PRIMARY OSTEOSARCOMA OF THE CALVARIA

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ABSTRACT

Two cases of calvarial osteosarcoma was presented using images of plain films and CT scan. The first case was a 22-year-old female patient who had bony density mass of osteoblastic osteosarcoma at left frontal bone. The involved bone showed a mixed osteolytic and permeative lesions . The second case was a 4-year-old boy who had bony density mass of chondroblastic osteosarcoma at right temporal region. The involved bone also showed a mixed osteolytic and permeative lesions.

INTRODUCTION

Primary osteogenic sarcoma of the skull is rare. A review of the records of more than 1,200 osteogenic sarcoma patients over a 60-year period discovered 19 cases(1.6%) arising in the skull.[1] Ten cases were primary de novo tumors, while six cases were superimposed on Paget disease, two occurred as a complication of previous irradiation, and one arose in association with fibrous dysplasia. The mean patient age was 26 years in the group with the primary neoplasms. Thirteen neoplasms involved the calvarium, while the other six occurred in the skull base. The spectrum of the images by radiologic examinations include purely osteolytic lesions, sclerotic lesions and permeative destructive lesions.

The primary calvarial osteogenic sarcoma in our institution is also unusual and two cases of this condition are presented.

CASE REPORTS

Case 1

A 22-years-old female patient from Srisakes province (north-eastern part of Thailand) presented to the plastic and maxillofacial division due to the palpable left frontal mass for 3 months. The mass was a hard , enlarging and non-tender one. No other symptoms were complained and the physical examination otherwise was normal.

Plain film of the skull in PA and lateral views (Fig. 1) showed a round shape osteoblastic area of left frontal bone. In lateral skull projection (Fig.1), and bone window of the CT scan (Fig.2) revealed that the destruction of the all table- layers of the frontal bone was obvious. The destruction was composed of osteolytic and permeative areas (Fig.2,3). Mild expansion of the medulla of the frontal bone was observed (Fig.3). Bony density occupied most area of the tumor was noted (Fig.4). Dense tumor matrix and tumor bone was probably responsible for this density (Fig. 5). The subgaleal tumor extension contains no tumor matrix (Fig. 6).

At operation, the 4.5 cm- diameter soft tissue mass at the outer cortex of left frontal bone was found and was biopsied. The pathological diagnosis was osteoblastic osteosarcoma, moderately differentiated. The patient received chemotherapy and was in the process of follow–up.

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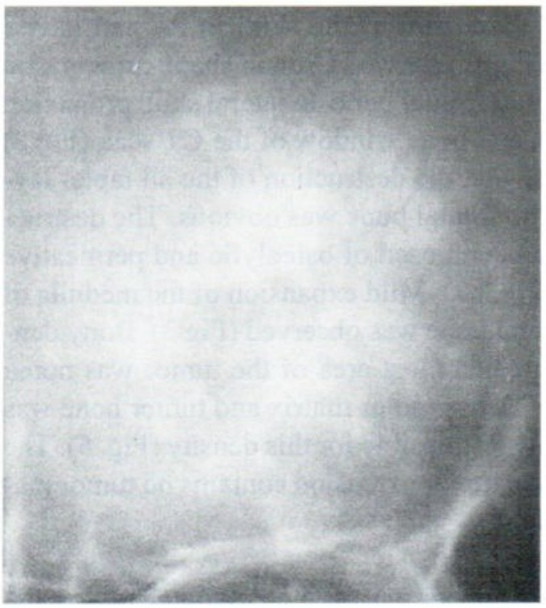
Case 2

A 4-year -old boy, from Pathumthani province (central part of Thailand) , had a lump at right temporal area for 3 years. The lump enlarged rapidly for 5 months. He had pain around the lump and had ear pain for 3 weeks. He noted limitation of the temporomandibular joint movement. On physical examination, a firm and fixed 12 cm - diameter mass was noted at right temporal fossa , displacing right ear laterally .The mass protruded into the external auditory canal. Limitation of the mouth opening and deviation of the jaw to the left was observed.

PA and lateral plain films of the skull revealed an ill defined border dense area in the right temporal bone with irregular outline of the tables (Fig.7). CT scan showed a mass at and around the right temporal bone. The subgaleal part of the mass had only minimal calcification (Fig.8). The intracranial part of the mass contained much of the bony density area(Fig.9) which was due to tumor bone and matrix. Bone window revealed a mixed permeative lesion and an expanding osteolytic areas with sclerotic rim at the right temporal bone (Fig. 10).

At operation the tumor involving posterior part of right temporal bone, auditory canal, infratemporal fossa, lateral orbital wall, zygomatic arch, ramus of the mandible and the temporal lobe was found. Accidental tear of the intracranial lateral sinus and massive bleeding(4000 cc) was encountered. Cardiac arrest occurred in the operative room and finally the patient passed away .At pathology, the tumor was found to be high grade, chondroblastic osteosarcoma .



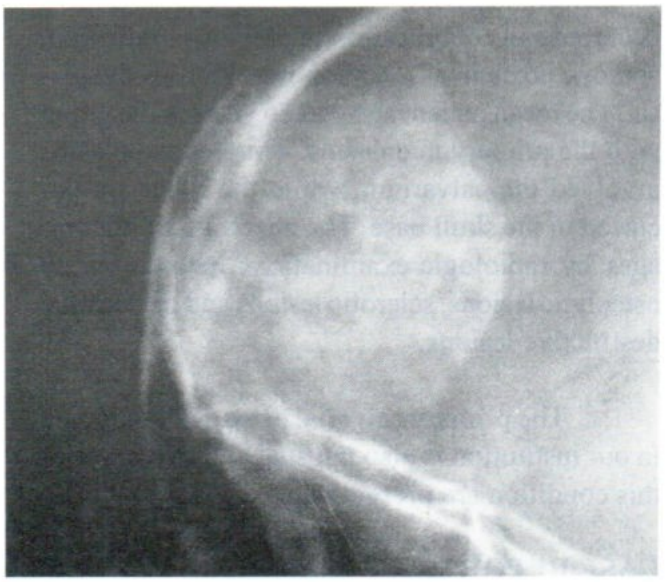


Fig. 1. Case 1. Plain film of the skull in PA and lateral views showed a round shape osteoblastic area of left frontal bone. All layers bony destruction was shown in lateral projection.

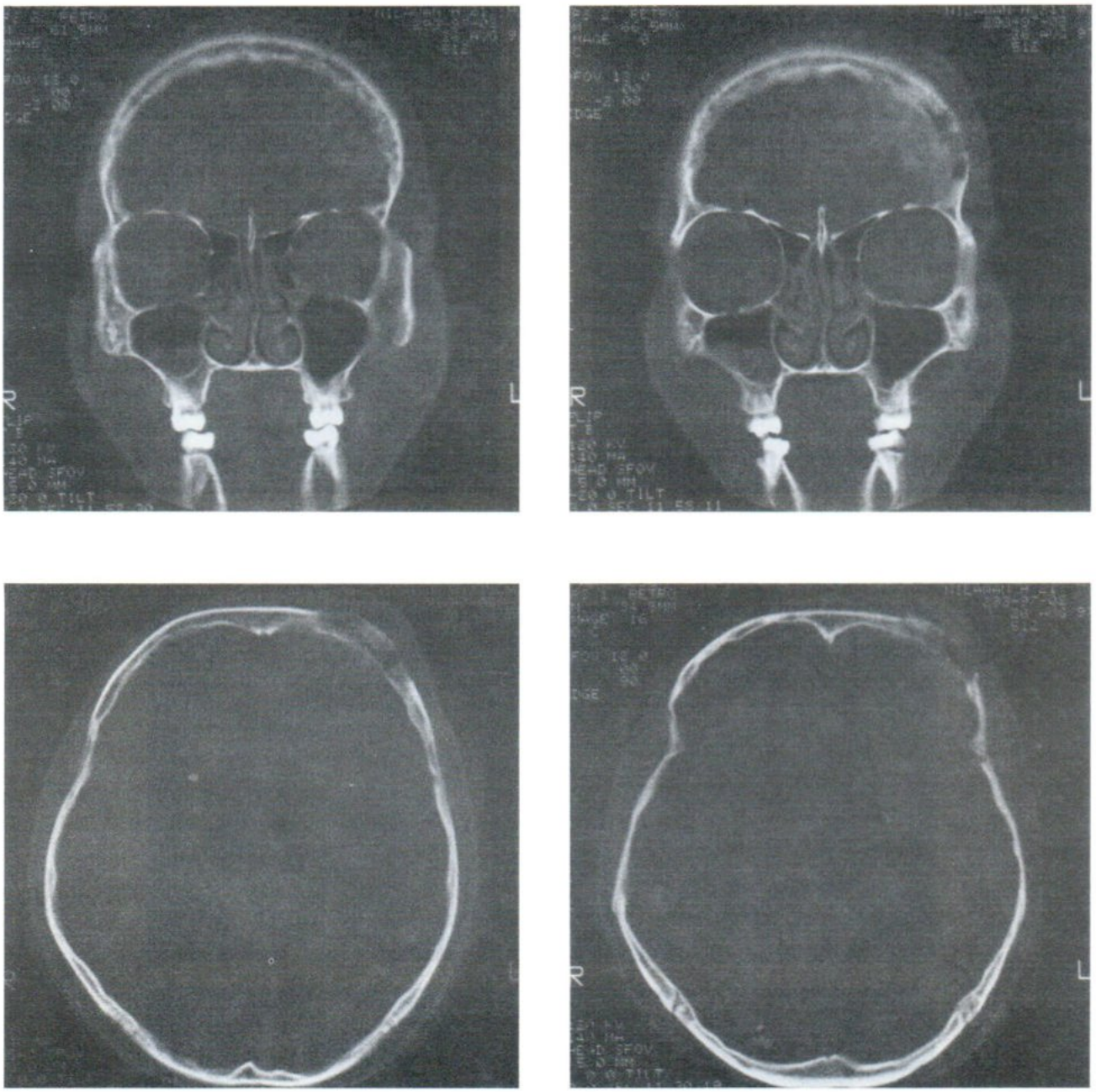


Fig. 2. Case 1. Bone window of the CT scan revealed that the destruction of the all table-layers of the frontal bone was obvious. The destruction was composed of osteolytic and permeative areas.

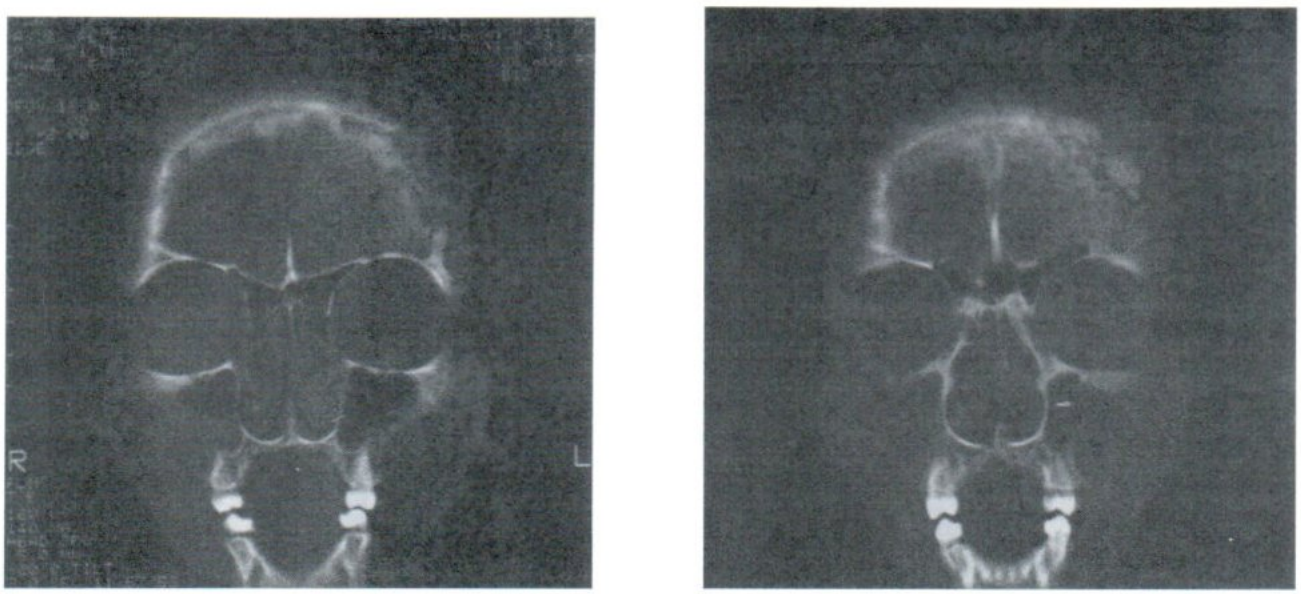


Fig. 3. Case 1. Bone window of the CT scan , mild expansion of the medulla of the frontal bone was observed.

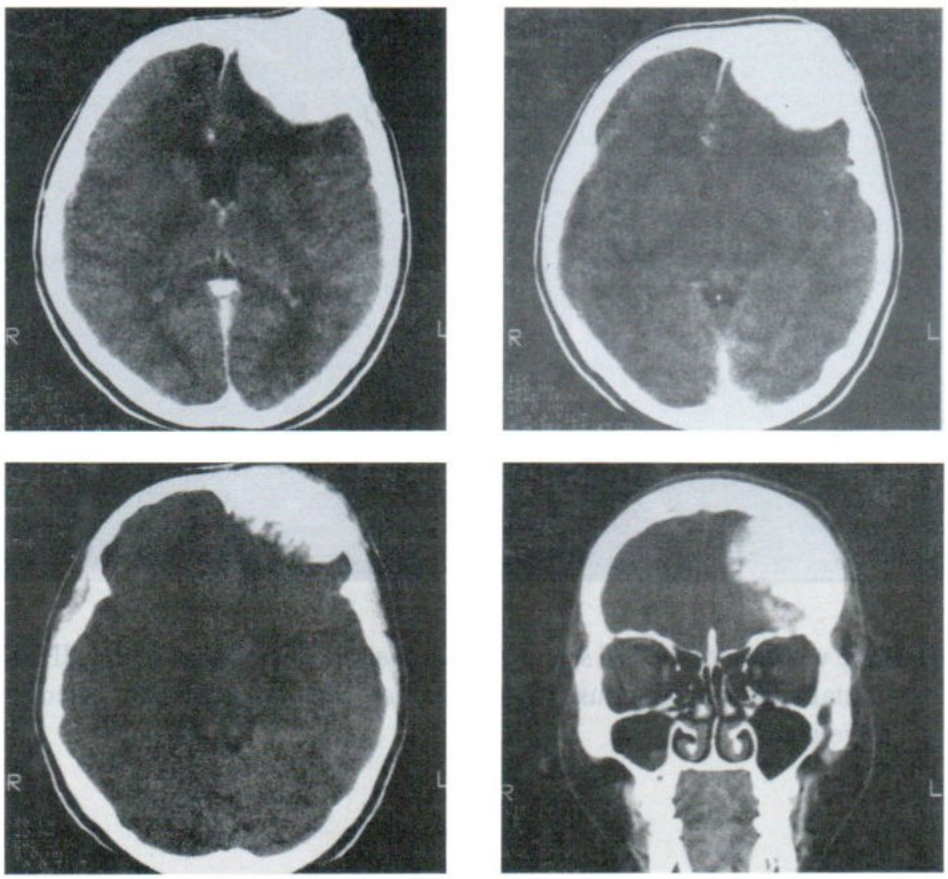


Fig. 4. Case 1. Soft tissue window of the CT scan of the lesion showed that the nearly entire part of the mass was bony dense.

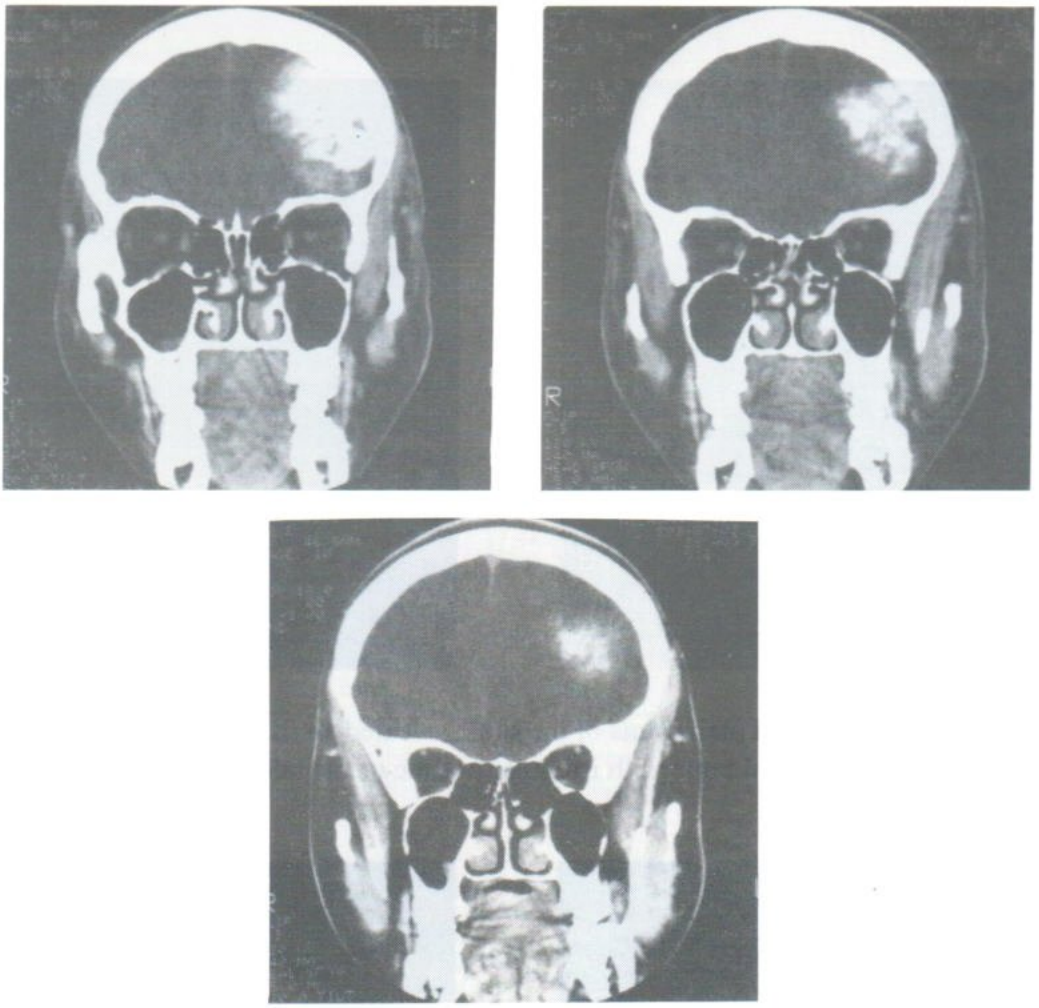


Fig. 5. Case 1. CT scan of the lesion showed that the lesion contained heavy calcification.

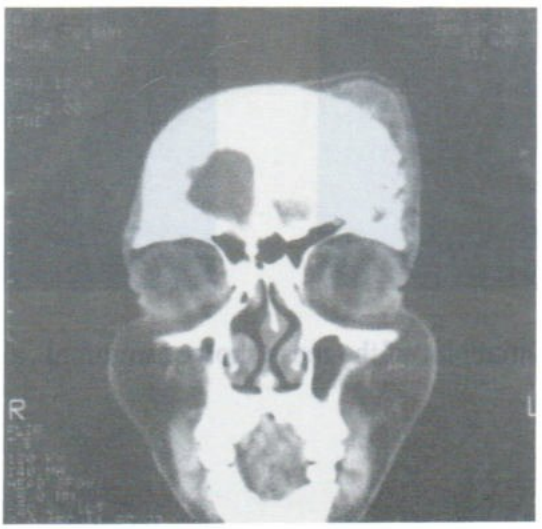


Fig. 6. Case 1. CT scan of the lesion showed that the extracranial subgaleal part of the lesion contained no calcification.

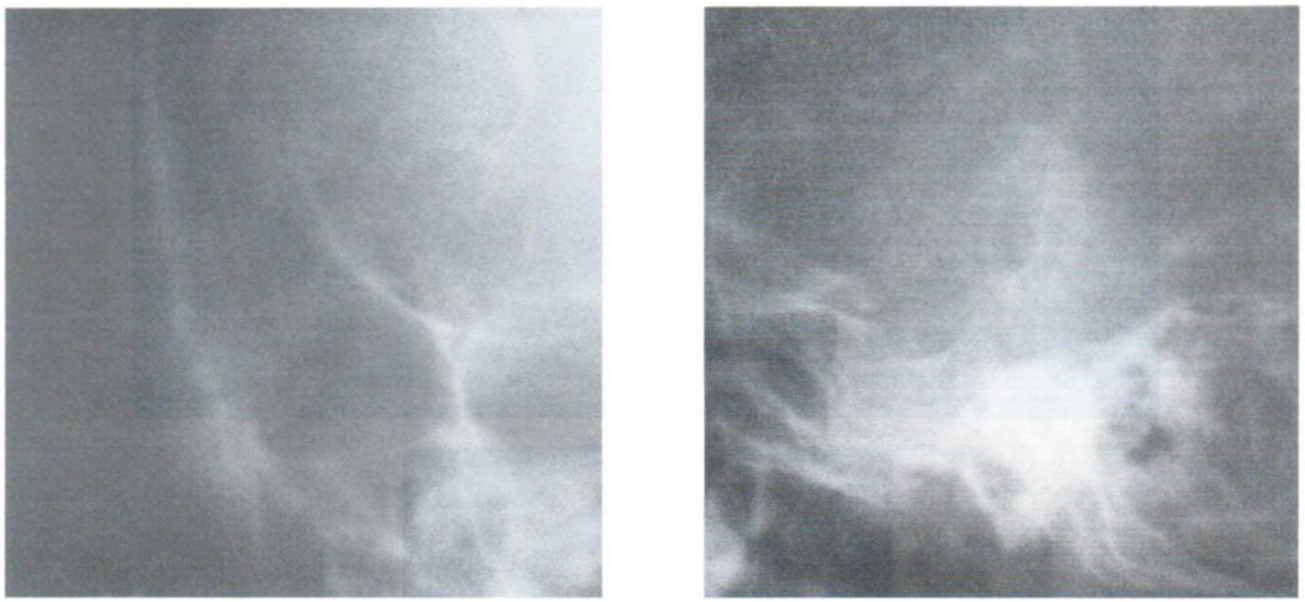


Fig. 7. Case 2. PA and lateral plain films of the skull revealed an ill defined border dense area in the right temporal bone with irregular outline of the tables.

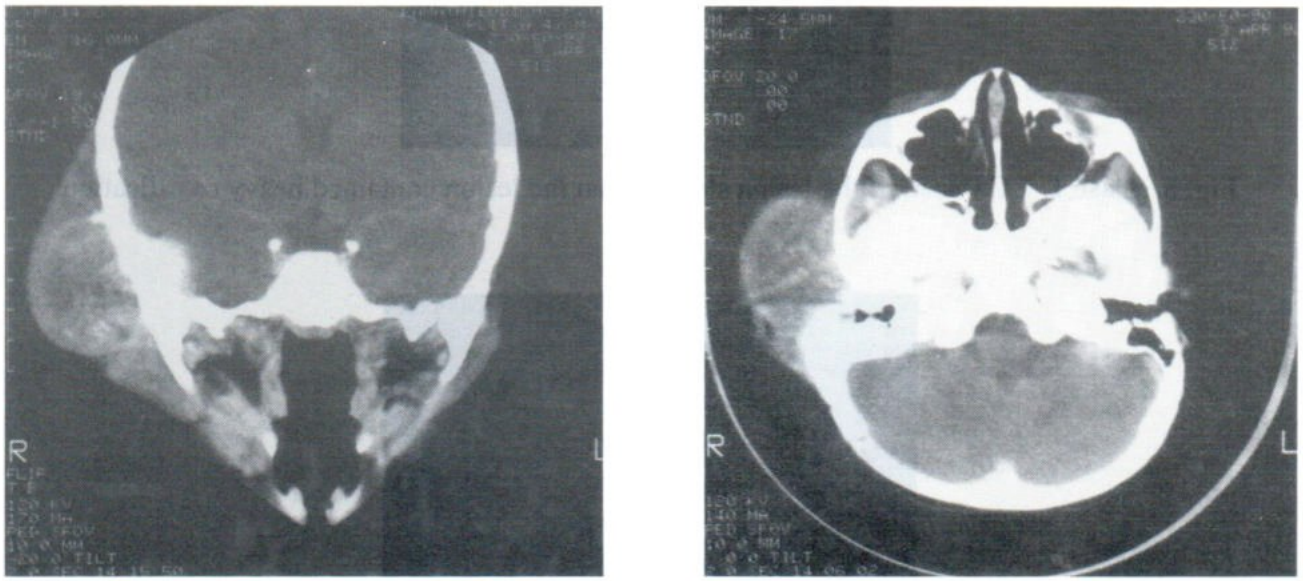


Fig. 8. Case 2. The extracranial part of the mass showed minimal calcification by CT scan.

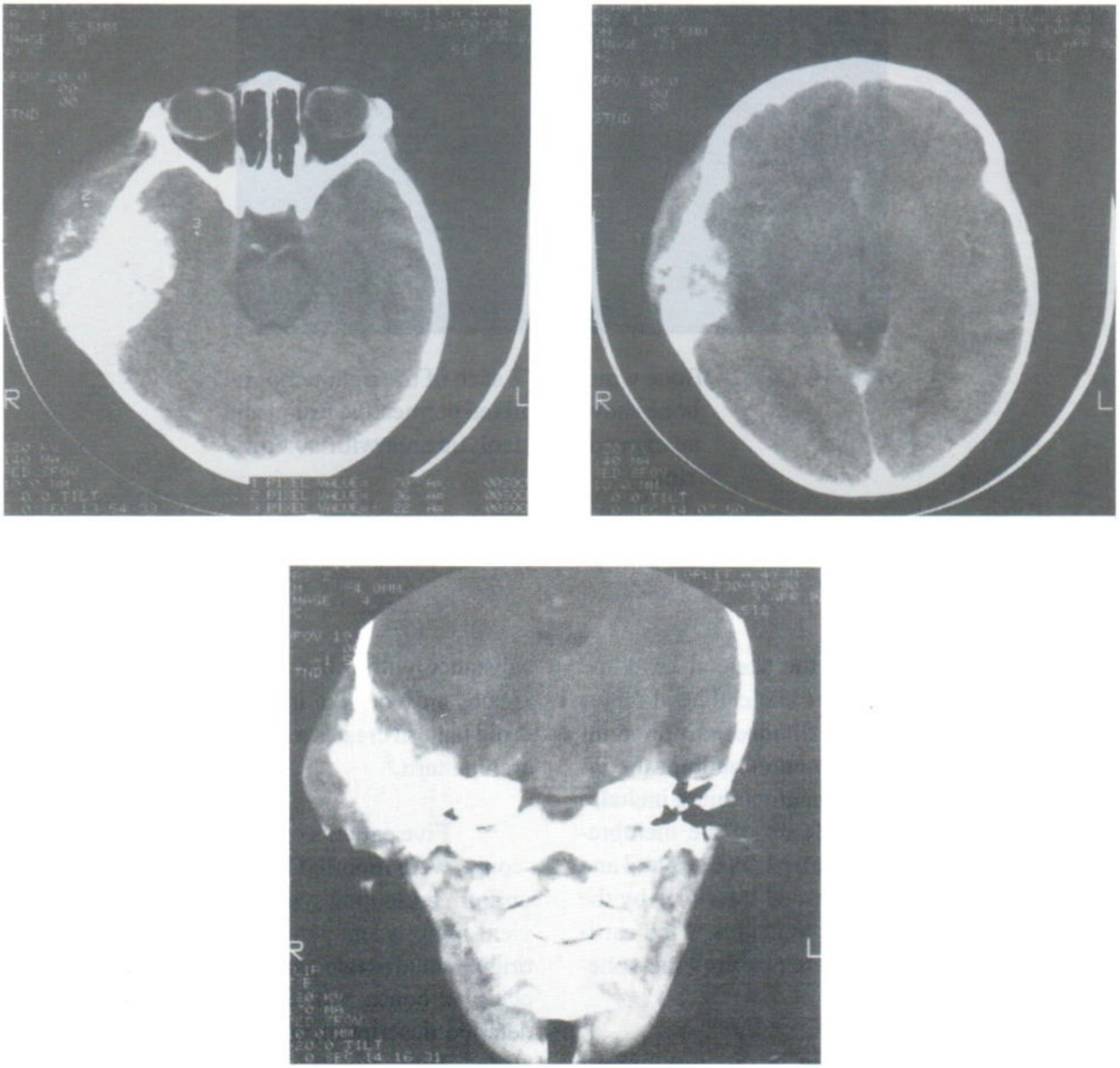


Fig. 9. Case 2. Most of the intracranial part of the mass showed dense calcification by CT scan.

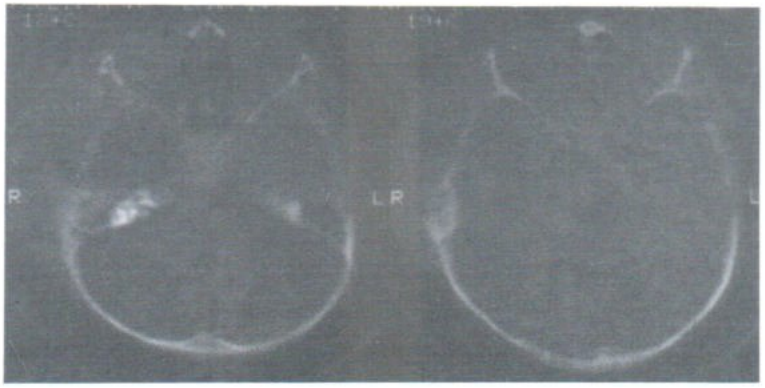


Fig. 10 Case2. Bone window CT scan of the right temporal bone showed a mixed permeative lesion and an expanding osteolytic area with sclerotic rim.

DISCUSSION

The skull includes the skeletal head and mandible: the cranium is the skeletal head minus the mandible. The skull is divided into three interconnected portions: the neurocranium, the facial area, and the base. The neurocranium includes the calvaria, which is made up of the membranous portions of the occipital, parietal, frontal and temporal bones, and is bounded inferiorly by the base of the skull , which is made up of the cartilaginous portions of these bones plus the sphenoid and ethmoid bones.[2]

Osteogenic sarcoma is the commonest bone neoplasm of the long bones, with the greatest predilection for the metaphyses, most frequently the distal femur and proximal tibia.[3] It primarily affects older children and young adults.[4] Very few osteosarcomas occur before age 5 or over age 30.[5] Radiographically, most long bone osteosarcoma (46%) demonstrate a mixed pattern, with osteoblastic and osteolytic type accounting for 32% and 22% respectively. Periosteal reaction is associated with 80% of long bone lesions. The majority of osteosarcomas have matrix mineralization, calcification of the osteoid or osteoid-like substance within the tumor; the osteoid pattern creates an ivorylike increased density, the chondroid pattern creates a stippled,flocculent, or ringarch pattern.[6]

Five patients of primary calvarial osteosar -coma were reported by Lee et al.[3] There was no uniform presenting symptom and there was no cervical lymphadenopathy. The bony site of tumor origin included two parietal, two temporal and one occipital bones. All lesions were osteolytic. The detailed description of each cases were not presented. Two patients of osteogenic sarcoma of the calvarium was described in children (12-year-old girl and 11- year-old boy) by Kornreich et al.[7] The described CT scan in one case showed a destructive lesion in all layers of the right parietal bone with irregular and expanding margins and an associated small non-calcified soft tissue mass without dural invasion. The skull (right parietal bone) of another case was involved as a part of multifocal osteosarcoma (another site was at distal femur). The CT scan of the latter case showed a remarkable extracranial soft tissue mass with hyperdense borders without bone destruction: but the inner

table was irregular with formation of internal body spicules. The tumor invaded the epidural space.

Shramek et al,[8] reported a case of osteogenic sarcoma at left occipitoparietal bone in an 8-year-old boy. The CT of this case showed a heavily calcified left occipitoparietal mass with extra-and intracranial components. The calvaria was thin at the region of the mass. MRI showed much of the soft tissue component of the lesion which was isointense to the brain on both T1-and T2-weighted images with signal of acute hemorrhage at the peripheral of the mass.

In conclusion, two cases of calvarial osteosarcoma have following characteristics:

1. Both patients were young, 4-year-old boy and 22- year-old woman.
2. Both masses had heavy calcifications or bony density in the portions that surrounded the originated pathological bones.
3. Associated extracranial masses were lumpy and contained small areas of calcification.
4. The involved calvaria (frontal, and temporal bones), showed mixed permeative and osteolytic areas.
5. The cases of the reviewed literatures also showed no osteoblastic lesions of the involved calvaria.

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