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Clinical report

Osteomyelitis of the maxilla in a patient with Malignant Infantile Osteopetrosis

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ABSTRACT

Osteopetrosis is characterized by a considerable increase in bone density resulting in defective remodeling, caused by failure in the normal function of osteoclasts, and varies in severity. It is usually subdivided into three types: benign autosomal dominant osteopetrosis; intermediate autosomal recessive osteopetrosis; and malignant autosomal recessive infantile osteopetrosis, considered the most serious type. The authors describe a case of chronic osteomyelitis in the maxilla of a 6-year-old patient with Malignant Infantile Osteopetrosis. The treatment plan included pre-maxilla sequestrectomy and extraction of erupted upper teeth. No surgical procedure was shown to be the best to prevent the progression of oral infection. Taking into account the patient's general condition, if the patient develops severe symptomatic and refractory osteomyelitis surgery should be considered. The patient and his family are aware of the risks and benefits of surgery and its possible complications.

La osteomielitis crónica en pacientes con osteopetrosis maligna infantil. Presentación de un caso

Palabras clave:
Osteopetrosis
Osteoclerosis frágil
Enfermedad ósea petrificante
Esplenomegalia
Osteomielitis

RESUMEN

La osteopetrosis se caracteriza por un aumento considerable de la densidad ósea que resulta en un remodelado defectuoso, causado por mal funcionamiento de los osteoclastos, de severidad variable. Usualmente se divide en 3 tipos: osteopetrosis dominante autosómica benigna, osteopetrosis recesiva autosómica intermedia y osteopetrosis infantil recesiva autosómica maligna, considerado el tipo de mayor gravedad. Los autores describen un caso de osteomielitis crónica en el maxilar superior de paciente de 6 años de edad con osteopetrosis infantil maligna. El plan de tratamiento incluyó secuestrectomía y exondoncia de los

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dientes superiores erupcionados. Ningún procedimento quirúrgico se ha comprobado que sea superior a otros en la prevención del avance de infecciones bucales. Tomando en cuenta las condiciones generales del paciente al desarrollar osteomielitis refractaria y sintomática severa, la cirugía debe ser considerada. El paciente y sus familiares deben ser conscientes de los riesgos y beneficios de la cirugía, así como de sus posibles complicaciones.

Introduction

Considered a rare inherited skeletal disease, osteopetrosis is characterized by a considerable increase in bone density resulting in defective remodeling, caused by failure in the normal function of osteoclasts, ranging in severity.[1] According to its severity it and can be asymptomatic to fatal[2] and it is often diagnosed by radiographic exams[3], not being essential a bone biopsy.[4]

It is usually subdivided into three types: benign autosomal dominant osteopetrosis; intermediate autosomal recessive osteopetrosis; 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; anemia and thrombocytopenia with subsequent extramedullary hematopoiesis and impaired vision and hearing caused by encroachment of the foramina and nerve canals.[5] 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.[6]

Some differential diagnosis to consider include fluorosis; beryllium, lead and bismuth poisoning; myelofibrosis; Paget's disease (sclerosing form); and malignancies (lymphoma, osteoblastic cancer metastases).[7]

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.[6,8]

Regarding patients with osteopetrosis, osteomyelitis is the most common and well documented maxillofacial complication and can be severe and difficult to treat.[2,3] We describe a case of chronic osteomyelitis in the maxilla of a patient with Malignant Infantile Osteopetrosis (MIOP).

Case report

A 6-year-old female patient, daughter of consanguineous phenotypically normal parents (cousins), attended the Maxillofacial Surgery Department of a hospital in Cuiabá (Mato Grosso, Brazil) accompanied by her grandmother with the story of fall from own height, hitting her mouth on the ground, resulting in the avulsion of two upper incisors. After 15 months, the wound had not healed.

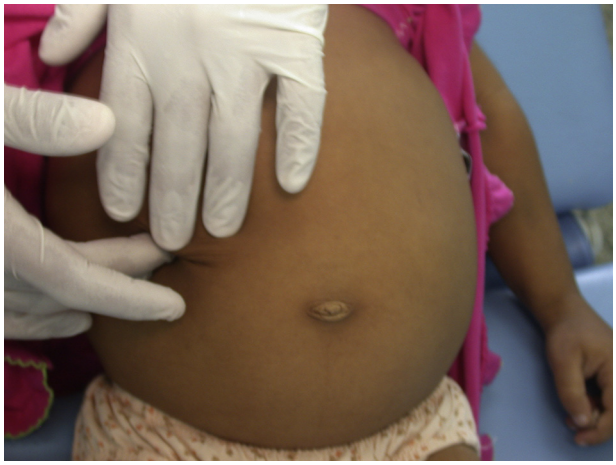

Fig. 1 – Distended abdomen with no soreness to palpation.

In the interview, it was found that at birth the child was diagnosed with normocytic and normochromic anemia, needing transfusion every 15 days. In the first year of life she was diagnosed with MIOP.

On physical examination it was found that her height, weight and facial appearance were not in accordance with her chronological age. She presented distended abdomen with no soreness to palpation (Fig. 1). Percussion revealed a massive sound at the space Traube and hepatosplenomegaly. The spleen reached the region of the left iliac fossa across the median line. The liver reached the region of the iliac fossa on the right side. The surface was hard, smooth, regular edges and blunt. Genu valgus was observed (Fig. 2). Patient also had hearing loss.

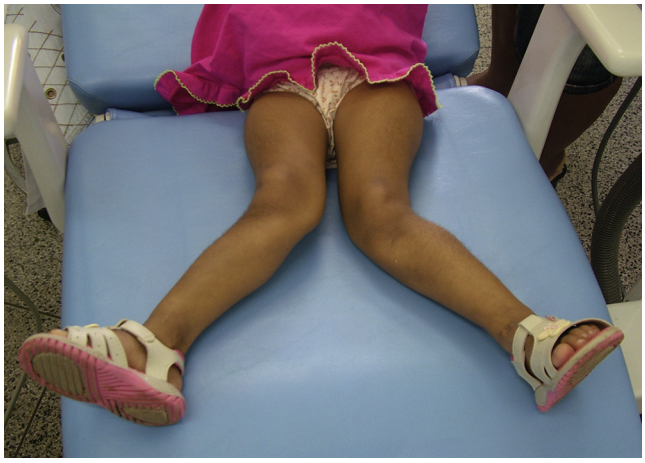

Fig. 2 – Genu valgus.