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Case Report

A Rare Case of Osteopetrosis with Dental manifestation in a Young Child

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ABSTRACT:
Osteopetrosis also known as "marble bone disease" is a group of rare genetic disorders caused by osteoclast failure, which ranges widely in severity. An increase in bone mass and density results in severe skeletal malformation and bone marrow supression, which may be fatal. We present here a rare case of osteopetrosis in 13 years old child reported with chief complain of bleeding gums.
Key words: Osteopetrosis, bone disorder.

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INTRODUCTION

Osteopetrosis is an extremely rare congenital bone disorder, It is also known as Albers schonberg disease. It occurs due to failed functioning of osteoclasts to resorb bone. This dysfunction results in thick cortical bone, causing suppression of marrow spaces. Abnormal expansion of bone interferes with medullary haematopoesis resulting in reduced blood cells, which further initiates extramedullary haematopoesis, resulting in expansion of its sites namely liver and spleen. It occurs one in every 250,000 individuals. Osteopetrosis can be subdivided into Autosomal recessive; malignant infantile type (ARO), Intermediate autosomal (IAO), Autosomal dominant; adult type (ADO).[1-4]

Autosomal recessive form that is malignant infantile variety is considered as severe and rare of all three. These forms present at birth or in early childhood or in young adolescence. Prognosis of this form is considered as poorest of all.[2] If untreated, infantile osteopetrosis usually result in early death. This condition is most commonly diagnosed soon after birth or within 1st year of life with characteristic presentation of macrocephaly and hepatosplenomegaly along with abnormal bone remodelling.[5] We present here a rare case of osteopetrosis in 13 years old child reported with chief complain of bleeding gums.

CASE REPORT

A 13 year old male reported to the Department of Pedodontics and Preventive Dentistry RIMS Ranchi with the chief complaint of frequent bleeding in oral cavity**.** Clinical examination of the child showed stunted growth, distended abdomen hepatosplenomegaly, pallor, flat feet, dolicocephalic head, frontal bossing, macrocephaly and extra oral swelling in lower left mandible area (Figure 1). There was a fresh wound, over left lower eye lid, to which he gave history of bicycle fall. Other sibling elder to child was of normal growth pattern and there was no such history in family. Past family history revealed that boy was alright in first 2-3 years after birth. Past dental history includes extraction of multiple deciduous lower teeth, 3 years back by local dentist.

Oral finding showed erupted 31, 36, 41, and 46 in lower jaw where as 34, 45 were partially erupted (Figure 2). Upper jaw showed highly convex palatal vault and partial eruption of premolars and molars along with bony sequestrum in posterior alveolar region (Figure 3). Deep pockets were associated with these teeth, which showed bleeding on probing.

OPG showed dense maxillary and mandibular bone with loss of usual trabecular pattern, along with mixed radiolucency and radio-opacity in premolar region of

mandibular jaw, various unerupted teeth, and teeth with malformed roots, lower permanent first molar shows taurodontism (Figure 4). X-Ray skull PA View showed highly dense maxillary bone with absence of frontal and maxillary sinus (Figure 5).

CT scan showed thickening and increased density of skull bone and immature closure of fronto- parietal suture (Figure 6).

Blood investigation showed abnormal increased level of serum PTH as 241.9pg/ml, whereas level of vitamin D was reduced (11.0mg/ml) than normal value. Haemoglobin showed the value of 4.2 gm.

Supragingival scaling and pocket curettage was done, to provide symptomatic relief from gum bleeding. Chlorhexidine gluconate mouth wash was advised. Child was sent to department of paediatrics for further investigations & treatment.

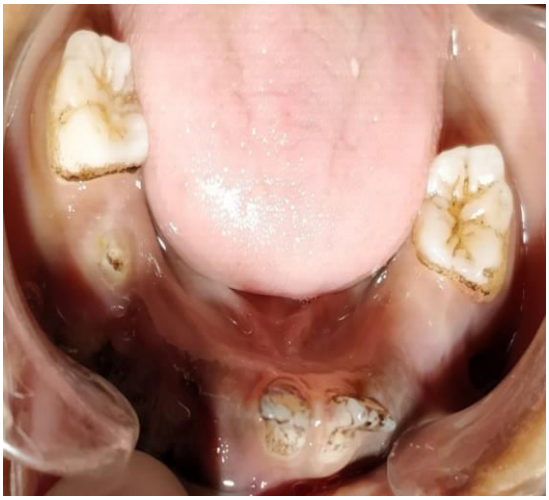

Figure 2: Intra-oral photograph of Mandibular Arch

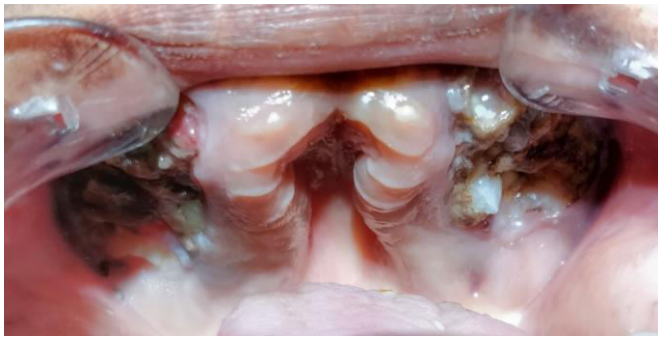

Figure 3: Intra-oral photograph of Maxillary Arch

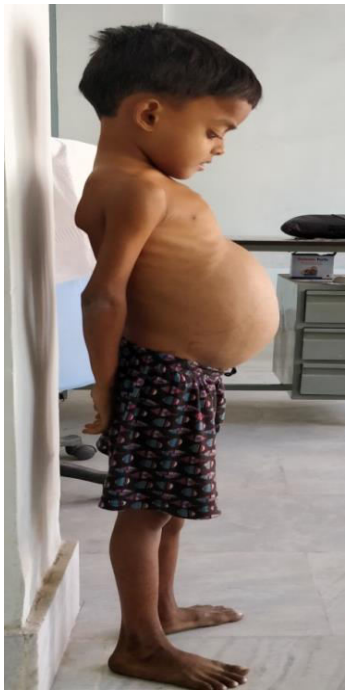

Figure 1- Child with dolicocephalic head, distended abdomen, flat feet

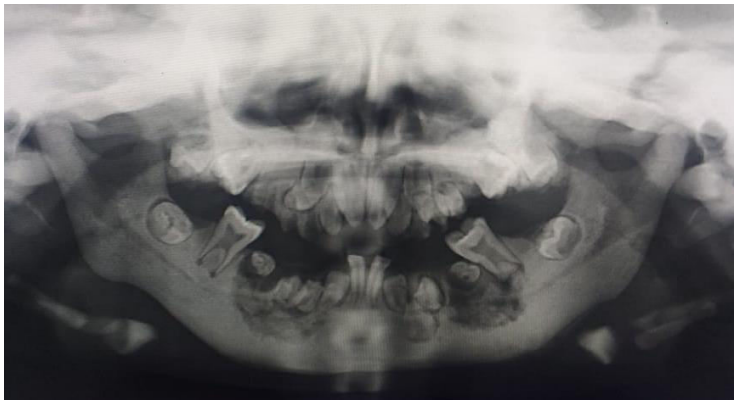

Figure 4: Panoramic radiograph showing bilateral radiolucency in premolar region, multiple impacted rotated teeth, tarodontism in lower permanent molars

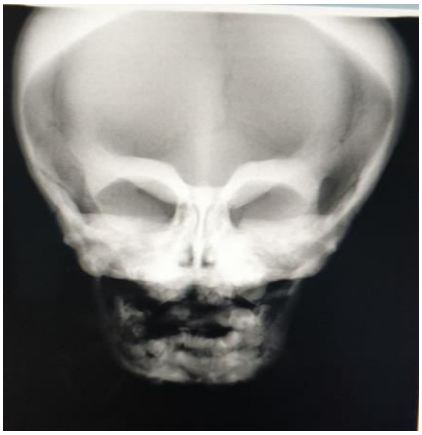

Figure 5: PA VIEW showing absence of frontal and maxillary sinus.

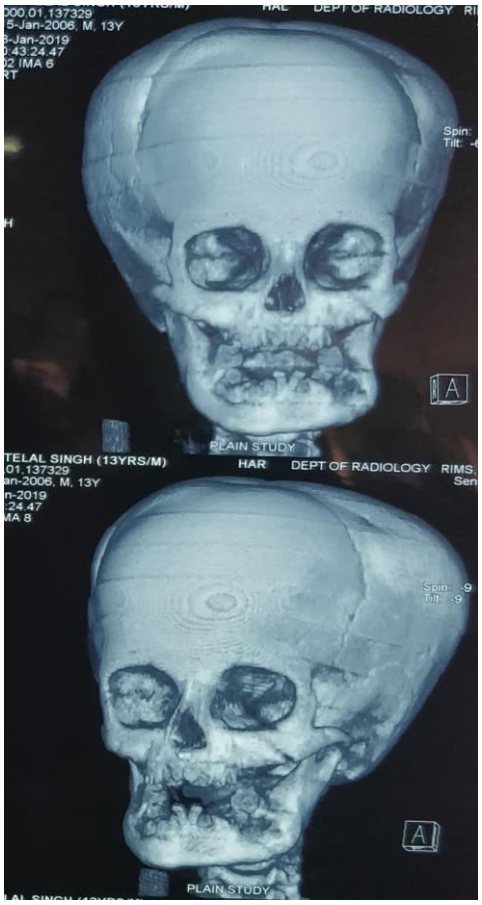

Figure 6: CT Scan showing thickening and increased density of skull bone and immature closure of fronto-parietal closure.

DISCUSSION

Osteopetrosis was first described by German radiologist Albers-Schonberg in 1904. Infantile osteopetrosis is diagnosed early in life, and considered to have poor prognosis. Prognosis for survival of this form of osteopetrosis is 30% at 6 years.[6] Clinically infantile osteopetrosis consists of primary and secondary pathologies such as reduced medullary haemopoetesis leads to hepatosplenomegaly and anaemia, osteomyelitis and poor development of dental structures due to reduced perfusion capacity, mental retardation due to expansion of cranium., visual and hearing loss due to compression of optic and auditory nerves by high density bones.[1,3] Growth retardation is a common symptom.[3] Bony defect occurs due to defective osteoclsatic activity. Medullary spaces of jaw is markedly reduced and osteomyelitis of mandible is common due to deficient blood supply.[5] Tooth removal should be limited due to expected bone fracture and osteomyelitis. Dental complications are enamel hypoplasia, teeth with malformed crown and short roots embedded teeth.[3] In present case child OPG showed multiple impacted teeth with malformed root and taurodontism of mandibular first molar Mandible showed mixed radiolucency and radio opaque area which is common feature of chronic osteomyelitis. Bone marrow suppression interferes with medullary haematopoiesis resulting in life threatening pancytopenia and secondary expansion of extramedullary haematopoiesis sites such as liver and spleen. Child showed distended abdomen, stunted growth, however visibility and hearing are not compromised and have normal intelligence. Haemopoetic stem cell transplantation is the only treatment that had shown good prognosis.[7] It can restore bone osteoclastic function and promote bone resorbtion. Corticosteroid, gama interferon are other treatment alternatives.[8] Patient was referred for higher centre for systemic treatment. It is extremely important that dentist should be aware of limitation presents in these patients due to severe systemic problems, particularly complication associated with extraction due to poor blood supply and overtly dense bone. Preventive programme should be initiated early

REFRENCES

1. Stark Z, Savarirayan R. Orphanet J Rare Dis. 2009 Feb 20;4:5. Doi: 10.1186/1750-1172-4-5.
2. Raghunath Reddy MH. Int J Clin Pediatr Dent. 2011 Sep-Dec; 4(3): 232-234.
3. Abbas Makarem, Nosrat Lotfi, Seyed Amir Danesh-Sani, Soudabeh Nazifi. Int J of Head & Neck surgery.May-August 2012,3(2):115-117.
4. C Wilson, A Vellodi. Arch Dis Child. 2000 Nov; 83(5): 449-452.doi: 10.1136/adc.83.5.449.
5. Neville, Damm, Allen, Bouquot (Editors). Oral and Maxillofacial Pathology. 2 nd ed. Elsevier Saunders 2004;1:148.
6. V.Luzzi et al. European journal of pediatric dentistry. 2006 Mar;7(1):39-44.
7. Chaudhary S, Sharma A. Journal of Indian academy of oral med & radio.oct-dec;2008/vol 20/issue4
8. Machado Cde V, da Rocha MC, Telles PD. BMJ Case Rep. 2015 Mar5;2015.