Case Report

Odontogenic Myxoma in the Pediatric Patient: A Literature Review and Case Report

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Abstract: *Odontogenic myxoma is a rare benign tumor of the jaws which, in most cases, grows slowly and asymptomatically. In general, the radiographic features are not pathognomonic of the lesion and the histologic characteristics are similar to the normal follicular and dental papilla tissue. Most reported cases involve noticeable expansile lesions in the jaws of individuals older than 10. The purpose of this report was to present the case of a maxillary odontogenic myxoma diagnosed in an asymptomatic 7-year-old girl on routine dental radiologic examination. The lesion's clinical, radiographic, and histological features and the treatment are discussed and compared to similar cases reported in the literature. (Pediatr Dent 2007;29:409-14) Received August 28, 2006 / Revision Accepted November 3, 2006.*

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Myxomas are benign soft tissue or bone neoplasms that may appear anywhere in the body. In the head and neck region they have been specifically found in the: (1) tongue; (2) nose; (3) cheek; (4) neck muscles; (5) larynx; (6) pharynx; and (7) parotid gland.[1] Most central myxomas occur in the jaws, where they are called odontogenic myxomas (OM) because of their presumably odontogenic origin—although their histologic origin is still controversial.[2] The notion of the dental origin of OM is based on: (1) its almost restricted localization in the jaws; (2)its occasional association with missing or unerupted teeth; (3) the resemblance of the tumor cells with cells from the dental papilla or follicle; and (4) the occasional presence of odontogenic epithelium.[3-5]

Besides the odontogenic ectomensechyme, the tumor cells have been postulated to originate from normal or transformed fibroblasts[6-9] or from other cells thorough messenchymal or myofibroblastic differentiation.[2,4] Furthermore, OM has been linked to a myxomatous change of an odontogenic fibroma or residual foci of embryonic nondental soft tissue.[10,11]

OM is a rare neoplasm with an annual incidence of 0.07 per million.[11] Among oter odontogenic tumors, however, OM is the second most common following ameloblastoma—with a relative frequency of less than 1% to 19%.[11] OM is most often seen in patients between 10 and 40 years of age.[12] Regarding sex bias, authors have reported a male-to-female ratio ranging from 1:1.5 to 1:4,[11-13] whereas others have found both sexes to be equally affected.[3,5,14,15]

OM occurs in both the maxilla and mandible, and its incidence has been found to be higher in the mandible by some but not all authors.[9,12,15-17] Nevertheless, most reports indicate that the jaws' posterior region is the most commonly affected site.[18] Besides the alveolar process, maxillary involvement may include the zygomatic processes. Mandibular involvement, on the other hand, may include the posterior body of the mandible, angle, and ramus.[18] Moreover, the OM is localized on one side of the jaw and rarely crosses the midline.[11] According to Kaffe, in only 5% of the cases is OM associated with an unerupted tooth.[12]

OM is a locally invasive lesion that grows slowly and generally without significant symptoms. For as long as the tumor remains inside the bone, the associated pain, if present, is of mild to moderate magnitude. The involved teeth may become mobile and malpositioned, but they remain viable.[19] More severe pain and other symptoms may appear upon bone perforation and invasion in the maxillary sinus, palate, orbit, and nasal cavity. In such cases, nasal obstruction, diplopia, pain, or paresthesia may develop.[11,20] Interestingly, although OM frequently spreads into the paranasal sinuses, it does not seem to extend in the cranial cavity.[13]

Radiographically, OM most commonly presents as a unilocular or multilocular, well-defined radiolucency. The internal trabecular pattern has been described as "honey-

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comb," "soap-bubble," or "tennis racquet."[21] The latter appearance is characterized by angular or straight trabeculations forming square or triangular compartments[22] and has been considered almost pathognomonic of OM.[21] The lesion usually remains well-defined even if it has perforated the cortex and has expanded into the soft tissue.[3,21] It may appear scalloped between the roots of the teeth or it can include teeth; it is associated with tooth displacement in 26% of the cases and/or root resorption in up to 50% of the cases.[3,11,12,23]

Upon gross examination, OM appears as a white or yellow, gelatinous, lobulated mass.[18] Histologically, it is rarely encapsulated and is composed of spindle-shaped and stellate cells interspersed in the loose mucoid background. Collagen fibers may also be seen scattered in the mucopolysaccharide ground substance, and their amounts determine the tumor's texture and whether it is called myxoma or myxofibroma. Odontogenic epithelium may occasionally be found, but its role as a tumor-inducing agent is controversial and, thus, its presence is not a requirement for the diagnosis of OM.[24]

The generally accepted treatment for OM includes resection of the tumor with a greater than 1.5 cm margin of surrounding tissue. Conservative excision of the lesion can be performed, but it is associated with a significantly higher recurrence rate.[18]

The overall prognosis for OM is generally good. Yet, the recurrence rate varies between 10% to 43%—with an average of about 25%.[15,17,25,26] This relatively high recurrence rate is ascribed to: (1) its local infiltration inside the cancellous bone, far from: the radiographic visible margins; (2) its gelatinous consistency; and (3) a usual lack of encapsulation.[4,21] The recurrence rate appears to correlate with the width of the surgical margin, with a range varying from 10% for hemimandibulectomy or hemimaxillectomy,[17] to 33% for curettage.[15] Although recurrence has been reported up to 15 years after treatment,[1] it usually occurs during the first 2 years. During this period, it is recommended that the patients be followed up very closely.[4,27] Importantly, malignant variants or malignant transformation of these tumors are extremely rare and metastasis has not been reported.[4,15]

Case report

An otherwise healthy 6-year, 11-month-old girl with an unremarkable medical history presented to the pediatric dental clinic of the School of Dentistry, University of Louisville, Louisville, Ky, for routine dental care.

Oral examination revealed a normal complement of teeth for her age, with no evidence of dental caries. The initial radiographic examination consisted of panoramic and bitewing radiographs. A bifurcation radiolucency and root resorption were observed on the bitewing radiograph in the area of the primary maxillary left second molar, but no pain, buccal, or palatal expansion was associated with the observed lesion.

The panoramic radiograph (Figure 1) revealed a poorly defined radiolucency in the furcation of the primary maxillary left second molar. The buccal roots were resorbed compared to the contralateral side. Furthermore, the successor second premolar was noticeably displaced superiorly.

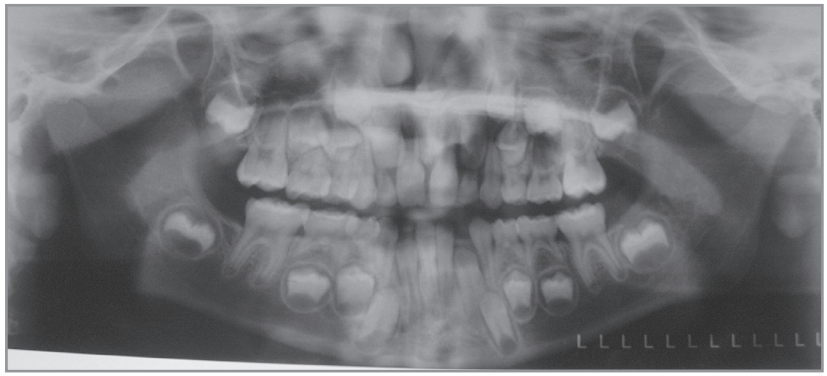

Figure 1. Preoperative panoramic radiograph showing radiolucency involving the trifurcation of and buccal root resorption in the primary maxillary left second molar. Superior displacement of the unerupted maxillary left second premolar can be also seen.

The provisional diagnosis included a cystic lesion or odontogenic tumor. Prior to definitive treatment, it was decided to extract the involved primary molar and perform an incisional biopsy. Microscopic examination of the excised mucoid soft tissue provided a histopathologic diagnosis of an odontogenic myxoma. Specifically, sections of the myxomatous tissue's lobular masses showed stellate-shaped fibroblastic cells interspersed in a matrix blue ground substance. Hyalinized connective tissue inclusions and mineralized bone trabaculae were seen in some areas, but no epithelial lining or odontogenic epithelium could be detected. Based on this diagnosis, cross-sectional imaging was recommended to better define the lesion's anatomical extent so that the preoperative surgical plan would be as accurate as possible.

Upon cone beam computed tomography (**CBCT**) imaging (iCAT, Imaging Sciences International, Harfield, Pa; Figures 2 and 3), the lesion presented as a single unilocular radiolucency with well-defined scalloped borders located superior to the primary maxillary left second molar's extraction socket. Orthogonal imaging (Figure 2) showed involvement of the pericoronal space of the second premolar and superior/palatal displacement of this tooth. Adjacent to the second premolar, the lesion had caused buccal expansion, thinning, and convexity of the inferior floor of the maxillary sinus in

this region. There was no evidence of perforation of the lesion into the maxillary sinus. Paracoronal reformation (Figure 3) demonstrated palatal cortical expansion, extension, and disruption of the crypt to the level of the palatal alveolar junction. The buccal involvement extended from the maxillary left first premolar's anterior surface to the first molar's mesial surface. The lesion extended palatally from the distal of the primary first molar to the palatal cusp of the permanent first molar, and internally there was evidence of spicules of trabeculae.

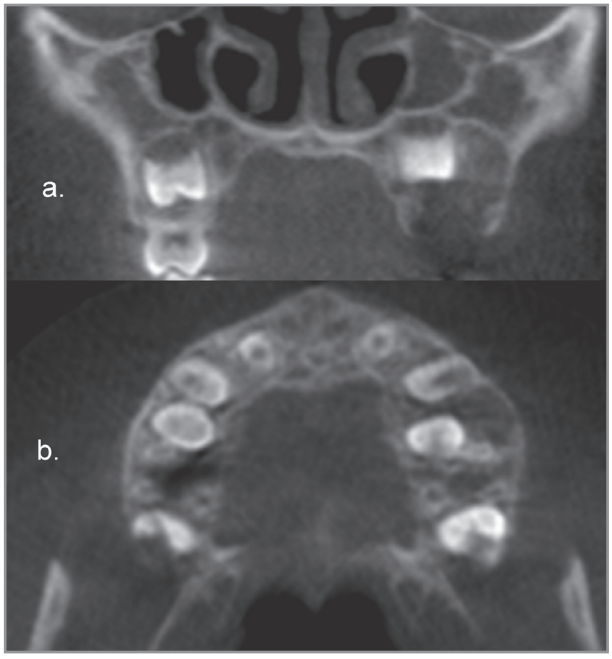

Figure 2. Coronal (a) and axial (b) 0.4-mm thick orthogonal slices of left maxilla region of interest located at the epicenter of the lesion (courtesy of Drs. Allan G. Farman/William C. Scarfe, Department of Surgical/Hospital Dentistry, School of Dentistry, University of Louisville).

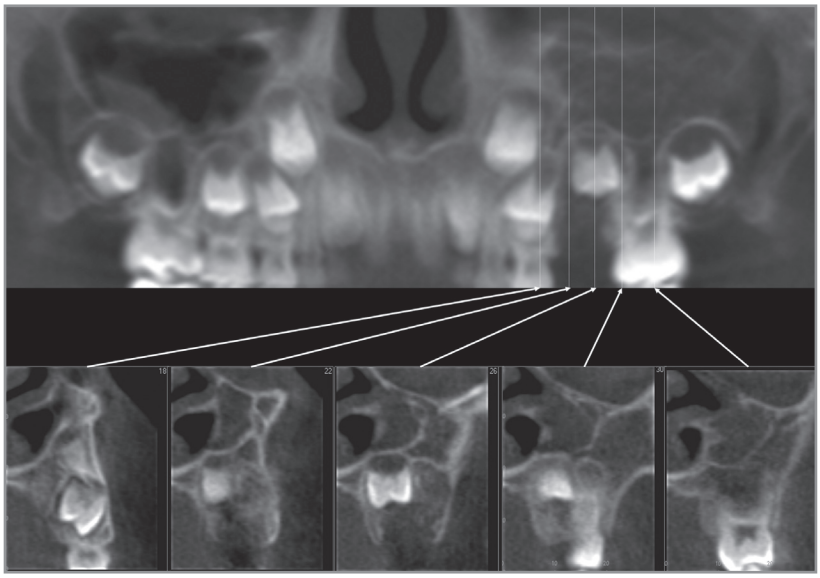

Figure 3. Reformatted cone beam CT panoramic reference image and selected 2-mm thick/4-mm interval paracoronal (cross-sectional) images of left maxilla region of interest (courtesy of Drs. Allan G. Farman/William C. Scarfe, Department of Surgical/Hospital Dentistry, School of Dentistry, University of Louisville).

The mass was surgically excised in the operating room (Figure 4). The buccal cortex surrounding the lesion was removed, and the tumor was enucleated without difficulty. The permanent first molar and the 2 premolars were extracted because they were found to be inside the tumor bed (Figure 5). The primary maxillary left first molar was also removed because of its proximity to the tumor bed. Although the tumor was surrounded by intact dense cortical bone, segmental maxillary resection was performed.

Subsequent histopathologic examination of the surgical specimen confirmed the previous diagnosis of OM (Figure 6). Nine months following the lesion's excision, the patient has continued to demonstrate no recurrence of the lesion on radiographic and clinical examination.

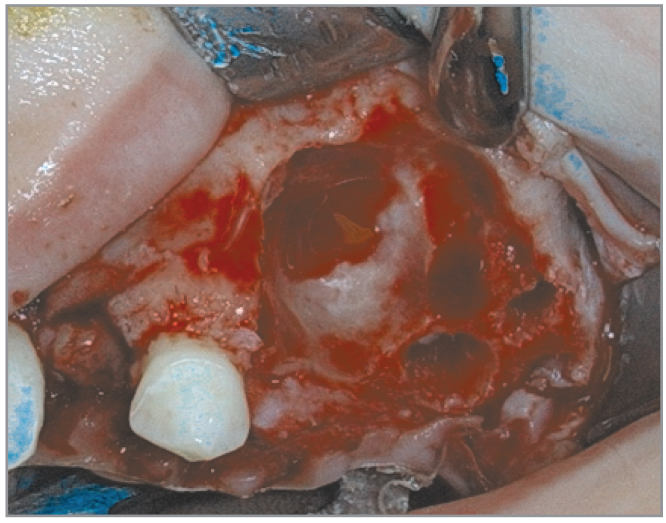

Figure 4. Removal of the tumor, involved teeth, and surrounding bone in the operating room.

Discussion

In 60% to 75% of cases, OM is diagnosed in the second or third decade of life.[12,18] Our patient was almost 7 years old when the diagnosis was made. Although OM cases in 3- to 19-month-old patients have been reported in the literature,[11,20] it is generally believed to be rather uncommon in childhood—with only about 7% of these lesions occurring in children younger than 10 years old.[28] Interestingly, Keszler observed a higher frequency of this neoplasm than other aggressive tumors in children. He concluded

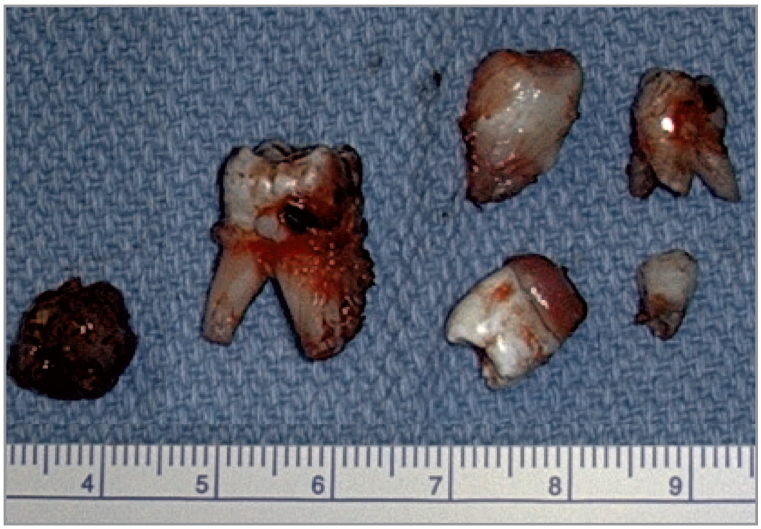

Figure 5. Excised lesion and associated teeth.

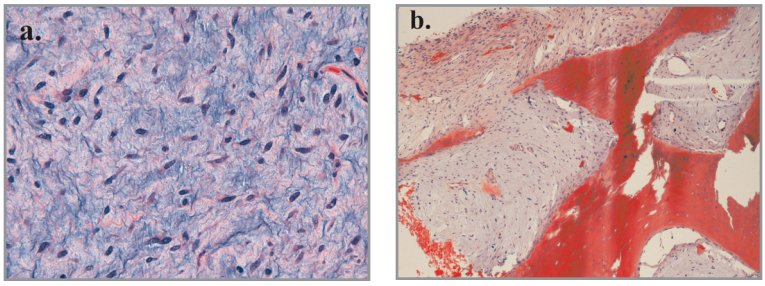

Figure 6. Tumor section showing spindle cells in the myxoid background (a, Hematoxylin Eosin x 100). Lower power magnification of a section from the tumor showing infiltration of the surrounding bone (b, Hematoxylin Eosin x 400) (courtesy of Dr. Mark Bernstein, Department of Surgical/Hospital Dentistry, School of Dentistry, University of Louisville).

that OM should be considered in the differential diagnosis of radiolucent lesions in children and adolescents.[28] In fact, because of its slow and asymptomatic growth, it is reasonable to consider that OM originates early in life, but is not discovered until later when signs or symptoms present.

It is impossible to estimate when the tumor of this patient started developing or how long it would have taken before the first signs or symptoms appeared. On average, there is a delay between 1 to 5 years from the lesion's onset to the first sign, which is usually a slowly growing facial or intraoral swelling, that causes the patient to seek medical help.[20] Exceptions of rapidly growing tumors are extremely rare and seem to occur in very young patients.[11] Notably, OM in the mandible is detected earlier than in the maxilla, where initial spreading of the tumor in the paranasal sinuses does not cause an intraoral noticeable expansion.[13,20]

Because of its asymptomatic growth, the discovery of the tumor can be an incidental finding on routine dental radiographic examination.[18] The radiographic findings that prompted the authors to further evaluate this patient were the:

1. root resorption of the primary maxillary left second molar compared to its antimere;
2. furcational radiolucency of the primary maxillary left second molar in the absence of an obvious etiology; and
3. displacement of the developing maxillary second premolar.

These tooth-related radiographic findings are reported in OM cases, but they are not pathognomonic features of this neoplasm. We did not observe the highly pathognomonic radiographic feature of OM, the so called "tennis racquet" appearance. Actually, this radiographic appearance is observed in only one third of the cases. The remainder show quite diverse manifestations ranging from unilocular, as in this case study, to multilocular with soap-bubble appearance to multiple radiolucent areas separated with curved or straight bony septa.[12-14,22,26] This diversity makes interpretation difficult and emphasizes that OM should be included in the radiographic differential diagnosis along with other lesions, such as: (1) dentigerous cyst; (2) odontogenic keratocyst; (3) ameloblastoma; (4) central giant cell granuloma; (5) central hemangioma; (6) traumatic bone cyst; (7) aneurysmal bone cyst; or (8) fibrous dysplasia.[1,3]

The ill-defined radiolucency in the area of the primary maxillary left second molar on the panoramic radiograph of this patient was more clearly visualized on (CBCT) examination. This underscores the limitations of conventional radiography in the assessment of lesions requiring broad surgical excision. For example, what appears to be a multilocular lesion on a panoramic film can be intralesional trabeculations projected on a 2-dimensional film.[22] Indeed, in this case study, CBCT helped establish the lesion's effects and degree of involvement within the alveolus and maxilla. Furthermore, the lesion's borders which have been found to be poorly defined or diffuse in 34% of the cases[12] are better delineated on the CBCT scan. Diffuse borders are seen more often with the maxillary than the mandibular lesions on the conventional radiograph, presumably because there are many bony structures that are superimposed in the maxilla.[22] Defining the tumor borders is essential for planning the extent of the resection, which seems to be associated with a high recurrence rate. The tumor's actual borders, however, are usually well beyond even the CBCT-scan borders and they are impossible to determine. This is due to the infiltration of the tumor cells within the normal bony trabeculations or into the soft tissue.[22]

Histologically, the structure and cellularity of the OM of our patient seemed comparable to dental follicular or dental papilla tissues.[29,30] The distinction of OM from dental follicles is based on its destructive nature and its being larger than the 3-mm typical follicular radiolucency.[30] On the other hand, histological distinction of dental papillae from OM is based on: (1) the presence of odontoblasts and eosinophilic dentinoid tissue around their well circumscribed elliptical myxoid tissue;[30] and (2) their small diameter, 1.5 cm or less.

Other lesions that can be included in the histopathological differential diagnosis of OM are the: (1) myxoid degenerated benign or malignant nerve sheath tumor; and (2) myxoid chondrosarcoma.[4] In this case study, as in most cases, there was little collagen interspersed within the ground substance. There is no evidence, however, that the collagenized variants (ie, fibromyxomas) behave differently.[1]

Treatment for our patient followed the current standard protocol, which consists of: (1) surgical resection of all clinically obvious tumor tissue; and (2) a healthy tissue margin or single tissue plane.[10] Curettage or chemical/electrical cauterization may be also added to reduce the recurrence.[18] The width of the excised clear margins or planes is a subject of controversy. Some surgeons support a conservative excision of narrow margins or planes, whereas others recommend radical resection to reduce the risk of recurrence.[18] Localization of the tumor, particularly for the pediatric population, may be an important factor in determining the excision's extent. Indeed, there is some indication that pediatric maxillary OM can be treated efficiently with conservative surgical treatment,[10] although this view awaits further support. Radiotherapy generally is not standard treatment for OM because:

1. the tumors are benign and easily excised; and
2. in young patients, radiation may induce long-term complications, including:
   a. cognitive disfunction;
   b. second malignant neoplasms; and
   c. dental anomalies, such as:
      i. tooth and root agenesis;
      ii. root shortening; and
      iii. localized enamel defects.[31,32]

Some surgeons support preoperative radiation, however, to shrink very large myxomas and/or more adequately define the surgical margins.[4,33]

In terms of reconstruction, because of the high recurrence rate, permanent bone and soft tissue rehabilitation and implants should be delayed 3 to 5 years after surgery or until there is confidence that the patient is safe from recurrence. Until then, prosthetic reconstruction by means of maxillary obturators may be necessary.[18] For our patient, we intend to provide eventual bone grafting with space maintenance for future endosseous implant reconstruction.

In conclusion, the maxillary odontogenic myxoma of this study's patient is a rare case of this benign tumor because it was diagnosed before it became symptomatic. Although the histolopathologic evaluation was crucial for the diagnosis, its nonpathognomonic radiographic appearance was the first indication of this lesion. Based on current knowledge, we treated this case with aggressive excision, beyond the visibly affected bony borders, and a close follow-up will be maintained for at least the first 2 years. Odontogenic myxoma should be included in the differential diagnosis of radiolucent as well as mixed lesions seen in the alveolar process area in the pediatric population.

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