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Dumbbell Shaped Giant Cell Tumor of the Temporal Bone: Case Report and Literature Review

Temporal Kemiğin Kumsaati Görünümlü Dev Hücreli Tümörü: Olgu Sunumu ve Literatürün Gözden Geçirilmesi

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ABSTRACT

We present a 29-year-old woman with a giant cell tumor of the temporal bone. The differential diagnosis is discussed with reference to the literature regarding giant cell lesions, especially of the cranium.

KEY WORDS: Giant cell tumor, magnetic resonance imaging, temporal bone

ÖZ

29 yaşındaki bir kadın hastada temporal kemiğin dev hücreli tümörü sunulmuştur. Hastalığın ayırıcı tanısı literatür ışığında özellikle kraniumu ilgilendiren dev hücreli lezyonlar açısından tartışılmıştır.

ANAHTAR SÖZCÜKLER: Dev hücreli tümör, manyetik rezonans görüntüleme, temporal kemik

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INTRODUCTION

Primary giant cell tumors rarely occur in the skull and make up less than 1% of all reported bone tumors (5, 11). The classical location is the epiphysis of long bones. These tumors are very rare in the skull and have been reported in the temporal, sphenoid, petrous and occipital bones (10, 21). We present a patient with a primary giant cell tumor of the left temporal bone.

CASE REPORT

A 29-year-old woman was admitted with a progressively expanding mass in the left temporal region of one-year duration. The patient had a history of trauma at the same localization. On physical examination there was evidence of local swelling on the left temporal region approximately two cm superior to the temporomandibular joint. Neurological examination revealed no cranial nerve deficits. All laboratory findings were normal. Enhanced axial CT scan (Figure 1) of the temporal bone showed an expansive, enhancing lesion involving the left temporal bone and extending into the temporal lobe and extracranial soft tissue. The MRI scan showed a soft tissue tumor of heterogeneous low intensity on T2-weighted non-contrast images (Figure 2) and heterogeneous enhancement was observed on the enhanced MRI scan (Figure 3A-B). Total removal of the tumor was performed via the transtemporal approach. A photomicrograph of the surgical specimen (Figure 4) showed osteoclastic multinucleated giant cells mixed with spindle-shaped stroma cells, compatible with a giant cell tumor.

The dumbbell-shaped tumor on the temporal bone that was expanding intracranially to the left temporal lobe and extracranially to soft tissue was not observed on the early postoperative enhanced CT scan (Figure 5). Post-operative radiotherapy was not planned as the tumor was resected with a radical operation. The patient was discharged on the ninth postoperative day without any neurological deficits.

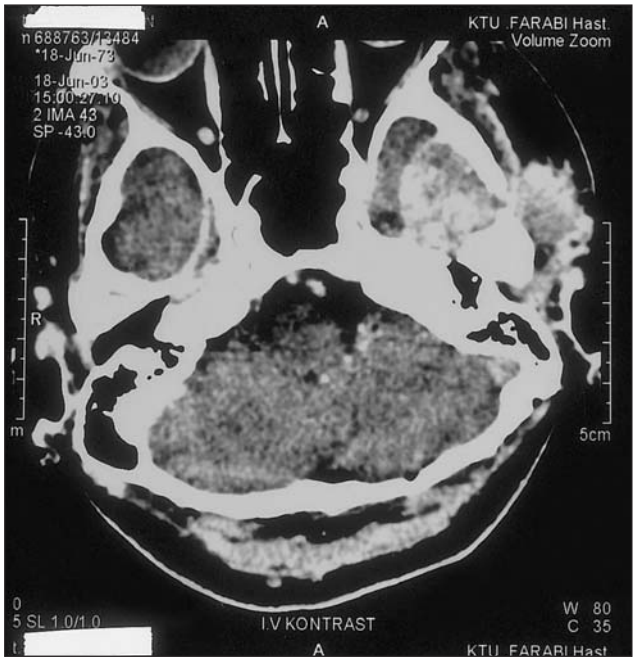

Figure 1: Postcontrast axial CT scan of giant cell tumor before operation showed a mass expanding the squamous and petrous sections of the left temporal bone.

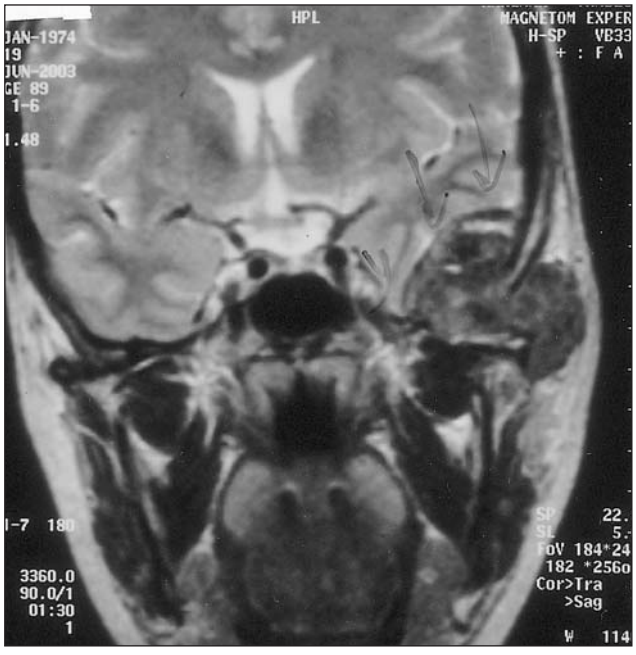

Figure 2: Heterogenous low-intensity soft tissue tumor was seen on T2-weighted MR image.

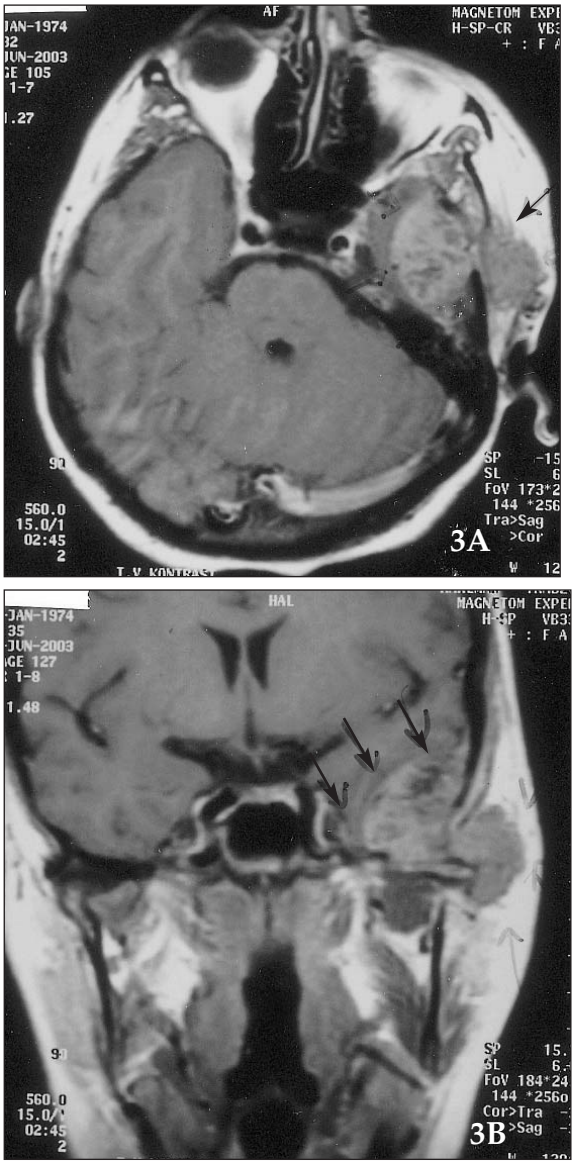

Figure 3: After contrast administration, there was heterogeneous enhancement within the tumor on axial T1-weighted (**A**) and coronal T1-weighted (**B**) MRI images.

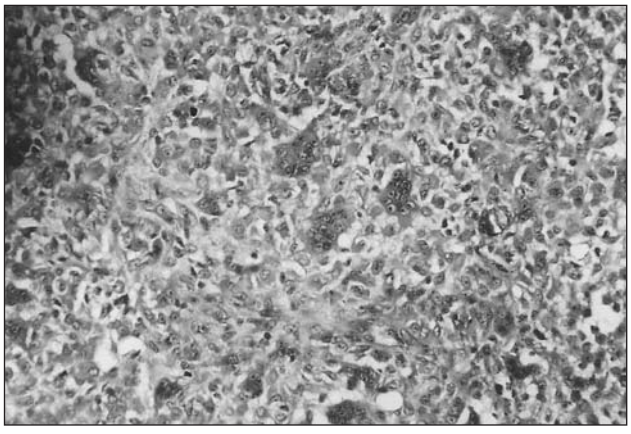

Figure 4: Osteoclastic multinucleated giant cells within the spindle cell stroma (H&E x200).

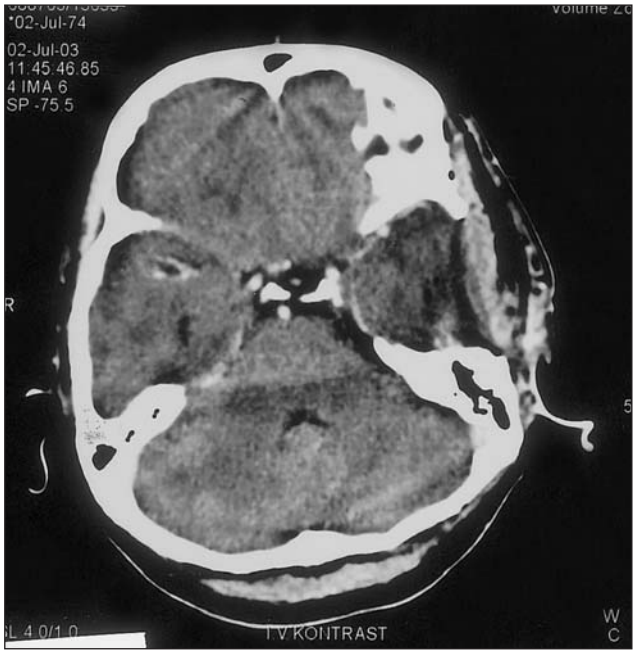

Figure 5: Early postoperative postcontrast axial CT scan showed no tumoral mass on the left temporal bone.

DISCUSSION

Giant cell tumor of bone is a rare tumor with an incidence of one per million (6). Giant cell tumors originate from the connective tissue within the bone marrow. They most commonly occur in the epiphysis of long bones (10), especially the distal femur, proximal tibia and distal radius (25). There is a high recurrence rate after resection (10-40%) (19). Giant cell tumors of the head and neck are uncommon and account for approximately 2% of all giant cell tumors (20). The most commonly involved sites are the sphenoid and temporal bone. Females are affected more frequently in all age groups (1, 5, 17). The usual symptoms and signs are local tenderness and swelling, or even pathological fractures. The age at presentation is usually the third ot the fourth decade (18). The clinical presentation depends on the site of origin. The symptoms are pain, swelling in the region of the affected bone, limitation of motion of the adjacent joint and weakness of the involved extremity (24). Tumors arising from the sphenoid bone usually manifest with headache, ophthalmoparesia, trigeminal hyperesthesia and loss of vision (9). Temporal bone tumors typically cause pain behind the ear on the affected side, deafness and facial weakness (22). Our patient presented only with increasing swelling. Histologically, the tumor is composed of a vascular network of spindle-shaped

or oval stromal cells and multinucleated giant cells (10). On imaging, giant cell tumor usually has the nonspecific appearance of an expansive, destructive soft-tissue mass (1).

The radiographic appearance may mimic that of a malignant tumor (24). Radiological differential diagnosis includes chondroblastoma, chondrosarcoma, aneurysmal bone cyst, dermoid cyst, eosinophilic granuloma, osteolytic metastasis, giant cell reparative granuloma and pigmented villonodular synovitis (PVNS) (6, 20, 21).

Chondroblastomas are benign tumors of immature cartilage cells and very rarely localize in the skull. The characteristic findings of these tumors are well-demarcated osteolytic areas, often with foci of calcification. The presence of calcification within the lesion rules out a diagnosis of giant cell tumor (23). Aneurysmal bone cysts are benign lesions composed of large vascular spaces with trabeculae of connective tissue and bone. Nearly 5% are located in the skull. These tumors usually expand both the inner and outer table. Radiologically they show an irregular 'soap bubble' appearance (23).

Dermoid cysts usually show a rind of host bone sclerosis and radiologically present as sharply circumscribed lesions. They can erode any area of skull bone and project either subcutaneously, intracranially or both (15). Dermoids are low attenuated lesions both before and after contrast medium injection (22). Eosinophilic granulomas are most commonly found in the skull (43-80%) and are benign tumors with eosinophils and mononuclear cells. The classical radiographic finding is a round or oval non-sclerotic pounced-out lesion, involving both the inner and outer tables (12). Computed tomography shows osteolytic, hypodense lesions eroding the calvarium, sometimes associated with a soft-tissue mass in metastatic conditions and sarcoma (3). Both the inner and outer table are destructed irregularly on most patients with metastasis. Imaging appearance of giant cell reparative granulomas are identical to that of giant cell tumor (19). Giant cell reparative granuloma is rare in the axial skeleton and at this site, and is also referred to as a 'solid' aneurysmal bone cyst (16). It is typically found in the maxilla and mandible, but has been reported in other sites such as the frontal lobe (8). It is seen more frequently in women with the peak age of occurrence in the second decade of life (14). This benign tumor is probably hormone-dependent (7). Giant cell reparative granulomas and giant cell tumors have some important histological differences. Perivascular hemorrhage with clustering of the giant cells around the hemorrhage and hemosiderin deposition are seen in giant cell reparative granulomas, whereas in giant cell tumors there is usually a uniform distribution of giant cells and hemosiderin deposits are rare. Giant cells are also usually smaller and have relatively fewer nuclei in giant cell reparative granulomas than giant cell tumor (13).

Pigmented villonodular synovitis (PVNS) is a rare tumor of the synovium or the tendon sheath. There are two forms; a diffuse form involving the entire synovium and a localized form presenting as a solid nodule (22). In the skull, the diffuse form usually involves the temporomandibular joint (4). On computed tomography scanning a rim of bone around the lesion and lack of erosive changes of the mandibular head can help to differentiate giant cell tumor from PVNS (22).

Giant cell tumors are considered to be benign but are very aggressive, and have a recurrence rate as high as 40-60% (19). The preferred treatment of a giant cell tumor is surgical. Surgery is recommended for all operable lesions (2). Radiotherapy is restricted to inoperable cases or those not undergoing radical surgery (2).

Nineteen cases of giant cell skull tumor have been reported in the literature. The most common complaints among these patients were hearing loss and vertigo. Peripheral facial paralysis was reported only in one patient. In our case, the only complaint of the patient was local swelling in the left temporal region. This article presents a case of a giant cell tumor in a very rare location, treated with complete surgical resection.

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