Osteosarcoma of the maxilla and the maxillary sinus: A case report

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Osteosarcoma of the jawbones is a rare malignant mesenchymal neoplasm with the tendency for new bone being directly formed by the tumor cells. Clinically, the tumor may be central or peripheral—periosteal—and histologically can be divided into three subtypes: osteoblastic, fibroblastic, and chondroblastic. This report presents a case of a central osteoblastic osteosarcoma of the left maxillary tuberosity and maxillary sinus. Problems related to definitive diagnosis and therapy are described and discussed. (Quintessence Int 2004;35:228-233)

Key words: diagnosis, histologic criteria, interdisciplinary treatment, maxilla, osteosarcoma

Osteosarcoma of the jawbones is a rare malignant mesenchymal neoplasm with the tendency for bone formation. Osteosarcomas of the jawbones are rare and account for 5% to 6% of all osteosarcomas. The estimated incidence is approximately 0.07 cases per 100,000 per year.

Clinically, the tumor may be central or peripheral—periosteal—and histologically can be divided into three subtypes: osteoblastic, fibroblastic, and chondroblastic. There seems to be a difference in the clinical behavior of osteosarcomas of the jawbones (OSJ) and osteosarcomas of the long bones (particularly distal femoral metaphysis, proximal tibia, and humeral metaphysis). OSJ are diagnosed 10 years later, usually in the third and fourth decade, show a minor tendency for metastasis, and generally have a better prognosis than osteosarcomas of the long bones. Their frequency is slightly higher in males, and they are more often located in the mandible. Typical clinical signs for OSJ are the following: intra- and/or extraoral swelling, loosening and change in position of teeth, proptosis, neurologic deficits, and limitation of mouth opening. Radiologically, a widened periodontal ligament in periapical radiographs of teeth located in the area of the malignoma often is seen. In many cases, these signs are present without pain; therefore, patients often seek medical care in a more advanced stage of the disease.

The current therapy of choice is total surgical removal of the lesion with safe margins. Radio- and chemotherapy seem to be less effective.

The following case report demonstrates and discusses the problems in establishing the final diagnosis and the subsequent interdisciplinary therapy of an osteoblastic osteosarcoma of the maxilla and the maxillary sinus.

CASE REPORT

A 50-year-old man was referred to the Department of Oral Surgery and Stomatology, University of Berne,
for evaluation and further treatment. The referring dentist had observed a hard and painless swelling in the region of the carious maxillary left third molar approximately 6 months prior. The tooth was assumed to be the reason for the swelling and was extracted. Subsequently, the lesion did not disappear but increased in size. The patient denied any significant medical history, had no known drug allergies, was not taking any medicine, and was not consuming alcohol or tobacco.

Extraoral examination revealed no swelling of the left side of the face. On palpation, there was a hard and painless induration below the left zygomatic bone. Mouth opening was not limited, with a distance between the incisors of more than 40 mm. The cervical and submandibular lymph nodes showed no pathologic findings.

Intraorally, there was a hard and painless swelling to the buccal and palatal aspect in the region of the maxillary left molars and maxillary tuberosity. The extraction socket of the maxillary third molar was still not healed and showed fibrin covering (Fig 1). At the junction of the buccal gingiva and the alveolar mucosa in the region of the left third molar, there was a mobile bony sequestra visible, measuring approximately 3 x 3 mm (Fig 2). The second premolar and second molar, supporting a fixed partial denture, showed increased mobility and probing depths up to 10 mm. The second premolar reacted positively to cold pulp testing. The second molar had a previous root canal treatment.

The panoramic radiograph showed inhomogeneous radiopacity in the region of the maxillary left molars and maxillary tuberosity. The floor of the left maxillary sinus was not clearly visible (Fig 3). The bony sequestra in the region of the maxillary left molar was removed without local anesthesia and sent for histologic examination (first biopsy). The clinical and radiologic findings indicated a malignant lesion; therefore, the patient was referred to the Department of Craniomaxillofacial Surgery, University Hospital of Berne, on the same day.

The histologic analysis of the bony sequestra (first biopsy) showed immature bone with necrosis, moderate cellular atypia, and signs of inflammation (Fig 4). These findings were consistent with an infected sequestra but gave no definitive conclusion about the diagnosis of the lesion.

Further radiographic imaging, including computed tomography scan (CT) and magnetic resonance imaging (MRI), was performed, and the MRI showed a large expansile lesion of the left maxilla involving the maxillary sinus without signs of infiltration or destruction of the surrounding tissues. In the CT scan, signs of calcification were evident in the susceptible lesion (Figs 5 and 6). Because the clinical, radiologic, and pathologic findings were not conclusive, a large bony
biopsy of the left maxilla and the maxillary sinus (second biopsy) was performed under general anesthesia.

The histologic analysis of this second biopsy showed hypercellular tumor components with immature bone (osteoid), atypical cells, atypical mitotic figures, little chondroid differentiation, and infiltration of adjacent soft tissue (Fig 7). The histomorphologic criteria were consistent with those of an osteoblastic osteosarcoma.

To exclude eventual distant metastasis, a three-phase skeletal scintigraphy (Fig 8) and a thoracic CT scan were performed. In both the scintigraphy and CT scan, no signs of metastasis could be found. As definitive treatment, radical surgical excision of the primary tumor with subsequent chemotherapy was performed.

En bloc removal of the lesion was performed by a partial maxillectomy under general anesthesia (Fig 9). Intraorally, a sublabial incision was made and the mucosa overlying the left lateral wall of the maxilla was elevated. The dissection of the soft tissue was extended posteriorly to the pterygoid plates. The initial osteotomy was made using a sagittal saw vertically at the left alveolar ridge between the canine and the first premolar. The vertical extent of the osteotomy was taken directly inferior to the infraorbital foramen preserving the nerve. The vertical osteotomy was extended posteriorly along the anterior and lateral walls of the maxillary sinus in a horizontal fashion to include the medial and lateral pterygoid plates. A cuff of the pterygoid muscles was taken with the specimen. A median sagittal osteotomy was also performed through the hard palate to connect to the lateral osteotomy site. A portion of the soft palate was resected to include a 1.5-cm margin around the tumor.

The primary reconstruction of the defect was performed using a pedicled temporalis muscle flap and free calvarium bone grafts with expectation of dental
implants in the future. Harvesting the flap and bone grafts was done through a hemicoronal incision. The incision was extended to the preauricular crease to expose the zygomatic arch. The size of the bony defect was carefully measured, and appropriate split calvarial bone grafts were harvested from the parietal skull. Four pieces of split calvarium measuring approximately 1.5 × 3.0 cm were harvested (Fig 10). The harvested split calvarium was used to reconstruct the alveolar ridge and fixed to the remaining wall of the maxillary sinus using a titanium plating system (Modus System, Medartis). Dissection of a temporalis muscle flap was performed in the deep layer of the superficial fascia. Anterior and posterior zygomatic osteotomies were done to allow for rotation of the flap. The muscle flap was rotated into the site of the bony defect through the site of the zygomatic osteotomies. Pedicled buccal fat was also mobilized into the defect to provide soft tissue coverage of the inner side of the bone graft. After release and rotation of the flap, the zygomatic osteotomies were fixed into position using a titanium miniplate (Modus System, Medartis). Finally, the entire defect including the bone graft was covered by the temporalis muscle flap. The flap was secured to the border of the oral mucosa with absorbable sutures.

After excision, the tumor specimen was analyzed histologically, and the definitive diagnosis by the pathologist was a predominantly osteoblastic high-grade osteosarcoma with focally chondroid differentiation. The histologic presentation was a hypercellular malignant tumor with immature bone (osteoid), produced by atypical cells, atypical mitotic figures, few focal chondroid differentiation, and infiltration of adjacent soft tissue (Fig 11). Excision borders were free of tumor. All around the block resection, a minimal distance of 5 mm was seen from the tumor to the excision borders. The specimen also showed an intact epithelium of the maxillary sinus.

The postoperative course was uneventful. Following surgery, the patient underwent an adjuvant chemotherapy consisting of eight cycles of adriablastin and ifosphamid/platinol agents. The patient also received concurrent assistance with neupogen (recombinant G-CSF) treatment.

In the first year after surgery, the patient was seen on a monthly basis according to the cancer protocol of the Department of Craniomaxillofacial Surgery, University Hospital of Berne.

**DISCUSSION**

Predisposing factors for OSJ discussed in the literature are Paget’s disease, trauma to bone, previous radiation therapy, existing bone pathology (for example, fibrous dysplasia), and/or genetic predisposition. In the present case, none of the predisposing factors mentioned above could be confirmed with the available anamnestic, clinical, or radiologic data. Trauma to the bone through the extraction of the carious left third molar can almost be excluded with certainty because the swelling in the left maxilla was already present before the extraction. The progression of the osteosarcoma could have been accelerated by the extraction of the maxillary third molar. Although clinical and radiographic signs indicated malignancy, it was first after an open biopsy of the left maxilla and the maxillary sinus (second biopsy) that diagnosis could be established.

In the same osteosarcoma, different histologic aspects such as osteoblastic, chondroblastic, fibroblastic, and teleangectatic areas can be seen. Typical histologic criteria for diagnosis of a high-grade osteosarcoma are the following: hypercellular malignant tumor, immature bone formation (osteoid) produced by atypical tumor cells without osteoblasts, atypical mitotic figures, infiltration of adjacent soft tissue, and
in this case, few focal chondroid differentiation. Almost decisive for the definitive diagnosis is the finding of tumor osteoid, which atypical mesenchymal cells have produced.\textsuperscript{4} The diagnosis of osteosarcoma has severe and invasive therapeutic consequences for the patient, and therefore, it is very important to confirm such a diagnosis by a reference institute. In the present case, the histologic specimens were presented to and discussed with Prof Dr med G. Jundt, DÖSAK reference registry, Institute of Pathology, University Hospital of Basel, Basel, Switzerland.

To successfully treat osteosarcomas with predictable long-term results, radical resection has been proven to be the most important factor.\textsuperscript{13,14} In OSJ, metastasis in regional lymph nodes is rare. A neck dissection in combination with the surgical excision of the primary lesion is only necessary if regional lymph nodes have been tested positively.\textsuperscript{17}

Adjuvant chemotherapy in the treatment of osteosarcomas of the long bones has a beneficial effect on the 5-year survival rate.\textsuperscript{15,16} For surgery alone, the 5-year survival rate was around 15%; for surgery in combination with chemotherapy it was 60% to 80%. In OSJ, significant improvement of the survival rate with the help of chemotherapy has been questioned.\textsuperscript{22-24} Additionally, it was shown that adjuvant radiotherapy has no positive effect on the survival rate.\textsuperscript{27}

The general practitioner has an important role in the detection of premalignant and malignant lesions. Malignancies are rare and in their initial stages often without symptoms. The so-called “patient delay” — the time from patient awareness of the lesion to the first consultation of a doctor — cannot be influenced by medical personnel.\textsuperscript{21} In the present case, the “patient delay” seems not to have been very long because the patient went to his practitioner immediately after noticing the swelling in the left maxilla. The “doctor/professional delay” — the time from the first scheduled visit to the finding of the definitive diagnosis and subsequent treatment — can clearly be reduced with improved education of medical and dental professionals. In the present case, the “professional delay” was quite long — approximately 6 months. Furthermore, the healing disturbance of the extraction socket was not considered as a warning sign. Only after further progression of the swelling in the left maxilla did the practitioner refer the patient to the Department of Oral Surgery and Stomatology for further diagnosis and treatment. With the delayed finding of the definitive diagnosis, the surgical resection was aggressive and reconstruction more difficult than it would have been in an earlier stage of the disease. The aim of medical and dental practitioners is to recognize potential malignant processes as soon as possible and to refer them for diagnosis and treatment at a medical center offering an interdisciplinary therapeutic approach between general practitioners, oral and maxillofacial surgeons, pathologists, radiologists, and oncologists. Only with this procedure can malignancies be diagnosed and treated in their early stages with predictable long-term success.

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**REFERENCES**


With candor and familiarity, this book tells the story of Per-Ingvar Bråemark’s discovery of osseointegration and the challenges he faced to find acceptance for this concept within the scientific community. The biologic and biomechanical principles involved in this approach to rehabilitation are summarized, and the various clinical applications for osseointegration—from oral and maxillofacial prostheses to limb and finger-joint replacements—are described. Accounts of some of Bråemark’s first implant patients depict how this technique can transform lives. Anyone interested in osseointegration will find this book a fascinating read.

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