OSTEOSARCOMA OF SKULL WITH METASTASIS IN DURAL VENOUS SINUSES: A COMMON TUMOR AT RARE LOCATION

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ABSTRACT

Osteosarcoma is an aggressive and most common primary bone tumor but rarely arise in skull. Most of reported cases are of high grade and without intracranial extension. We are presenting a case of a young male with osteosarcoma arising from skull with intracranial extension as well.

Introduction

Osteosarcoma is aggressive neoplasm composed of bone forming spindle cells.[1] It is the most common primary bone malignancy and is more common in males than females.[1,2] Osteosarcomas commonly occur in metaphyseal regions of long bones and rarely arise in skull particularly after radiotherapy, chemotherapy or associated with pre-existing disease like Paget's disease, fibrous dysplasia, chronic osteomyelitis.[1] Its incidence in skull is 1-2%.[1] These lesions arise from outer calvarial surface without intracranial extension unless differentiated.[3] Most of skull osteosarcomas are high grade tumors but they usually present late.[2,3]

Case Report

A 16 years old boy presented in emergency department with history of seizures for the first time. On clinical examination, there was no neurological deficit. Rest of the examination was unremarkable apart from incidental finding of a 4 x 5 cm rubbery hard non tender lump in left frontal region. The patient had not noticed it before. There was no history of trauma. His previous history was unremarkable without any history of radiotherapy, pre-existing disease like chronic osteomyelitis. The patient had no family history of cancer. On laboratory investigations, alkaline phosphatase was high.

CT scan was performed which revealed localized permeative destruction of left frontal bone in parasagittal location with associated soft tissue mass measuring 4 X 5 cm. This soft tissue was extra-axial with amorphous calcification in it. Underlying brain parenchyma was unremarkable apart from mild edema. Contrast enhanced MRI showed left frontal bone destruction with enhancing extra axial soft tissue mass. This was adherent to superior sagittal sinus and had adjacent meningeal enhancement. A small extension into left frontal lobe was also noted with adjacent vasogenic edema. This was hypointense on DWI and ADC mapping, likely due to internal calcification. Subsequent radionuclide bone scan showed intense uptake in this region with no evidence of remote disease.

Based on imaging and laboratory findings, preliminary diagnosis of osteosarcoma was made. CT of chest was negative for pulmonary secondaries. Case was

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discussed in multidisciplinary meeting and decision was made for biopsy and possible subsequent resection. Biopsy confirmed radiological diagnosis of osteosarcoma.

Radical excision of this osteosarcoma was done. Histopathological diagnosis of resected tumor was high grade osteosarcoma. Resection margins were clear. On microscopic examination, there was meningeal and brain parenchymal invasion.

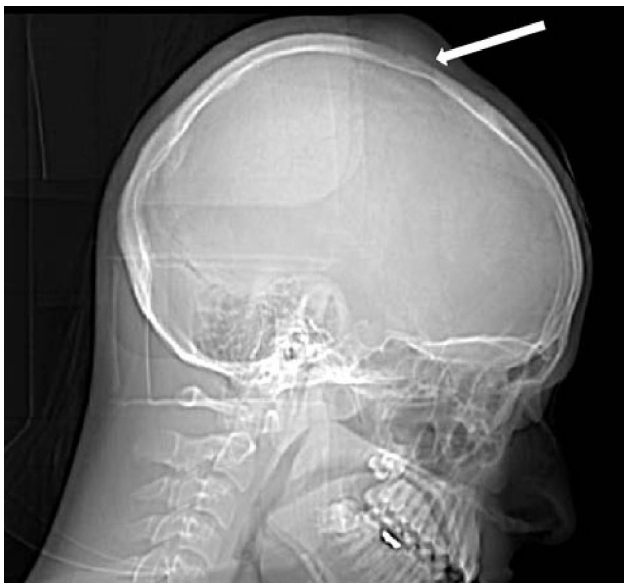

Figure 1: CT Scanogram.Ill defined lytic lesion (arrow) in skull vault with soft tissue component.

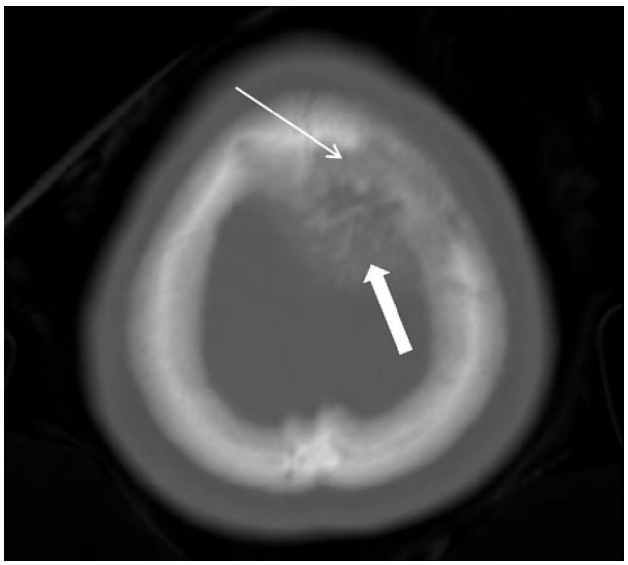

Figure 2: CT scan brain, bone window.Permeative lytic lesion in left frontal bone (thin arrow).Associated soft tissue mass and internal calcification (thick arrow).

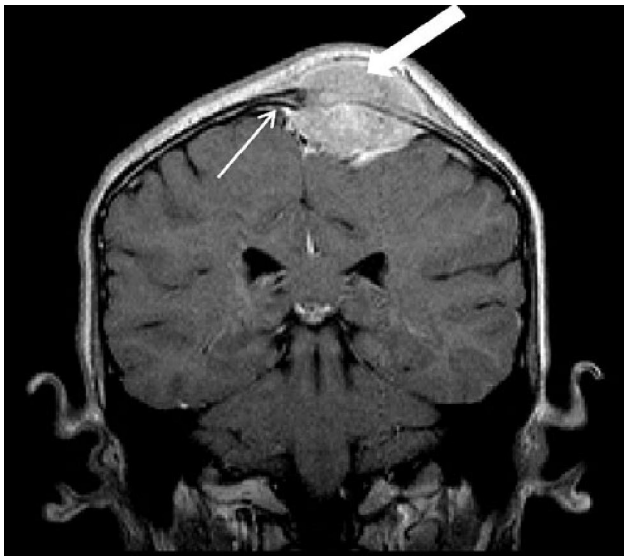

Figure 3: Coronal post contrast T1W image. Destructive lesion in left frontal bone with large viable soft tissue component (thick arrow) and meningeal involvement (thin arrow).

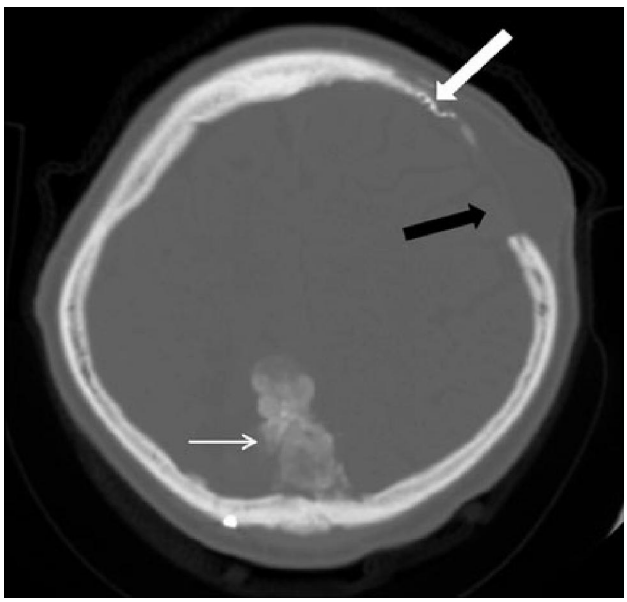

Figure 4: Recurrent osteosarcoma in superior sinus (thin arrow) and surgical site (thick black arrow) with mesh reconstruction (thick white arrow).

Patient was started on chemotherapy. 7 months after surgery, patient again presented with left hemiparesis. CT scan detected lumpy calcified soft tissues along falx and posterior superior sagittal sinus. On biopsy this turned out to be metastatic osteosarcoma. This rapidly progressed to involve torculaherophili, right transverse sinuses, sigmoid sinus extending into proximal internal jugular vein. Luckily patient so far has no complication of sinus thrombosis.

Discussion

Osteosarcoma is most common primary bone tumor (35%) followed by chondrosarcoma (25%) and Ewing's sarcoma (16%).[5] 80% of osteosarcomas occur between 10-30 years of age while 10% of cases are old in 60-70 years of age.[2,5] Most common site for primary osteosarcoma is metaphysis of long bones of arms and legs while 10 % cases are located in flat bones of pelvis, ribs, sternum and clavicle. 5-6% arise in craniofacial bones with vast majority located in zygomatic bones.[1,6] Cranial vault is exceedingly rare site for primary osteosarcoma i.e. 1-2 %.[1] Caron et al. reported only 37 cases of calvarial osteosarcoma in their 37 years study.[7]

Etiology of osteosarcoma is not well known. In later age group, the calvarial osteosarcomas are more likely to be secondary commonly related to Paget's disease.[8] Other well known risk factors include prior radiotherapy, multiple hereditary exostosis, fibrous dysplasia, chronic osteomyelitis, bone infarcts, myositis ossificans and sites of metallic implants.[9,10] Despite the fact that most of calvarial osteosarcomas are high grade, they usually present late often after history of trauma.[2] Common presentation is localized head swelling which is frequently tender on palpation.[1,2] Calvarial osteosarcoma can metastasize via Batson's venous plexus or marrow sinusoids or lymphatic spread.[4]

Initial radiological investigations include CT scan and MRI. Usual sunburst pattern may not be visible on in flat bones but these modalities are good in detection of tumor calcification and intracranial extension as in our case. Macrometastasis are present in 15-20% of cases at the time of presentation and chest CT scan is mandatory to rule out pulmonary metastasis in preoperative workup.[2] Radionuclide bone scan is used to rule out unsuspected polyostotic disease.

Treatment modalities for craniofacial osteosarcomas include wide surgery with clear margins, radiotherapy and chemotherapy.[3] Negative tumor margins on surgery are the most significant factor for good outcome.[1] Radiotherapy is known to be effective in peripheral osteosarcomas but calvarial osteosarcomas are usually radioresistant and same is the case with chemotheraphy.[1,3]

Prognosis is poor even when surgery is adequate, because of early intra and extra cranial recurrence. 5 year survival rate is 25-37% with high local recurrence rates of 26-69%3. Local recurrence is the major cause of death in calvarial osteosarcomas.

Conclusion

Primary osteosarcoma of skull is one of the rare tumors. Awareness of this fact can help in early diagnosis. Unfortunately its aggressive behavior has resulted in early recurrence and grave prognosis despite aggressive management strategies.

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