Osteosarcoma of the calvarium with intracranial extension- Case Report

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Abstract:
We describe the radiological findings of a primary osteogenic sarcoma of the calvarium in an adult male, confirmed on histopathological examination. The radiological, clinical features and the management of these tumors are discussed.

Keyword:
Calvarium, Osteosarcoma, Skull

Introduction:
Osteosarcoma, a common bone tumor, involves the craniofacial bones rarely.¹ The maxilla and mandible are the most common craniofacial bones affected but this tumor has been reported on the calvarium in fewer than 150 cases.²,³ Thus osteosarcomas of the calvarium are exceedingly rare. We report a case of a giant left frontoparietal osteosarcoma and describe the radiological, clinical features and the management of these tumors.

Case Report:
This 25-year-old male presented with headache and vomiting for six months, right sided hemiparesis for three months and progressive swelling on the left side of the scalp for one month. He had no seizures, loss of consciousness, cognitive decline or memory impairment. There was an ill-defined bony swelling measuring 8 cm in maximum dimension, predominantly over the left parietal region but extending across the midline to the right parietal region. There was no local tenderness or warmth. The skin over the swelling was stretched, red in the center with no breach in its continuity. The patient had spasticity with grade 2 in the right upper and lower limbs. Plain radiographs of the skull (Figure 1) showed a focal lytic and sclerotic lesion in the left parietal region, speculated sunray type of periosteal reaction and associated large soft tissue in the midline and across the parasagittal regions of the parietal regions. MRI brain (Figure 2) showed a large transcalvarial mass measuring about 12 x 9.5 x 1.5 cm, lobulated in nature. The extracranial soft tissue mass involved the scalp and subcutaneous region on either side of the
midline. The involvement in the bone itself was small along with large periosteal reaction. The tumor was isointense to hypointense in T1 weighted images and heterogeneously hyperintense in T2 weighted images. There were certain T2 weighted hypointense areas without calcification representing fibrosis within the tumor. There was remarkable enhancement with Gadolinium with a few non-enhancing areas suggestive of necrosis or cysts. Associated large perilesional edema around the parasagittal part of the tumor was present with a midline shift of 1 cm to the contralateral side. The adjoining superior sagittal sinus was invaded. The adjacent cortical veins, torcula and the medial two thirds of the left transverse sinus were also thrombosed. There was scalloping of the inner table adjoining the lesion with altered signal of the involved bone. There was no restriction in the diffusion weighted sequences. We did a trucut biopsy of the lesion. The histopathology (Figure 3) showed fragments of a cellular tumor composed of cells with moderately pleomorphic nuclei and indistinct cytoplasm separated by narrow acellular eosinophilic seams. There were occasional mitotic figures including atypical forms. A chondroid appearance was seen in focal areas. There was no evidence of necrosis. The tumor cells were strongly positive for vimentin and immunonegative for EMA, S100 protein and cytokeratin. The impression was that of an osteosarcoma. As the patient was not willing for surgery he was advised to take chemotherapy and radiation.

Discussion:
Osteosarcoma is a malignant neoplasm where spindle cells produce osteoid tissue or immature bone. 4 Only about 2% of all osteosarcomas 4 and 11% of the craniofacial osteosarcomas affect the calvarium. 5 The peak incidence of calvarial osteosarcomas is in the third or the fourth decade. 5 and unlike skeletal osteosarcoma, calvarial osteosarcoma tends to affect the mature bone. 4 The etiology of craniofacial osteosarcoma is unknown. Risk factors for their occurrence include exposure to radiation, retinoblastoma, Li-Fraumeni syndrome and Paget's disease, osteochondromatosis, chronic osteomyelitis, myositis ossificans and trauma. 5 Our patient did not have any of the risk factors or familial conditions described above. The diagnosis of osteosarcoma is based on the accurate identification of immature bone or osteoid. 5 Small cell osteosarcoma is characterized by an abundance of small round cells with scarce cytoplasm, and small amounts of bone or osteoid matrix. Giant cell rich osteosarcoma contains multinucleated osteoclast type giant cells along with spindle cells. In the epithelioid type the tumor cells form large cohesive sheets. The less frequent variants of osteosarcoma are malignant fibrous histiocytoma like, parosteal and osteoblastoma like. Other varieties of intramedullary osteosarcoma are telangiectatic osteosarcoma which has characteristic blood-filled cyst like spaces, and well-differentiated intramedullary osteosarcoma. 5 Based on the histopathological features seen in our patient, the differential diagnoses considered were osteosarcoma, chondrosarcoma, chordoma and chordoid meningioma. The immunohistochemical panel for categorization consisted of vimentin, S-100, EMA and cytokeratin. Chondrosarcomas are positive for S-100 while chordomas are positive for S-100 and cytokeratin. 6 Meningiomas are positive for EMA. 6 Osteosarcomas are positive for vimentin, sometimes S-100 and negative for EMA and cytokeratin. 6 The tumor in our patient showed strong
positivity for vimentin and was negative for S-100, EMA and cytokeratin which was suggestive of an osteosarcoma. The most common clinical presentation is that of a localized swelling based on the anatomical location. The mean duration of symptoms is generally less than one year. They are painless or only mildly tender. These patients also present with headache, cranial nerve palsies, exophthalmos or visual impairment. On examination these tumors are firm to hard in consistency, usually non tender and appear fixed to the underlying bone. The skin over the tumor is stretched but generally not adherent to it. They are more common in the skull vault than in the skull base. Serum Alkaline phosphatase and lactate dehydrogenase may be elevated in 30-40% of patients with osteosarcoma.

Our patient had presented with features of raised intracranial pressure and right hemiparesis. His scalp swelling however was painless but rapidly increasing in size. The general radiological features are not specific. The tumor may be osteolytic, osteoblastic or mixed. The bony changes seen on the plain skull x-rays may show a 'sun ray' appearance or Codman triangle. CT scans can be used to look for calcification within the tumor and assess the extent of intracranial extension. New bone formation within soft tissues and the characteristic bone matrix are strongly suggestive of osteosarcoma. On MRI's these tumors are hypointense in T1 weighted images. On T2 weighted images they are hypointense if the tumor is mineralized and hyperintense if the tumor is non mineralized and has a soft tissue component. However, the differential diagnosis of osteochondroma and chondrosarcoma have to also be considered. Transcalvarial masses include meningioma, hemangiopericytoma, metastases and lymphoma other than the above mentioned differentials.

The differentiating features of an osteosarcoma from meningioma and hemangiopericytoma are the periosteal reaction and the bone changes; meningioma usually causes expansion of the diploic space, hemangiopericytoma shows intense neovascularity. Expansile metastases from primary such as renal and GI tract have been reported. Lymphoma of the skull is known to have significant soft tissue replacement of the bone, is restricted on DWI but does not cause periosteal reaction. The treatment protocol for management of primary calvarial osteosarcoma has not been standardized due to the rarity of the disease. According to Federman et al the most important prognostic factors for the disease are metastatic disease status, the ability to completely resect all sites of tumor, and histological response to chemotherapy. Osteosarcomas are treated with surgical resection followed by multidrug chemotherapy with doxorubicin, cisplatin, methotrexate and ifosfamide. Preoperative chemotherapy makes conservative surgical resections possible by causing tumor shrinkage through necrosis. Kassir et al have reported better survival in patients with osteosarcoma of the maxilla and mandible as compared to those with the tumor in other parts of the cranium probably due to the fact that the former could be more radically excised. Gross total excision involves excision of the involved dura as well. In cases where gross total resection cannot be achieved, we need to add on adjuvant chemotherapy and radiation to maximize the overall survival.
Radiation therapy is reserved for cases where the tumor is not resectable, or for metastatic tumors. Guadagnolo et al published a retrospective study of 119 patients with craniofacial osteosarcoma who underwent macroscopic total resection between 1960 and 2007. They found that radiation therapy significantly improved local control, disease-specific survival, and overall survival in patients with positive or uncertain resection margins after surgery. However, for patients with negative resection margins, radiotherapy did not improve survival.

According to Shinoda et al, of 13 patients with intracranial invasion between 1945 and 1992, only 4 patients survived more than 1 year and none of the patients survived more than 2 years. Our patient had a large intracranial component to the tumor. Metastasis from craniofacial osteosarcomas is relatively uncommon and occurs in the lungs and brain. Metastatic spread of tumor to the brain generally occurs after multiple recurrences and has a poor prognosis. Despite aggressive surgery, chemotherapy and radiation, about 30-40% of patients with localized osteosarcoma will develop recurrence. Patients with solitary pulmonary metastasis should have the lesion removed. Salvage adjuvant chemotherapy using ifosfamide with or without etoposide may be added in these patients. The 3-year overall, disease-specific, and recurrence-free survival rate for calvarial osteosarcoma was 73%. Conclusion: To conclude, complete surgical excision of the tumor continues to remain the mainstay of treatment in patients with calvarial osteosarcoma. Negative surgical margins are the most important predictors of overall and disease specific survival. Tumors measuring more than 4 cm in greatest dimension and positive margins of surgical excision were found to be associated significantly with local recurrence.

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Fig 1: X-ray skull lateral and AP views showing a lytic and sclerotic lesion in the midline and left parasagittal region of the parietal bone associated with sun-ray spiculated periosseal reaction and soft tissue mass.

Figure 3: Spindle cell tumor osteoid matrix with focal chondroid appearance. The tumor cells showing strong positivity for vimentin, in the second image. (Courtesy Dr. Geetha Chacko, Department of Neuropathology, Vellore)

Fig 2: Brain MRI images (a) T2w images showing a heterogeneously hyperintense transcalvarial mass with hypointense areas with mass effect on the regional brain, compression of the left lateral ventricle, midline shift to the right (b,c) T1w post Gadolinium coronal and sagittal scans showing intense enhancement of the scalp mass, focal involvement of the bone with large intracranial component also showing intensely enhancing and non enhancing areas within. Invasion of the superior sagittal sinus is also seen.