Case Report

Primary Calvarial Osteosarcoma: A Case Report

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Abstract

Osteosarcoma (OS) is a malignant tumor of mesenchymal origin and composed of osteoid tissue or immature bone production. OS of the skull is exceedingly rare and devastating tumors. The overall prevalence of primary craniofacial OS is around 2-9% in published series, while the prevalence of pure cranial localization is 1-2%. It usually develops secondary to preexisting bone diseases or after the radiation therapy. Local recurrence rates are 26-69% and five-year survival rate is 25-37%. Amongst primary craniofacial OS, skull lesions appear have the worst prognosis. In this report, the clinical, radiologic, pathological features, recommended treatment and prognosis of a 26-year-old female patient diagnosed with primary calvarial osteosarcoma has been discussed.

Keywords: Primary cranial osteosarcoma, skull neoplasm, primary tumor

INTRODUCTION

An osteosarcoma (OS) of the skull is rare, particularly as a primary tumor. The overall prevalence of primary cranial OS has been reported to be 2% to 9% in published series; it accounts for 1-2% of overall OS of the skull cases.(1,2) These lesions primarily originate from pericranial or outer calvarial surface and do not show intracranial extension unless differentiated(1,3). OS is extremely uncommon and is more prevalent in the second and third decades of life. Most cases of OS are sporadic(1-6).

OS of the skull is often asymptomatic, present late. The common clinical presentation of the tumor is growing protuberance on the head that may or may not be tender(2,5,7-9). Evaluation methods include plain radiographs, computerized...
tomography (CT) and magnetic resonance imaging (MRI). The general radiographic features are not specific\cite{1,8}. Chondroblastic type OS is also common in the skull, but fibroblastic type OS is uncommon\cite{5,10}.

OS's behavior and treatment has grown almost exponentially over the past few years. Treatment modality of all craniofacial osteosarcoma involves a combination of aggressive surgery with good margins, radiotherapy and multi-agent chemotherapy. Surgery is the mainstay of the therapy. However, because of the anatomy of the head and neck, complete resection may be difficult to achieve surgically without causing a significant functional impairment or cosmetic defect. Complete tumor resection with negative histological margins is associated with long-term survival. Prognosis is poor even in patients who received adequate therapy. The 5-year survival rate for head and neck OS is between 25-37\%. Local recurrence is the major cause of death in skull bone OS\cite{2,3,5,7-13}. We present a case and discuss the diagnostic and therapeutic implications of a primary osteosarcoma of the skull.

**CASE PRESENTATION**

A 26-year-old female patient presented to the Neurosurgery polyclinic with headache for a period of two years and a painless lump over her scalp existed for 3 months. The patient had been tenderness at the site of the lump for about two months before being seen by the neurosurgeon. No abnormality except for a lump in the calvarium was detected on her physical examination. Her personal history was unremarkable without any history of previous radiation therapy or a preexisting disease such as Paget's disease or osteomyelitis. The patient had no family history of cancer. Cranial computerized tomography scan, with and without contrast, showed an expansile, lytic, destructive, extra-axial mass with hazy contour measuring 6x5x4 cm including amorphous calcifications; it was starting from the parasagittal region on the right lateral aspect of the occipital bone and extending to the superior aspect over the surface of the occipital bone (Figure 1). Cranial MRI revealed extra-axial lesions of bone origin with lobulated contour and with hypointense character in the T1A and T2A series starting from the posterior parasagittal region on the right lateral aspect of the occipital bone and extending to the superior aspect over the occipital bone surface. The lesion crossed over the dura, and a mass with an intra-axial extension was detected (Figure 2). She underwent gross total resection of the mass with a negative margin. Postoperative MRI showed gross total removal (Figure 3). The pathological examination, together with clinical history, radiological and histopathological findings, was consistent with "primary osteosarcoma" (Figure 4). After surgery, the patient was seen by oncology specialist to consider adjuvant radiotherapy or chemotherapy or both. But she refused further treatment and follow-up.

The patient, re-admitted seven months later with recently developing lumps in hairy skin and with dizziness. On cranial MRI, post-contrast axial T1A images obtained from infra- and supratentorial levels revealed a contrast uptake consistent with a relapse mass adjacent to the cerebrum, cerebellum and torcula (Figure 5). The patient only could have a subtotal resection performed. Pathological examination confirmed recurrence of high grade osteoblastic OS with invasion into the adjacent dura and cerebral parenchyma. She was admitted to the hospital for chemotherapy and radiotherapy. Since the patient was also pregnant for 22 weeks, chemotherapy was postponed after delivery. Totally 60 Gy with 200 cGy fractions were given to the relapsed area with 6 MV photons with Linac device. She gave birth with section cesarean (c/s) after radiotherapy. No metastatic focus was detected with control thoracic CT, and abdominal ultrasonography. A bone scan
showed a defect only in the right occipital region of the skull. Following the c/s, the patient received four courses of Cisplatin 100 mg/m² (1 day) and Doxorubicine 25 mg/m² (1-3 days). Relapse was detected on cranial MRI during the follow-up, surgery was not considered, therefore second line chemotherapy consisted of Ifosfamid 3000 mg/m² (1-4 days), Mesna 3000 mg/m² (1-4 days) and Etoposide 75 mg/m² (1-2 days) were given for four courses with disease stabilization. She was followed at 3 month intervals. On her MRI after 3 months, no significant change was observed in lobulated fluid collection, whereas an increase in dural contrast and a new intraparenchymal nodular contrast were detected. Re-operation was not considered for the patient and she underwent radiotherapy in another center and her status is unknown.

Figure 1a, 1b: Expansile, lytic, and destructive extraaxial mass is observed on the contrast enhanced CT, which begins from the parasagittal region on the right lateral aspect of the occipital bone and extends to the superior aspect over the surface of the occipital bone.

Figure 2a, 2b: The lesion has hypointense character on the axial plan T2A and T1A MRI. Note the signals confirming the infiltration into the adjacent cerebral parenchyma.
Figure 3: Postoperative CT scan with contrast showing gross total resection of the tumor.

Figure 4: Malignant osteoblasts and osteoid production are observed on the light microscope (Hematoxyline eosine x100).

Figure 5a,5b: On post-contrast axial T1A MRI scan obtained from supratentorial levels in postoperative period, it is seen that the majority of calvarial lesion in the occipital region has been excised and secondarily meningocele has occurred. However, contrast uptake consonant with relapse is seen adjacent to the cerebrum, cerebellum and torcula (arrows).
DISCUSSION

OS is one of the most common bone tumors. OS tends to occur in the long bones of the extremities, whereas only 2% to 9% arise in the craniofacial bones. The vast majority of cranial OSs is located in the zygomatic bone, whereas tumors affecting the skull are rare. OS primarily originates from pericranial or outer calvarial surface and does not show intracranial extension unless differentiated(1-5). Huvos et al. reported that of 1200 patients found to have osteogenic sarcomas over 60 years' time, only 19 cases (1.6%) of calvarial origin were detected(5). Shinoda et al in 1993 provided a review of primary osteogenic sarcomas of the skull. The authors performed a meta-analysis of 19 cases of primary osteogenic sarcomas of the skull(13).

OS appear to have a different epidemiology and may either be primary or secondary. Secondary OSs have been seen more frequently and develop either on the base of preexisting bone diseases such as Paget's disease, fibrous dysplasia and chronic osteomyelitis, or after the radiation therapy(5,7,8,11,12,14). No precipitating factor was identified in our patient.

OS is often asymptomatic and presents late. The most common complaint associated with this type of tumor is a growing protuberance on the head and may or not tender(3,4,7,10,12,13). Likewise, the present case presented with painless lump in the occipital region, developing in a short time.

The general radiographic features are not specific, but may be osteolytic, osteoblastic or mixed. The radiological appearance of OS resembles the sarcomas seen in appendicular skeleton. While evaluating the radiological findings, the age of the patient, the presence of concomitant diseases such as Paget's disease and osteomyelitis, history of previous radiotherapy, invasion into the cerebral parenchyma, being lytic-sclerotic, the presence of calcification, and causing expansion or destruction should be taken into consideration for the diagnosis. CT and MRI are complementary methods in showing tumor spread and in planning surgical intervention. CT scan allows good detection of tumor calcification and accurate evaluation of intracranial extension of tumors. Evaluation with MRI shows the vascular channels and soft-tissue involvement of the tumor more clearly(2,6,11,13,14). In our patient, based on CT and MRI evaluations preoperatively, we expected to find an aggressive type tumor at the time of surgery.

OS is characterized by the presence of malignant osteoblasts that show histological osteoid production and it has osteoblastic, condroblastic and fibroblastic subtypes. Osteoblastic subtypes tumors generally belong to the high-grade tumors (grade 3 or 4), whereas and fibroblastic OSs are usually either grade 1 or 2. A majority of skull bone OSs is of the chondroblastic variety(1,5,8,13). Accurate diagnosis requires correlation of clinical, radiographic, and histologic features as in our case.

Since OS is rarely encountered, it is difficult to diagnose and treat them. Prognosis and treatment are influenced by histological variant, tumor localization and biological behavior(4,5,8,12,14). OS's behaviour and treatment has grown almost exponentially over the past years. The recommended treatment has not been established. Treatment modality of all craniofacial OS involves a combination of aggressive surgery with good margins, radiotherapy and multi-agent chemotherapy. Complete tumor resection with negative histological margins is the most significant factor contributing to a good outcome. Patients with primary OS even after radical surgery, chemotherapy have been found to contribute to the survival in the patients with OS(11). RT is known to provide local control for OSs.
Although there are reports showing RT is beneficial, calvarial osteosarcomas are usually radio-resistant. Highly conformal RT techniques such as intensity-modulated RT and/or proton beam RT are likely to be important in the management of lesions of the skulls, head-and-neck region\(^{[3,4,6,7,12,14]}\). In this present case, additional therapy had been recommended since the mass was gross totally excised during the first surgery but the patient refused adjuvant treatment. However, early relapse was observed. Since the case was pregnant, adjuvant therapy was postponed after the delivery. Both on post-radiotherapy and post-chemotherapy evaluations, the disease failed to improve completely. Recurrence within the first year was consistent with the literature. It was observed during the second operation that intraparenchymal involvement has negatively affected the prognosis.

The prognosis is poor even in patients who have received adequate therapy. Local recurrence rates are 26-69% and five-year survival rate is 25-37%. The majority of the patients die after 12-18 months due to the intracranial and extracranial spread. In such patients, local recurrence is the main situation that determines poor prognosis. Whereas pulmonary metastasis is well known in primary long bone sarcomas or in those developed after radiotherapy, OS does not display such a propensity\(^{[5,7,8-13]}\). In our case local recurrence that affected the prognosis has developed in the 7\(^{th}\) month. Furthermore, no metastatic focus was detected also in the present case during the screenings.

In conclusion, primary calvarial OS is fairly uncommon. A majority of skull bone OS is of the condroblastic variety. CT and MRI are complementary methods in determining the nature of the calvarial lesions. Appropriate radiological approach is important while making a decision for biopsy and surgical therapy regarding the lesion. Accurate diagnosis requires correlation of clinical, radiographic, and histological features. The rarity of OS of the skull makes it difficult to arrive at a definitive treatment plan. With adequate surgical excision with a negative margin, there is defined role for chemotherapy and/or radiation as adjuvant treatment. These treatments may be affective in prolonging survival.

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