Oral and Maxillofacial Surgery Cases 4 (2018) 146–150

Contents lists available at ScienceDirect

Oral and Maxillofacial Surgery Cases

journal homepage: www.oralandmaxillofacialsurgerycases.com

Early diagnosis of maxillofacial polyostotic melorheostosis due to altered dental eruption: A case report

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Introduction

Melorheostosis is a rare disorder that causes deposition of dense cortical bone in limbs [1]. It has also been reported in the maxillofacial complex in rare cases [2]. Ethunandan et al. reported a case of a facial polyostotic melorheostosis in a 66 year old female [3]. However, to our knowledge there are not cases of Polyostotic Melorheostosis in pediatric patients reported in the literature. We present a case of a patient that presented for evaluation of mandibular bony projections and failure of teeth to erupt. These clinical findings are consistent with this uncommon entity.

Case description

We present a rare case of polyostotic melorheostosis diagnosed in a 4 year old female referred to the pediatric dentist initially for evaluation of a bony mass on the lower right arch (Fig. 1).

This patient has contributory medical history previously diagnosed of Melorheostosis. The patient underwent multiple biopsies during early childhood consistent with Melorheostosis of the maxilla, nasal bones and frontal bone. The patient continued to follow up with the pediatric dentist as well as her orthodontist to monitor the eruption of the impacted teeth. She presented to the Oral and Maxillofacial Surgery Clinic at Nova Southeastern University, referred by her orthodontist for evaluation of mass on left lower arch. At this point close follow up was established.

Panoramic radiographic examination teeth # 22 and 23 appear to be impacted and a radiopaque lesion is evident (Fig. 2). The patient underwent surgical exposure of impacted teeth # 22 and 23 with excision of soft and hard tissue overlying the mentioned teeth. The excised tissue was submitted to pathology for microscopic evaluation.

Histopathology evaluation showed bony and soft tissue changes consistent with Melorheostosis, these characteristics include absence of increased cellular divisions in osteoclasts and osteoblast with constricted harversian canals (Fig. 3).

After two years from initial surgery the patient returned to the Oral and Maxillofacial Surgery department at Nova Southeastern University for continued follow up. Upon evaluation, teeth # 9,11,22, and 23 failed to erupt and remained impacted and tooth #10 erupted into position (Fig. 4).

A CBCT was taken showing generalized cortical hypertrophy with dense irregular bone formation in the anterior region of the maxillofacial complex. Teeth #9,11,22,23 have failed to erupt (Fig. 5).

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https://doi.org/10.1016/j.omsc.2018.08.002
Received 29 May 2018; Accepted 12 August 2018
Available online 14 August 2018

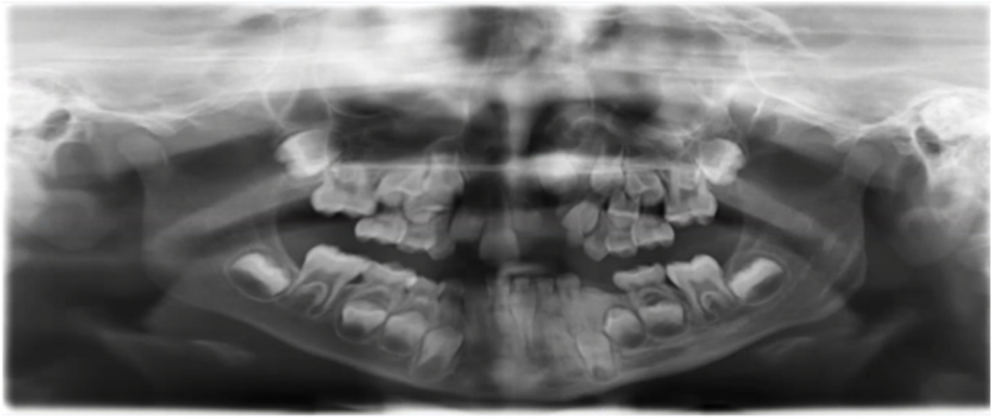

Fig. 1. Initial presentation at age 4.

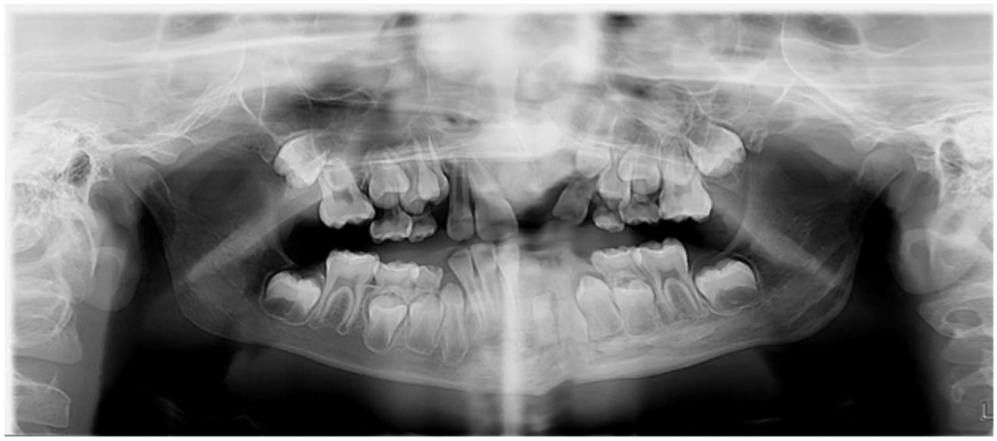

Fig. 2. Follow up Panorex at age 6 demonstrating radiopaque lesion and impacted left lower incisors.

Discussion

Melorheostosis, is a non - hereditary disorder that causes deposition of dense cortical sclerotic bone on the appendicular skeleton [1]. This disease is mainly seen on long bones of limbs, but craniofacial and mandibular manifestations have also been identified [2]. Melorheostosis was first defined by Leri and Joanny in 1922 and affects both males and females equally [3]. The estimated incidence for this rare disorder is low, 9 per million [4]. Seven cases of craniofacial origin have been reported since 1922 and a total of four hundred cases described from 1922 to 2006 [2].

Even though the cause for melorheostosis is unknown, histologically there are a few prominent characteristics. Microscopic histological studies show enlarged bone trabeculae, with constricted haversian canals, that lacked osteoclastic activity. On a cellular level, there is an absence of increased cellular atypia or mitotic figures, which leads to normal levels of osteoclastic and osteoblastic activity. Histologic diagnosis of melorheostosis includes, absence of increased cellular divisions in osteoclasts and osteoblasts along with constricted haversian canals. In some instances fatty tissue formation within the medullary cavity can occur, and become a source of pain for patients [3,5].

Radiographically, cortical hyperostosis in melorheostosis presents as a “flowing candle wax” appearance. The candle wax appearance is due to the flowing nature of sclerotic bone deposition along the cortex of long bones or joints. Subperiosteal or segmented endosteal bone deposition can develop as monostotic, polyostotic, or monomelic [1,7,14]. While melorheostosis is more common in lower limbs, it can affect any bone in the body such as the skull and face [6,7]. Appendicular skeleton bone deposition appears segmented, while cranial bone deposition appears more homogenous with an increase in volume [3].

Melorheostosis is usually asymptomatically present in early childhood, but doesn’t become apparent until early adulthood. This is due to the emergence of symptoms, usually pain which prompts the patient to undergo further testing [8]. Fifty percent of patients develop symptoms by the 2nd decade [2,8]. Signs and symptoms vary, the most common include pain in affected limbs. Other symptoms include; joint contractures, joint swelling, soft tissue alterations, skin hardening, pigmentation, fibrosis, erythema, hard non-tender bony projections, muscle weakness, and reduced range of motion [1,9]. However, nerve alterations have not been proven in the maxillofacial