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Case Report

Large chondrosarcoma of skull base: an unusual intracranial tumor in a young male

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Abstract. A 25-year-old male presented with headache. He was noted to be neurologically intact on physical examination. He had normal laboratory findings. CT and MR imaging showed a large right temporal mass. Radiological diagnosis was cartilaginous tumor arising from the skull base. The patient underwent surgery at another hospital. The histopathologic features were supporting the radiologic diagnosis being evaluated as mixoid type chondrosarcoma. Although intracranial cartillaginous tumors are rare, the diagnosis can be possible with careful radiological analysis.

Key words: skull base, chondrosarcoma, intracranial

1. Introduction

Chondrosarcomas are tumors that typically arise from long bones and rarely take origin in cranial bones. They are rare, slow-growing, highly infiltrating tumors and represent 0.15% of all primary intracranial lesions (1). Most are located at the base of skull, where they are thought to arise from cartilage of the synchondroses. Histologically, three variants of chondrosarcoma have been described: myxoid, mesenchymal, and classic chondrosarcoma (2). Few examples of intracranial chondrosarcomas arising above the skull base have been reported (3) and most of these tumors are highly malignant mesenchymal variants. We report the computed tomography (CT) and magnetic resonance imaging (MRI) findings of a large intracranial myxoid type chondrosarcoma in a young patient.

2. Case report

A 25-year-old previously healthy male presented with a severe headache. His significant history was the occurrence of severe headaches. He was noted to be neurologically intact.

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Physical examination and laboratory findings were normal. CT scan revealed a well marginated, 6x5 cm in diameter isodens mass on the right temporal lobe. Petrous bone destruction was seen on CT scan. Curvilineer calcifications were present throughout the mass and there was no apparent contrast enhancement (Figure 1A,B). The tumor was displaying a high signal on T2-weighted images, resembling a cyst. However, FLAIR images showed high signal and T1-weighted MR images displayed slightly low signal intensities. T2 weighted MR images showed minimal peritumoral eadema. While no apparent contrast enhancement was observed in first scans, heterogeneous enhancement with a delay of 5 minutes was shown after the contrast administration (Fig 2 A-D).

A pre-operative angiogram noted no significant vascularity arising from the internal and external carotid arteries. However, intracranial arteries were deplaced by the lesion. The lesion was in extraxial location. Cartilaginous tumor (chondrosarcoma or chondroblastoma) was thought radiologically.

The patient underwent surgery at another hospital. Histopathologic diagnosis was myxoid type chondrosarcoma.

3. Discussion

Intracranial cartilaginous tumors are rare lesions that generally present as a solitary mass.

Chondrosarcomas comprise less than 15% of

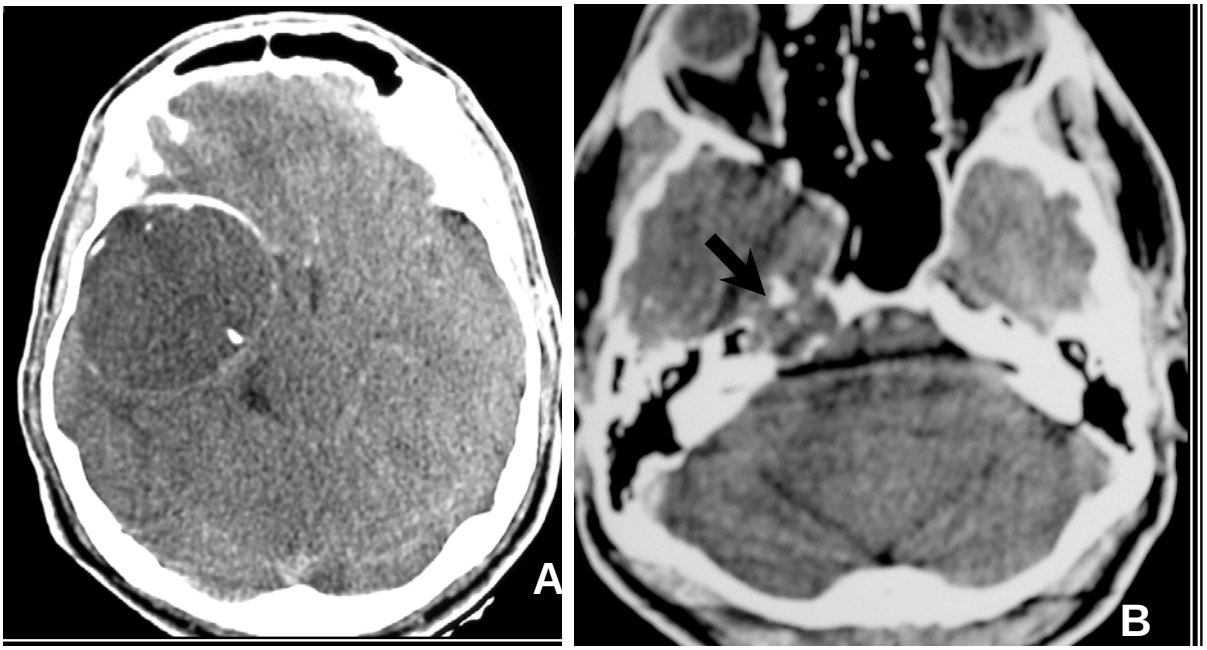


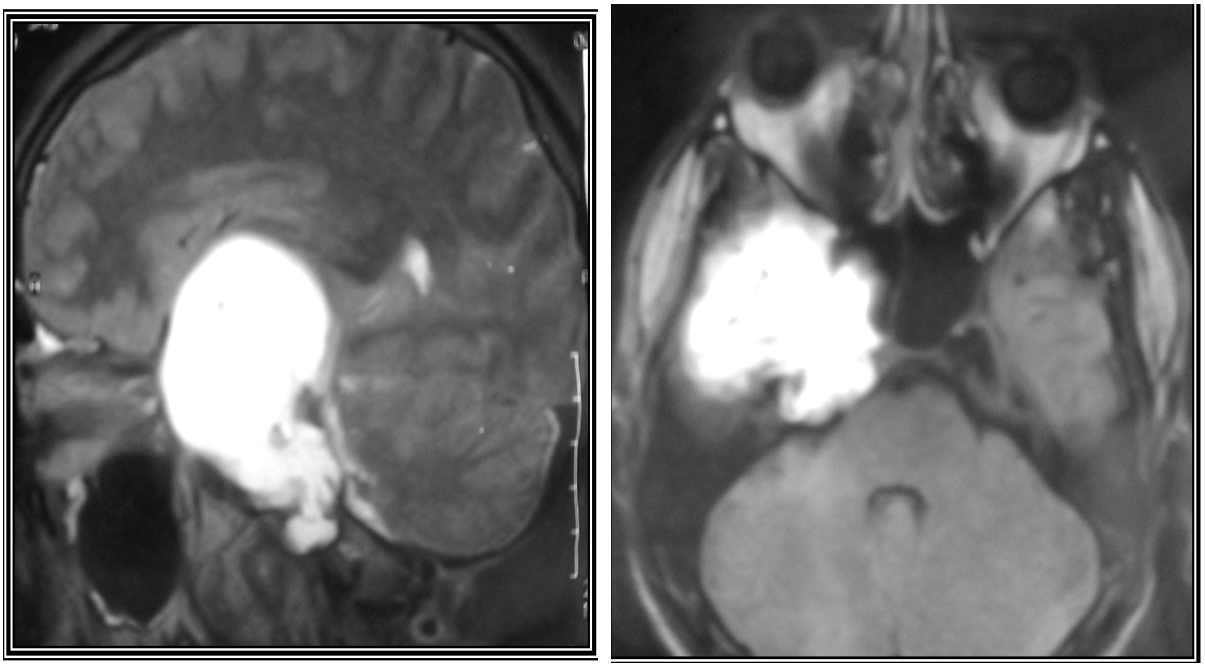
Fig. 1. A,B Axial contrast-enhanced CT shows hypodens mass extending to the right temporal lobe. Curvilinear calcified wall, punctate calcifications in the lesion with moderate mass effect are seen (A). Petrous bone destruction is seen on CT scan at the level of skull base (arrow) (B).

intracranial chondromatous tumors and generally arise from the base of the skull (3). Intracranial chondrosarcomas typically arise during the fourth and fifth decades, but our case was a young male. Patients usually present with a long-standing history of headaches as in our case, and with signs related to increased intracranial pressure (4).

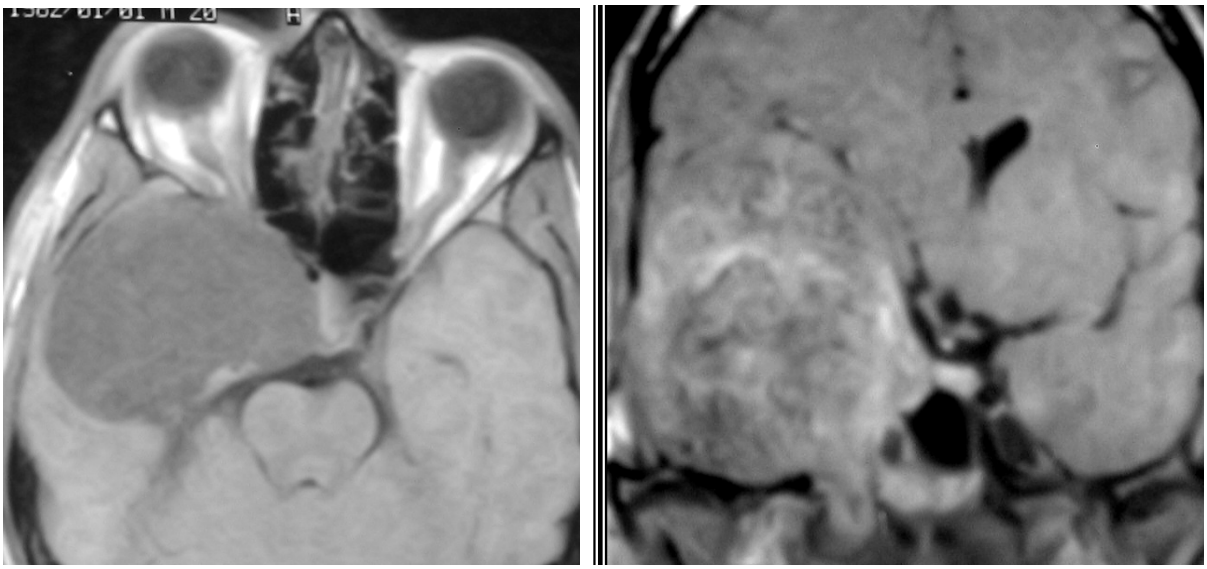
Chondrosarcoma and chondroblastoma may have very similar radiographic appearances (5). The mean age of tumor occurrence in chondrosarcoma is one decade later, but the overlap of the age range complicates the differential diagnosis (6). A precise diagnosis is important since the therapy of chondrosarcoma is more aggressive than that of chondroblastoma. The contrast enhancement is heterogeneous in clear cell chondrosarcomas and chondroblastomas. Myxoid chondrosarcoma is the rarest variant of chondrosarcomas in intracranial location. Histological diagnosis of this entity must also include other myxoid lesions such as chordoma or chondromyxoid fibroma (7).

Chondrosarcomas have increased T2 signal like cystic lesion due to the low-cellularity and high water content of mature hyaline cartilage. The lobulated internal architecture helps to distinguish these tumors from cystic masses. However, MRI signal characteristics of chordomas and chondrosarcomas may be similar: low to intermediate signal intensity on T1-weighted and high signal on T2-weigthed images, with heterogeneous contrast enhancement (8). The lesion in our case showed low signal intensity on T1-weighted images and high signal on T2-weighted images. FLAIR images displayed high signal due to the hyalin cartilage. However, bone destruction is very important for the differential diagnosis and there was destruction of petrous bone in our case.

Chordoma usually affects clivus and appears at the midline. The mass in our patient had been placed laterally. The clivus was intact. Epidermoid tumors and chondrosarcomas display similar high signal on FLAIR images and have a lobulated pattern of growth. Diffusion MRI could differentiate the lesion from epidermoid tumor, but it could not be performed due to the fact that the MRI system was 0.3 Tesla and diffusion sequence was absent in this system. Bone destruction seen in chondrosarcoma can help in the differential diagnosis from epidermoid tumor. Angiography can also be helpful in the diagnosis of intracranial chondrosarcomas. Depending on the sub-type of chondrosarcoma, the lesion can show varying degrees of vascularity. Classical chondrosarcomas and about half of mesenchymal chondrosarcomas demonstrate avascularity. The remaining portion of mesenchymal and virtually all myxoid chondrosarcomas display an intense



A **B**



C **D**

Fig. 2. A-D Sagittal T2-weighted image shows a lobulated mass with high-signal intensity resembling a cystic lesion. The tumor is compressing the lateral ventricle superiorly (A). Axial FLAIR image shows no suppression and high-signal (B). Axial T1-weighted image shows low signal intensity mass (C). Heterogeneous enhancing mass is seen on coronal T1-weighted image after the gadolinium administration. Right Meckel's cave and cavernous sinus were invaded. The mass was extending exracranially through foramen ovale (D).

hypervascularity that is similar to vascular malformations and hemangiopericytomas. However, the tumor in our case showed no significant vascularity arising from the internal and external carotid arteries. The preferred treatment of choice for intracranial chondrosarcoma is radical surgical resection (9).

Reports describing imaging studies of intracranial chondrosarcomas arising above the skull base are primarily CT based with few reports including MRI findings. We report a rare

case of a huge chondrosarcoma of skull base extending intracranially. CT features of a broad-based, bone destructive mass with cord-like internal architecture and scattered-curvilineer calcifications, and MRI features of high signal on T2-weighted images like cystic lesion, high signal on FLAIR images and heterogeneous enhancement should suggest the diagnosis of this rare tumor.

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