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Chondroblastoma of the Temporal Bone
: A Clinicopathologic Study of Five Cases

Chondroblastoma is a rare benign bone tumor. It commonly affects the epiphysis of long bones during the second and third decades of life. Chondroblastoma of the temporal bone is extremely rare. We reviewed five cases of chondroblastoma arising in the temporal bone. Four cases were female and one was male. The ages ranged from 41 to 60 years (mean, 53.6 years). All cases involved the temporal bone. Three involved the left side and two the right. Chief complaints were long-standing localized pain and hearing difficulty. A sharply demarcated lobulated mass was the main radiological finding. Microscopic findings were those of chondroblastoma of usual locations. Two cases showed aneurysmal bone cyst-like areas. Immunohistochemical studies for CD34, CD99, S-100 protein and cytokeratin were performed. Tumor cells were diffusely positive for S-100 protein in three cases and weakly positive for cytokeratin in one case. CD34 and CD99 were negative in all cases. In summary, chondroblastoma of the temporal bone is rare and occurs in older age group than reported cases of chondroblastoma of the usual location in the literature.

Key Words: *Chondroblastoma; Temporal bone; Immunohistochemistry*

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INTRODUCTION

Chondroblastoma is an unusual benign neoplasm of the bone. It consists of less than 1% of all bone tumors. It was first described by Codman in 1931 and was given its name by Jaffe and Lichtenstein (1-3). Classically chondroblastoma occurs at the ends of long bones including proximal tibia, proximal humerus and distal femur. It commonly occurs in the second decade of life (3). Chondroblastoma arising in the temporal bone is rare. We report five cases of chondroblastoma arising in the temporal bone with the results of histological, immunohistochemical, radiological and clinical findings.

MATERIALS AND METHODS

Five cases with the diagnosis of chondroblastoma of the temporal bone were retrieved from bone tumor files of two institutes. Radiological, clinical and follow-up data were obtained from the letters of referring pathologists.

Four unstained slides were also obtained from each case by referring pathologists. Immunohistochemical studies of cytokeratin (mouse monoclonal antibody, Dako., Glostrup, Denmark; 1:20 dilution), S-100 protein (rabbit polyclonal antibody, Dako.; 1:200 dilution), CD 34 (mouse monoclonal antibody, Dako.; 1:20 dilution), and CD 99 (mouse monoclonal antibody, Novocastra Lab. Ltd., Newcastle upon Tyne, UK; 1:50 dilution) were performed on formalin-fixed, paraffin-embedded tissue using the avidin-biotin-complex method in each case.

RESULT

Clinical findings

The clinical findings were summarized in Table 1. Four cases were female and one was male. Ages of the patients ranged from 41 to 60 years (mean, 53.6 years). Temporal bone was involved in all 5 cases. Three involved the left side and two involved the right. Information on symptoms was available in all cases. Pain of the temporomandibular joint and tinnitus were the most common chief complaints of the patients. The patients also complained of ear plugging sensation, hearing impairment, mass sensation, facial nerve palsy, headache, and neck pain and swelling. Duration of the symptoms

Table 1. Clinical data of patients with chondroblastoma of the temporal bone

Case	Sex	Age (yr)	Symptoms	Duration	Site	Initial treatment	Further treatment	Follow-up
1	F	41	Pain in TM joint	20 mo	right	Curettage	Chemotherapy Radiation therapy	27 mo NED
2	F	52	1. Ear plugging sensation 2. Decreased hearing 3. Pain in TM joint 4. Tinnitus 5. Mass sensation	20 yr 10 yr 10 yr 10 yr 10 yr	left	Excision	No treatment*	29 mo NED
3	F	57	1. Neck pain and swelling 2. Facial nerve palsy	20 day	left	Curettage	Radiation therapy	27 mo (recur at 26 mo)
4	F	58	Pain in TM joint	24 mo	right	Excision	No treatment	LAF
5	M	60	1. Hearing loss 2. Tinnitus 3. Headache	25 mo	left	Excision	Radiation therapy	37 mo NED

TM joint, temporomandibular joint; NED, no evidence of disease; LAF, lost at follow-up
*Chemotherapy for breast cancer detected after excision of chondroblastoma of the temporal bone

varied ranging from 20 days to 20 years.

Radiological findings

Preoperative radiological data were available in four of five cases. Each case showed similar characteristic findings. Computed tomography (CT) scan showed an irregularly lobulated expansile mass with bony destruction. The mass was heterogeneously enhanced after contrast administration (Fig. 1). The mass showed a heterogeneous low-signal intensity on T1 weighted images (Fig. 2A) and a high-signal intensity on T2-weighted images by magnetic resonance (MR) images. The mass was heterogeneously enhanced on post-contrast T1 weighted images (Fig. 2B).

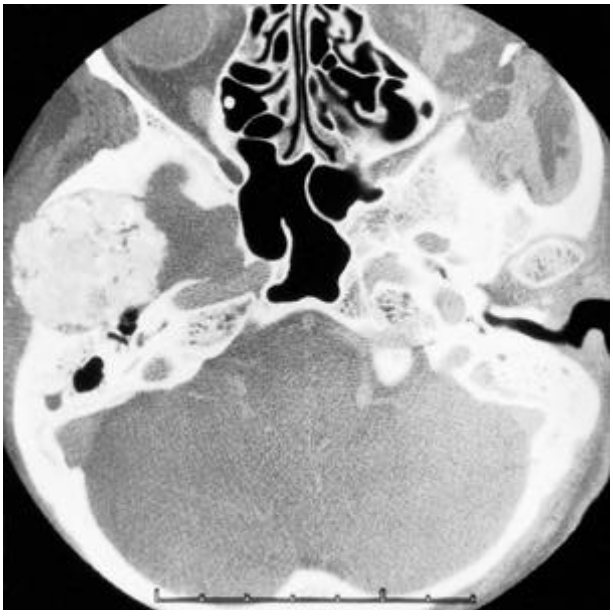

Fig. 1. Post-contrast CT scan shows a heterogeneously enhanced, irregularly lobulated mass in the right temporal bone (case 4).

Histological findings

All five cases showed histological findings of chondroblastoma of the usual location. With low power examination, multinucleated osteoclast-like giant cells were diffusely distributed throughout the mononuclear tumor cells (Fig. 3A). The tumor cells had eccentrically located oval to polyhedral nuclei with relatively abundant eosinophilic cytoplasm. Many of the tumor cells showed grooved nuclei (Fig. 3B). Mitotic figures were rare and atypical mitoses were absent. Distinct cartilage differentiation or formation of an immature cartilaginous matrix was occasionally observed within the tumor. In one case, the tumor was heavily calcified with areas of coarse granular calcification. Another case showed needle-shaped crystalline calcific deposits. The fine linear calcification between and around mononuclear cells showed a characteristic chicken wire pattern of calcification in three cases (Fig. 3C). One case showed focal spindle cell differentiation of mononuclear tumor cells. Two cases had aneurysmal bone cyst-like areas.

Immunohistochemical findings

The results of immunohistochemical study were summarized in Table 2. In three cases (case 1, 3, and 4), the mononuclear tumor cells were focally positive for S-100 protein. Some cells in the chondroid matrix were

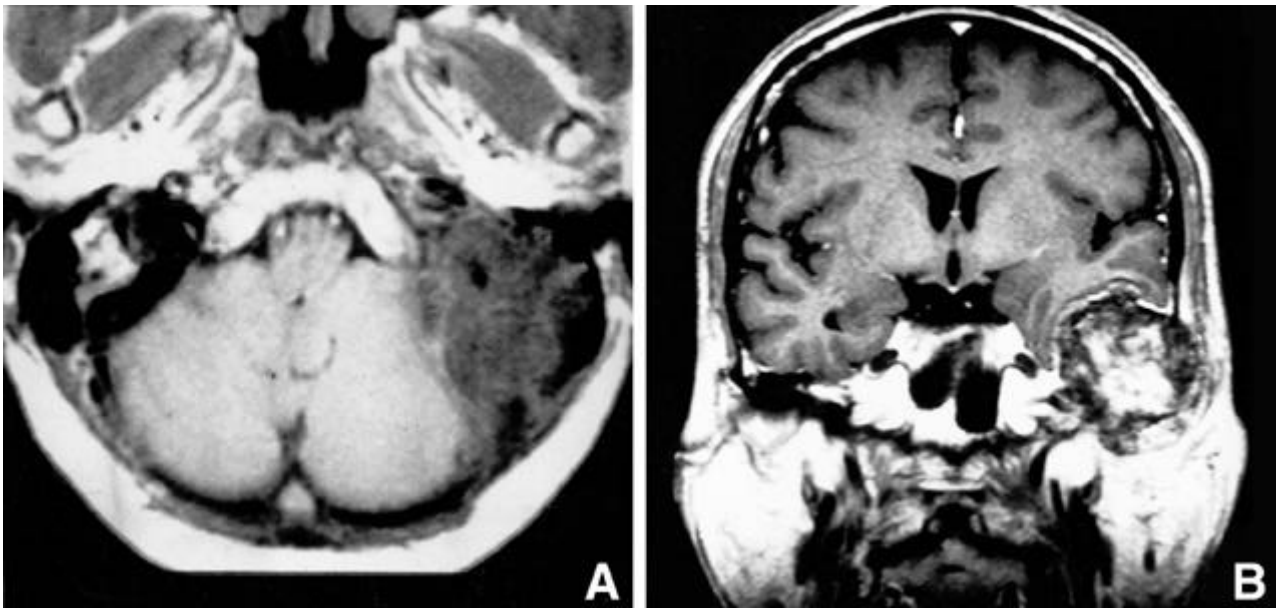

Fig. 2. A: Tumor in the left temporal bone shows a heterogeneous, low-signal intensity on T1 weighted image (case 3). B: Tumor in the left temporal bone is heterogeneously enhanced on post-contrast T1 weighted images (case 5).

more strongly positive (Fig. 4A), whereas, multinucleated giant cells were negative. In one case (case 4), the mononuclear tumor cells were weakly positive for cytokeratin (Fig. 4B). Cytokeratin was negatively stained in the multinucleated giant cells and in the chondroid matrix. CD 34 and CD 99 were negative in all five cases.

Treatment and follow-up

Data on treatment and follow-up results were summarized in Table 1. Sufficient follow-up data were available on four patients. One patient was lost at follow-up one month after surgery. Initial treatment was curettage in two patients and excision in three patients. Three patients had received further treatment: two patients received radiation therapy and one patient had combined chemotherapy and radiation therapy. One other patient had received chemotherapy for the treatment of breast cancer detected after excision of chondroblastoma of the temporal bone (case 2). Tumor recurred in one patient (case 3) 26 months after surgery. The other three patients had no recurrence, more than 27 months after surgery.

Table 2. Immunohistochemical findings of chondroblastoma of the temporal bone

Case No.	Antibody			
	Cytokeratin	S-100 protein	CD 34	CD 99
1	−	+	−	−
2	−	−	−	−
3	−	+	−	−
4	+	+	−	−
5	−	−	−	−

+, positive; −, negative

DISCUSSION

Chondroblastoma is a rare benign tumor of the bone. It accounts for less than 1% of all bone tumors. It usually affects epiphysis of the long tubular bones, including proximal tibia, proximal humerus, and distal femur. It also occurs in pelvis, scapula, spine, and ribs. Less frequently, it involves patella and calcaneus or other tarsal bones. Occasionally, chondroblastoma involves the craniofacial bones including temporal bone and mandible. The peak incidence of the chondroblastoma is during the second decade of life (1-3).

Chondroblastoma arising in the temporal bone is rare. Including one series of clinicopathologic study of 30 cases (5), only 38 cases have been reported and 31 of them had detailed pathologic and clinical data (4-10). The ages of the patients ranged from 2.9 years to 70 years. If one patient who was 2.9 years of age was excluded, the mean age was 43.5 years (5). The age of patients with chondroblastoma of the temporal bone is older than that of patients with chondroblastoma of usual sites, who are in their second decade of life. In our study, mean age was 53.6 years and it was older than that of previously reported cases of chondroblastoma of the temporal bone. It was also older than that of chondroblastoma of the usual sites.

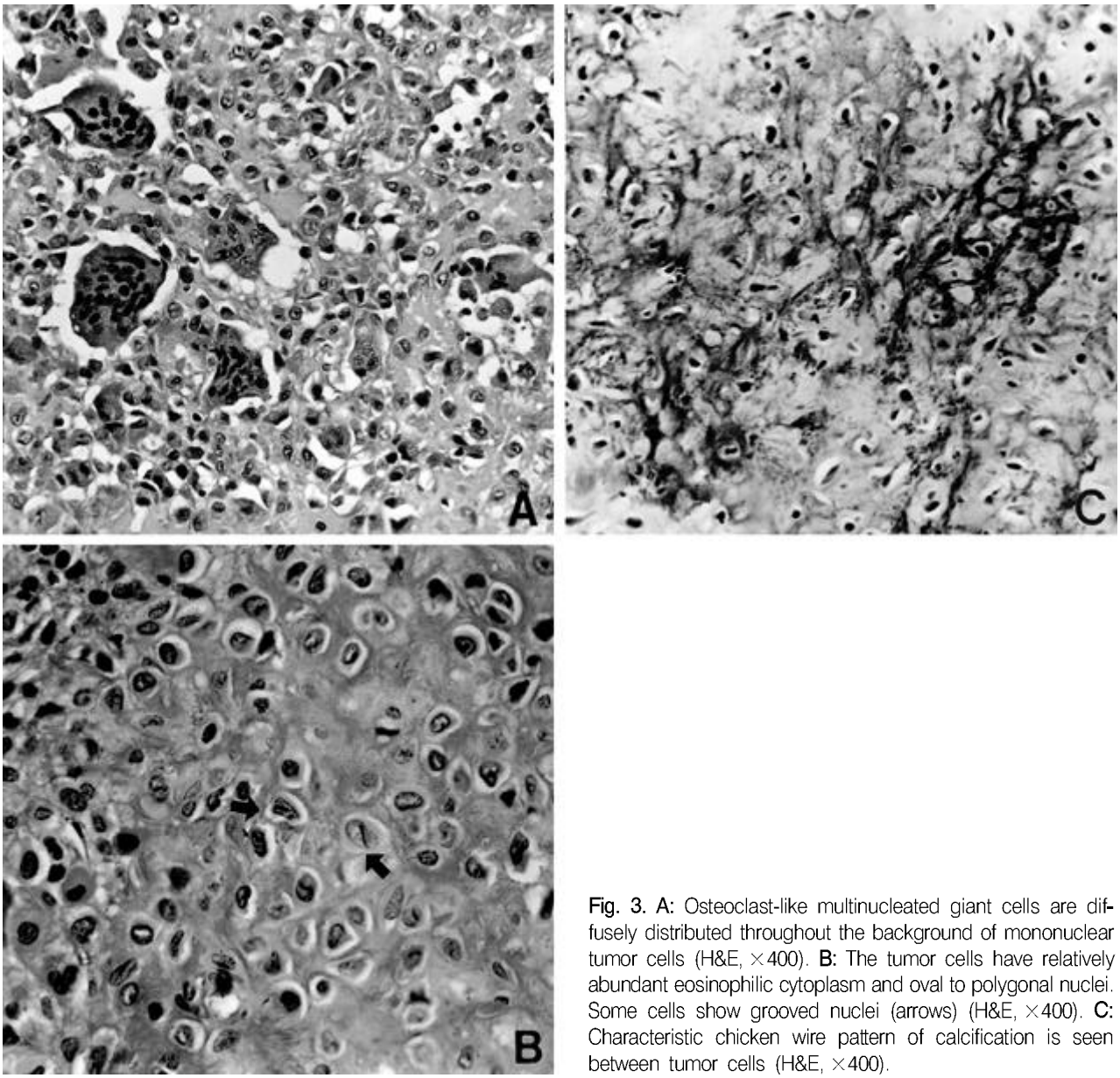

Fig. 3. **A:** Osteoclast-like multinucleated giant cells are diffusely distributed throughout the background of mononuclear tumor cells (H&E, ×400). **B:** The tumor cells have relatively abundant eosinophilic cytoplasm and oval to polygonal nuclei. Some cells show grooved nuclei (arrows) (H&E, ×400). **C:** Characteristic chicken wire pattern of calcification is seen between tumor cells (H&E, ×400).

Our CT scan findings revealed an irregularly lobulated expansile mass with bony destruction that was enhanced after contrast administration. These findings were similar to those of the previous report (10). However, in our study, the mass was peripherally enhanced but the central portion was not enhanced revealing a mass with heterogeneous enhancement. The MR findings of our cases were a mass with heterogeneously low signal on T1-weighted images and a high signal on T2-weighted images. These findings were compatible with those of previous report (10). However, the MR appearance of post-contrast T1-weighted images of chondroblastoma has not been described in the past. In our cases, the mass was heterogeneously enhanced on post-contrast T1-weighted images.

There have been immunohistochemical studies (11-16) done on chondroblastoma of usual sites. However, the immunohistochemical results of chondroblastoma of the temporal bone have not been described in the literature.

S-100 protein is an acidic calcium-binding protein that is present in central and peripheral neural tissue and neural tumor. It has been shown to be present in non-neural cells, such as the stellate cells of adenohypophysis, interdigitating reticulum cells of the lymphoid system, melanocytes and Langerhans cells of the skin, adipocytes and chondrocytes (11, 12). Weiss et al. (12) demonstrated immunoreactivity for S-100 protein in cartilaginous cells in both benign and malignant tumors, including enchon-

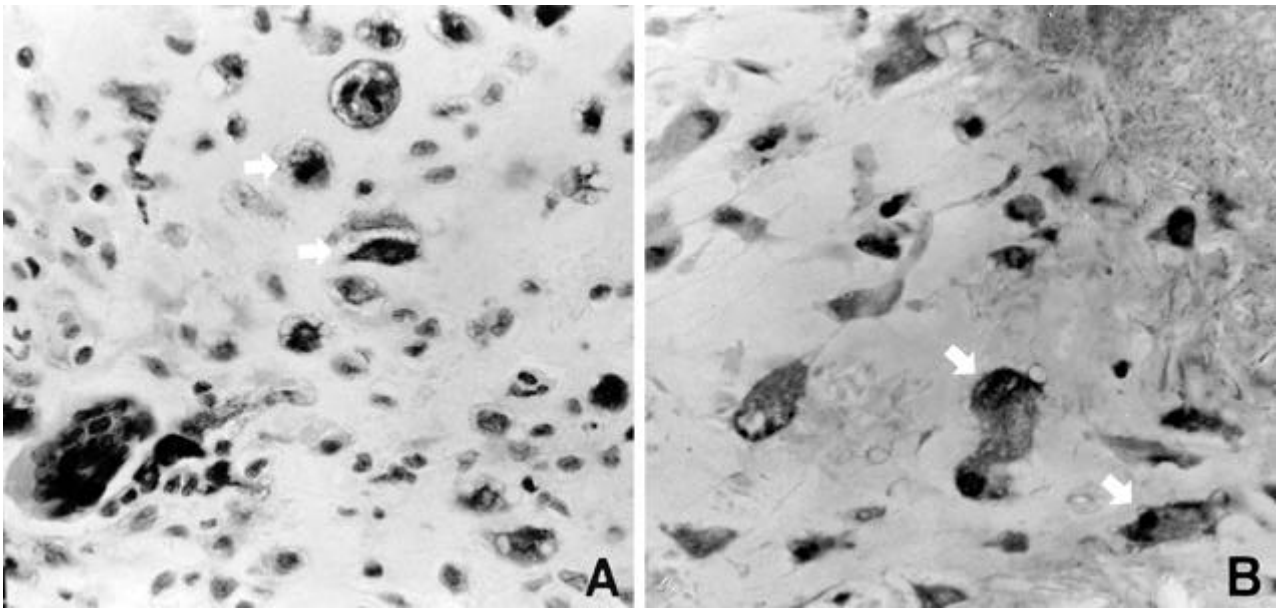

Fig. 4. A: The tumor cells are strongly positive for S-100 protein (arrows), whereas multinucleated giant cells are negative (ABC, ×400). B: Some tumor cells are weakly positive for cytokeratin (arrows). Background shows needle shaped crystalline calcific deposits (ABC, ×400).

droma, osteochondroma, chondromyxoid fibroma, chondroblastoma, and conventional and clear cell chondrosarcomas. Chondroblasts in the chondroblastoma were less reactive than chondrocytes in areas of chondroid matrix production in their study. In our study, the mononuclear tumor cells were focally positive and some cells in the chondroid matrix were more strongly positive for S-100 protein in three cases. However, multinucleated giant cells were negative. Nakamura et al. (13) found that immunoreactivity for S-100 protein in the anaplastic chondrocytes of mesenchymal chondrosarcoma and cartilaginous foci of osteosarcoma. Karabela-Bouropoulou et al. (14) also demonstrated immunoreactivity of the cells of the chondroblastoma with S-100 protein, while negative in giant cell tumor of bone. S-100 protein immunostaining may be helpful in differentiating chondroblastoma from giant cell tumor in giant cell rich neoplasms.

In our study, one case of the chondroblastoma was weakly reactive for cytokeratin in mononuclear tumor cells but not in multinucleated giant cells. This aberrant expression of cytokeratin in chondroblastoma was described by Semmelink et al. (15) and Edel et al. (16). However, the significance of this finding is unclear. The cytokeratin expression in chondroblasts may be an aberrant phenomenon or an indication of expression in early differentiation stages. Edel et al. (16) speculated that some chondroblasts in developing cartilaginous tissues might also express some types of cytokeratin if cytokeratin expression of chondroblasts indicates expression of early differentiation stage. But in their study, the fetal chondroblasts were negative with cytokeratin antibodies on paraffin embedded tissue. The findings of Edel et al. (16) further support that cytokeratin expression in chondroblastoma is most likely an aberrant phenomenon.

CD 34 and CD 99 immunostaining were performed in this study to differentiate other mesenchymal tumors as well as to demonstrate staining patterns in chondroblastoma. In chondroblastomas, these findings have not been studied. CD 34 has been shown to react with cells in variety of mesenchymal neoplasms (17). CD 99 immunoreactivity has been demonstrated in some bone tumors, including Ewing's sarcoma/primitive neuroectodermal tumor, small cell osteosarcoma, and mesenchymal chondrosarcoma (18, 19). Because of uniformly negative staining, these stains appeared to have minimal or no diagnostic utility for the diagnosis of chondroblastoma.

Chondroblastoma must be differentiated from giant cell tumor, chondrosarcoma, aneurysmal bone cyst, and pigmented villonodular synovitis. However, they could be differentiated from chondroblastoma by histological and immunohistochemical findings that were described above.

Different treatment modalities in the previously reported cases were as follows: total en bloc excision, curettage, irradiation, and irradiation combined with surgical excision. In our cases, one received curettage and adjuvant combined chemotherapy and radiotherapy. Two received curettage and adjuvant radiotherapy. One received excision and chemotherapy due to treatment of breast cancer that was detected after excision of chondroblastoma of the temporal bone. In one patient, the tumor

recurred 26 months after surgery. The other three patients showed no evidence of disease after more than 27 postoperative months. The recurred case had aneurysmal bone cyst-like areas. The previous study of Huvos and Marcove (20) found that there was a higher recurrence rate when chondroblastoma contained areas of aneurysmal bone cyst than chondroblastoma without aneurysmal bone cyst. In our series, we could not demonstrate any positive relationship between the presence of aneurysmal bone cyst component and the frequency of the recurrence due to the small number of cases.

In summary, chondroblastoma of the temporal bone is rare and occurs in an older age group than chondroblastoma of usual sites. It shows the same histological and immunohistochemical features as seen in chondroblastoma of usual sites. The chondroblastoma of the temporal bone is an irregularly lobulated expansile mass which shows a low-signal intensity on T1-weighted images and a high signal on T2-weighted images. It is heterogeneously enhanced after contrast administration. Although rare, chondroblastoma should be included in differential diagnosis when a well-demarcated, lobulated bony lesion occurs in the temporal bone.

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