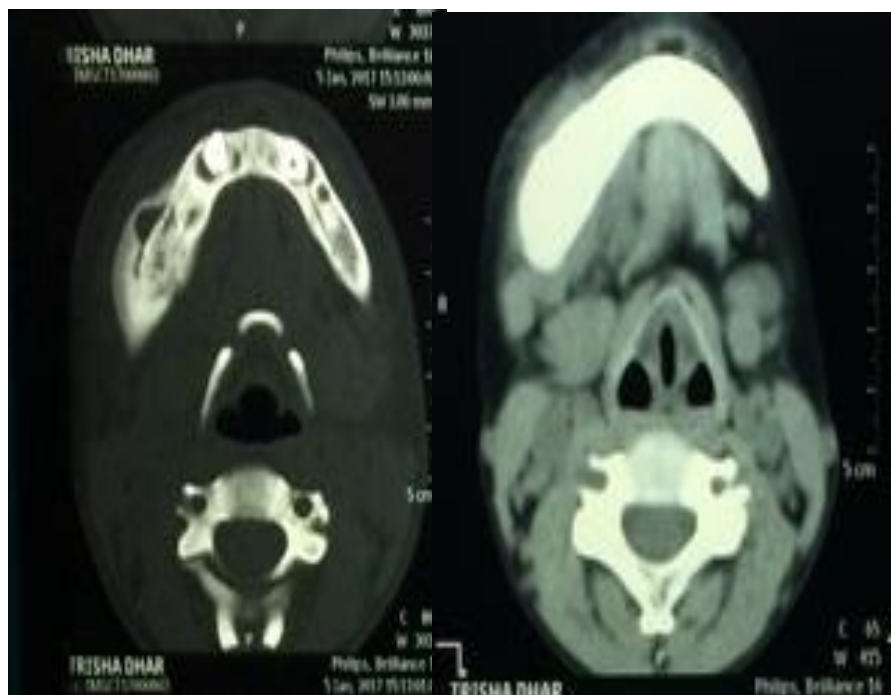


Fig 3: CT scan suggestive of destruction along with cortical breach multiple bone bits and significant periosteal reaction noted in the angle of the mandible on right side. Cortical breach noted in both outer and inner cortex with enlarged submandibular and submental lymph nodes.



Fig 4:“Shepherd crook” deformities of upper ends of femurs and “Erlenmeyer flask deformity” with excessive callus formation and lateral bowing.



Fig 5: Diffuse sclerosis and ‘bone within bone appearance’ seen in iliac wings. Bilateral ‘coxa vara’ deformity was also noted.

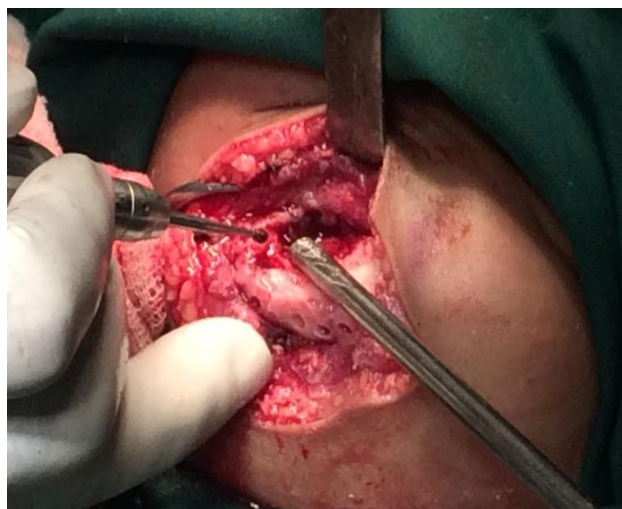
History, clinical examinations and all the radiographs indicated it’s a case of osteopetrosis with superadded chronic osteomyelitis of right body of the mandible. Upon admission, the RBC count was $4.22 \times 10^6/\mu\text{l}$, the leukocyte count was $8.2 \times 10^3/\mu\text{l}$, the haematocrit was 31.4%. and the haemoglobin was 10mg/dl. The platelet count was $360 \times 10^3/\mu\text{l}$. The red blood cells showed mild microcytic hypochromic anaemia. The neutrophil count was 45% and lymphocyte count 50%.

Treatment:

Primarily intravenous antibiotic therapy (clindamycin) was started and one unit of packed red blood cell was transferred to maintain the haemoglobin level. Corticotomy and sequestrectomy with debridement of the necrotic bone were performed. Trephination was also performed over the lower border around the margin. Vancomycin powder mixed with distilled water (in paste form) was applied locally before the closure and the taken specimen was sent for biopsy.



(A)



(B)



(c)

Fig 6: (A)Incision (B)After corticotomy trephination was being done (c)Local application of vancomycin to fill the dead space

III. Discussion

OP is a rare metabolic bone disease characterized by a generalized increase in skeletal mass. This inherited disorder results from congenital defects in the development or function of the osteoclast.¹⁰ OP classification was stated before. The consequent impairment of bone resorption prevents formation of bone marrow cavities. The pathogenetic defect may be intrinsic either to the osteoclast lineage or to the mesenchymal cells that constitute the microenvironment supporting the development and activation of osteoclasts. Three clinically distinct forms of OP are recognized-infantile malignant autosomal recessive form, fatal within first few years of life (in the absence of effective therapy); intermediate autosomal recessive form, appears during the first decade of life but does not follow the malignant course and the adult benign dominant form with full life expectancy but many orthopedic problems.¹² The disease represents a spectrum of clinical variants because of the heterogeneity of genetic defects resulting in osteoclast dysfunction. OP complicated by the development of osteosarcoma has been reported.¹³ Survival after 20 years of age is rare.

There are always some chances of hazardous intubations in case of autosomal recessive MIOP¹⁴, actually occurred in our case. A MIOPatient commonly have mandibular abnormalities, with hypoplasia reported in younger patients¹⁵ and narrowing of the area between the base of the tongue and the posterior pharyngeal wall¹⁶. A high and narrow hard palate is believed to contribute to airway obstruction¹⁷, and nasal obstruction and congestion are almost universal in infants with MIOP¹⁵.

Orthopedic problems in the intermediate and autosomal dominant forms include bone pain (26% of patients), increased fractures (40%) coxa vara (FIG 5), long bone bowing (FIG 4), hip and knee degenerative arthritis and osteomyelitis. Dental abnormalities may be attributed to the pathological changes in OP. Patients with the disease seem to be especially susceptible to caries. Constriction of canals housing neurovascular bundles that supply teeth and jaws, along with obliteration of the marrow cavities and the dental pulp chambers, is the most likely contributing factor to bone necrosis and dental caries¹⁸. Other dental changes may include delayed eruption and early loss of teeth, enamel hypoplasia, malformed roots and crowns and thickening of the lamina dura.¹⁹ Osteomyelitis, due to dental caries (10%), is well recognized hazard in OP due to reduced blood circulation to bone as a result of obliteration and fibrosis of the marrow. It is a potentially severe infection that runs a protracted course, due to the accompanying severe anaemia and neutropenia.²⁰

Radiography show uniform increase bone density without corticomedullary demarcation. The long bones have a dense chalk like appearance. They may have an 'Erlenmeyer flask' deformity at their ends due to failure of metaphyseal remodelling, giving gross distal under tubulation, and the presence of dense bone, vertical fine lucencies extending to the metaphysis are present probably due to vascular channels being better seen against dense bone. Bowing of the long bones as well as coxa vara may be present due to multiple fractures.

There is also an increase in density of bone at the base of skull especially prominence in the floor of anterior cranial fossa. The sphenoid and frontal sinuses and mastoids may be underpneumatised. A 'bone within a bone' or endobone phenomenon may be seen in small bones of the hands but, with increased density around the periphery. In a review of 57 cases of osteomyelitis resulting from OP, most cases were found to be chronic and resistant to treatment²¹. Unfortunately, there seems to be no definitive treatment for OP of the maxilla or mandible without complete removal of the affected bone. In order to minimize such problems, patients with OP should be encouraged to maintain good dental care and oral hygiene, because there is a potential risk of promoting osteomyelitis if surgical procedures are performed.²⁰

Palliative treatment, including nerve decompression and debridement, seems to be the best course of action. The best treatment appears to be preventive management with routine dental care. Teeth should be endodontically treated, if possible, rather than extracted, because periosteal stripping of bone may predispose asymptomatic bone to become necrotic and to sequester. Any debridement should be as conservative as possible, removing only grossly necrotic bone through limited flap dissections. Because the disease process is systemic, there is often no clear delineation between affected and unaffected bone²². That is why in this case we tried to make it as conservative as possible and trephination is very useful here to increase the periosteal blood supply, originating from endosteum and high concentration of local vancomycin application was not only do they deliver high tissue levels of antibiotics but they also help obliterate the dead space that occurs after bone debridement.²³ (FIG 5) Recently, bone marrow transplantation has been successfully used in the treatment of malignant OP, offering hope to those so afflicted;²⁴ it has been curative in a significant percentage of patients but an acceptable donor can be found in only about 40%²⁵. **Consent:** Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Conflicts of interest: None

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