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CASE REPORT

A Rare Case of Occipital Region Bony Calvarial Primary Osteosarcoma

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ABSTRACT

Rationale: Primary osteosarcoma of calvarium is considered one of the rarest neoplasm of skull and accounts for 1 to 3% of all skull neoplasms. It is an aggressive bone neoplasm and is characterized by presence of spindle cell leading to a generally poor prognosis.

Case report: A female patient aged appox 55-years came to our neurosurgery outpatient department having complain of progressively enlarging mass in occipital region of her skull. MRI revealed a lytic, expansile soft tissue lesion involving the occipital bone which was excised surgically and tissue was sent for histopathology which confirmed the final outcome as osteosarcoma of skull. Decompression surgery was done to alleviate symptoms.

Conclusion: Primary calvarial osteosarcoma is considered as one of the rarest neoplasm of skull, emphasizing need for prompt identification and accurate diagnosis to improve patient outcome. Clinician must be vigilant in recognizing this condition to facilitate timely intervention. A comprehensive treatment approach involving surgical resection followed by chemoradiation has been shown to yield optimal results.

Keywords: *Decompression Surgery, Occipital Region Bony Calvarial, Spindle Cells*

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INTRODUCTION

Bony calvarial primary osteosarcoma is a prevalent bone malignancy which has predilection for extrimities.[1] However it may also occur in region of head and neck which is responsible for 5–15% of all osteosarcoma & 1-5% of neoplasms occurring in head and neck region. Primary bony calvarial osteosarcoma typically manifest in adults between age group of 30–40 years. It often occurs as consequence of prior chemotherapy, radiotherapy or other predisposing conditions such as Paget's or Fibrous dysplasia. Calvarium and base of skull are common occurring sites.[3,4] Primary osteosarcoma of skull is less prevalent entity. This study highlights the case of primary calvarial osteosarcoma with associated classical presentation and positive response after proper management which includes complete removal by surgery which is followed by chemoradiotherapy which increases patient survival.

There are various histopathology based categories of osteosarcoma which includes mainly Osteoblastic, Chondroblastic, epitheloid, small cell, telangiectetic, fibrohistiocytic like & giant cell rich type variants.

CLINICAL DETAILS

A female patient aged appox 55-years came with complain of progressively enlarging mass in occipital region of her skull, accompanied by moderate to severe pain for $2^{1/2}$–3 Months. Her medical history was unremarkable, with no prior radiation exposure or family history of similar conditions. On external examination, it presents as painful, soft consistency space occupying lesion in occipital area of bony calvarium having no sensorineural deficiency. All necessary diagnostic lab investigations including routine blood examination were normal. Radiological investigations which include CT & MRI demonstrated a lytic lesion with surrounding soft tissue mass, measuring appox 82 X 72 X 48 mm, involving occipital bone. The lesion was displaying hyperintensity on both T1 & T2 MRI sequences with permeative bony lesions & cystic regions suggestive of necrosis. The lesion abutted the bilateral cerebral hemispheres and occipital lobes, with loss of intervening fat planes and oedema in the adjacent cerebral hemispheres. A differential diagnosis of metastasis and Paget's disease with secondary sarcoma was considered based on radiological findings. The patient underwent an occipital craniotomy, during which surgical removal of the mass lesion done from underlying duramater with a margin of healthy tissue surrounding it and surgical repair of cranium done with mesh made up of Titanium. Postoperative CT scans showed no residual tumour.

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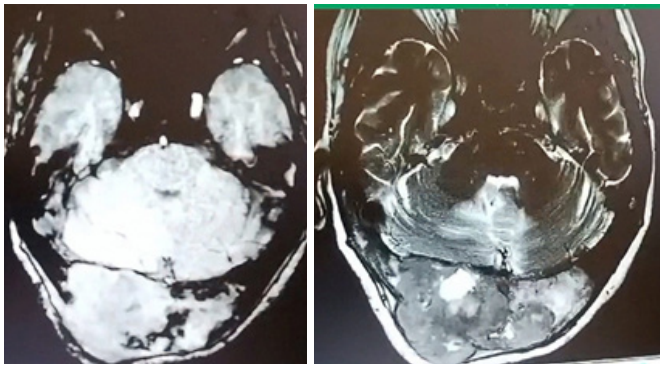

Figure 1: Presurgical MRI revealed well defined extracerebral SOL (Space occupying lesion)~ appox 8.2X7.8X4.2 cm in Occipital region which was hyperintense on T1-weighted (1A) and T2-weighted (1B) series.

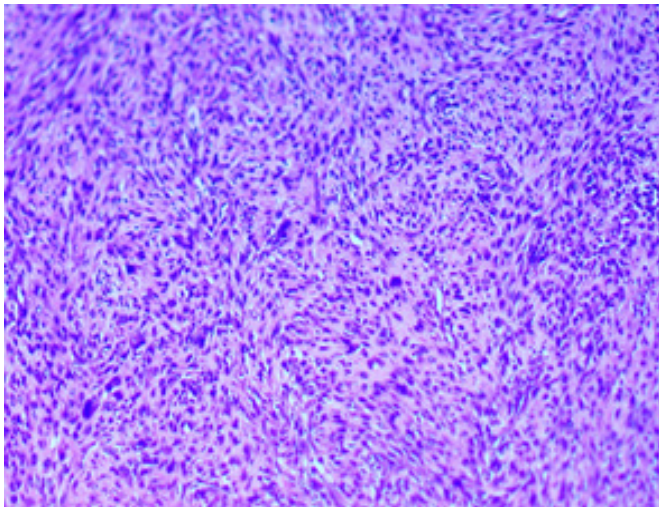

Figure 2: Photomicrograph showing proliferation of fascicles of mixed spindle and multinucleate giant cells (H & EX 100).

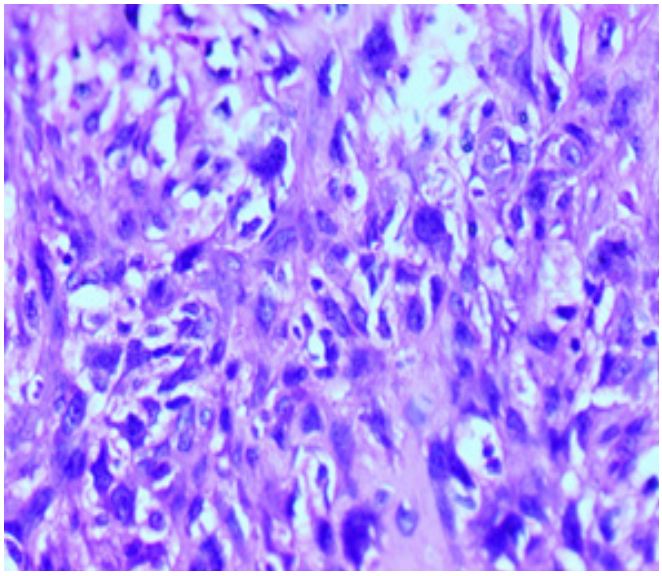

Figure 3: Photomicrograph showing multinuclear giant cells (H & EX 400).

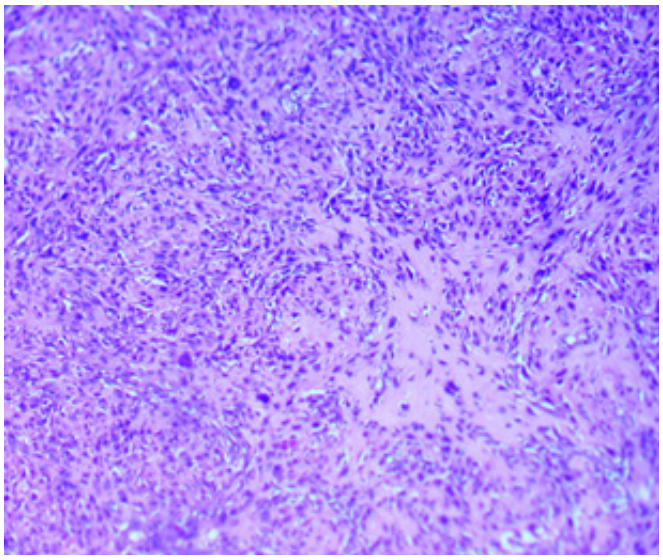

Figure 4: Photomicrograph showing plump cells with prominent eosinophilic extracellular material (osteoid) (H & EX 100).

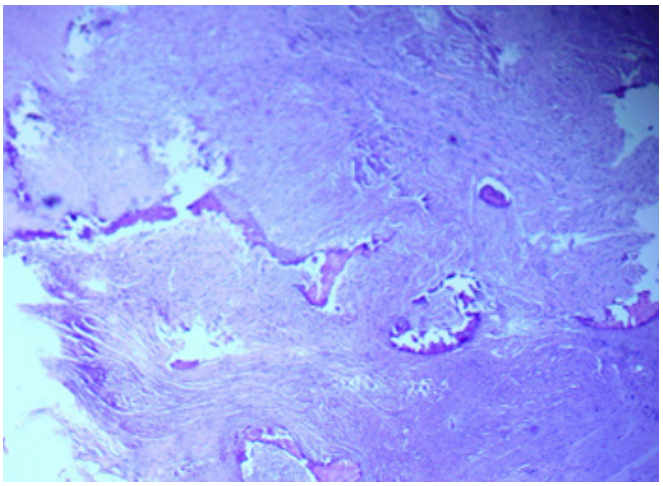

Figure 5: Photomicrograph showing dead and necrotic bone(H & E-40).

Preoperative MRI revealed claearly outlined extracerebral SOL (Space occupying lesion) which measured appox 8.2 X 7.8 X 4.2 cm in occipital region. The lesion exhibited hyperintensity on T1(1A) and T2 (1B) W.I. sequences. Excised tissue was submitted to our laboratory for histopathological evaluation. Upon gross examination, multiple grayish white soft tissue fragments measuring 5 X 3.5 X 2.5 cm were observed. The tissue samples underwent standard histopathological processing. Standard staining protocols followed using Hematoxylin & Eosin (H&E) staining. Microscopy revealed a cellular tumor comprising spindle cells fascicles which are arranged in cartwheel like configuration exhibiting hyperchromasia, moderate pleomorphism, prominent nucleolar structures with very less amount of cytoplasm (Figure 2). Furthermore, mitotic count of 1 per high power field (HPF) & contained multinucleate giant cells (Figure 3). Additionally, focal areas of delicate, lace like osteoid material were observed (Figure 4).

The background of the tumor exhibited desmoplasia and mixed chronic and acute inflammatory infiltrates. Based on these findings, a histopathological diagnosis of high grade sarcoma, with ostesarcoma being the favoured diagnosis was rendered.

DISCUSSION

Primary osteosarcoma of calvarium is one of the rarest neoplasm which accounts for appox <2% of all skull neoplasms. Causative factor of skull osteosarcoma is uncertain, however several risk factors have been identified, including radiological exposure, osteitis deformans, Sarcoma, breast, leukemia & adrenal gland (SBLA) syndrome & childhood retinal neoplasm.[10,12-14] Notably osteitis deformans induced osteosarcoma has predilection for skull & are more prevalent as compared to primary bony calvarial osteosarcoma.[6,12] Additional potential risk factors[12] include osteofibrous dysplasia, multiple hereditary exostosis (MHE), recurrent bone infections, calcific myonecrosis and injury. Primary bony calvarial osteosarcoma occurrence is particularly having very less prevalent in pediatric population as reported by Kirky *et al*. in 2011.[7] According to Mars *et al,* bony osteosarcoma of facial region is one of the least common tumor seen in pediatric population. A comprehensive review by Huvos in 1979 idebtified more than 400 issued cerebrofacial region cases in a span of appox 60-years (1921-1981). Notably, patients with newly diagnosed osteosarcoma occurs in young populations as compared to secondary osteosarcoma. In contrast, this patient was diagnosed at age of appox 55-years. A study by Smith *et al*. analysing appox 500 cases of calvarial and cervical region osteosarcoma found similarities between various categories of osteosarcoma occuring in calvarial and maxillofacial region. However, high grade tumors were more commonly found in the skull and other craniofacial bones.

Patients with primary bony calvarial osteosarcoma generally having a history of progressive increasing swelling, often accompanied by minimal or no pain.[6,12.15] Clinical features of skull osteosarcoma patients are pain in craniofacial region, dysfunction of cranial nerves, ophthalmological problems, bulging eyes, Intracranial hypertension.[6,16] In contrast to osteosarcoma of extrimities, which frequently metastasize to cerebropulmonary region, metastatic spread of bony calvarial osteposarcoma is relatively rare but local recurrence is a common issue in skull osteosarcoma cases.[17]

Osteosarcoma are generally composed of fusiform or spindle shaped cells accompanied by excess disorganized primitive bony tissues.[12] These tumors can be categorized into 3 main subtypes-Osteoblastic, Chondrblastic and fibroblastic.[18] Less common variants of osteosarcoma comprises of small cell, periosteal, parosteal & telangiectatic osteosarcoma.[9]

Osteosarcoma severity ranges from low to high risk and depends primarily on degree of abnormal cell morphology and tissue architecture.[19] However, morphologic chracterstics of osteosarcoma sometimes not necessarily correspond to prognosis.[4,13,14] Diagnosis can be confirmed through immunohistochemical staining,with SATB2 and MDM2 being useful IHC markers.

CT scan with bone windows are crucial for diagnois, typically revealing bony outgrowth having lytic areas with alteration in shape of bone.[4,6,20] Contrast enhanced MRI plays a key role in assessing infiltration of soft tissue. On MRI,high grade osteosarcoma usually appear isointense on T1W.I . and hypointense on T2 W.I. & also shows homogenous enhancement having sharply defined outlines on contrast enhanced images.[21] To assess the metastatic disease, thoracic region tomography and radioscintigraphy are the other options available. These tests help identify potential lung and skeletal metastasis respectively.[22,23]

Surgical removal of the primary calvarial osteosarcoma still remains mainstream treatment option.[24] Several studies confirmed the improved prognosis with complete excision of the neoplasm, also involving some part of surrounding healthy tissue.[9,10,12] However chemotherapy also plays a key role in inceasing postsurgery survival rate of patient due to complexity of skull anatomy.[25] Radiotherapy has a pivotal role in cases of incomplete or indeterminate resection. Conversely, radiation therapy does not appear to provide additional survival benefits in patients who have undergone surgery with removal of tumor having margin of healthy tissue surrounding it.[26]

CONCLUSION

Skull osteosarcoma is an uncommon condition that poses diagnostic and therapeutic chanllanges. As a result, possibility of skull osteosarcoma always kept in consideration when evaluating skull lesions. A multidisciplinary approach, involving complete surgical excision and adjuvant chemoradiation, has shown promise as a primary treatment strategy for this rare disease.

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