Scholars Journal of Applied Medical Sciences (SJAMS)

Sch. J. App. Med. Sci., 2013; 1(6):981-984

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Case Report

A Rare Case Report of Primary Osteosarcoma of the Left Parietal Bone Barhate Anand^{1*}, Athawale Kedar², Lakhkar Dilip³

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Abstract: The common sites for primary osteosarcoma include arm, legs and pelvis. Male are more affected than female. Primary osteosarcoma of skull is rare 1- 2% of all osteosarcomas. Osteosarcoma may present as solitary or multifocal lesion. Most of them are high grade intramedullary, about 5% are lower grade and others are secondary to radiation therapy or Paget's disease. We here present a rare case of skull bone osteogenic sarcoma in 70 year female who presented with swelling left parietal region and extension of the soft tissue in left extraconal compartment. **Keywords:** Osteogenic sarcoma, parietal bone, orbit, infratemporal fossa, extraconal lesion

INTRODUCTION

Primary osteosarcoma of the skull represents 1-2 % of all osteosarcoma of the bones. Secondary osteosarcomas of the skull are secondary to Paget's disease, fibrous dysplasia, chronic osteomyelitis and post radiation therapy. Clinically they present as protuberance in head with tender soft tissue swelling. CT and MRI are useful in evaluation of intracranial and extra cranial extent of the lesion and involvement of neurovascular bundles. The overall prognosis of the disease correlates with histological type of tumour and the intracranial extent of it. Treatment modalities include surgery, radiotherapy or chemotherapy.

CASE REPORT

A 70 yr female came to the ophthalmology department with complaints of sudden loss of vision in the left eye .She was congenitally deaf and mute. On local examination she was found to have decreased vision of hand movement, chemosis, and swelling over left periorbital region. The swelling was firm in consistency and tender. There was no local rise of temperature. The overlying skin was intact.

Scan showed extraconal soft tissue laterally displacing the eyeball on medial aspect. The lateral rectus appeared separate. No invasion of the eyeball was noted. Incidentally noted was soft tissue in the left temporal region which showed increased vascularity.

CT scan was done for further evaluation of swelling over the parietal bone. Contrast enhanced CT scan showed erosive lesion in the left parietal bone. The lesion had wide zone of transition. The margins were not well appreciated. The cranial part showed linear onion peel like periosteal reaction. There was associated well defined soft tissue component in the intracranial as well as extra cranial compartments. The intracranial component of the soft tissue was seen to cause mass effect over the underlying cerebral hemisphere with associated cerebral edema. The soft tissue was also seen to extend in the left orbit as well as in left infra temporal fossa. The soft tissue in the orbit was extraconal and was displacing the eyeball inferomedially. No obvious invasion of the optic nerve or sclera was seen .The soft tissue showed moderate heterogeneous contrast enhancement.

The CT findings indicated neoplastic etiology. As patient was 70 years a strong possibility of osteosarcoma was considered. It was proved on biopsy. Patient was referred to radiotherapy department for further management.



Fig. 1: A 70 years female with swelling over left parietal region with proptosis of left eyeball

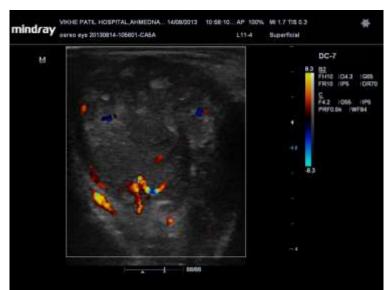


Fig. 2: Doppler study of the mixed echogenic solid mass in parietal region showing increased vascularity

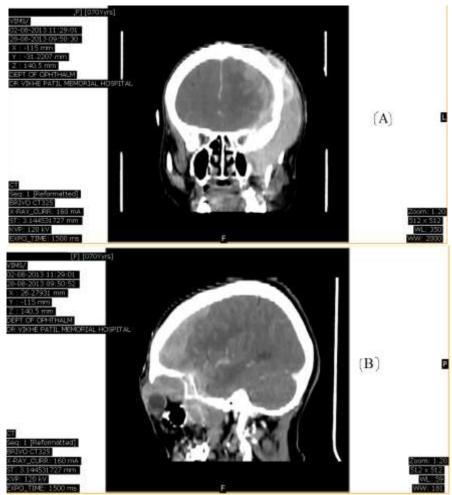


Fig. 3: Contrast enhanced CT images: (A) Coronal image – Soft tissue mass in extracalvarial region and in extradural space. (B) Sagittal image shows mass displacing left globe medially and inferiorly. The mass is seen to extend in left infra temporal fossa.

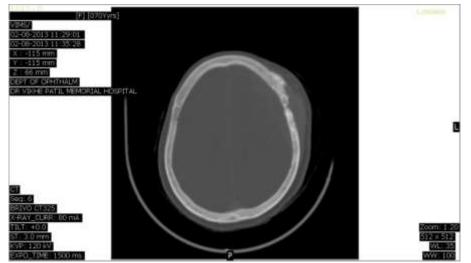


Fig. 4: CT in bone window showing erosive lesion involving left parietal bone with associated soft tissue swelling

DISCUSSION

The common sites for primary osteosarcoma include arm, legs and pelvis. Male are more affected than female. Primary osteosarcoma of skull is rare 2% of all osteosarcomas [1].

Primary osteosarcomas arise from the metaphysis of the long bones. About 10 % are located in the flat bones of pelvis, ribs, sternum and clavicle [2, 3]. There are many corresponding studies that reported the rarity of de novo osteosarcoma of the skull. For instance, Nora *et al.* [4] reported that 21 of 1,000 osteosarcoma cases had tumor in the skull, and only 14 out of 21 cases (1.4%) were de novo tumor; Huvos *et al.* [5] reported that only 10 out of 1,200 osteosarcoma cases (0.8%) over a 60 year period were de novo osteosarcoma of the skull.

The etiology of osteosarcoma is not well known. Various association of the disease are seen with genetic mutations causing changes in osteoblasts [6]. Risk factors include prior radiation therapy, paget's disease and other lesions like osteochondroma, enchondroma, multiple hereditary exostoses, fibrous dysplasia, chronic osteomyelitis, bone infarcts and sites of metallic implants [7].

Clinically osteosarcoma presents as localized pain with swelling. The important finding is a soft tissue mass, which is mostly large and tender [1].

Pericranial or outer calvarial surface are the main site of origin of these tumors and they show intracranial extension when differentiated [5, 8, 9]. The radiographic feature of these lesion show osteolytic, osteoblastic or mixed pattern. Osteosarcoma originating from such a rare site poses a diagnostic challenge to the radiologist, pathologist, and the surgeons [10]. The typical 'sunburst' radiological pattern observed in the osteosarcoma of the long bones may not be evident in the osteosarcoma of the flat bones.

CT and MRI scans allow good detection of tumour calcification, vascularity and accurate evaluation of intracranial extension of tumours [11]. CT scan gives accurate evaluation of intra and extra cranial extension of the tumour. CT scans are best suited to evaluate the bony involvement and chest for metastatic deposits, (osteosarcoma is seen in lungs in 80% of cases). MRI is superior to CT for defining soft tissue extension, vascular channels and nerve involvement.

Histologically most of the skull osteosarcoma is of chondroblastic type [5, 8, 12]. The diagnosis of this disease requires correlation of clinical, radiographic and histologic feature as it was done in our case [13].

The 5-year survival rate for head and neck osteosarcoma is between 25-37%. Local recurrence is the major cause of death in skull bone osteosarcoma [13].

CONCLUSION

We report this case of osteosarcoma of skull due to its rarity and presentation. Imaging modalities like CT and MRI play an important role in diagnosing the disease and its extension. Biopsy and histological confirmation is needed for suspected sites of metastatic disease. Chemotherapy can successfully eradicate primary deposits if initiated at a time when disease burden is low. The prognosis depended mainly on the degree of intracranial involvement at the time of the diagnosis is rather than the mode of therapy.With adequate surgical treatment there is role of chemotherapy and radiotherapy as adjuvant therapies.

ACKNOWLEDGEMENT

Authors acknowledge the great help received from the scholars whose articles cited and included in references of this manuscript. The authors are also grateful to authors / editors / publishers of all those articles, journals and books from where the literature for this article has been reviewed and discussed. Authors are grateful to editorial board members and team of reviewers who have helped to bring quality to this manuscript.

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