**PRIMARY OSTEOSTEOGENIC SARCOMA OF THE SKULL**

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**BACKGROUND**

An osteogenic sarcoma of the skull is rare, particularly as a primary tumor. The incidence of primary osteogenic sarcomas of the skull is about 1 to 2% of all skull tumors.

**CASE DESCRIPTION**

A 21-year-old male was initially evaluated because of a large mass that had been growing for 7 months. The patient had been experiencing frequent headaches and tenderness at the site of the lump for about a month before being seen by the neurosurgeon. A computed tomography scan revealed a large mass, 12 cm × 7 cm, involving the scalp extending from the right temporal region to the vertex. A magnetic resonance imaging (MRI) scan showed a large mass arising from the posterolateral aspect that was 90% extracranial and 10% intracranial on the right side of the skull. The MRI showed marked vascularity and neovascularity of the tumor. An angiogram was performed, which demonstrated that the mass was fed by the branches from the right external carotid artery. The patient subsequently underwent surgery for embolization of the right occipital and superficial temporal arteries and removal of the mass. Pathology evaluation of a specimen revealed a high-grade osteoblastic osteosarcoma.

**CONCLUSION**

We review the literature of reported cases of primary osteogenic sarcomas of the skull to discuss the common clinical presentation, evaluation methods, and recommended treatment plans. © 2002 by Elsevier Science Inc.

**KEY WORDS**

Osteosarcoma, skull neoplasm, primary tumor.

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An osteogenic sarcoma of the skull is rare, particularly as a primary tumor. Huvos et al reported that of 1200 patients found to have osteogenic sarcomas over 60 years’ time, 19 patients (1.6%) had tumors of the skull, and only 10 of those were de novo tumors [3]. Nora et al. reported an incidence of 2% (21 of 1000) for skull osteosarcomas, and in their series, 14 patients had de novo tumors [6]. Because of the rarity of this type of tumor, we present another case of a primary osteogenic sarcoma of the skull.

**Case Report**

A 21-year-old male was initially evaluated because of a large mass on his head and a headache. The patient stated the mass had been growing for 7 months. He had seen a primary care physician who believed the lump was an abscess. The physician performed cystic aspiration, which revealed a negative result. For the past month, the patient had been experiencing frequent headaches and tenderness at the site of the lump. The patient had no family history of cancer, but the patient stated he used tobacco and alcohol and occasionally used cocaine.

A computed tomography (CT) scan, with and without contrast, was obtained of the brain, which revealed a large mass involving the scalp extending from the right temporal region to the vertex. Its maximum anteroposterior dimension was 12 cm and in the transverse dimension, the mass was 7 cm. Without contrast, the CT showed irregular calcification and low attenuation areas, which represented necrosis (Figure 1A). With contrast administration, the mass was found to be eroding the right parietal bone, and there was extension through the bone to the right parietal dural surface. There was some compression of the adjacent brain parenchyma (Figure 1B).

A magnetic resonance imaging (MRI) scan was performed, and it showed a large mass arising from the posterolateral aspect of the skull on the right side. The mass was 90% extracranial and 10% intracranial (Figure 2). There was a calcified hemorrhagic part of the mass. The MRI showed marked...
vascularity and neovascularity of the tumor. An angiogram was performed, which demonstrated that the mass was fed by the branches from the right external carotid artery (Figure 3). The patient subsequently underwent surgery for embolization of the right occipital and superficial temporal arteries.

The patient underwent surgery to remove the mass. An incision was planned along the edges of the mass, extending 1.5 cm superiorly and posteriorly from the edges. The incision extended from the posterior occipital area to the anterior temporal area. The flap, which was densely adherent, was retracted anteroinferiorly. Another incision was made in the sterile neck, making it a T-shaped incision. The flap was dissected off the tumor. The extracranial tumor along with surrounding muscle and galea were excised in multiple fragments using Bovie, curettes and Leksell rongeur. A frozen section was sent to the pathology lab for evaluation. Necrotic bone around the edges of the tumor was also removed until healthy bone was obtained. Under microscopic magnification, the intradural extension of the tumor was removed. Meticulous hemostasis was obtained. The patient’s skull was reconstructed with bone cement, and a Z-plasty dural reconstruction was performed. Postoperative MRI showed gross total removal (Figure 4).

Pathology evaluation of the specimen revealed a high-grade osteoblastic osteosarcoma. After surgery, the patient was seen by oncology specialists to consider adjuvant radiation therapy or chemotherapy or both. Radiation therapy is not known to provide high rates of local control for osteoblastic sarcomas; however, high doses of chemotherapy can be effective.
T1-weighted MRI showing hemorrhagic mass (A); T2-weighted MRI with gadolinium, axial (B) and coronal (C) sections showing an enhancing, predominantly extracranial mass.
At 3 weeks after surgery, the patient had extensive recurrence with intracranial extension of the tumor. He had multiple masses in the right parietal region. The large lesion was $11 \times 9$ cm and smaller lesions were 4 to 2.5 cm. Radiographs and CT of the chest showed no evidence of pulmonary metastases. A bone scan showed a defect only in the right parietal region of the skull. He was admitted to the hospital for chemotherapy. The patient received adriamycin 37.5 mg IV drip daily for three doses, ifosfamide 4625 mg IV drip daily for 3 doses, and mesna 4635 mg IV drip concurrently with ifosfamide for 3 days. After completion of the adriamycin, mesna, and ifosfamide treatment, cisplatin 185 mg IV drip was given. The patient developed significant nausea and vomiting, which was treated with Neupogen 480 mg given daily.

Follow-up evaluation showed good response to chemotherapy treatment. However, the patient refused further follow-up or treatment and his status is unknown.

### Discussion

The rarity of primary osteogenic sarcomas of the skull makes it difficult to arrive at a definitive treatment plan. The common clinical presentation of the tumor is a growing protuberance on the head that may or may not be tender [4]. The tumors are most common in the second and third decade of life [1,7–9], and the cranial vault is affected more than the base of the skull [2,7]. Evaluation methods include plain radiographs, CT, and MRI. An angiogram is useful for determining the blood supply of the tumor and planning the surgical approach. CT scan
allows good detection of tumor calcification and accurate evaluation of intracranial extension of tumors [3,5,9]. Evaluation with MRI does not show calcification of the tumor as precisely as CT but may show the vascular channels and soft-tissue involvement of the tumor more clearly [9]. The presence of bone destruction and mineralization of the tumor are indications of an osteogenic sarcoma, but the differential diagnosis includes osteochondroma and chondrosarcoma [1].

Shinoda et al in 1993 provided a review of primary osteogenic sarcomas of the skull that were reported in the literature from 1945 to 1992 [8]. The authors performed a meta-analysis of 99 cases of primary osteogenic sarcomas to determine clinical features, therapeutic treatment methods, and survival rates. The analysis did not distinguish the histotype of the tumors (such as osteoblastic, fibroblastic, telangiectatic, or mixed), but the osteoblastic variety is the most common [3,6,7]. In this review, the mean age of the patients was 30.0 years and the male to female ratio was 32:28. The authors
were able to analyze data regarding the extent of tumor invasion and the outcome in 35 patients. Of 13 patients with intracranial invasion, only 4 patients survived more than 1 year and none of the patients survived more than 2 years. Of 13 patients without intracranial invasion by the tumor, 10 survived more than 1 year and 5 patients survived more than 2 years. The authors concluded that the prognosis depended mainly on the degree of intracranial involvement at the time of the diagnosis rather than the mode of therapy. It is interesting to note, however, that all of the 13 patients with intracranial invasion by the tumor were reported before 1985. The report did not include an analysis of outcome based on treatment regimens (surgery, chemotherapy, irradiation, or a combination of these methods). It is probable that patients since 1985 have been evaluated and treated differently, making the outcome more favorable in recent years.

Chemotherapy has been found to improve the survival rate of patients with craniofacial sarcomas [7,10,11]. Sundaresan et al described a “neoadjuvant chemotherapy” approach for 8 patients in whom preoperative chemotherapy was used to reduce the bulk of the tumor before surgery [11]. Five patients had undergone previous surgery elsewhere, so the authors had the advantage of knowing the diagnosis before considering their treatment choices. The authors stated that, even with CT evaluation, a diagnosis of osteogenic sarcoma was rarely considered. This neoadjuvant chemotherapy approach resulted in 4 of 6 patients with primary tumors surviving longer than 3 years.

Salvati et al, in 1993, also used a neoadjuvant chemotherapy approach for 5 of 19 patients with skull osteosarcomas [7]. The diagnosis was established preoperatively in the 5 patients by obtaining a bone biopsy. The study analyzed the outcome of patients from two groups, those patients who received chemotherapy (9 patients) and those patients who did not receive chemotherapy (10 patients). The mean survival length was 16 months in patients who did not receive chemotherapy, whereas 5 of 9 patients who received chemotherapy were still alive at 2 years after the diagnosis. Of note, 4 of the 5 patients who received neoadjuvant chemotherapy remained free of illness at the time of the report.

Based on our review of the plain radiographs, CT, and MRI evaluations preoperatively, we expected to find an aggressive type of tumor at the time of surgery. Because of the rarity of osteogenic sarcomas of the skull, however, other diagnoses were considered. The patient in this case report responded well to the initial treatment of chemotherapy. Given that this patient refused further evaluation or treatment, we cannot obtain information that could be helpful for establishing an effective treatment regimen. The report by Salvati et al indicated that they routinely obtained a bone biopsy to provide an accurate diagnosis [7]. This approach combined with chemotherapy (adjuvant and neoadjuvant) and surgery may increase survival [7,11].

REFERENCES


COMMENTARY

Bose presents a rare case of primary osteogenic sarcoma of the skull. In our own clinical experience of over 25,000 cases of central nervous system tumors, only one such case was noted and verified by pathological examination: a 9-year-old boy who presented with a rapidly growing mass in the right frontotemporal region. Surgical resection of the mass, which involved the middle fossa, was performed. However, tumor recurrence one month later interrupted radiotherapy and made chemotherapy impossible. The boy died 2 months after surgical resection. A literature review reveals the