PAROSTEAL OSTEOSARCOMA: CASE REPORT AND REVIEW OF THE LITERATURE

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Abstract: Background. The majority of osteosarcoma cases of the head and neck are high-grade lesions. We present a case and discuss the diagnostic and therapeutic implications of a rare low-grade parosteal osteosarcoma of the maxilla.

Methods. A 32-year-old man presenting to the Head and Neck Surgical Oncology clinic with a 1-year history of a firm palatal mass. Evaluation clinically and radiographically raised the suspicion of an osteosarcoma.

Results. A partial maxillectomy revealed a parosteal osteosarcoma with negative margins. No adjuvant therapy was recommended, and the patient remains without evidence of local recurrence after 3 years.

Conclusions. Parosteal osteosarcomas of the head and neck region are rare, low-grade variants of osteosarcoma, but have the potential to recur with simple local excision. Clinical and radiographic features are diagnostically helpful. Definitive diagnosis comes from histopathology, and wide local resection should be employed as the optimal treatment.

Keywords: parosteal osteosarcoma; juxtacortical osteosarcoma; maxilla

Sarcomas of the head and neck region are rare tumors, accounting for only 1% of all head and neck malignancies.1 Although osteogenic sarcomas represent the most common nonhematopoietic primary malignant bone tumor with an overall incidence of 1:100,000 per year,2 osteosarcomas of the head and neck region represent only 6% to 10% of all osteosarcomas.3,4 Classic osteosarcomas, also termed central osteosarcoma, are still 1 of the most common sarcomas of the head and neck region.5,6 Osteosarcomas of the jaws, when compared with long bones, tend to develop in older patients. Osteosarcomas are more frequent in the mandible than in the maxilla (49% vs 37%, respectively) and are usually of high grade.7 A separate class of osteosarcomas has been termed juxtacortical osteosarcoma, which includes intermediate grade periosteal and low-grade parosteal variants. Because of less aggressive biological behavior, it is important to recognize these subtypes of osteosarcomas to plan treatment appropriately and avoid overtreatment. We present a case of parosteal osteosarcoma of the maxilla along with a review of the relevant literature.
CASE REPORT

A 32-year-old man presented to the Head and Neck Surgical Oncology clinic, the University of Utah’s Huntsman Cancer Institute, complaining of a 1-year history of a firm, right palatal mass. The onset of this mass was gradual in duration. Over the ensuing month, it increased in size and became painful, and the overlying mucosa began to ulcerate. He did not have hypesthesia or mobility of his maxillary dentition. The patient was otherwise healthy, taking no medications except occasional antacids, and claimed only occasional alcohol use and no tobacco use.

Physical Examination. On examination, an approximately 2.5- to 3.0-cm bony hard mass was found to overlay the palatal portion of the right maxillary alveolar ridge in the region of the molar teeth. It extended to but did not include the midline of the hard palate. The teeth were not mobile. There was a small area of ulceration of the overlying mucosa that appeared to be due to pressure from the mass, but no evidence of obvious epithelial neoplastic changes was noted. There was no evidence of cervical lymphadenopathy. Because osteosarcoma was highest on the differential diagnostic list, a CT scan with bone windows was ordered in conjunction with a chest CT. No evidence of pulmonary spread was noted on the lung CT. He was also referred to our prosthodontic colleague for fashioning of an obturator in preparation for surgical intervention.

Radiographic Evaluation. A CT scan was performed, with 2.5 mm axial, noncontrasted bone, and standard algorithm images, with reconstructions in both the sagittal and coronal planes. This study demonstrated a 1.2 cm × 2.4 cm × 2.2 cm focal mass, with dense ossification medial to the right maxillary ridge. The mass appeared well circumscribed without a soft tissue component or destructive element. There were no surrounding aggressive changes to suggest inflammation or osseous invasion seen on the CT. There was not a suggestion of continuity with the medullary cavity as well as benign appearing edges overhanging a bony stalk (Figure 1). This radiographic appearance suggested a low-grade malignancy or a benign process, such as an osteoma.

Surgical Intervention. On the basis of these findings, the patient was counseled regarding the need for surgical excision. The patient underwent a partial maxillectomy with a midline cut through the central incisors across the maxillary face just above the alveolus to the lateral buttress and along the hard palate. The defect was packed with Xeroform gauze, and an obturator was utilized to keep the packing in place. On his 1-year follow-up examination, he was without evidence of disease recurrence.

Pathologic Findings

Gross Pathology. The specimen was a 6.5 cm × 3.5 cm × 2.5 cm resection of the right maxillary alveolus and hard palate, including dentition from the central incisor to second molar. There was a 1.6 cm × 1.3 cm defect in the palatal alveolar mucosa. A round bony mass measuring 2.5 cm × 2.5 cm × 2.5 cm protruded from the hard palate and extended into the interdental space between the first and second molars.

Microscopic Pathology. Histologic sections revealed that the bony mass was superficial to the cortical bone and beneath the mucosa of the maxillary alveolus and hard palate. In foci, the bony mass was in contact with the cortical bone, but the mass was mostly separated from the cortical bone by a thin layer of fibrous periosteal tissue (Figure 2). There were no areas in which the bony mass penetrated...
into medullary bone of the maxilla. Where the bony tumor extended into the interdental space, it was superficial to the crestal alveolar bone (Figure 2). The tumor mass was composed of well-developed bone trabeculae of varying thickness in a moderately to lightly cellular stroma (Figure 3). In some areas, the tumor formed a nearly solid bony mass. Lacunae within the tumor bone were moderately numerous, and osteoblasts lined many of the trabeculae. The intertrabecular stromal cells were spindled and rounded. Cellular atypia varied from slight to focally moderate. The tumor bone in the area of the ulcerated mucosa had been exposed directly to the oral cavity and was necrotic. All surgical margins were clear of tumor.

**Discussion/Review of Literature.** Geschickter and Copeland provided the first description of a parosteal osteoma in 1950. They described lesions that occurred primarily in the long bones of young or middle-aged adults.8 These tumors were thought to begin as ossifying fibrous tissue of the periosteum with a tendency toward malignant differentiation resembling sclerosing osteogenic sarcoma. The first report of a parosteal osteosarcoma in a craniofacial site was reported in 1961.9 This mandibular parosteal osteosarcoma experienced a number of local recurrences, which eventually was definitively treated with a hemimandibulectomy resulting in 7 years without evidence of a recurrence. In 1970, Roca et al7 reported 2 cases of parosteal osteogenic sarcoma, 1 of the mandible and the first reported involvement of the maxilla. These lesions have also been termed juxtacortical osteogenic sarcomas, which some authors separate into the low-grade parosteal and the high-grade periosteal osteosarcomas.10 To date, including this report, only 17 cases of maxillary or mandibular juxtacortical osteosarcomas have been reported. Only 8 of these rare cases were parosteal osteosarcomas, and of the 9 cases of parosteal osteosarcomas (this case included), 5 originated in the maxilla (Table 17,9,11–22).

The clinical presentation of the parosteal variant of osteosarcomas is as an expansile lesion on the involved bone, with a nonlobular outer surface, and a potential for overlying mucosal ulceration. The majority of the reported cases lacked demonstrable paresthesia or anesthesia, and no report found any evidence of lymphadenopathy.

Utilizing the panorex for radiographic analysis, parosteal osteosarcomas demonstrate a lack of continuity with cortical bone, but medullary involvement may be seen. This is in contrast with periosteal lesions, which are distinguished by an intact cortex with no marrow involvement radiologically. Additionally, parosteal osteosarcomas usually overgrows its base of origin, whereas the periosteal variant tends to remain within the confines of its base.17 Bianchi et al23 found the CT scan to be helpful for characterization of the lesions when compared with conventional radiologic studies. They found characteristics consisting of a hyperdense, homogeneous mass similar to compact bone attached to the cortex by a short pedicle, with sharply defined posterior margins separated from the maxillary cortex by a layer of hypodense tissue. The anterior margin was described as an irregularly thickened overlying mucosa and no local infiltration.23 In our case, the...
CT scan features could not differentiate between a benign process and a low-grade osteosarcoma.

Histologically, parosteal tumors are ill-defined, exophytic tumors with a regular external surface and no cartilaginous cap, a tendency to overgrow the base of origin, and a spindle-cell stroma with mild atypia. These lesions are composed of intermixed bone, fibrous, and cartilaginous tissue involving the periosteum. The bland histologic appearance may lead to misdiagnosis as osteoma, osteochondroma, heterotopic ossification, or myositis ossificans. Periosteal osteosarcomas tend to have a lobular and well-defined periphery composed of more poorly differentiated malignant cartilaginous tissue. Accurate diagnosis requires correlation of clinical, radiologic, and histologic features.

Parosteal osteosarcomas of the craniofacial area behave similar to the long bone counterparts with slow growth as a low-grade malignancy that do not tend to metastasize but can recur after local excision. The consensus is that these lesions must be recognized and differentiated from benign lesions as they have a propensity for recurrence with limited local excision, and necessitate a wider resection with negative margins. In fact, recurrent or long-standing lesions may develop a more aggressive appearance and behavior. However, they are not as aggressive as central osteosarcoma. With adequate surgical excision with a negative margin, there is no defined role for chemotherapy or radiation as adjuvant treatment.

CONCLUSIONS

Parosteal osteosarcomas of the mandible or maxilla are rare, low-grade variants of osteosarcoma. Although these lesions are low-grade malignancies with minimal potential to metastasize, they can recur with simple local excision. The clinical features combined with the radiologic characteristics presented are diagnostically helpful. Definitive diagnosis comes from histopathology, and wide local resection should be employed as the optimal treatment.

REFERENCES


Table 1. Parosteal and periosteal osteosarcoma cases of the maxilla and mandible.

<table>
<thead>
<tr>
<th>Author</th>
<th>Sex/age, y</th>
<th>Site</th>
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<th>Follow-up, y/status</th>
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<td>Som and Peimer</td>
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<td>Multiple resections, hemimandibulectomy</td>
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<td>Midline maxilla</td>
<td>Local resection</td>
<td>9/NED</td>
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<td>M/63</td>
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<td>Right mandible</td>
<td>Several local resections, hemimandibulectomy</td>
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Abbreviations: NED, no evidence of disease; DOC, died of other causes; NP, not provided; DOD, died of disease.
trunk: an analysis of 1,702 patients treated on neoadjuvant cooperative osteosarcoma study group protocols.


