Expansive osseous dysplasia: report of 9 lesions in an African population sample and a review of the literature

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Objectives. The aim of this study was to report on the clinicopathologic features of osseous dysplasias (ODs) that manifest with gross expansion in a black South African hospital population sample.

Study design. The files of 9 histology-verified expansive ODs in 8 patients were reviewed. The clinical records and radiographs were analyzed and compared with reports in the literature.

Results. The main complaint of all patients was related to expansion of the jaw. All except 1 were female, with ages between 26 and 71 years and sizes of the lesions ranging from 4 to 15 cm. No history of similar lesions in family members was obtained. All lesions occurred in the mandible and 6 crossed the midline. The expansive growth pattern was associated with persistence of radiolucent areas in the lesions which microscopically consisted of cellular fibro-osseous tissue. Resorption of mineralized deposits by osteoclasts was prominent in the radiolucent parts of the lesion. Maturation with enlargement of the radiodense component was associated with a decrease in osteoclast activity and the formation of lobular bone masses and confluent psammomatous mineralized deposits.

Conclusion. We propose the term “expansive osseous dysplasia” for the rare albeit important clinical subcategory of ODs that manifest with progressive jaw expansion. Suspension of osteoclast activity plays an important role in the maturation of the lesions into dense mineralized masses. (Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2011;111:e35-e41)

The osseous dysplasias (ODs) are idiopathic nonneoplastic conditions characterized by fibro-osseous replacement of bone in the periapical region of teeth. After a period of inconsistency in the application of nomenclature and the use of controversial terms in designating the mineralized deposits as “cementum,” the World Health Organization recommended in 2005 a subclassification of OD into 4 subtypes.1 Focal OD is a solitary lesion with a predilection for the posterior jaw quadrant,2 periapical OD affects the periapical regions of several mandibular incisors, and florid OD is more widely spread and involves the posterior mandible bilaterally or often all 4 posterior quadrants simultaneously.1 The fourth subtype, familial gigantiform cementoma (FGC),1,3 is a rare autosomal dominant disorder with variable phenotypic expressivity4,5 that is histologically indistinguishable from the more common florid ODs. It is noteworthy that although “cementoma” was no longer recommended as a descriptive term for the nonexpansive ODs, it was retained for FGC. Persistent growth with jaw expansion is reported to be the distinctive feature of FGC. Jaw expansion seldom occurs and, if ever, to a lesser extent in florid OD.6 Sporadic (nonfamilial) cases of ODs with gross expansion were diagnosed in the past as “gigantiform cementoma.”7 A rare association between FGC, brittle bone disease, pathologic fractures, and osteosarcoma was described by Rossbach et al. in 20058 and Moshref et al. in 2008.9 Florid ODs and all periapical and focal ODs mature through an early radiolucent stage, an intermediate mixed stage, and a late radiopaque stage after which the lesions cease to enlarge with a resultant failure to expand the cortical plates and displace teeth. Management of the nonexpansive cases entails the prevention of infection.1 Due to the risk of infection during the taking of a biopsy, asymptomatic cases are diagnosed on radiographs only. Once infected, ineffectiveness of antibiotic therapy6 owing to the avascular nature of the mineralized masses makes sequestrectomy the procedure of choice. The purpose of the present study was to report on ODs with an expansive growth characteristic in an African population sample.

MATERIAL AND METHODS

The files of the units of Radiology and Oral Pathology at the University of Limpopo were reviewed for cases with histologically verified ODs that manifested with jaw expansion between 2000 and 2010. These units render histopathologic and radiologic services to clinics in the
rural and periurban areas of the northern sector of South Africa and serve mainly a rural black population sample.

RESULTS

A total of 2,245 cases of OD were diagnosed over the 10-year period of which 9 lesions in 8 patients showed signs of gross jaw expansion (Fig. 1, A and B). Jaw expansion was the only complaint recorded in the files of the 8 patients, and no histories of similar lesions in family members were obtained. The ages and genders of the patients, sites of involvement, clinical sizes measured in centimeters, and descriptions of the radiologic appearances are presented in Table I. All patients presented with a single expansive lesion in the mandible except case 1 that had 2 expansive lesions (Fig. 2). Teeth at the site of the lesions were displaced but not resorbed. Out of 9 lesions, 6 crossed the midline. All lesions were radiologically well defined, showed gross expansion, and were of mixed radiodensity. The radiodensities in the lesions were spherical in shape and coalesced to form larger lobular radiodense masses (Fig. 3). In lesions with a high radiopaque content, the masses were surrounded by a radiolucent periphery of varying widths (Figs. 4 and 5). Two lesions occurred in patients with late-stage florid OD (Fig. 6) and 1 with early-stage florid OD (Fig. 7). Two lesions showed simple bone cyst formation (Figs. 2 and 3). Microscopic examination of the radiolucent areas in all cases showed areas of cellular fibrous connective tissue containing foci of active woven bone formation and psammomatoid deposits (Fig. 8, A). Osteoclastic activity was prominent in the radiolucent areas (Fig. 8, B). The radiodense parts of the lesions showed an increase in mineralized tissue with a reduction of the cellularity of the fibrous component and absence of osteoclasts. Two types of mineralized tissue that manifested on radiographs as radiodense were recorded: dense, relatively acellular lobular deposits resembling bone (Fig. 8, C) and confluent acellular psammomatous masses (Fig. 8, D). The former were identified as well defined lobular gingery root-like mineralized masses in the excision specimen, and the latter presented as more diffuse areas with a gritty consistency upon sectioning. In all specimens that contained teeth, we were unable to demonstrate a microscopic interface between dental cementum and the lesion. The treatment of choice was surgical removal by enucleation, although some were resected due to reconstructive considerations. No recurrences were reported over the period of the study.

DISCUSSION

Expansive ODs comprise only 0.35% of the total number of ODs diagnosed in our African population sample, underscoring the extreme rarity of the condition. The main complaint of all our patients was related to the unesthetic appearance of the painless swelling of the jaw. Tooth displacement was recorded in all dentate subjects. The majority of cases reported as FGC in the literature occurred in caucasians despite the common occurrence of the classic ODs in African patients. Coleman et al. reported the first FGCs involving 3 African family members in 1996, and Rossbach et al. described the condition in 4 members of an African-American family in 2005. It is interesting to note that in Coleman et al.’s sample of 3 FGCs (reported as familial florid cemento-osseous dysplasias) in a black South African family, both genders were affected and a predilection for the anterior area of the mandible was noted. The lack of cases with a familial history in our
sample and the scarcity of reports on FGC in people of African origin suggest that FGC is less common among Africans than in other population groups. Our sample of sporadic nonfamilial expansive ODs demonstrates a predilection for women, a wide age range (26-71 years), and involvement of the anterior mandible by the majority of cases, unlike FGC, which has an equal gender distribution,11 predilection for young individuals, and manifestation in all quadrants of both mandible and maxilla.6

The main radiologic differential diagnosis of expansive ODs is the classic and more common florid, periapical, and focal ODs. Before expansion sets in, the distinction between the classic types and expansive OD may be difficult. The classical ODs show maturation into dense lobular mineralized masses resembling ginger roots macroscopically1 and fail to enlarge after complete mineralization has occurred. Quantification of the extent of mineralization in relation to the size of an OD may be an important indicator of the potential of a lesion to expand. The biopsy samples and excision specimen of our cases show the presence of a significant proportion of fibrocellular tissue (presenting radiologically as radiolucent) between the confluent radiopaque mineral deposits, many of which were in the process of transforming into ginger root like mineralized masses. The radiolucent or mixed radiolucent-radiopaque fibro-osseous proliferation in the early stages of ODs may resemble ossifying fibroma radiologically. Confluent lobular radiopaque mineralized deposits are, however, the hallmark of advanced ODs and differentiate them from a diagnosis of ossifying fibroma. In ossifying fibromas, the connective tissue increases at the expense of the mineralized component

Table 1. Age, gender, site, size and radiological appearance

<table>
<thead>
<tr>
<th>Case</th>
<th>Age, y</th>
<th>Gender</th>
<th>Site</th>
<th>Size, cm*</th>
<th>Radiologic appearance</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>42</td>
<td>F</td>
<td>21-32 and 17-20</td>
<td>9 and 5</td>
<td>Two expansive lesions with mixed density, tooth displacement, 3 simple bone cysts</td>
</tr>
<tr>
<td>2</td>
<td>48</td>
<td>F</td>
<td>20-29</td>
<td>8</td>
<td>Buccal and lingual expansion, mixed density, 2 simple bone cysts</td>
</tr>
<tr>
<td>3</td>
<td>71</td>
<td>F</td>
<td>21-26</td>
<td>4</td>
<td>Buccal and lingual expansion, central radiopacity with lucent periphery, tooth displacement</td>
</tr>
<tr>
<td>4</td>
<td>47</td>
<td>M</td>
<td>29-32</td>
<td>5</td>
<td>Buccal expansion, lobular radiopacity with core of mixed density, lucent periphery</td>
</tr>
<tr>
<td>5</td>
<td>26</td>
<td>F</td>
<td>20-30</td>
<td>15</td>
<td>Buccal and lingual expansion, mixed density, tooth displacement, associated with late-stage florid OD</td>
</tr>
<tr>
<td>6</td>
<td>35</td>
<td>F</td>
<td>21-28</td>
<td>5</td>
<td>Buccal and lingual expansion, radiopacity with broad lucent periphery, tooth displacement, associated with early florid OD</td>
</tr>
<tr>
<td>7</td>
<td>46</td>
<td>F</td>
<td>21-28</td>
<td>5</td>
<td>Buccal expansion, well defined mixed density, tooth displacement, associated with late-stage florid OD</td>
</tr>
<tr>
<td>8</td>
<td>42</td>
<td>F</td>
<td>29-31</td>
<td>4</td>
<td>Buccal and lingual expansion, fusiform mixed density, tooth displacement</td>
</tr>
</tbody>
</table>

*OS, Osseous dysplasia.

*Largest dimension of the lesion.

Fig. 2. Case 1: panoramic radiograph showing 2 expansive lesions located in the left and anterior-to-right mandible, respectively. Note their mixed radiolucent-radiopaque appearance and lobular radiopacities. Three simple bone cysts (arrows) appear round, radiolucent, and with smooth contours.
and the neoplasm often becomes less radiodense with increasing size.\textsuperscript{12}

The radiologic appearance of the second lesion in the anterior right mandible of case 1 created the impression of a collision growth between 2 separate lesions with the site of contact in the premolar area. It can be speculated that case 1 originally had 3 ODs which presented in the early stage as bilateral mandibular lesions reminiscent of florid OD and anterior mandibular lesions presenting as periapical OD. In a recent case report, the presence of hereditary osteomatous jaw lesions in a case of Gardner syndrome caused diagnostic confusion with FGC.\textsuperscript{13} This may cause interpretive problems in patients with >1 expansive lesion, as demonstrated in 1 of our cases. Other radiologic differential diagnoses include chronic sclerosing osteomyelitis and benign cementoblastoma. Benign cementoblastomas are unlike ODs attached to the surface of the root of the involved tooth (most commonly a mandibular molar) and are usually surrounded by a radiolucent periphery.\textsuperscript{14} Chronic sclerosing osteomyelitis is generally nonexpansive, poorly demarcated, and associated with an infective etiology.

The microscopic differential diagnosis includes all the fibro-osseous lesions and may be challenging in the early stages of growth. In small lesions, the well demarcated mixed radiologic appearance may prompt a diagnosis of ossifying fibroma. The earlier fibro-osseous appearance of OD is progressively transformed into

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Fig. 3. Case 2: axial computerized tomography view demonstrating the expanded mandible. Note the mixed radiolucent-radiopaque appearance, the lobular radiopacities and round-to-oval smooth contoured radiolucent areas corresponding with simple bone cyst formation (arrows).

Fig. 4. Case 3: panoramic image showing a lobular radiopacity surrounded by a radiolucent periphery. Note displacement of the incisors.

Fig. 5. Case 4: radiograph of the resection specimen. Note the central mixed radiolucent-radiopaque appearance, lobular radiopacity, and peripheral radiolucent rim (arrows).
lobular hypocellular mineralized masses making a microscopic distinction possible. The transformation of psammomatous deposits into acellular globular masses and growth of trabecular bone into hypocellular lobular structures with a characteristic macroscopic “ginger root” appearance take place at the expense of the cellular fibrous tissue. This distinction can also be made on the specimen received in the pathology laboratory. Ossifying fibromas are submitted to the laboratory as either encapsulated firm fibrous tumors or larger fibrous masses with calcifications on cut surface, whereas ODs typically consist of small gritty fragments of tissue in the earlier stages of growth or large mineralized masses resembling ginger roots in the mature stage.1 The prospect for expansion diminishes as the lesions become increasingly radiodense. Growth is therefore dependent on the maintenance of the cellular fibrous zones in a lesion. This implies that our case 2, despite its larger size, was not as close to maturity and cessation of growth than case 3, which was smaller, more extensively mineralized, and surrounded by a narrower radiolucent rim, indicating termination of its growth stage. Microscopic evidence suggests that retained activity of osteoclasts may play a role in maintenance of the fibrous (radiolucent) areas, because the enlargement of the mineralized component was found to be associated with a distinctive reduction in the numbers of osteoclasts. The occurrence of simple bone cysts was histologically confirmed in 2 of our lesions. This phenomenon has been reported previously in association with florid OD.6,15 Care should be taken, however, not to interpret the radiolucent fibrocellular areas in expansive ODs as simple bone cyst change. The latter is readily distinguished from the irregular radiolucent fibrocellular areas by its smooth contoured round-to-oval radiolucent appearance.

To group the expansive ODs with expansive growth potential in 1 category, the term “expansive osseous
"dysplasia" is suggested which should include the familial- and nonfamilial (or sporadic) subtypes. Separation of this category of ODs from the nonexpansive types is necessary, because unlike the more common nonexpansive ODs, they are treated differently. The treatment of choice for expansive ODs is total surgical removal. Shave-down surgical procedures to improve esthetics are not successful, because the lesions rapidly regrow. This is in contrast to the nonexpansive ODs, which require no treatment unless secondarily infected.

Some of our lesions were, however, resected owing to reconstruction-related complications resulting from diminished residual bone at the site of the lesion. The term "gigantiform cementoma" should fall into disuse, because the international trend is to move away from the term "cementoma" (referring to a benign neoplasm of cementum) unless the mineralized growth is attached to a root surface, thereby satisfying the definition of cementum. In the present sample, no attachment to a tooth surface could be demonstrated, a disqualification for using "cementum" in diagnostic terminology. The word "gigantiform," which refers to the exceptional...

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**Fig. 8.** A, Microscopic view of a radiolucent area in case 2. Note the fibro-osseous appearance and psammomatous (black arrows) and trabecular (white arrows) mineralized deposits (hematoxylin-eosin [HE] stain, ×100). B, Radiolucent area in the anterior lesion of case 1, showing a resorptive facet with osteoclasts (arrows) on the surface of a mineralized deposit (HE stain, ×200). C, Microscopic appearance of the lobular radiodense area in the right side of case 1. Note the dense bone with absence of the fibro-osseous proliferation except for the small areas indicated with arrows (HE stain, ×40). D, Cortical bone (right), fibro-osseous proliferation representing the radiolucent rim (between arrows), and radiodense core (left) composed of confluent psammomatous mineralized deposits (case 4, HE stain, ×40).
size of a lesion, is also inappropriate, because only a small percentage of neglected expansive ODs achieve this status.

The present manuscript describes a rare, albeit important, subtype of OD for which the term “expansive osseous dysplasia” is suggested. The potential for gross disfigurement and a different therapeutic approach warrant separate categorization in the group of ODs. No recurrences have been recorded in our study, indicating the nonaggressive nature of expansive OD.

REFERENCES