Melorheostosis and central giant cell granuloma of the mandible in a 15-year-old girl

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Melorheostosis is a nonhereditary bone dysplasia primarily affecting the appendicular skeleton. Because clinical and histologic features are often nonspecific, the diagnosis is often based on the radiographic presentation. Involvement of the craniofacial skeleton is rare. We describe a case of a 15-year-old girl with appendicular and craniofacial melorheostosis with adjacent central giant cell granuloma. We discuss the possible significance of this previously unreported finding. (Oral Surg Oral Med Oral Pathol Oral Radiol 2013;116:e399-e404)

Melorheostosis is a nonhereditary bone dysplasia that primarily affects the appendicular skeleton and may ossify adjacent soft tissues.[1-3] It was initially described in 1922 by Léri and Joanny; the name was derived from the Greek words for limb (*melos*) and flow (*rhein*), because the radiographic appearance resembled melting wax dripping down the side of a candle.[4] Because the clinical and histologic features are nonspecific, the diagnosis is often based on the radiographic presentation.[2]

The typical radiographic presentation of melorheostosis includes an irregular hyperostosis affecting the outer cortical bone. Frequently, the hyperostosis extends into the cancellous bone and may be either completely radiopaque or a mixed pattern. Common locations include the diaphysis of long bones, the pelvis, the ribs, and the bones of the hands and feet. Reports of changes in the craniofacial complex are less common.[1] Although 4 distinct types of melorheostosis are described in the literature, the condition is more practically classified as monostotic or polyostotic.[5] We report a case affecting several areas including the facial bones and jaws of a 15-year-old girl. A detailed description of the radiographic and histologic features is provided, including a possible association of central giant cell granuloma with this condition.

CASE REPORT

Clinical findings

A 15-year-old white girl presented for evaluation of right mandibular swelling of unknown duration and failure of proper eruption of tooth 31. On examination, palpable expansion of the right posterior mandible was noted. Intraorally, the expansion was seen extending from the premolar area to the ramus. The expansion was asymptomatic. No lymphadenopathy was noted, and blood studies were within normal limits. No significant family history of disease was known.

Radiographic findings

A computed tomography (CT) scan of the head and neck was obtained to evaluate the extent of structural changes. Axial and coronal images were acquired using a helical CT with exposure settings of 100 kilovolt peak, 150 mA, slice thickness of 0.8 mm, and resolution of 0.468 mm^2. Bone changes were noted in the right parietal, temporal, zygomatic, sphenoid, ethmoid, maxillary, and mandibular bone (Figure 1). The lesion presented with mixed density but mostly as ground-glass changes with irregular but well-delimited margins and cortical expansion. In the right parietal and temporal bones, thickening of the cortices with sclerosis was observed. In the sphenoid, the same irregular ground-glass sclerosis was noted, including thickening of the medial and lateral pterygoid plates. The sphenoid sinus was displaced to the left and diminished in size. Constriction of the right superior and inferior orbital fissures was also observed. In the maxilla, 3-dimensional reconstructions clearly exhibited involvement of only the right side, with expansion of the outer maxillary cortex showing the classic dripping-wax appearance of melorheostosis (Figure 2). In the nasal cavity, the inferior nasal concha was hypoplastic and superiorly displaced. Owing to cortical expansion, the right nasal cavity was partially obliterated. Other radiographic findings in the maxilla included superior displacement of the floor of the right sinus and impaction of the fully developed right second molar tooth and developing third molar. In the mandible, the same ground-glass changes with cortical expansion extended posteriorly from the periapical region of the right mandibular canine to the posterior third of the ramus. Changes extended superiorly from the lower border of the mandible to the sigmoid notch and coronoid process. The right condyle was not involved. The mandibular canal was inferiorly displaced and constricted in some areas. The mandibular right second molar was buccally displaced, and the third molar was lingually displaced. The molar teeth of the right jaws exhibited a somewhat abnormal morphology, with underdeveloped and stunted roots, when compared with the left side. The mandibular right second

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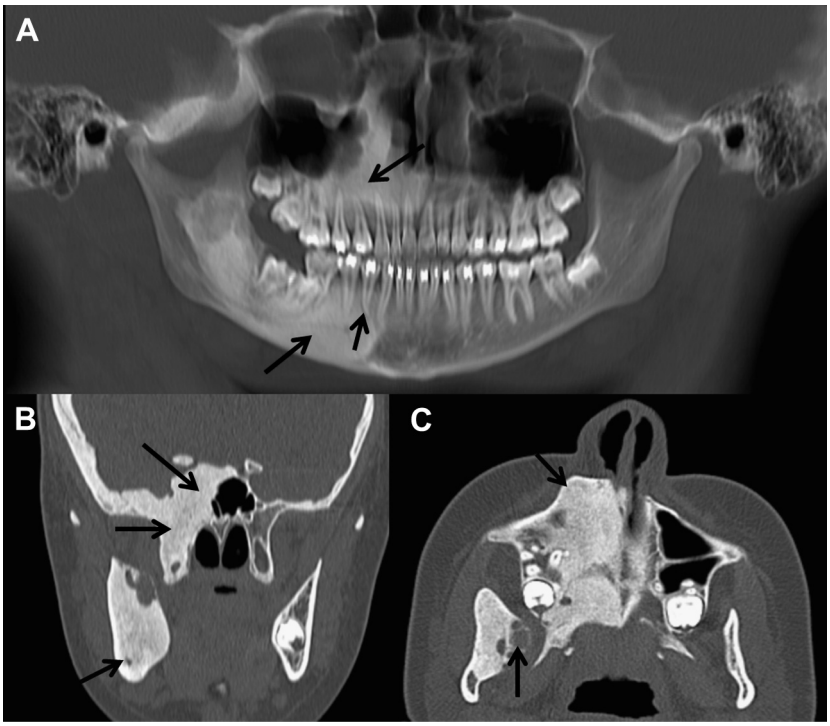

Fig. 1. Computed tomography (CT) examination. **A**, Panoramic reconstruction from CT data. Note ground-glass bone, dental displacement, and widening of periodontal ligament spaces in the right maxilla and mandible. **B**, Coronal CT reconstruction depicts extension of lesion in the sphenoid and mandible. Note lateral displacement of the right sphenoid sinus and thickening of the pterygoid process and mandible, with cortical expansion and inferior displacement of the mandibular canal. **C**, In the axial CT scan, unilateral involvement is noted with typical ground-glass bone and cortical expansion of the anterior maxilla, as well as similar changes of the right mandible. Note multilocular radiolucency in the medial aspect of the right mandible.

molar was impacted, with root dilaceration. A multilocular radiolucency with scalloped margins and cortical thinning with expansion was noted in the right posterior mandible. All involved maxillary and mandibular teeth exhibited irregular widening of the periodontal ligament spaces and absence of lamina dura.

In addition to the CT scan, radiographic plain projections of the lower limb were available and demonstrated additional characteristic findings associated with appendicular melorheostosis. In the lateral projection of the left femur (Figure 3, *A*) and anteroposterior projection of the lower limb (see Figure 3, *B*), endosteal hyperostosis, linear ground-glass sclerosis, and mild expansion were present in the diaphysis of both the femur and tibia. In the epiphyses of these bones, rounded areas of hyperostosis with irregular contours characteristic of melorheostosis were present. Dorsiplantar and lateral projections of the left foot showed similar irregular endosteal hyperostosis involving the first and second distal, intermediate, and proximal phalanges; metatarsals I and II; the medial and intermediate cuneiforms; and the navicular bones. Lateral deviation of the second toe was also evident (see Figure 3, *C*).

Histopathologic findings

Microscopic examination of the radiolucent lesion removed from the right posterior mandible showed a benign proliferation of ovoid to spindle-shaped cells within a background of dense fibrous connective tissue. Scattered throughout the tissue were numerous multinucleated giant cells, with prominent erythrocyte extravasation and hemosiderin deposition (Figure 4). Based on these features, a diagnosis of central giant cell granuloma was made. A subsequent biopsy of the mandibular bony changes was later done, with a diagnosis of dense cortical bone, consistent with melorheostosis.

At the 3-year follow-up in March 2010, no recurrence of the giant cell granuloma was seen clinically or radiographically. However, some clinical deformity was noted involving the right temporal region as the patient's melorheostosis continued to progress. Ocular symptoms, consisting of partial blindness, had also developed, necessitating optic nerve decompression the previous year. Additional sensory disturbances were not reported.

DISCUSSION

Melorheostosis is a rare, benign bone disorder characterized by hyperostosis that, in later stages, radiographically resembles melting candle wax. It is classified among the "sclerosing bone dysplasias," which also include disorders such as craniometaphyseal and diaphyseal dysplasias, osteopetrosis, pyknodysostosis, osteopoikilosis, and osteopathia striata.[6]