

An interesting case of osteomyelitis of mandible in a child with osteopetrosis

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Section: Musculoskeletal system

Area of Interest: Musculoskeletal joint Head and neck

Procedure: History

Procedure: Diagnostic procedure

Procedure: Computer Applications-Detection, diagnosis

Imaging Technique: CT

Special Focus: Genetic defects Case Type: Clinical Cases

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Patient: 7 years, male

Clinical History:

A 7-year-old boy, a known case of osteopetrosis, came to the ER department presenting with fever along with swelling of the face. Clinically, an infectious process was suspected. CT of the face was done for confirmation of the diagnosis and to rule out any complication and progression of this disease.

Imaging Findings:

Past medical history of osteopetrosis was present. The previous chest radiograph showed increased bone density with clear lung fields. The radiograph of the pelvis with both thighs revealed fracture of the left femur with generalized increased bone density.

Recent non-enhanced CT showed dense thickened bones with loss of corticomedullary differentiation.

Fractures are noted in medial and lateral walls with floor of orbit and zygomatic arch on the right side. Irregularity with erosion of ramus and body of mandible is noted on the left side. Irregular periosteal reaction is noted in the body and ramus of the mandible on the left side with minimally enhancing adjacent soft tissue component.

Multiple enlarged homogeneously enhancing nodes are seen in bilateral submandibular region, in submental region and in upper jugular region on either side. The largest one measures approximately 28 x 17 mm.

Discussion:

Osteopetrosis, (also known as “Marble bone disease” and “Albers-Schönberg disease”), comprises a clinically and genetically heterogeneous group of conditions that share the hallmark of increased bone density on radiographs. [1] The increase in bone density results from abnormalities in osteoclast differentiation or function. Four forms of the disease have been identified, (a) an autosomal dominant benign heterogeneous form, (b) an autosomal recessive severe malignant form, (c) an intermediate form that is a recessive type, and (d) a recessive type with renal tubular acidosis (also known as carbonic anhydrase II deficiency syndrome) [2]

Clinical features includes severe anaemia, repeated bleeding episodes and infections, hepatosplenomegaly, lymphadenopathy, frequent pathological fractures from minor trauma, and failure to thrive. [2]

Radiological findings are classical and include generalized sclerosis of the skeleton with homogeneously increased density of all the bones with little or no differentiation between cortical and medullary regions. While the bones may

appear radiopaque, they are actually brittle and subject to pathological fractures, which are characteristically transverse. Multiple striations producing a bone within a bone appearance may be noted. The skull shows basilar and calvarial thickening with increased density, and poorly developed sinuses. [5]

Dental abnormalities include caries, delayed eruption and early loss of teeth, enamel hypoplasia, malformed roots and crowns and thickening of the lamina dura. Constriction of the canals housing neurovascular bundles supplying the teeth and jaws as well as obliteration of the marrow cavities and dental pulp chambers lead to bone necrosis and dental caries and ultimately develop osteomyelitis in 10% of cases. [1] Osteopetrotic patients are prone to develop infections and are susceptible to jaw bone fractures, hence surgical dental procedures need to be planned to avoid complications. [3] Associated osteomyelitis is a potentially severe infection that runs a protracted course, due to the accompanying severe anaemia and neutropenia. Surgical resection should be planned with caution as osteopetrotic bone has less capacity to heal and these children are at risk of adverse respiratory events and increased perioperative morbidity and mortality as anaesthetic complications. Treatment regimens include high-dose systemic antibiotics coupled with thorough debridement of necrotic bone and primary closure of soft tissues. Hyperbaric oxygen (HBO) has also been used for the treatment of osteomyelitis. [4] As the facility of hyperbaric oxygen therapy was not available in our setup, the patient was advised to undergo reconstruction surgery, which his parents refused. So the patient was put on systemic antibiotic therapy and local debridement of the affected jaw was done.

Differential Diagnosis List: Osteomyelitis of mandible with osteopetrosis, Associated neoplasm, in osteopetrosis

Final Diagnosis: Osteomyelitis of mandible with osteopetrosis

References:

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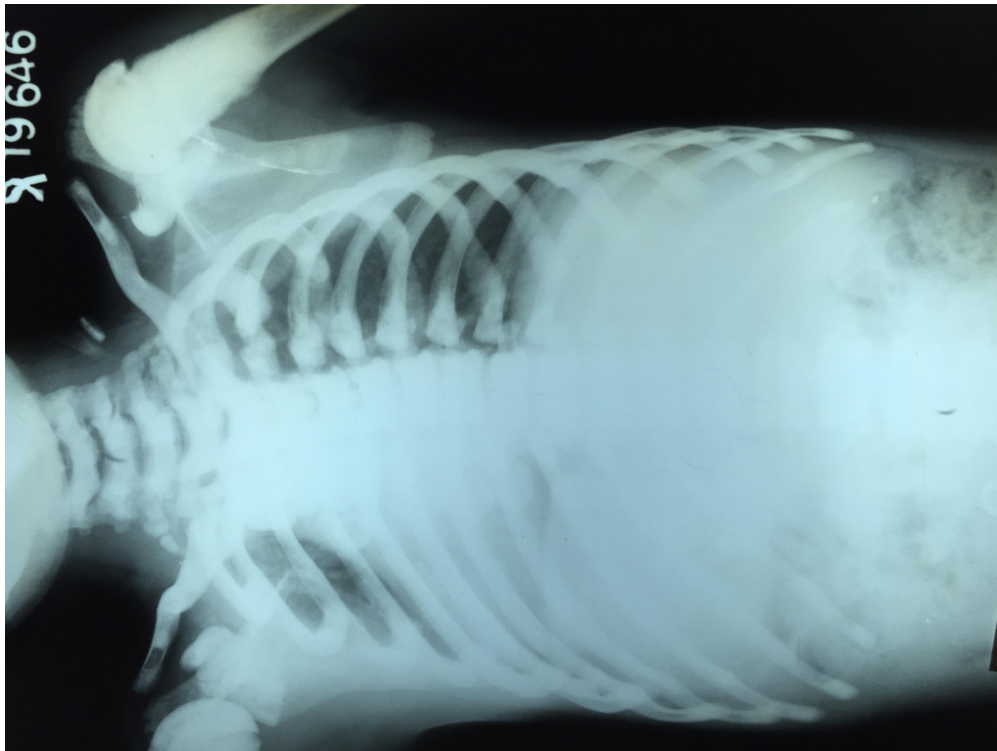
Figure 1

a



Description: Radiograph of pelvis and hip shows increased density of bones with fracture of left femoral shaft. **Origin:** PDU Medical college, Rajkot

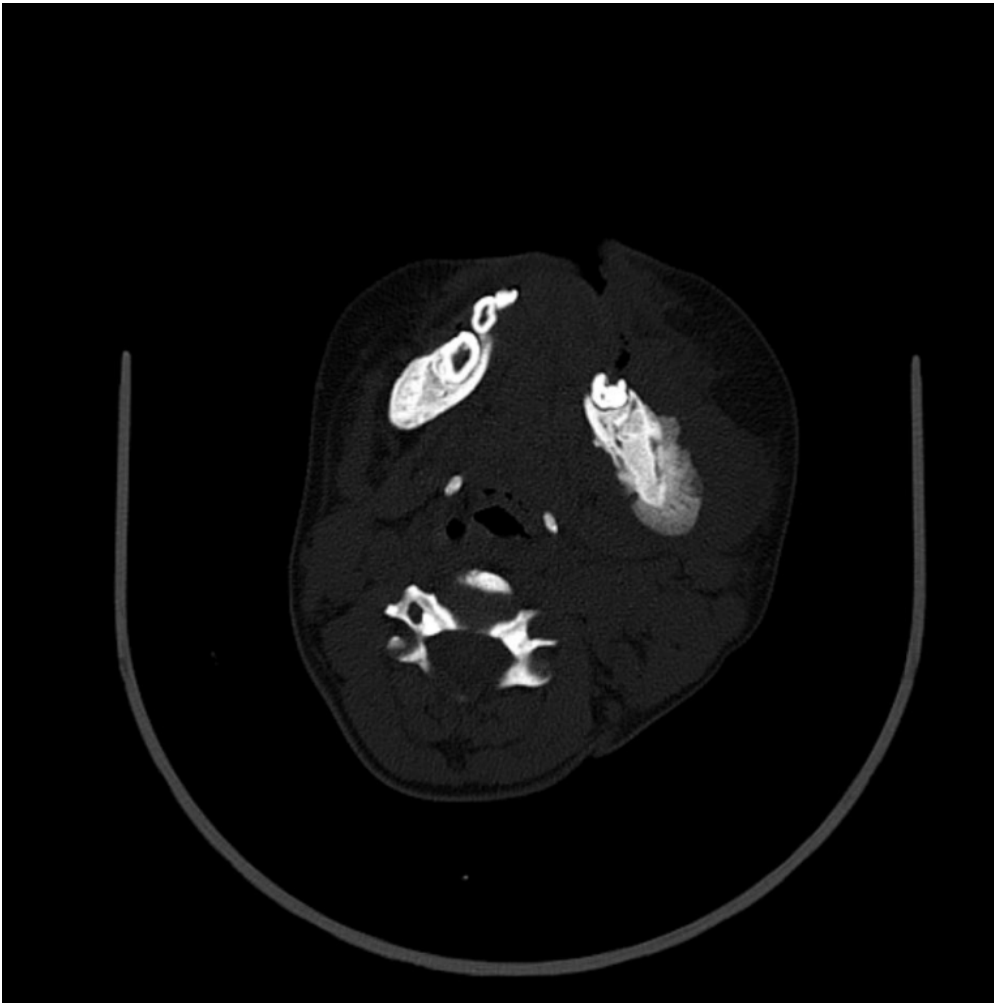
b



Description: Chest radiograph shows increased density of bones. **Origin:** pdu medical college

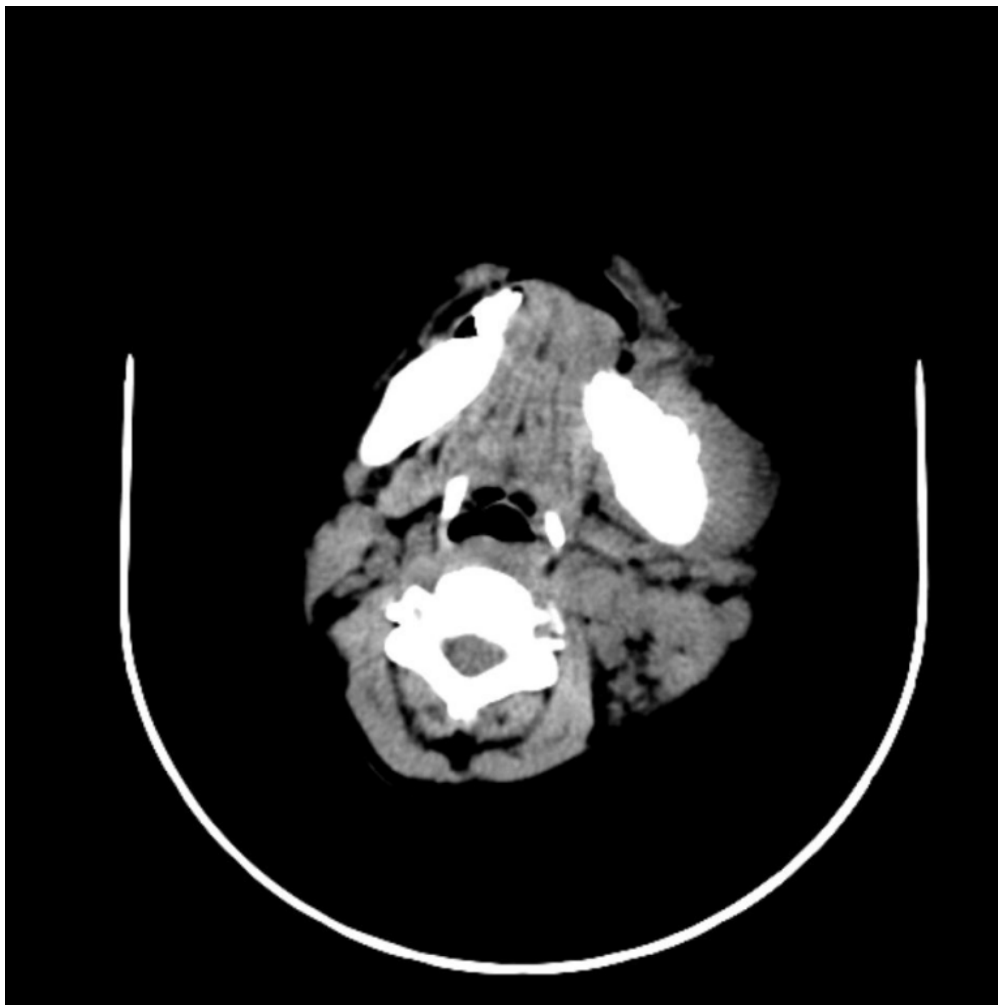
Figure 2

a



Description: Irregularity, erosion of body of mandible, overlying periosteal reaction is noted on the left side. **Origin:** PDU MEDICAL COLLEGE RAJKOT

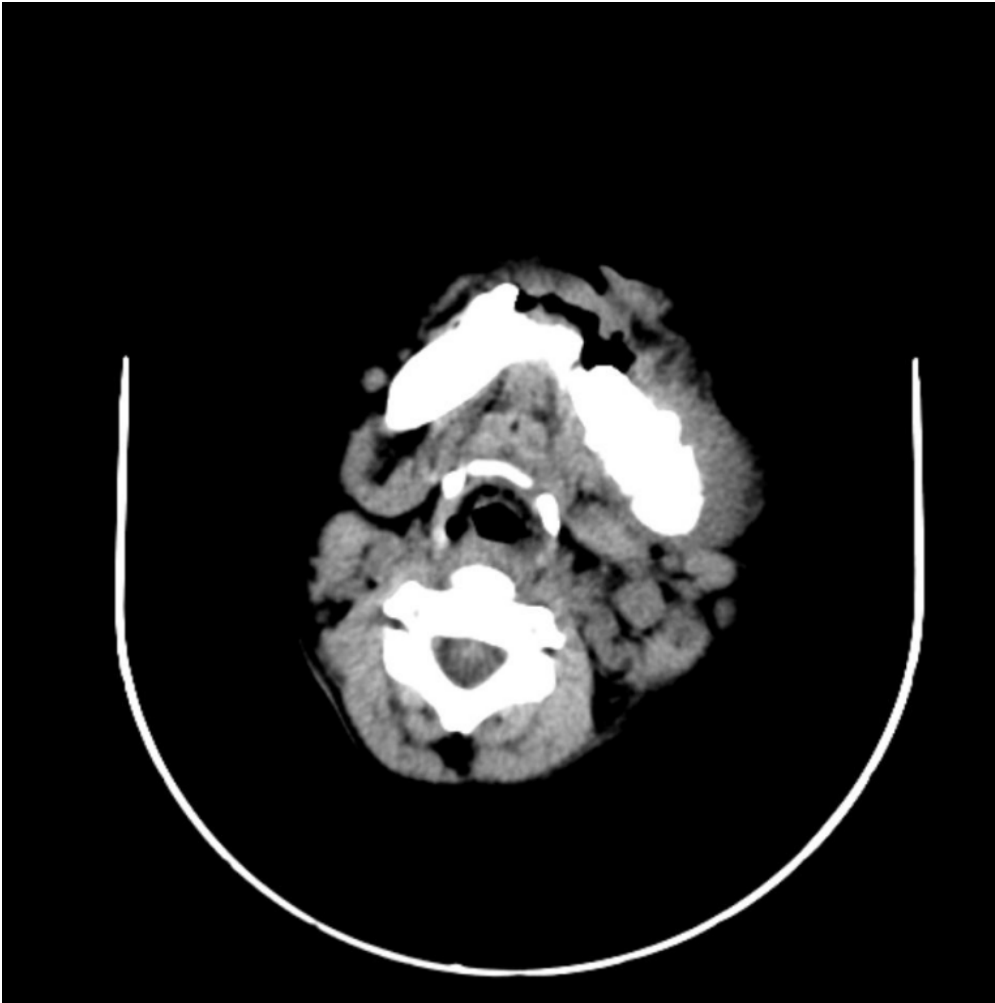
b



Description: Minimally enhancing soft tissue is seen adjacent to body of mandible on the left side.

Origin: pdu medical college

c



Description: Enlarged submandibular and jugular lymph nodes on both sides. **Origin:** pdu medical college

d



Description: Fractures of medial and lateral walls of orbit with zygomatic arch fracture on the right side.

Origin: pdu medical college