CRANIAL AND FIBULA CHONDROSARCOMA:A CASE REPORT

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ABSTRACT

Cranial chondrosarcomas were discovered in a 62-year-old female patient who presented with neurological symptoms. CT scan showed a large mass at right temporal fossa, extending to near-by structures and another smaller lesion at left temporal fossa. The masses were invasive and destructive, containing no calcification. T1W MRI study showed low to iso-signal mass and on T2WI, the signal was iso to the gray matter and the contrast enhancement was dense. There was no neovasculature at angiography. Bone survey was later performed after obtaining histology of the mass from craniotomy. The primary lesion was detected at the left fibula with classic appearance of of chondrosarcoma.

INTRODUCTION

Chondrosarcomas comprise a group of tumors that have a broad spectrum of clinical and pathologic findings. The feature common to all is the production of neoplastic cartilage. It morphologically subclassified into conventional, intramedullary and juxtacortical, clear cell, dedifferentiated, and mesenchymal variants.[1] Chondrosarcomas commonly arise in the central portions of the skeleton, including the pelvis, shoulder and ribs. The clear cell variant is unique in that it originates in the epiphyses of long tubular bones. Chondrosarcoma rarely involves the distal extremities. These tumors usually present as painful, progressively enlarging masses. 70% of grade 3 tumors disseminated. When chondrosarcomas metastasize, they preferentially spread to the lungs and skeleton.

The reported patient had predominated neurological problems and intracranial lesions were earlier detected than the primary site.

CASE REPORT

A 62-year-old female patient admitted to Prasat neurological institute due to intermittent headache, right temporal mass, decreased visual acuity of right eye and progressive weakness of left extremities for three months. On physical examination, a 5 cm-solid mass at right temporal area was detected. Left lower extremity showed mild swelling. The patient was drowsy and confused. Right pupil was 3.5 mm and fixed while left pupil was 2 mm and reacted to light. Right optic atrophy and left papilledema was noted. Right total ophthalmoplegia and left lateral rectus palsy was seen. Left hemiparesis, grade I and right sided motor power was grade IV.

Cranial CT scan was performed and revealed two inhomogeneous enhanced masses. The first mass was at right temporal area, extending to the right orbit, squamosal temporal bone, medially to obliterate right lateral ventricle and cause 2 cm midline shift to the left, to the middle cranial fossa and destroy the sphenoid bone (Fig.

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1). The second mass was at the left temporal area with extension inferiorly to destroy the sphenoid bone. No intratumoral calcification was detected (Fig.1).

Signal of the mass on T1W MRI study showed low to iso-signal and on T2WI study showed iso-signal to the grey matter. Marked enhancement of the lesion by Gd-DTPA was noted (Fig.2).

Intracranial angiography showed no neovasculature from both internal and external carotid arteries (Fig.3).

The right fronto-temporal craniotomy with subtotal tumor removal was then performed. The gross specimen revealed light brown, firm tissue which was measured to be 8 cm in diameter. The microscopic examination showed moderately cellular tumor with fragments of bone, dense fibrous tissue and striated muscle. The tumor cells were round in shape, medium to large in size and were in cartilage lacunae. Multinucleated tumor cells and mitotic figures were frequently noted. The pathological diagnosis was chondrosarcoma.

The bone survey was later performed and the lesion of extensive chondrosarcoma of left fibula was detected (Fig. 4).

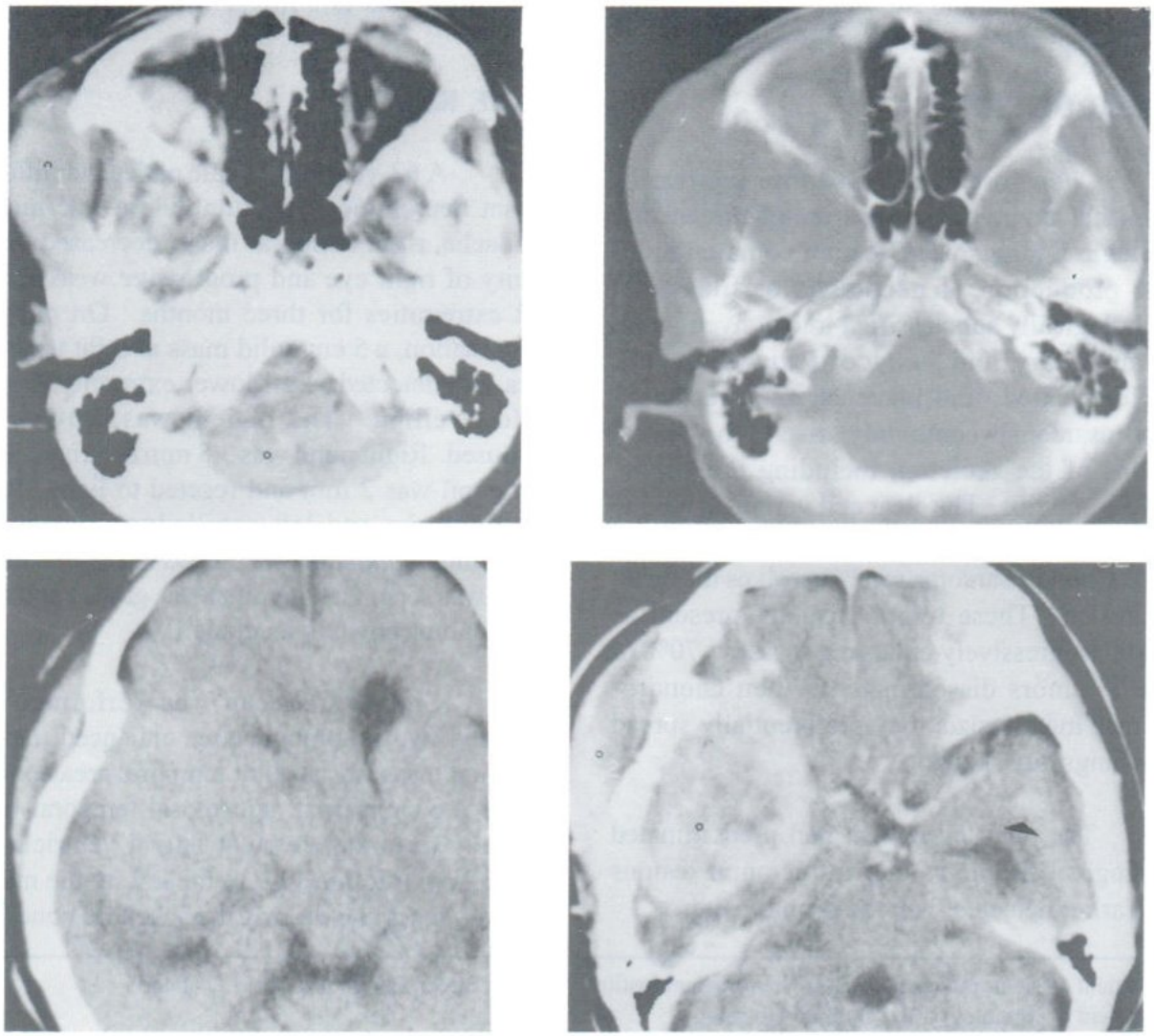
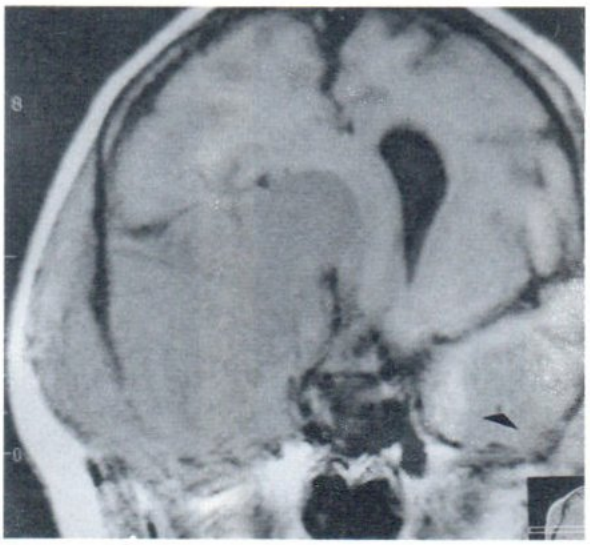


Fig.1 CT scan showed metastatic chondrosarcomas at both temporal fossae with invasive nature.



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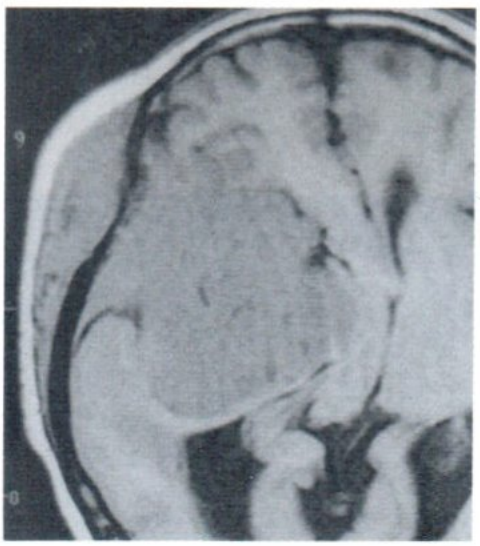

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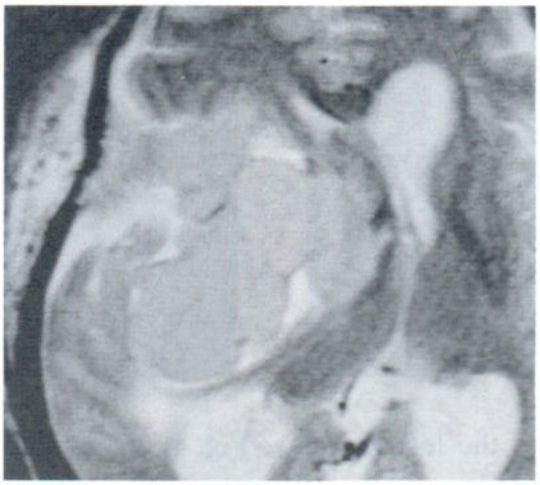

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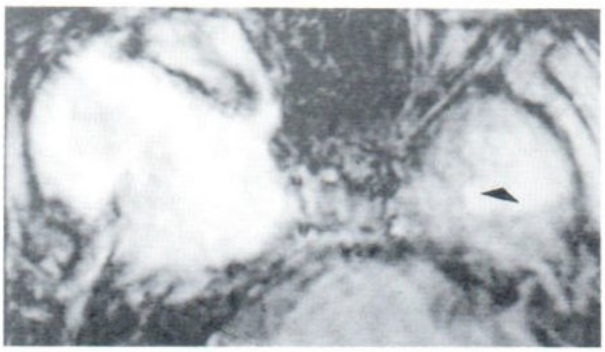

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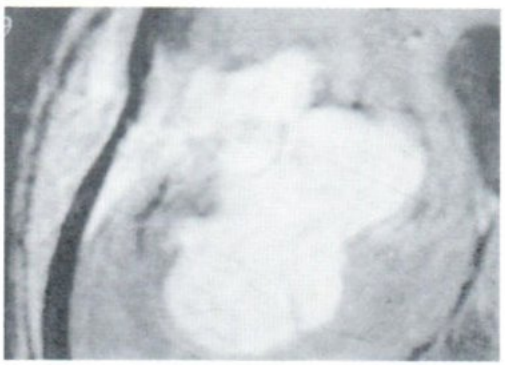

(2c)

Fig. 2 MRI images of the lesion
(a) T1WI, (b) T2WI, (c) post gadolinium enhancement

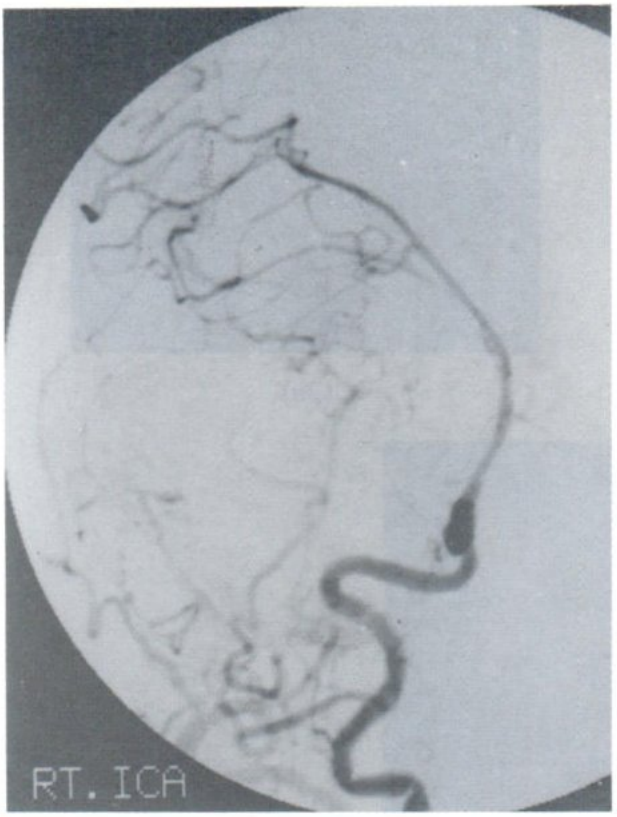

Fig. 3 No neovasculature to the lesion was noted at angiography

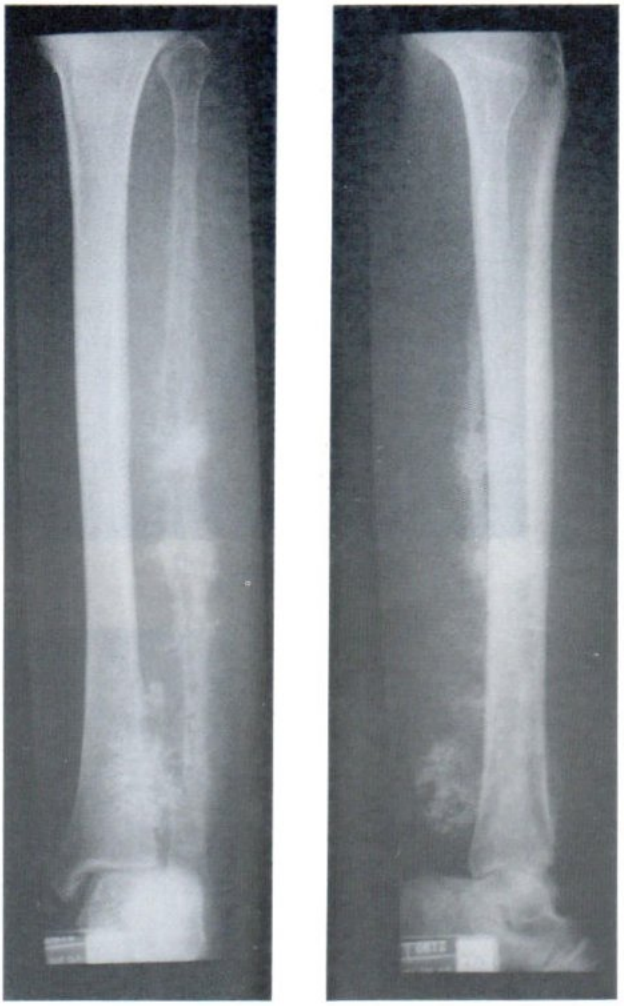

Fig. 4 Classical chondrosarcoma at the left fibula was shown by radiography

DISCUSSION

Chondrosarcomas of the bone represent a range of malignant tumors of cartilage that includes the classic or conventional chondrosarcoma, dedifferentiated chondrosarcoma, clear cell chondrosarcoma, mesenchymal chondrosarcoma, and malignant chondroblastoma.[1]

Classic or simply chondrosarcoma is the second most common malignant neoplasm of bone after osteosarcoma. It is subclassified as central (medullary) if it is located within the medullary cavity or peripheral (juxtacortical) if its origin is on the surface of the bone. It may arise de novo or secondarily from preexisting benign cartilage lesions such as the osteochondromas in multiple osteochondromatosis or the enchondromas of Ollier's disease. Chondrosarcoma is a malignant tumor of hyaline cartilage that arises in the medullary cavity or on the surface of a bone. It is a tumor of adults generally between 20 and 70 years of age. Males are affected more often than females. The most common sites of involvement are the pelvis, femur, scapula, humerus, and the ribs. Radiographic findings indicate the cartilaginous nature of a tumor when typical calcifications are present. Matrix calcifications have been variously described as annular, punctate, popcorn-like, ringlet-like, stippled, or small ill defined radiodense formations. These features do not discriminate benign from malignant tumors.

Dedifferentiated chondrosarcoma has the worst prognosis. Postulation was formed that low grade cartilaginous lesions may transform into a high-grade sarcoma[2] or that these tumors are formed by synchronous differentiation of two separate clones of cells.[3] Most cases occur in the sixth decade of life.[4] The patient distribution by sex is about equal. The site of tumor is most often the bones of the pelvis, followed by the femur.[5] Radiologically, these tumors may show foci of typical cartilaginous calcifications, denoting the low-grade component, and a large lytic component, which usually has a significant soft-tissue mass component. However, a good number of tumors do not present the calcification of the low-grade component, but only the large noncacified mass. Glossly these tumors are large infiltrative lesions that not only involve the bone but also extend into the adjacent soft tissues. Foci of cartilage may be seen on gross inspection.

Mesenchymal chondrosarcoma is rare, accounting for 0.33% of all primary malignant neoplasms of bone. Eighty per cents of cases are diagnosed before 40 years of age. The peak incidence is in the second decade of life. Radiologically there is nothing characteristic for this tumor. If calcified cartilage matrix is present, the tumor may look like a conventional chondrosarcoma. Any bone may be involved. Huvors reported 31% of it in the lower extremity, 17% in the pelvic girdle, and 14% in the cranial bones.

Clear cell chondrosarcoma has a relatively good prognosis and is extremely rare. The patients ages ranged from 14 to 84 years, with a slight peak in the third and fourth decades of life. There was a male predominance; the male-to female ratio was 2.5 to 1.[6] Radiologically this lesion is seen in the epiphysis of the long bones, especially the head of the femur which is the most common location; most of these tumors contain typical cartilaginous calcifications. Some small tumors may be difficult to differentiate from chondroblastoma.[7]

Chondrosarcomas of the skull occur from childhood to old age. No sex predilection has been noted. They are locally aggressive but grow slowly with tumor frequently leading to death five or more years after onset of symptoms.[8,9] Metastases have been reported[10] but are uncommon. Chondrosarcomas occur most often along the base of the skull, the most frequent site being parasellar.[11] The predilection of intracranial chondrosarcomas for the base of the skull is not surprising since embryologically the base arises from a cartilaginous

matrix. Atypically located chondrosarcoma may arise from cartilaginous rests other than at the base of the skull or from multipotential cells within the meninges.

On MR images obtained with short TR/TE sequences, chondrosarcomas generally had low to intermediate signal intensity and were slightly hyper-or isointense to muscle and iso-or hypointense to gray matter. On MR images obtained with long TR/short TE and long TR/TE sequences, chondrosarcomas of the skull base generally had high signal intensity and were predominantly hyperintense to muscle and gray matter.[11] Chondrosarcomas showed a marked degree of contrast enhancement.

Cranial involvement in our case was most probably metastatic lesions due to multiplicity of the lesions. Though the primary site contained abundant typical cartilaginous calcifications, the metastatic components had absent calcification. The signal on T2WI in our case was not similar to those reported.

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