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Periosteal Osteosarcoma of the Mandible: Case Report and Review of the Literature

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Abstract

Purpose—Periosteal osteosarcoma is an intermediate-grade chondroblastic osteosarcoma, accounting for less than 2% of all osteosarcomas and it occurs extremely rarely in the jaws. We present a rare case of mandibular periosteal osteosarcoma.

Patients and Methods—A 39-year-old man presented with a four months history of a firm swelling of the mental region. The clinical, radiologic, and histologic findings pointed to a diagnosis of periosteal osteosarcoma.

Results—The patient underwent a partial mandibulectomy. After ten months the lesion recurred locally. We removed the tumor together with the residual right mandible, and the patient remained without evidences of local recurrence after 22 months.

Conclusions—Periosteal osteosarcoma may give rise to local recurrence and distant metastasis. It is important to differentiate the periosteal osteosarcoma from other types. Accurate diagnosis requires correlation of clinical, radiographic, and histologic features, and wide surgical margins are mandatory for successful therapy.

Keywords

periosteal osteosarcoma; juxtacortical osteosarcoma; mandible; recurrence; surface osteosarcoma

Osteosarcoma (OS) is the most common primary malignancy of bone, with a reported incidence of 1:100,000 per year.¹ These tumors typically originate in the extremities and the pelvis, with only 2.7–6.6% of OSs were located in the jaws.^{2,3}

Although conventional OS arises intramedullary, some OSs occur on the outer surface of bone, this is called surface or juxtacortical OS and exhibits distinct radiographic, histologic features and biological behavior that is obviously different from conventional OS. OSs arising on the surface of bone can be classified into three types according to the nature of the tumor: parosteal, periosteal, and high-grade surface OS.⁴ Parosteal OS is a low-grade OS, with an incidence of about 4% of all OSs. Periosteal OS is an intermediate-grade chondroblastic OS, accounting for less than 2% of all OSs. High-grade surface OS is the most rare subtype, accounting for less than 1%. Periosteal type tends to have a better

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prognosis than that of conventional OS or high-grade surface OS, but poorer than that of parosteal OS and it is capable of local recurrence and distant metastasis.⁵

Periosteal OS occurs extremely rarely in the jaws. To our knowledge, including this report, only ten cases have been reported. We report a case of periosteal OS located at the mental region of the mandible and review the literature with respect to clinical, radiological and histopathological criteria, we also discuss the differential diagnosis.

Report of a case

A 39-year-old man was admitted because of swelling of the mental region and numbness on the overlying skin. His symptoms started four months earlier when he initially noticed a gradually enlarging mass in the mental region. Since his lower incisors had been mobile for several months, he consulted a countryside dentist, who recommended removal of his mobile teeth. Consequently, the lower central incisors and lower right lateral incisor were removed by the dentist but the mass failed to resolve. During the next 4 months, the lesion continued to increase in size gradually. The patient was subsequently referred to our hospital for further management. Before seeking treatment, he developed numbness of the mental region. His past history and family history were not contributory, and no apparent history of trauma around the mental region was noted.

On examination, an approximately 4×5 cm mass was found to overlay the mental region (Figure 1). The mass was rubbery, nontender, and fixed on palpation. There was no palpable cervical lymphadenopathy. Intraoral examination revealed mandibular vestibule and the mouth floor swelling with expansion extending from the lower right first premolar to the lower left lateral incisor. The swelling was covered with intact, normal appearing mucosa. The lower incisors extraction sockets had healed. The other teeth were not mobile. Laboratory data including alkaline phosphatase level were within normal limits. Because osteogenic neoplasms were higher on the differential diagnostic list, a mandible CT scan was ordered.

CT scan of the mandible revealed a well circumscribed homogeneous and radiolucent mass measuring 30mm × 46mm in size, with non-homogeneous contrast enhancement. The tumor was found to encircle the cortex of the bone partially. There was no evidence of cortical destruction or medullary involvement (Figure 2).

On the basis of these findings, the patient was counseled regarding the need for an incisional biopsy. At the time of surgery, after elevation of a mucoperiosteal flap, the lesion was found to be closely attached to the cortical plate of the mandible, but the cortex had remained intact. Histopathological examination of an incisional biopsy specimen revealed that the tumor consisted exclusively of lobules of cartilaginous-appearing tissue with small foci of osteoid formation. The chondroid matrix component was composed of poorly differentiated, anaplastic chondroblastic cells (Figure 3A). In addition, mitotic figures were readily seen among the chondrocytes and osteocytes (Figure 3B). Immunohistochemically, the tumor showed diffuse reactivity with antibodies against vimentin and focal reactivity with antibodies against the S-100 protein in the chondroid containing fields and the spindle cells (Figure 3C). On the basis of clinical, radiological, histopathological features, a diagnosis of periosteal OS of the mandible was established. After the diagnosis of periosteal OS had been made, the patient underwent a partial mandibulectomy from the lower right second premolar to the lower left canine with the overlying skin and mucosa and immediate reconstruction with a titanium plate and bilobed radial forearm free flap. No residual tumor was identified. After ten months the lesion recurred locally, which was located at the lingual bone cortex surface of the residual mandible on the right side (Figure 4A). We removed the tumor together with the residue of the right mandible (Figure 4B). The resected primary and

recurrent lesions demonstrated histopathologic features identical to those seen in the initial biopsy specimen. At no time was radiation or chemotherapy given. 22 months after the second surgery, he was without evidence of disease recurrence or distant metastasis.

Discussion

Periosteal OS is an uncommon neoplasm that comprises less than 2% of all OSs⁴ and is rarely located in the jaws. To date, including this report, only 10 cases of periosteal OS of the jaws have now been reported in the English-language literature.^{5–12} There were 5 males and 5 females with an age range from 16 to 65 years and a median of 33 years. Our case was in this age range. Three tumors arose in the maxilla and seven in the mandible. Our case arose in the mandible. All patients underwent surgery to excise the tumor. The median follow-up is 30 months (range 8–72 months). With the exception of only 2 (this case included) of the 10 cases reported, there have been no recurrences of tumor. In the first case, described by Piatelli and Favia,⁵ tumor recurred locally 18 months later. The patient underwent a radical right maxillectomy followed by several cycles of adjuvant chemotherapy. Lung metastases appeared, and the patient died 5 years after the first diagnosis.

The definition of periosteal OS is very precise. Accurate diagnosis requires correlation of clinical, radiographic, and histologic features. Importantly it is essential to exclude intramedullary OS with periosteal extension, high-grade surface OS and parosteal OS from this group. The clinicopathologic characteristics of periosteal OS arising in the jaws include the following: (1) peak occurrence is over 30 years of age, the median age of periosteal OS of the jaws is about 15 years later than that in the long bone counterparts;¹³ (2) the tumor is most often lobulated and well defined; (3) radiologically, periosteal OS tends to be more radiolucent than the other types of OS since it is predominantly cartilaginous,^{2, 6} the cortex is intact and sometimes thickened, and there may be minimal tumor invasion into the cortex, without medullary involvement;¹² (4) the tumor is predominantly composed of poorly differentiated malignant cartilaginous tissue;^{11, 12} (5) the tumor may give rise to local recurrence and distant metastasis;⁵ (6) prognosis in periosteal OS seems to be excellent with surgical treatment² only and is considerably better than that of conventional OS or high-grade surface OS, but poorer than that of parosteal OS.⁶ The clinical presentation of the parosteal variant^{14, 15} of OS is as an expansile lesion on the involved bone, with a nonlobular or lobular outer surface, and a potential for overlying mucosal ulceration. Radiologically, parosteal OS appears as a hyperdense, homogenous mass similar to compact bone attached to the cortex by a thin layer of hypodense tissue. Histologically, this tumor is well differentiated and characterized by a spindle-cell fibroblastic-like proliferation containing well-developed bony trabeculae. Our patient's tumor was dominated by aberrant chondroid with a lesser component of osteoid. The CT images of this case revealed that the tumor was a homogeneous and radiolucent mass with no evidence of involvement of the marrow cavity. High-grade surface OS¹⁶ is characterized by a rapid-growing sarcoma arising from periosteal tissue. Radiologically, the tumor appears as dense to moderate mineralization with a fluffy, immature appearance. Histology reveals a highly pleomorphic, predominantly osteoblastic, lesion that lacks the cartilaginous lobules seen in periosteal OS. The criteria for a diagnosis of high grade surface OS are that the lesion had arisen on the surface of bone, that histologic findings of the tumor are identical to those of conventional intramedullary OS and that, even if medullary involvement is present, most of the tumor is on the surface of bone.^{17, 18} Conventional OS typically is of a higher histologic grade than its periosteal counterpart. Radiographs of most conventional OSs of the jaws show a unicentric bone destructive lesion with indefinite margins. The radiological appearance usually will be sclerotic, lytic or mixed, in which areas of calcification and of lysis are intermingled. Histologically, conventional OS is characterized by tumor osteoid and/or

osseous tissue arising directly from a sarcomatous stroma without passing through a cartilaginous phase. The stromal cells may be predominantly osteoblastic, chondroblastic or fibroblastic.¹⁹ The key differences in four types of OSs are summarized in Table 1. In our patient's case, an intramedullary OS with periosteal extension and a high-grade surface OS were ruled out because the tumor originated and initially proliferated outward from the osseous surface and without intramedullary extension.

Revell et al¹³ described the tumor was found to encircle between 5 and 100% of the cortex of the bone in their series of periosteal OS arising in the long bone, similar to what was observed in our case. This is the first documented case of a periosteal OS of the mandible with partial cortex encirclement. The overlying skin numbness in the present case could be the result of the right mental nerve involved. The local recurrence in our patient was found after ten months later. We performed a radical en bloc resection of the right mandible. At the 32 month follow-up appointment after the first diagnosis, there was no evidence of recurrent disease or distant metastasis.

In the jaws, OS has different clinical and biologic characteristics than its counterpart in the long bones.^{1-3, 20, 21} OS of the jaws tends to occur at an older mean age, and the prognosis for jaw OS is better than that of OS arising in other sites. The more favorable prognosis may be due to lower mitotic activity of tumor cells and because cellular anaplasia is found less often in OS of the jaws. In addition, jaw OSs have less of a tendency to metastasize than OSs of the long bones. However, the survival rate of periosteal OS of the jaws cannot be accurately assessed because of the limited number of cases and lack of long-term follow-up. The most effective therapy for periosteal OS of the jaws is wide surgical excision of either the mandible or the maxilla with a tumor-free margin but the role of chemotherapy in this condition is still debatable.^{1, 5-13}

The clinical, radiologic, and microscopic features of our case fulfill the criteria proposed for periosteal OS. Periosteal OS may give rise to local recurrence and distant metastasis. It is important to differentiate the periosteal OS from other types. Accurate diagnosis requires correlation of clinical, radiographic, and histologic features, and wide surgical margins are mandatory for successful therapy.

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Figure 1.
Clinical photograph of a 39-year-old man with a 4×5 cm swelling on the mental region.

Figure 2. This axial contrast-enhanced (soft tissue window) CT image shows a well-defined, radiolucent, with peripheral contrast enhancement mass, encircling the cortex of the bone partially (arrow). There is no signs of cortical destruction or medullary involvement.

Figure 3.

A. Photomicrograph showing lobules of atypical chondroid admixed with small foci of malignant osteoid formation (arrow A). The tumor consists of highly atypical malignant chondroblastic cells (arrow B). (H&E, original magnification $\times 100$).

B. Some mitotic figures are present among the chondrocytes (arrows). (H&E, original magnification $\times 100$).

C. Some tumor cells show positive staining for antibodies against the S-100 protein. (original magnification $\times 100$).

Figure 4.

A. Axial contrast-enhanced (soft tissue window) CT scan demonstrating a recurrent lesion (arrow) on the outer surface of the lingual cortex of the right posterior mandible.

B. Photograph of the resected recurrent tumor together with the residue of the right mandible.

Table 1

The key differences in four types of osteosarcomas

Characteristic	Intramedullary OS	High grade surface OS	Periosteal OS	Parosteal OS
Clinic				
Chief complaint	Swelling, pain	Swelling, pain	Swelling	Swelling
Progress	Rapid	Rapid	Slow	Slow
Prognosis	Poor	Poor	Relatively good	Good
Radiologic				
Appearance	Sclerotic, lytic or mixed	Fluffy, immature ossification	Radiolucent mass	Radiodense mass
Origination	Medullary	Juxtacortical	Juxtacortical	Juxtacortical
Medullary involvement	Yes	None or minimum	None	None
Histologic				
Main component	Tumor osteoid, osteoblastic, chondroblastic, or fibroblastic stroma.	Identical to intramedullary OS. predominantly osteoblastic stroma	Cartilaginous tissue, chondroblastic stroma	Bone tissue, fibroblastic stroma
Grade	High	High	Intermediate	Low
Growth pattern	Highly aggressive	Highly aggressive	Expansile, locally aggressive	Expansile, locally aggressive