Case Report: a Pediatric Case of Cementoblastoma with Histologic and Radiographic Features of an Osteoblastoma and Osteosarcoma

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Abstract One case of an expansile mass involving the roots of a mandibular left first molar in a nine year old male is presented. The lesion has features of a cementoblastoma (CB), osteoblastoma (OB), and osteosarcoma (OS) both radiographically and histologically. Radiographically, a “sunburst” appearance is present. Histologically, the lesional tissue is intimately involved with the root, not the periosteum. The clinical, radiographic, and histopathologic characteristics of CB, OB, and OS are discussed as well as the difficulties associated with distinguishing between these lesions in the gnathic region.

Keywords Cementoblastoma · Osteoblastoma · Osteosarcoma · Pediatric

Report of a Case

The patient, a 9-year-old male, presented to the University of Pittsburgh School of Dental Medicine with pain and swelling of several months duration. Significant buccal and submandibular left sided facial swelling was noted (Fig. 1). The child was initially seen by a dentist who noted a carious tooth in the region and presumed that the swelling was the result of infection based on periapical radiographs.

When the swelling persisted, a panoramic tomogram was obtained and the family sought consultation with an oral and maxillofacial surgeon.

On a panoramic radiograph, the lesion appeared to be a well-defined radiopacity with a thin peripheral radiolucent band or “halo” around the roots of the left mandibular first molar (Fig. 2). A radiopaque “wheel and spoke” or “sunburst” pattern was seen emanating from the root surface. No root resorption was noted. The radiopacity extended coronally slightly beyond the cemento-enamel junction. Bone in the area of the lesion appeared to be sclerotic. The mandibular canal was displaced inferiorly. Computed tomography illustrated the extent of the tumor, showing thinning of the cortical plate secondary to buccal-lingual expansion. Distinct separation was seen between the tumor and the adjacent bone on both the panoramic radiograph and on computed tomography. The surrounding soft tissues showed edema and reactive changes. A number of radiographic interpretations were considered as differential diagnoses with osteosarcoma and cementoblastoma being strongly favored.

An intraoral incisional biopsy of the left mandibular lesion was performed and was reviewed by a number of pathologists from a variety of subspecialties and institutions. No consensus was reached on the histological diagnosis. On gross examination, the biopsy consisted of dense, irregular, red-tan bony fragments. Hematoxylin and eosin staining revealed dense sclerotic bone with osteoblastic rimming of the bony trabeculae. Fibroblastic proliferation was noted with nuclear pleomorphism, rare mitotic figures, and focal areas of necrosis. The final interpretation of the initial incisional biopsy was low grade osteosarcoma (OS).

Two large swellings, considered to be lymphadenopathy, were found on clinical examination in the area of the mass. A fine needle aspiration was performed on the left

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Fig. 1 Frontal view of patient upon initial presentation with marked left facial swelling

Fig. 2 Panorgraphic X-ray revealed a radiopaque lesion encompassing the roots of the mandibular first molar with a spoke and wheel or stellate appearance (*arrow*)

neck on the two areas of palpable nodularity; two Diff-Quik and Papanicoulau stains were prepared from the smears of each site. These specimens were benign salivary tissue with a focal lymphocytic infiltrate and a benign reactive lymph node. Therefore, no metastatic disease was evident.

Given the concern for malignancy, the tumor was formally resected with margins to avoid undertreating the pathology. Since no metastatic disease was noted and the tumor was judged to be low grade, no adjuvant therapy was planned. A comprehensive surveillance program was planned after the initial resection.

Fig. 3 Medial view of the resection specimen. The tumor is well circumscribed with displacement of the inferior alveolar nerve and cortical expansion without perforation

Fig. 4 Lateral view of the resection specimen. The thickened tumoral edges can be appreciated with a space separating it from the surrounding bone

An en bloc resection was performed extending from the central incisor to the mandibular condyle incorporating the overlying mucosa. A selective neck dissection was not utilized as it is generally not felt to be helpful in staging or treating osteosarcoma and the palpable neck swellings were found to be benign. A microvascular fibula graft was placed in the left mandibular region immediately after the resection. Grossly, the tumor measured $4.0 \times 3.5 \times 3.5$ cm (Figs. 3, 4). It appeared red-tan, well-circumscribed, and the crown of the mandibular left first molar protruded from the specimen. On sectioning, areas of cavitation with bony walls having a spiculated appearance and containing a gelatinous material were appreciated (Fig. 5). Histological examination showed many of the same features as in the preceding biopsy. Within the dense, sclerotic osteoid, rare atypical mitotic figures were seen within the osteoblasts (Fig. 6). No tumoral infiltration of the surrounding bone was seen, and the osteoblasts were more densely aggregated in the peripheral aspect of the

Fig. 5 On sectioning, the relationship between the coronal aspect of the tooth is seen, as the crown is partially embedded in the tumor

Fig. 6 Hematoxylin and eosin $\times 10$. At low power dense bony trabeculae are seen with a well-defined tumoral border

Fig. 7 Hematoxylin and eosin $\times 40$. Rare mitoses (arrow) were seen within the bony trabeculae

lesion in a radiating pattern (Fig. 7). An intimate association was noticed at the interface of the tooth apices and the tumor (Fig. 8). Intrapulpal calcification was seen, a process

Fig. 8 Hematoxylin and eosin $\times 20$. Interface of tooth apex and the tumor

that is abnormal at this patient's age and indicative of a reactive process. Ki-67 immunostaining illustrated a low level of proliferative activity. All excised lymph nodes were negative for metastatic disease. The tumor was diagnosed as an osteoblastoma-like low grade OS, with the comment that the tumor had features of a cementoblastoma (CB). Prior to this diagnosis, the histologic specimen was reviewed by multiple pathologists, including Pediatric, Head and Neck, and Oral pathologists and the impressions ranged from atypical CB, to OB, to OS.

Postoperatively, the patient showed no signs of recurrence at 2 years follow-up. Despite the possibility that this lesion may have represented a CB, this lesion was managed as if an OS, although, no adjuvant therapy was administered. Aggressive surgical intervention is accepted as the mainstay of treatment for patients with localized disease [1]. Many dental and medical sub-specialties including pathology, radiology, surgery, and oncology were involved in the diagnosis, treatment, and rehabilitation of this patient.

Discussion

Cementoblastomas are uncommon odontogenic tumors derived from neoplastic cementoblasts. They have a predilection for the mandible, most often in association with the mandibular first molar. They are more commonly seen in males with a mean age of 21 years [2]. They can present with pain, swelling, cortical expansion of the mandible, tooth mobility, paresthesia, or be entirely asymptomatic. Radiographically, they are well circumscribed mixed or

radiopaque lesions with a lucent border and appear contiguous with the root of a tooth [3]. Histologically, the fusion of the tumor with the apices of the tooth, referred to as replacement resorption, is a distinguishing feature, although expansion of OB may result in this fusion. Treatment for CBs range from en bloc resection to root amputation with tumor removal to curettage without tooth extraction or root amputation. The CB is regarded as a benign lesion. However, a recurrence rate as high as 37% has been described [2].

Osteoblastoma are primary bone tumors of which approximately 11% are seen in the jaws [4]. No clear gender predilection is seen with a peak in the second to third decade of life [4, 5]. In the jaws, they often affect the mandible and present with painful swelling [6]. Radiographically, they begin as a well-circumscribed radiolucency progressing to a mixed lesion; at times they are associated with expansion of the cortical bone [7]. Histologically, the appearance can be variable with bony trabeculae in a loose fibrovascular stroma, plasmacytoid to large epithelioid osteoblasts with vesicular nuclei containing eosinophilic cytoplasm, prominent osteoid production with osteoblastic rimming, and minimal mitotic activity [4, 6]. At times, atypical cells that are large, multinucleate, with unusual degenerative nuclear features, and prominent nucleoli can be seen [8, 9]; thus, differentiating this lesion from OS can be difficult. In fact, one histological variant, with multiple nidi of epithelioid osteoblasts, a pattern difficult to discern from OS, has a predilection for the jaws [10]. Treatment for atypical OB is surgical resection rather than simple curettage.

Primary OS of the head and neck are seen in only 6–10% of cases [11]. Generally, there is no sex predilection, but they present later in life than OS of the long bones, with a peak in the third to fourth decade of life [12, 13]. Craniofacial OS is seen within the mandible in 30–40% of cases [14, 15]. Gnathic OS often presents with pain, swelling, or sensory deficit [13]. Radiographically, the tumor often has a sunburst periosteal pattern, and

causes “spiking” resorption of the involved tooth roots. The stromal cells often exhibit marked pleomorphism, atypical mitotic activity, increased nuclear-to-cytoplasmic ratio, and dense osteoid with osteoblastic rimming.

The three traditional subtypes of OS are osteoblastic, chondroblastic, and fibroblastic and are identified by a composition of greater than 50% of the tissue defining the histologic type [16]. Osteoblastic OS are characterized by a fine unmineralized or osteoid matrix between tumor cells. Chondroblastic OS contains a malignant cartilaginous component. In fibroblastic OS a spindle cell pattern with minimal osteoid matrix is present. While the histology is often mixed, in the head and neck, the chondroblastic subtype is most common [1].

It has been proposed that CB and OB are the same entities only differentiated by anatomic location; CBs are found only in the tooth bearing areas of the jaws. Differentiating OB from OS can be extremely challenging given that the former can display locally aggressive behavior that is not histopathologically predictable. Aggressive OB may have more epithelioid osteoblasts, increased mitotic activity, and more contiguous areas of osteoid; the question has been raised whether aggressive OB is a distinct clinicopathologic entity [9]. Features that distinguish OS from OB include poor circumscription with infiltration of normal bone by the neoplastic cells, nuclear pleomorphism, and increased mitotic activity with abnormal figures (Table 1).

This tumor, best described as an atypical cementoblastoma, represents the epitome of a diagnostic dilemma that resulted in aggressive treatment of a lesion that in certain circumstances may have been effectively treated with a more conservative approach. Unfortunately, certain histologic features more concerning for malignancy usually dictate a more aggressive approach, and these features were present in this child’s case. Nuclear pleomorphism and rare atypical mitotic figures were seen. The radiographic features were most consistent with those of a cementoblastoma. The computed tomography of this tumor lacked the sharp, severely tapered, root resorption often seen with OS.

Table 1 Comparison chart

Features	Cementoblastoma	Osteoblastoma	Gnathic osteosarcoma
Age	Peak incidence in the 1st and 2nd decades of life	Peak incidence in the 2nd and 3rd decades of life	Peak incidence in the 3rd and 4th decades of life
Sex predilection	Male predilection, 2:1	No clear gender predilection	
Presentation	Commonly presents with painful swelling		
Radiographic	Continuity between tooth and tumor with effacement of the periodontal ligament space	No radiolucent halo around the lesion	Spiking root resorption, starburst or sunray periosteal appearance
Histologic	Radiating osteoid (cementoid) pattern with fusion of tumor to cementum	Low rate of mitotic activity, pushing growth pattern, plump osteoblasts	Mitoses more common with atypia, radiating pattern, no fusion with tooth seen, poorly defined borders

However, the radiopacity extends beyond the cemento-enamel junction—a feature not often seen in CBs. However, this child's tumor is intimately associated with the tooth apices—a feature classically seen in CBs. Making a definitive diagnosis of this type of lesion was difficult given current classification systems and nomenclature. Differentiating between atypical CB, aggressive OB, and OS even with radiographic correlation was not straightforward. This case illustrated some of the difficulties that can be seen in gnathic osseous lesions of tooth bearing regions.

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