CASE REPORT

Gigantiform cementoma in a child

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The objective of this report is to present an unusual case of a gigantiform cementoma manifesting with gross expansion of the maxilla in a 6-year-old black female. No history of similar lesions in the family of the patient was obtained. Radiographic examination revealed a well-circumscribed lobular radio-opaque mass surrounded by a radiolucent margin. The macroscopic specimen showed lobular mineralized masses with a "ginger root"-like appearance. Microscopic examination showed a lobular calcified mass with a peripheral zone of fibroproliferative tissue corresponding with the radiolucent margin. The differential diagnoses, which include osteoma, ossifying fibroma and cementoblastoma, are discussed.

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Case report

A 6-year-old female presented with a main complaint of a painless maxillary swelling which her parents noticed 8 months earlier. No history of similar lesions in family members could be obtained. Furthermore, the medical history of the child was uneventful. An examination revealed a bony hard swelling in the left maxilla (Figure 1). The swelling depressed the lower eyelid with exposure of the sclera, and expanded the palate. The skin over the lesion was freely movable and no nerve fall-out was noted. A panoramic radiograph showed a well-defined round lobular radio-opacity in the left maxilla extending from the maxillary alveolus to the inferior orbital border and left maxillary tuberosity (Figure 2). All teeth posterior to the first primary molar were absent, owing to previous extractions, and the remaining tooth germs were displaced. A CT image showed a spherical lobular radio-opaque mass measuring 4.2 cm in diameter surrounded by a narrow radiolucent rim (Figure 3). Obliteration of the maxillary sinus and expansion of the maxillary bony cortex was noted. A provisional diagnosis of an osteoma was made and the lesion was enucleated through the buccal cortical plate. The macroscopic specimen was densely calcified and consisted of several well-defined lobular bony masses, aptly described as "ginger roots"

(Figure 4). Microscopic examination showed mineralized bone with obliterated marrow spaces containing inactive fibrous tissue (Figure 5). The radiolucent peripheral margin showed cellular fibrous connective tissue with psammomatoid and trabecular bony deposits. A diagnosis of a gigantiform cementoma was made.

Discussion

Osseous dysplasias (ODs) are generally slow growing, non-expansive, fibro-osseous jaw lesions of unknown aetiology.¹ The 2005 World Health Organization classification² divides ODs into focal, periapical and florid OD and familial gigantiform cementoma (FGC). Focal OD is a solitary lesion affecting a single area in the jaw bone and mainly the posterior quadrant of the jaws. Periapical OD occurs in the anterior mandible and involves the periapical regions of a few incisor teeth, while florid OD is more extensive and can involve the mandible bilaterally or all four jaw quadrants. FGC is uncommon, occurs predominantly in young patients and has the potential to cause considerable jaw expansion.^{1,2} Not all cases are familial and sporadic cases have been reported.³ FGC has an equal gender distribution and although FGC appears to have a predilection for white people, a few cases have been reported in black people^{4,5} and Jamaicans of African descent.¹ The early radiological features of FGC resemble the radiolucent

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Figure 1 Clinical features demonstrating expansion of the left maxilla by a firm mass

and mixed radiolucent–radio-opaque appearance of the other more common OD types. During maturation, significant expansion, tooth displacement and progressive opacification occur, all of which are features which distinguish FGC from florid OD. Mature FGCs cease to enlarge and manifest as a lobular radio-opacity with a radiolucent rim.¹

The radiological differential diagnosis of a well-defined, expansive mineralized mass in the jawbone includes benign cementoblastoma, osteoma, ossifying fibroma and FGC. Benign cementoblastoma is a benign odontogenic neoplasm of cementoblastic origin, occurs most commonly in the mandibular first molar area, is attached to the root of an associated tooth and is surrounded by a well-defined radiolucent margin.⁶ The radiological differentiation between cementoblastoma and FGC is difficult because both lesions may occur in the periapical regions of molar teeth and exhibit a radiolucent rim around the periphery of the radiodense mass. Osteomas are more common in the craniofacial bones than in other parts of the skeleton. They may

Figure 3 Axial CT image showing the radiodense lobular mass surrounded by a radiolucent margin. The maxillary sinus and nasal passage are obliterated and the midline is slightly deviated to the right

develop on the surface of the involved bone when they originate from the periosteum (periosteal osteomas) and present as a polypoid mass or they may be located in the cancellous bone where they are often referred to as endosteal osteomas.⁷ Multiple osteomas may be a sign of Gardner's syndrome, which in its classical form consists of the clinical triad of epidermal cysts of the skin, pre-malignant adenomatous colonic polyps and osteomas.⁸ Radiologically, an osteoma appears as a radiodense mass and is distinguished from periapical idiopathic osteosclerosis (enostosis) and foci of chronic sclerosing osteomyelitis through its circumscription and tendency to enlarge.⁸ Osteomas lack the lobular radio-dense appearance of mature FGCs. A characteristic of all the lesions categorized as ODs (including FGC) is their fibro-osseous appearance in the early radiolucent and mixed-density stages of growth. As ODs mature, they progressively exhibit mineralization, and in the late stage consist of characteristic lobular calcified masses of

Figure 2 Panoramic radiograph showing the highly mineralized lobular lesion in the left maxilla. A radiolucent margin is present around most of the lesion

Figure 4 Macroscopic features of the enucleated specimen. Note the "ginger root"-like appearance of the mineralized masses

bone-like tissue which macroscopically resemble "ginger roots".⁹ These unique features are demonstrated clearly by our case. Out of the group of lesions categorized as ODs, only FGC presents clinically with significant jaw expansion.^{1,10} FGC occurs at a younger age than conventional ODs, a characteristic compatible with our case. The non-expansive ODs are not treated when asymptomatic and biopsy is ill-advised owing to the risk of infection. Infected lesions and those presenting with progressive expansion are generally removed surgically through enucleation. Incomplete removal or shave-off contouring are ill advised, as the lesions enlarge more rapidly in the early radiolucent stage.

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Figure 5 Micrograph demonstrating the bony mass showing a lobular appearance with small marrow spaces (arrows). The peripheral fibroproliferative zone (asterisk) corresponds with the radiolucent margin (H&E stain, × 200)

However, the management of the expansive FGCs entails complete surgical removal at an early stage of growth in order to limit the extent of surgery and prevent tooth displacement and loss of residual bone.

In our patient the radiological, macroscopic and microscopic appearances are characteristics of a late-stage OD. Although our patient is young, the absence of a family history does not fulfill the qualification for the diagnosis of a FGC type of OD. The term "familial gigantiform cementoma" is inappropriate and the authors suggest "expansive osseous dysplasia" as an alternative.