Unicystic Ameloblastoma In a Child: A Differential Diagnosis From the Dentigerous Cyst and the Inflammatory Follicular Cyst

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ABSTRACT

Unicystic ameloblastoma (UA) is a benign epithelial odontogenic tumor of the jaws with an aggressive potential that commonly occurs in children. This cystic odontogenic neoplasm is generally asymptomatic and found during routine radiographs. The purposes of this report were to describe a case of UA involving the crown of the unerupted right mandibular second premolar in an 11-year-old girl under orthodontic treatment, and discuss its diagnosis and radiographic and microscopic findings, emphasizing its distinction from the dentigerous cyst and the inflammatory follicular cyst. (J Dent Child 2007;74:245-9)

KEYWORDS: UNICYSTIC AMELOBLASTOMA, ODONTOGENIC TUMOR, DENTIGEROUS CYST, INFLAMMATORY FOLLICULAR CYST, CHILDREN

The unicystic ameloblastoma (**UA**) is a benign cystic neoplasm arising from the tooth-producing apparatus or its remnants.[1-5] This lesion represents 5% to 23% of all ameloblastomas[1,5-8] and has a considerable incidence in children.[5,9,10] Odontogenic tumors are uncommon lesions in the Brazilian population (2%). The ameloblastoma, however, is the most frequent of these tumors (45%).[5]

The term "unicystic ameloblastoma" was adopted in the second edition of the *Histological Typing of Odontogenic Tumors*.[2] Other terms not generally used today are "mural ameloblastoma"[11] and "cystic ameloblastoma."[12]

UA in children is an asymptomatic lesion that is generally found in routine radiographs.[10] It is preferentially located in the mandible,[5,10] frequently associated with unerupted teeth,[7,10] and often misdiagnosed as a dentigerous cyst.[10]

According to Ackerman et al,[6] there are 3 microscopic types of UA:

- Type I is a single cyst lined by ameloblastomatous epithelium with no infiltration into the fibrous cyst wall, which may often be seen in focal areas.
- Type II includes the features of type I, plus intraluminal proliferations.
- Type III includes the features of type I, plus invasion of epithelium into the cyst wall in either follicular or plexiform patterns (intramural proliferations).

The microscopic pattern that exhibits mural invasion in UA suggests a more aggressive potential.[6,10]

This report's purposes were to describe a case of UA involving the crown of the unerupted right mandibular second premolar in an 11-year-old Caucasian girl under orthodontic treatment, and discuss its diagnosis and radiographic and microscopic findings, emphasizing its distinction from the dentigerous cyst and the inflammatory follicular cyst.

CASE REPORT

In October 2002, an 11-year-old Caucasian girl was referred to the Oral Medicine Center of Goiás State, School of Dentistry, Federal University of Goiás, Goiânia, Goiás, Brazil, for evaluation of a cystic lesion detected during routine panoramic radiograph examination as a follow-up of orthodontic treatment.

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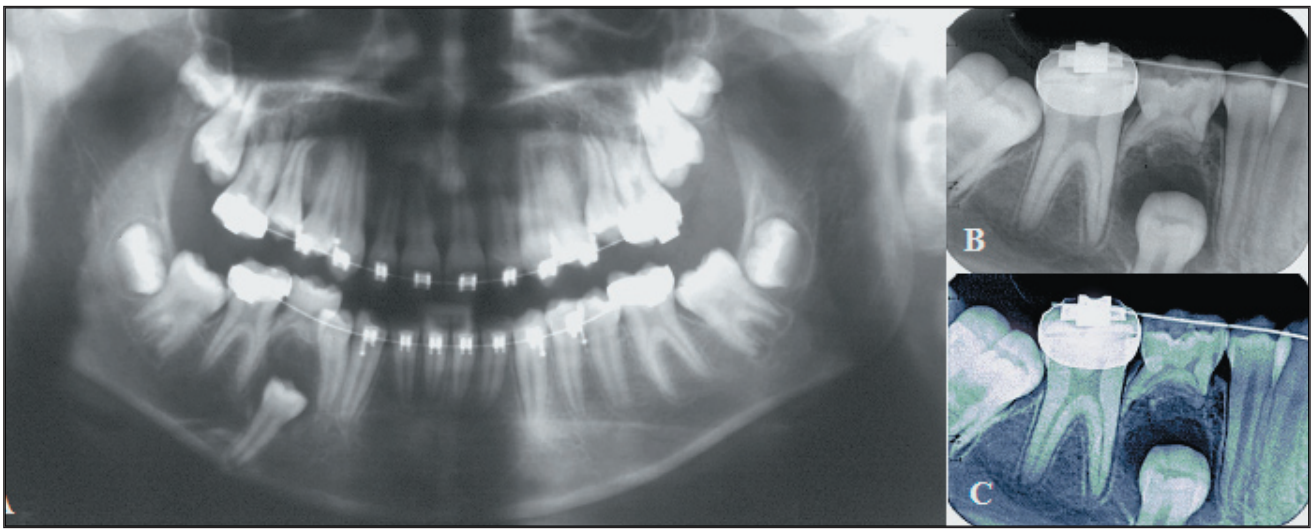

Figure 1. Panoramic (a), periapical conventional (b), and digitized electronically processed (c) radiographs revealed a unilocular, well-defined, radiolucent lesion surrounding the unerupted right inferior second premolar's crown and involving the right inferior primary second molar's apex. Note the radiolucent area showing an undefined margin.

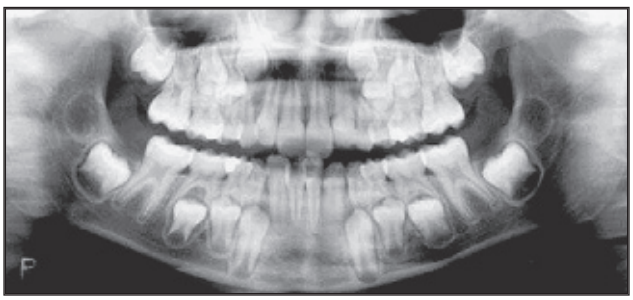

Figure 2. Radiographic evaluation showing a radiolucent area surrounding the second premolar's crown, suggestive of a normal dental follicle.

RADIOGRAPHIC AND CLINICAL FINDINGS

Panoramic and periapical radiographs revealed a unilocular, well-defined, radiolucent lesion, with an undefined margin, surrounding the crown of the unerupted right inferior second premolar with a delay in the normal eruption (Figures 1A, 1B, and 1C). These radiographs demonstrated that the right mandibular second premolar was located near the mandibular inferior border (Figure 1A). This lesion also involved the apex of the right mandibular primary second molar that exhibited resorption of the mesial and distal roots (Figures 1B and 1C). In addition, no carious lesion in tooth no. T was observed (Figures 1B and 1C). Figure 2, taken at the beginning of the orthodontic treatment 1 year before. At this time, the radiographic evaluation showed a radiolucent area surrounding the second premolar's crown, suggesting the presence of a normal dental follicle. An extraoral examination revealed no signs of abnormality. The intraoral examination demonstrated that tooth no. T contrasted with teeth nos. S, L, and K that were replaced by corresponding successors. Clinical examination also revealed no caries and positive response to pulp sensitivity test (thermic) in the right inferior primary second molar. The mucosa around the involved site appeared clinically normal.

On the basis of the radiographic and clinical features, it was assumed that the lesion was a dentigerous cyst.

MACROSCOPIC AND MICROSCOPIC FINDINGS

Treatment consisted of enucleation of the lesion and extraction of the right mandibular primary second molar. The surgical area was curetted and the specimen was submitted to microscopic examination. Macroscopic evaluation revealed cyst-like soft tissue with fibrous consistency, irregular shape, yellow in color, and a 10x9x3 mm size.

Microscopical examination showed the cavity to be lined by ameloblastic epithelium (Figure 3A), presenting columnar basal cells with hyperchromatic nuclei, nuclear palisading with reverse polarization, and cytoplasmic vacuolation (Figures 3B and 3C). The overlying epithelium presented characteristics mimicking stellate reticulum (Figure 3C). In addition, the following were observed: the presence of strands/cords with an plexiform arrangement in the connective tissue wall, islands or follicles composed of a peripheral layer of ameloblastomatous epithelial cells, and reticulum-like stellate in the center (Figures 3A and 3D).

The lesion was microscopically diagnosed as unicystic ameloblastoma with intramural proliferations.

FOLLOW-UP

A follow-up examination at 12 months postoperatively showed good healing with significant bone repair and substantial eruptive movement of the right permanent second premolar. There was no evidence of any residual or recurrent intraosseous tumor (Figure 4). To date, after a 4-year-period of follow-up, the second premolar is in correct occlusion and no evidence of recurrence of tumor has been observed (Figure 5). Due to the conservative nature of the operation performed, the patient remains under close observation.

DISCUSSION

UA is an odontogenic tumor of a developmental origin and may present diagnostic difficulties, particularly if the presentation mimics other odontogenic osseous pathologies.[2,4,5,13,14] In this case study, taking into consideration

clinical and radiographic findings, the differential diagnosis included an inflammatory follicular cyst, a radicular cyst, or a dentigerous cyst.

Although the inflammatory follicular cyst occurs at the apex of a deciduous tooth and involves the permanent tooth successor's dental follicle and crown, it is always associated with an inflammatory etiology.[13] The association between persistent and prolonged inflammation of a primary tooth and the development of a follicular cyst involving the permanent successor has been extensively discussed in the literature.[13,15-17] In the present case, there was no clinical or radiographical evidence of chronic infection (caries, pulpitis, or periodontal disease) in the primary tooth associated with a lesion. Thus, a diagnosis of inflammatory follicular cyst was disregarded. In addition, vital pulp and no caries in the right inferior primary second molar ruled out a chronic radicular infection.

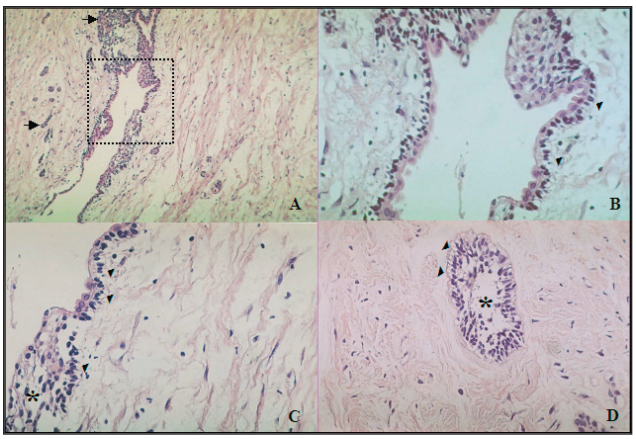

Figure 3. Photomicrographs of unicystic ameloblastoma (a to d) with intramural proliferations (arrows) showing a cavity lined by ameloblastous epithelium. This presents columnar basal cells with hyperchromatic nuclei, nuclear palisading with reverse polarization, and cytoplasmic vacuolation (arrowhead; b and c). B and c show close-up views of the detached area in a. The overlying epithelium showed features of mimicking stellate reticulum (; c). Note that the connective tissue stroma, the presence of islands or follicles is composed of a peripheral layer of ameloblastomatous epithelial cells (arrowhead) and, in the center, a reticulum-like stellate (*, d; hematoxylin-eosin, original magnification X25 [a], X100 [b], X100 [c], and X100 [d]).*

In contrast to the inflammatory follicular cyst and the radicular cyst, UA associated with an unerupted tooth's crown may share identical clinical and radiographic features with the dentigerous cysts.[1,10,14,18-20] Although UA closely mimics a dentigerous cyst, radiographically the microscopic distinction between UA and the dentigerous cyst has been well established, as in the present case.[14] In accordance with Dunsche et al,[14] the dentigerous cyst may reveal associated odontogenic cell nests in some cases, but does not lead to the detection of formerly missed ameloblastic cells. In the current case, the presence of ameloblastomatous epithelium[21] was observed and the microscopic diagnosis was consistent with UA with mural proliferation.

UA is considered a variant of the solid or multicystic ameloblastoma.[1,5-8] This benign lesion occurs in a younger age group, with slightly more than 50% of cases occurring in patients in the second decade of life.[5,22] The majority of ameloblastomas in children are unicystic, encountered in patients younger than 18 years of age.[10,22] In more than 90% of child cases, UA is located in the mandible with 77% located in the molar ramus region.[22]

The relative infrequency of recurrence of UA suggests that this lesion exhibits a less aggressive biological behavior than those of solid or multicystic ameloblastomas.[10] Furthermore, in children, more cancellous bone exists, allowing the lesion to grow more rapidly with extensive destruction, making surgery more difficult and demanding.[23] Considering the good prognosis of UA, some authors advocate only surgical enucleation.[24-26] Other authors, meanwhile, have recommended that the simple subtype, with and without intralumenal proliferations, may be treated conservatively enucleation. Subtypes showing intramural growths, however, must be treated radically (ie, as a solid or multicystic ameloblastoma).[1,6,7,10,12,19,27] Recently, systematic review of treatment modalities for UA demonstrated that recurrence rates were 4% for resection, 31% for enucleation alone, 16% for enucleation followed by application of Carnoy's solution, and 18% for marsupialization with/without other treatment in a second phase.[28]

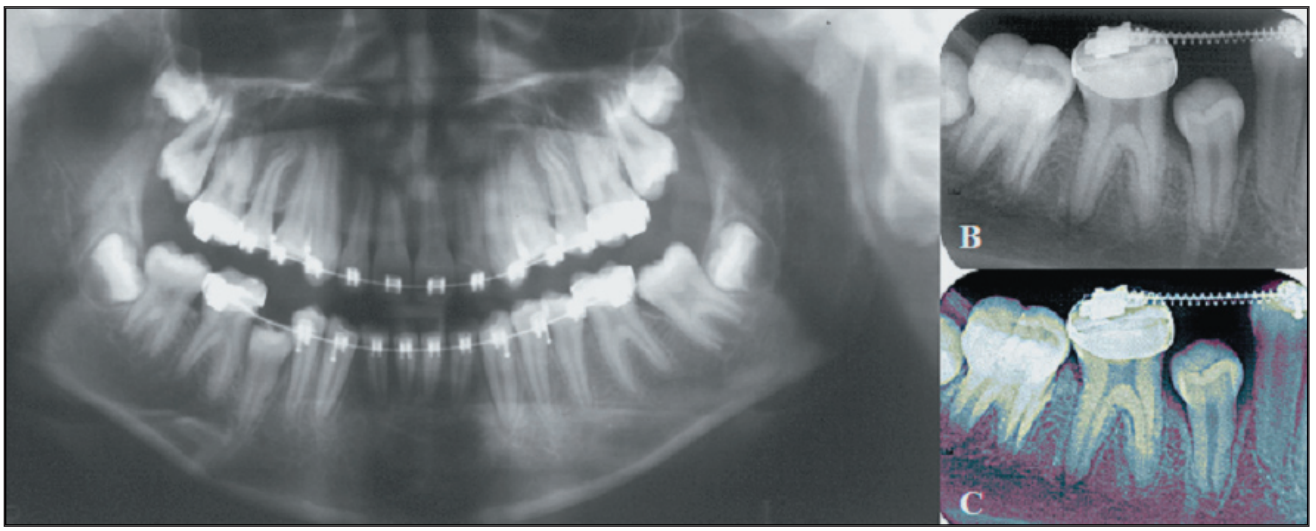

Figure 4. One-year postoperative panoramic (a), periapical conventional (b), and digitized electronically processed (C) radiographs demonstrating good bone formation and substantial eruptive movement of the right inferior second premolar.

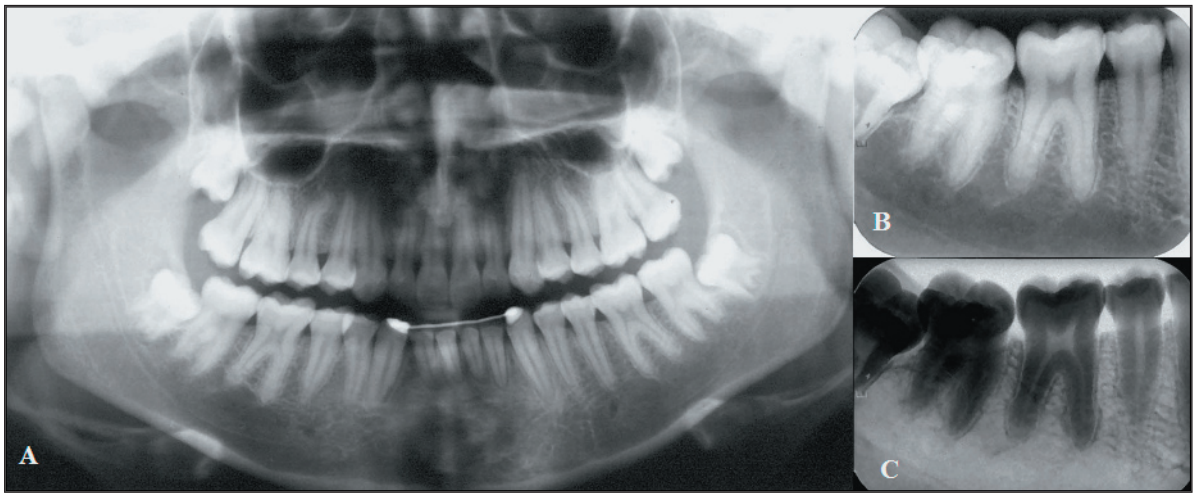

Figure 5. Panoramic (a), periapical conventional (b), and digitized electronically processed (c) radiographs demonstrating the second permanent premolar in correct occlusion and no recurrence of the tumor during the following 4 years.

Ghandhi *et al*, however, studied recurrence percentage of ameloblastomas over 20 years at 2 different centers, Glasgow, Scotland, and San Francisco, Calif, and demonstrated a higher recurrence risk (80%, range=55% to 90%) for unicystic lesions.[29]

A conservative treatment (enucleation) with the maintenance of the permanent tooth associated with the lesion was indicated in the present case. This was based on the clinical and radiographical characteristics and considering the authors' first diagnosis hypothesis of a dentigerous cyst. Therefore, a conservative surgical excision of UA with careful follow-up rather than partial or complete jaw resection appears to constitute appropriate therapy.[1,22,26]

Considering that UA may have a similar appearance to other common jaw lesions such as the dentigerous cyst, the inflammatory follicular cyst, and the radicular cyst[13,14,30] the diagnosis of this cystic neoplasm should be based on detailed clinical, radiographical, and microscopic evaluation. The authors also strived to maintain a careful follow-up of the patient whose unicystic ameloblastoma was treated by conservative surgery.

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