Clinical–Pathological Conference: Case 4

Kristin K. McNamara · Mikelle L. Kernig ·
Kelly S. Kreuter · John R. Kalmar ·
Carl M. Allen

Received: 16 June 2010 / Accepted: 10 July 2010 / Published online: 30 July 2010
© Humana 2010

Clinical Presentation

A 9-year-old male with a non-contributory medical history presented with left mandibular swelling. His mother had first noticed swelling in the area 2 months earlier, but stated that it seemed to resolve, only to develop again over the last 2 weeks. The boy had no complaints of pain, tenderness or altered sensation of his lower lip.

Extraoral examination revealed mild facial asymmetry with enlargement of the body region of the left posterior mandible. The overlying skin lacked erythema or warmth and no anesthesia or paresthesia of the left lower lip was detected. Intraoral examination showed marked buccolingual expansion of the mandible centered on tooth #19 with vertical enlargement to the height of the occlusal plane; tooth #19 appeared “engulfed.” The mucosa was intact and of normal color (Fig. 1).

The panoramic radiograph showed a normal pattern of tooth development and eruption. A solitary 3.5 cm mixed radiolucent/radiopaque lesion was noted surrounding the roots of tooth #19 (Fig. 2a, b). The lesion was composed of a central opacity exhibiting a fine, radiating trabecular pattern and a relatively well-defined radiolucent periphery. Expansion of the inferior border of the mandible as well as posterior displacement of teeth #18 and #19 was observed, without evidence of root resorption. Vertical extension of lesional tissue above the height of the surrounding alveolar

bone was seen. A computed tomography (CT) image revealed prominent expansion of the left mandible with discontinuity of the buccal and lingual cortices but no evidence of soft tissue involvement (Fig. 3).

Differential Diagnosis

Based on the clinical history and radiographic findings, a diagnosis of osteoblastoma was favored. The differential diagnosis included osteosarcoma, juvenile active ossifying fibroma, and calcifying epithelial odontogenic tumor.

Osteoblastoma is a rare primary bone tumor that typically occurs in patients under the age of 30, with a slight female predominance and a predilection for the mandible, particularly the posterior segments [1, 2]. Pain and swelling with bone expansion are common clinical features [1, 3]. While no discomfort was reported in the present case, recent literature indicates that significantly fewer patients experience pain associated with this neoplasm than was previously suggested [1]. The radiographic features of osteoblastoma can vary from a well-demarcated to ill-defined osteolytic lesion with variable internal structure that can range from entirely radiolucent to our finding of a central radiopacity with radiolucent periphery [1–3].

Osteosarcoma is a rare primary malignancy of bone that most often affects the long bones around the knee in teenagers or young adults. While patients with jaw lesions are usually older, all age groups have been affected [4]. A painful swelling is the most common clinical presentation, with paresthesia and loosening of teeth also frequent findings [5, 6]. Radiographically, the radiating pattern of trabeculation noted within the current case supported consideration of osteosarcoma, although only 25% of gnathic osteosarcomas demonstrate a radiating or “sunburst”

K. K. McNamara (✉) · K. S. Kreuter · J. R. Kalmar ·
C. M. Allen
The Ohio State University, 305 West 12th Ave., Columbus,
OH 43210, USA
e-mail: mcnamara.189@osu.edu

M. L. Kernig
USAF, Wright Patterson AFB, Dayton, OH, USA

Fig. 1 Intraoral appearance of *left* mandible exhibiting marked expansion centered on tooth #19

Fig. 2 **a** Panoramic radiograph demonstrating a solitary lesion of the *left* mandible. **b** Closer view showing a mixed radiolucent/radiopaque expansile lesion surrounding the roots of tooth #19

pattern of osteoid production [4]. Formation of calcified material above the alveolar crest was also considered suspicious for this entity. On the other hand, another important radiographic hallmark of gnathic osteosarcoma, widening of the periodontal ligament space [5, 6], was absent. In contrast, in our patient the periodontal ligament space of tooth #19 appeared obliterated by the process.

Fig. 3 Axial CT image shows mixed density lesion exhibiting prominent expansion of the *left* mandible with discontinuity of the buccal and lingual cortices

Juvenile active ossifying fibroma (JAOF) is a fibro-osseous tumor that, as the name suggests, typically occurs in young individuals and may exhibit rapid growth, similar to the current case. JAOF, however, more commonly arises in the maxilla, sinonasal region, or orbit compared to the mandible [7]. Radiographically consistent with the present case, JAOF are typically radiolucent and may contain central radiopacities. They tend to be well-demarcated and lack continuity with the adjacent normal bone [7]. Finally, calcifying epithelial odontogenic tumor (CEOT) was also a diagnostic consideration, however this lesion is frequently associated with an impacted tooth and typically occurs in patients between 30 and 50 years of age [8]. The radiographic features of CEOT are variable, consisting of a radiolucency with well-defined to poorly-defined borders, and calcification ranging from prominent, dense, and centrally located to more diffuse, fine, and radiating [8] as noted in the current case.

Diagnosis and Discussion

The patient was referred for incisional biopsy with extraction of tooth #19. A gingival flap was created by performing a sulcular incision from the primary canine to the distal of #19 with a vertical release anteriorly. Intra-operative evaluation confirmed perforation of buccal and lingual cortices and revealed an intimate association between the lesion and tooth #19, extending from the cemento-enamel junction apically. The tumor moved with luxation of the tooth and extraction of the tooth/tumor complex was performed (Fig. 4).

Histopathologic examination of H&E decalcified sections revealed a benign neoplasm consisting of sheets and

Fig. 4 Gross specimen demonstrating intimate association between the tumor and tooth

thick interlacing trabeculae of eosinophilic calcified material that resembled cellular cementum. The trabeculae were lined by plump angular, ovoid, and spindle-shaped cells supported by a fibrovascular stroma. Occasional benign multinucleated giant cells were seen, although there was no evidence of significant mitotic activity or necrosis. The tumor was well circumscribed and focally encapsulated. At its periphery, the lesional trabeculae were arranged in radiating columns (Fig. 5). The calcified tumor mass appeared fused to the tooth root, with microscopic evidence of root resorption (Fig. 6). These histopathologic features were consistent with a diagnosis of cementoblastoma.

Cementoblastoma is considered by many to be the only true odontogenic neoplasm of cementum. It is a rare neoplasm, accounting for 0.8–2.6% of all odontogenic tumors, and is most commonly seen in children and young adults, with no obvious sex predilection recognized [9]. Cementoblastomas typically arise in the posterior mandible and

Fig. 5 Tumor trabeculae arranged in radiating columns with a well circumscribed and focally encapsulated margin ($\times 10$)

Fig. 6 Cellular calcified mass fused to tooth root with evidence of root resorption ($\times 40$)

characteristically appear radiographically as a circumscribed sclerotic mass which is confluent with one or more tooth roots, resulting in loss of root outline, root resorption, and obliteration of the periodontal ligament space, and is surrounded by a radiolucent rim [10].

The tumor in the present case was associated with the roots of a fully developed mandibular first molar, however, cementoblastoma was not considered in the differential diagnosis due to the uncharacteristic radiographic features. The lesion demonstrated a central opacity, exhibiting a fine, wispy quality as opposed to a dense sclerotic mass. Interestingly, variable density has been reported previously, with rare cases of purely radiolucent lesions [10]. In addition, although the periodontal ligament space of tooth #19 was obliterated, the tumor did not appear to be confluent with the tooth roots. Radiographic evidence of root resorption, a characteristic feature, was also absent, although microscopic evidence of resorption was subsequently identified. Most authorities consider cementoblastoma to be an innocuous neoplasm; however, the current case, along with many in the literature, exhibits locally aggressive features such as dramatic bone expansion, cortical perforation, and displacement of teeth [10]. Vertical tumor growth above the alveolar crest, resulting in the clinical appearance of an “engulfed” tooth, was peculiar in the present case. While not typical, it was considered to be another aggressive feature of this tumor.

Cementoblastoma is histopathologically similar to osteoblastoma, but distinct due to fusion with a tooth root. Additionally, in contrast to the basophilic trabeculae often seen in osteoblastoma, cementoblastoma is characterized by sheets of eosinophilic mineralized tissue resembling cellular cementum containing plump cells within irregular lacunae [3, 9, 10]. The current diagnosis was supported by the identification of these features, in addition to focal

Fig. 7 Panoramic radiograph shows evidence of bone fill 4 months post-operatively

encapsulation and the presence of the classic histopathologic pattern of peripheral radiating trabeculae [9, 10].

Surgical extraction of the tooth together with the attached calcified tumor is standard care and was performed in this case. While historically this neoplasm has been understood to have a very low rate of recurrence, a recently reported series from the files of the Armed Forces Institute of Pathology suggests that recurrences may be greater than previously thought, with an overall recurrence rate as high as 22% [10]. Whether this increased recurrence rate reflects referral bias to the AFIP, which normally sees more difficult cases, remains to be determined. Conservative therapy, such as surgical excision of the mass with root amputation and endodontic therapy should be considered with caution, as inadequate removal is most closely related to tumor recurrence [10]. Bone expansion and cortical perforation, as seen in the present case, are also factors associated with increased risk for future recurrence [10], and necessitate long-term patient follow-up. At 4 months post-operatively, the current patient was asymptomatic with complete resolution of his facial asymmetry. Normalization of the alveolar ridge at the surgical site was noted clinically, along with radiographic evidence of bone fill (Fig. 7).

Cementoblastoma is a rare tumor that may be easily omitted as a diagnostic consideration in the absence of classic radiographic features. The present case represents an unusual presentation for this neoplasm and highlights the potential for variable clinical and radiographic findings.

References

1. Jones AC, Prihoda TJ, Kacher JE, et al. Osteoblastoma of the maxilla and mandible: a report of 24 cases, review of the literature, and discussion of its relationship to osteoid osteoma of the jaws. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 2006;102(5):639–50.
2. Capodiferro S, Maiorano E, Giardina C, et al. Osteoblastoma of the mandible: clinicopathologic study of four cases and literature review. *Head Neck.* 2005;27(7):616–21.
3. Rawal Y, Angiero F, Allen CM, et al. Gnathic osteoblastoma: clinicopathologic review of seven cases with long-term follow-up. *Oral Oncol.* 2006;42(2):123–30.
4. Garrington GE, Scofield HH, Cornyn J, et al. Osteosarcoma of the jaws: analysis of 56 cases. *Cancer.* 1967;20(3):377–91.
5. Givol N, Buchner A, Taicher S, et al. Radiological features of osteogenic sarcoma of the jaws. A comparative study of different radiologic modalities. *Dentomaxillofac Radiol.* 1998;27(6):313–20.
6. Nakayama E, Sugiura K, Ishibashi H, et al. The clinical and diagnostic imaging findings of osteosarcoma of the jaw. *Dentomaxillofac Radiol.* 2005;34(3):182–8.
7. Barnes EL, Eveson JW, Reichart P, Sidransky D, eds. Pathology and genetics of head and neck tumours. In: Kleihues P, Sobin LH, editors. World Health Organization classification of tumours. Lyon: IARC Press; 2005.
8. Kaplan I, Buchner A, Calderon S, et al. Radiological and clinical features of calcifying epithelial odontogenic tumour. *Dentomaxillofac Radiol.* 2001;30(1):22–8.
9. Ohki K, Kumamoto H, Nitta Y, et al. Benign cementoblastoma involving multiple maxillary teeth: report of a case with a review of the literature. *Oral Surg Oral Med Oral Pathol.* 2004;97(1):53–8.
10. Brannon RB, Fowler CB, Carpenter WM, et al. Cementoblastoma: an innocuous neoplasm? A clinicopathologic study of 44 cases and review of the literature with special emphasis on recurrence. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 2002;93(3):311–20.