Case Report

Osteosarcoma of Maxilla with Unusual Image Findings in Child

Aya Yamamoto, Junichiro Sakamoto, Takashi Muramatsu*,
Sadamitsu Hashimoto*, Takahiko Shibahara**, Masaki Shimono*
and Tsukasa Sano

*Department of Oral and Maxillofacial Radiology, Tokyo Dental College,
1-2-2 Masago, Mihama-ku, Chiba 261-8502, Japan*

**Department of Pathology, Tokyo Dental College,
1-2-2 Masago, Mihama-ku, Chiba 261-8502, Japan*

***Department of Oral and Maxillofacial Surgery, Tokyo Dental College,
1-2-2 Masago, Mihama-ku, Chiba 261-8502, Japan*

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Abstract

Osteosarcoma of the head and neck is relatively rare and accounts for less than 10 percent of all osteosarcomas in general. We report a case of osteosarcoma in which imaging and histopathology of the hard palate of an 11-year-old boy yielded atypical findings. An approximately 8 × 15 mm lesion found in the center of the palate was hard and healthy in color. Subsequent biopsy resulted in a diagnosis of nonepithelial malignant tumor. No abnormalities were observed in the maxillary bone or tooth on panoramic or occlusal radiographs. Computed tomography images revealed a mass lesion approximately 7 × 9 × 9 mm in size on the hard palate extending into the maxilla. The cortex of the maxilla adjacent to the lesion was unclear in parts. The internal structures were slightly inhomogeneous and its density was lower than that of muscle. On magnetic resonance images, the lesion was represented by low signal intensity on T1-weighted (T1W) images and high signal intensity on T2-weighted images with fat-suppression. The margin of the lesion was a little unclear and the internal structures were slightly inhomogeneous. The lesion was enhanced homogeneously on post-contrast T1W images with fat-suppression. The histopathological diagnosis was fibrogenesis-type osteosarcoma. No findings specific to osteosarcoma such as localized enlargement of the periodontal ligament space alongside the root, cortical destruction, periosteal ossification or osteogenesis were found in this case.

Key words: Osteosarcoma—Hard palate—Teens—
Computed tomography imaging—Magnetic resonance imaging

Introduction

Osteosarcoma, the most frequent malignant tumor of bone, is characterized by produc-

tion of variable amounts of osteoid tissue by neoplastic cells^{4,5}. It develops mainly in the young, and the median age at diagnosis is the teens⁵. Osteosarcoma of the head and neck is

Fig. 1 Panoramic radiograph showed no abnormal findings in maxilla or hard palate

relatively rare and accounts for less than 10 percent of all osteosarcomas in general¹⁰⁾, with the jaw being the most frequently encountered site. The peak incidence for osteosarcoma of the jaw is in the fourth decade of life, approximately 10–15 years later on average than that of the long bone¹⁰⁾. The main symptoms are swelling and pain. Imaging findings in osteosarcoma of the jaw have been reported in several previous studies. The lesion is usually radiolucent with an ill-defined border¹⁵⁾. If the lesion involves the periosteum, whether directly or by extension, typical sunray spicules may be observed. The appearance of Codman's triangle at the edges is also well documented.

The characteristic feature of osteosarcoma is the presence of osteoid tissue within the tumor. Tumor cells are extremely pleomorphic and produce osteoid tissue described as irregular trabeculae with or without central calcification¹¹⁾. Tumor cells are included in the osteoid matrix¹¹⁾. Osteosarcomas may exhibit multinucleated osteoclast-like giant cells¹¹⁾.

In this paper, we report a case of osteosarcoma occurring in the hard palate of an 11-year-old boy.

Case Report

Approximately 1 month prior to being referred to the Chiba Hospital of Tokyo Dental College for further testing and treatment

by his family dentist, an 11-year-old boy had noticed a stomatitis-like lesion on his palate which was continuing to grow in size. Although his family medical history showed no remarkable findings, the patient himself had a history of afebrile convulsions.

At the first medical examination, a hard lesion approximately 8×15 mm in size and healthy in color was observed in the center of the palate. Clinically, non specific stomatitis was suggested. Fine-needle aspiration biopsy was performed. No evidence of atypical cells was found in the specimen, and cytologically it was diagnosed as Class I. After aspiration biopsy, the lesion exhibited rapidly growth, and malignant neoplasm was suggested.

By open biopsy, the lesion was diagnosed as non-epithelial malignant tumor.

Evaluation of the palate on panoramic radiographs was difficult, and there were no abnormal findings in the maxillary bone or tooth (Fig. 1). Occlusal radiographs also revealed no evidence of a mass lesion on the palate or abnormal findings in the roots of the incisors (Fig. 2).

After the biopsy, computed tomography (CT) and magnetic resonance (MR) imaging were carried out.

Images were obtained using a multidetector-row CT scanner (Somatom Plus 4 Volume Zoom, Siemens, Erlangen, Germany). The CT images revealed a mass lesion approximately 7×9×9 mm in size on the hard palate extending into the maxilla (Fig. 3). The

Fig. 2 Occlusal radiograph of maxilla showed no abnormal findings

cortex of the maxilla adjacent to the lesion was unclear in parts. The internal structures were slightly inhomogeneous and its density was lower than that of muscle (Fig. 3c). Cross-sectional images revealed that the lesion was not in contact with the nasopalatine canal (Fig. 4).

MR images in the axial and coronal planes were obtained using the 1.5-Tesla MR unit (Magnetom Symphony Maestro Class, Siemens, Erlangen, Germany) with a head and neck coil. The lesion was represented by low signal intensity on T1-weighted (T1W) images (Fig. 5a, 5d) and high signal intensity on T2-weighted (T2W) images with fat-suppression (Fig. 5b, 5e). The margin of the lesion was a little unclear, and the internal structures were slightly inhomogeneous. The lesion was enhanced homogeneously on post-contrast T1W images with fat-suppression. The MR images also suggested that the lesion had not invaded the nasal cavity.

The lesion was excised surgically and examined histopathologically. The resected material was about 10-mm in diameter. The cut surface of the tumor revealed a regular-shaped, grey-white, solid mass without necrosis and hemorrhage (Fig. 6). Microscopically, the tumor mass consisted of prominent prolifer-

(a)

(b)

(c)

Fig. 3 CT images

- (a) Axial image showed smooth bone destruction by mass lesion (arrow).
- (b) Reconstructed coronal image showed smooth bone destruction by mass lesion (arrow). No abnormalities observed in floor of nasal cavity.
- (c) Axial image using soft tissue algorithm showed slightly inhomogeneous low density mass lesion on palate.

Fig. 4 Cross-sectional CT images of anterior maxilla

erating spindle-cells, with areas of a herring-bone or storiform pattern. The cellularity of the parenchyma was high, but cellular atypism was not so severe. Also, osteoid formation was observed, focally (Fig. 7). Immunohistochemically, the spindle tumor cells were strongly positive for vimentin, alpha smooth muscle actin and desmin. The intensity of the Ki-67 index was high exclusively in the tumor cells. The final diagnosis was fibroblastic-type osteosarcoma, including small amounts of osteoid tissue formation.

Discussion

Approximately 6.5% of all osteosarcomas arise in the jaw⁴⁾, and the mandible is involved almost twice as often as the maxilla¹³⁾. In its diagnosis, image evaluation is important, as the clinical symptoms are not specific.

In this case, the growing lesion was located on the palate and clinically no pain was reported. Conventional radiographic images, including panoramic radiographs, revealed neither the outline of the lesion nor any abnormalities in the maxillary structures. However, the anomalies adjacent to the lesion were detected easily on CT and MR images, which revealed that its internal structures were slightly inhomogeneous. Bone destruction adjacent to the lesion was smooth. The epicenter of the lesion was intra-maxillary bone and, morphologically, the cortex adjacent to the lesion was spiny (Fig. 4 arrowhead). This led us to believe that the lesion might have arisen from the maxillary bone and be malignant. Osteosarcoma, however, was not included in the differential diagnosis.

A number of studies have reported image findings in osteosarcoma of the jaw. Cavalcanti *et al.*¹⁾ reported that localized enlargement of

Fig. 5 MR images

(a) Axial T1W image (TR/TE: 500/15), (b) Axial T2W image with fat-suppression (TR/TE: 4,600/90), (c) Axial post-contrast T1W image with fat-suppression (TR/TE: 500/15), (d) Coronal T1W image (TR/TE: 500/15), (e) Coronal T2W image with fat-suppression (TR/TE: 4,600/90), (f) Coronal post-contrast T1W image with fat-suppression (TR/TE: 500/15).

the periodontal ligament space alongside the root was strongly suggestive of osteosarcoma, and Garrington *et al.*⁴⁾ reported that radiological evidence of a symmetrically widened periodontal ligament space was a significant early finding in osteosarcoma. We also reported that this finding was very useful for imaging diagnosis of osteosarcoma¹⁶⁾. However, in this case, no enlargement of the periodontal ligament space was observed alongside the left central incisor adjacent to the lesion on CT images. Furthermore, evaluation of the left lateral incisor was difficult as it was still under development.

Van der Heul and von Ronnen reported that cortical destruction and periosteal ossifying reactions (sunburst striations, Codman's triangle) are characteristic of osteosarcoma¹⁴⁾. Moreover, these findings are shown in highly aggressive neoplasms typically composed of spindle cells producing osteoid tissue⁸⁾. The usefulness of these findings was also reported in our previous report¹⁶⁾. However, no such reactions were observed in this case.

deSantos *et al.* reported that conventional radiographic findings associated with periosteal osteosarcoma have included thickening of the diaphyseal cortex with scalloping and a

Fig. 6 Gross appearance of tumor
Tumor shows expansive and well-circumscribed growth (arrows) resembling lesion seen on cross-sectional CT image.

Fig. 7 Tumor was mainly composed of spindle cells, often in herringbone or storiform pattern. Focal osteoid production by neoplastic stromal cells observed (Hematoxylin & Eosin staining, Original magnification $\times 200$).

perpendicular periosteal reaction extending into a broad-based soft-tissue mass³). In this case, however, no thickening of cortical bone or scallop-shaped border was observed (Fig. 4).

Osteosarcoma lesions are also characterized by bone destruction and high biological virulence apparently caused by active osteoclasts⁵). In this case, pressure-type bone destruction adjacent to the lesion was shown on CT

images (Figs. 3 and 4).

Finally, in terms of osteogenesis, Clark *et al.* has classified the image finding patterns of an osteosarcoma into three types: “lytic”, “sclerotic” and “mixed”²). This case showed a “lytic” pattern, as the CT images revealed none of the high density structures characteristic of osteogenesis (Figs. 3 and 4). The lesion in the cross-sectional CT image resembles the cut section of the resected specimen in the photograph (Fig. 6). On the other hand, the pathological findings were not reflected in the CT or MR images microscopically.

In terms of MR findings, some researchers have reported that osteosarcoma shows low signal intensity on T1W images and heterogeneous high signal intensity on T2W images^{6,7}). In this case, the lesion showed low signal intensity on T1W images and a little heterogeneous high signal intensity on T2W images with fat-suppression, which is in agreement with these earlier findings. Furthermore, MR images were evaluated for the presence and extent of mineralization (appearing as nodular foci of low signal intensity on images obtained at all MR pulse sequences) in the soft-tissue mass⁹). In this case, no low signal region indicating osteogenesis was observed. The extent

of bone marrow was observed most clearly on T1W images¹²⁾. In this case, the marrow signal in the tumor area could be differentiated clearly from that in the normal area on T1W images (Figs. 5a, 5d). Furthermore, in this case, the tumor was enhanced strongly on post-contrast T1W images with fat-suppression (Figs. 5c, 5f).

None of the above-mentioned specific image findings for osteosarcoma were observed in this case. This may have been because the lesion was small and still in the early stage of development.

In conclusion, we report a case of osteosarcoma with atypical images and histopathological findings.

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Reprint requests to:

Dr. Aya Yamamoto
Department of Oral and
Maxillofacial Radiology,
Tokyo Dental College,
1-2-2 Masago, Mihama-ku,
Chiba 261-8502, Japan
Tel: +81-43-270-3961