The Role of Imaging in the Diagnosis of Giant Cell Tumor of the Skull Base

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Summary: An expansile temporal bone lesion above the temporomandibular joint (TMJ) in a 55-year-old woman was initially diagnosed as pigmented villonodular tenosynovitis. Preoperative embolization of the tumor had resulted in excessive tumoral hemorrhagic pigment. The surgical and CT findings suggested that the TMJ was not involved, and a final diagnosis of giant cell tumor was made.

Index terms: Skull, base; Skull, neoplasms

The histologic differential diagnosis of skull base lesions containing osteoclastic giant cells include giant cell tumor, giant cell reparative granuloma, pigmented villonodular tenosynovitis (PVNS), "brown tumor" of hyperparathyroidism, and giant cell–rich osteogenic sarcoma. Previous reports have emphasized the difficulty in differentiating giant cell tumor from giant cell reparative granuloma, both radiographically and histologically (1, 2). Several reports have also described cases of PVNS that were initially diagnosed as giant cell tumor on the basis of findings at aspiration biopsy or frozen section but that later were confirmed to be PVNS on final pathologic examination (3–5). This article describes a case of a large giant cell tumor involving the lateral skull base and temporal bone that was preoperatively embolized with polyvinyl alcohol particles. The embolization caused excessive intratumoral hemorrhage and focal areas of necrosis. The abundant hemosiderin deposition, which resulted from embolization, plus the proximity of the lesion to the temporomandibular joint (TMJ), led to an initial histologic diagnosis of PVNS. However, a review of the preoperative computed tomographic (CT) scans showed a rim of bone around the lesion and an unequivocally uninvolved mandibular head. This, in conjunction with the surgical finding of an intrinsically normal TMJ, led to a corrected final pathologic diagnosis of giant cell tumor of the skull base.

Case Report

A 55-year-old woman presented with a 1-year history of right-sided otalgia and facial pain, which had been diagnosed as TMJ syndrome. She reported no history of trauma or other significant medical history. There was no evidence of hyperparathyroidism. Her left TMJ was asymptomatic. During the 3 months preceding admission, she had noted an enlarging right scalp/facial mass. Physical examination showed evidence of local swelling in the right temporozygomatic region just superior to the TMJ and mouth opening, limited to 2 cm. The intraoral and remaining portions of the head and neck examination were unremarkable. Neurologic examination revealed no cranial nerve deficits.

CT scans of the temporal bones (Fig 1A–C) showed an expansile lesion involving the anterior portion of the right temporal bone. There was extension to the surface of the glenoid fossa; however, the mandibular head appeared normal. A preoperative right external carotid angiogram revealed a moderate degree of vascularity supplied from branches of the internal maxillary artery. Embolization was performed about 48 hours before surgery with polyvinyl alcohol particles (150 to 250 μm in size) (Contour Emboli, Interventional Therapeutics Corp, Fremont, Calif).

The surgical approach was via the preauricular infratemporal fossa. There was significant thinning of the squamous portion of the temporal bone and erosion caused by the tumor of the superior semicircular canal. Although the tumor extended to the glenoid fossa, there was no involvement of the TMJ. The tumor was completely excised, and the patient did well after surgery with no complications.

Histologic examination of the surgical specimen revealed multiple osteoclastic multinucleated giant cells (Fig 1D) and evidence of hemorrhage and hemosiderin deposition (Fig 1E). The extensive hemosiderin deposition as well as the tumor's proximity to the TMJ led to an initial diagnosis of PVNS. However, a review of the initial CT

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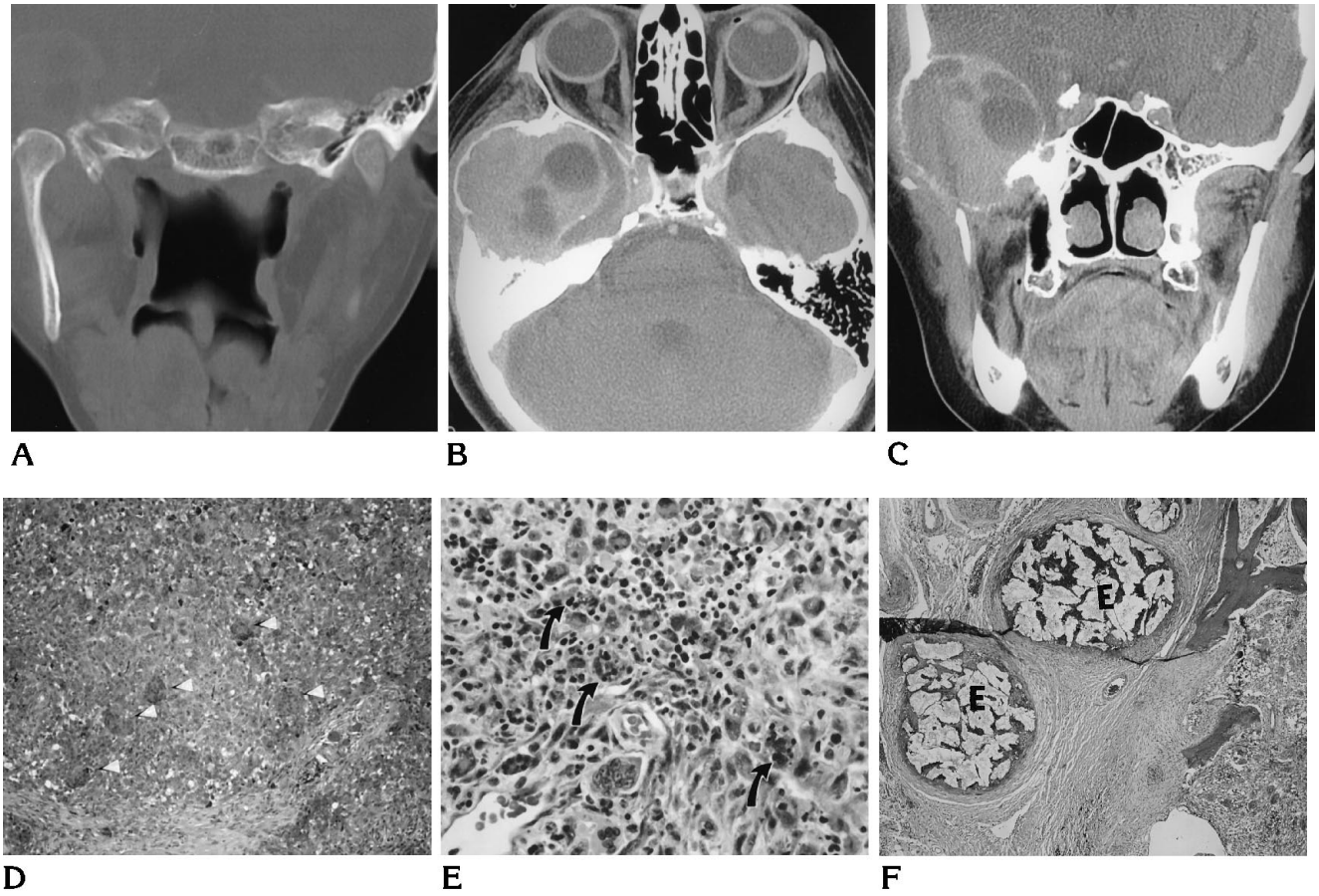

Fig 1. Fifty-five-year-old woman with TMJ syndrome.

Contrast-enhanced coronal CT scan (*A*) with a bone window setting shows partial destruction of the right temporal bone, including a portion of the glenoid fossa; however, the mandibular head is preserved. Axial (*B*) and coronal (*C*) contrast-enhanced CT scans at soft-tissue window settings show an expansile lesion involving the middle cranial fossa, with a surrounding thin rim of bone.

Low-power photomicrograph (*D*) shows multiple multinucleated osteoclastic giant cells (*arrowheads*) scattered uniformly throughout the tumor. High-power photomicrograph (*E*) shows coarse hemosiderin deposition within phagocytic cells (*arrows*). Low-power photomicrograph (*F*) shows polyvinyl alcohol emboli (*indicated by E*) in large arteries within the resected tumor.

scans, together with the intraoperative findings of no involvement of the synovium of the TMJ and the presence of polyvinyl alcohol emboli and hemorrhage within the resected tumor (Fig 1F), led to a final diagnosis of giant cell tumor. There was no histologic evidence of malignancy.

Discussion

Giant cell tumors, which most commonly occur in the epiphysis of long bones, are benign expansile tumors that are rich in osteoclastic giant cells. There is a high recurrence rate (40% to 60%) after resection (6). Giant cell tumors of the head and neck are uncommon, accounting for approximately 2% of all giant cell tumors (7). The most commonly involved sites are the sphenoid and temporal bones. There is a female predominance, and the age at presentation is usually in the third and fourth decades (8).

The clinical presentation depends on the site of origin. Giant cell tumor arising from the sphenoid bone usually manifests as headache, opthalmoparesis, trigeminal hypesthesia, and vision failure (9). Temporal bone giant cell tumor typically causes pain behind the ear on the affected side, deafness, and facial weakness. Histologically, the tumor shows a predominance of large multinucleated giant cells uniformly dispersed among the field (10). The nuclei of giant cell tumor are bland and variable in number, with approximately 10 to 20 nuclei per cell, although some cells may have 100 or more nuclei. These giant cells are dispersed within spindle-shaped stromal cells, which are short and plump with nuclei identical to those within the giant cells, unlike the fibroblastic spindle cell background of giant cell reparative granu-

loma. Fibroblasts are generally larger and more tapered than the stromal cells of giant cell tumors. They have abundant eosinophilic cytoplasm as they produce collagen. Hemorrhage and hemosiderin deposition, as was seen in this case, are not usually prominent features of a giant cell tumor.

On imaging, giant cell tumor usually has the nonspecific appearance of an expansile, destructive soft-tissue mass (11). There are no radiographically distinguishing features of a giant cell tumor of the skull base, and the differential diagnosis usually includes chondrosarcoma, osteolytic metastasis, and other fibroosseous lesions, as well as giant cell reparative granuloma (7).

The giant cell reparative granuloma may have an identical imaging appearance as that of a giant cell tumor (6). Giant cell reparative granulomas are rare in the axial skeleton (extragnathic giant cell reparative granuloma) and, at this site, are also referred to as "solid" aneurysmal bone cysts (12). In the head and neck, giant cell reparative granuloma is typically found in the mandible and maxilla, but has been reported in other sites, such as the frontal bone (13). In the jaws, giant cell reparative granuloma has also been referred to as *central* (referring to intraosseous) *giant cell granuloma*. Jaffe (14) originally suggested that giant cell reparative granuloma was not a true tumoral process but a proliferative lesion in response to hemorrhage or trauma. On the other hand, given its expansile, nonresolving nature, it is not truly a "reparative" lesion. Reports of accelerated growth, recurrence, or persistence (despite curettage) during pregnancy and in the postpartum period raise the possibility that giant cell reparative granuloma is hormone-dependent (15). There is a female:male ratio of 2:1, and the peak age of occurrence is in the second decade of life (16).

Historically, there has been difficulty distinguishing giant cell reparative granuloma from giant cell tumor. Hirschl and Katz (1) reviewed 23 cases that had been diagnosed as giant cell tumor arising in the temporal bone and concluded that 18 of these were more likely giant cell reparative granuloma. There are important clinical and histologic differences between giant cell tumor and giant cell reparative granuloma. Histologically, giant cell reparative granuloma has perivascular hemorrhage with clustering of giant cells around the hemorrhage and hemosiderin deposition, whereas in giant cell tumor, there is usually uniform distribution of giant cells and hemosiderin deposits are rare. Also, in giant cell reparative granuloma, the giant cells are generally smaller and have relatively fewer nuclei than do those in giant cell tumor. There may be new reactive bone and chondroid elements seen within giant cell reparative granuloma, which are not present in giant cell tumor. The background stroma of giant cell reparative granuloma consists of fibroblastlike, tapered spindle cells and dense scarlike collagen compared with the usual plump, oval cells in giant cell tumor (1).

There are also important clinical differences between giant cell tumor and giant cell reparative granuloma. Giant cell reparative granuloma occurs in a younger age group (second decade of life) and has a propensity for the jaws as compared with giant cell tumor (third and fourth decades), which has a predilection for the skull base. Giant cell reparative granulomas usually have a benign course, with surgical curettage resulting in a complete cure, although there is a 10% to 15% recurrence rate after incomplete surgical removal (2). This is in contrast to giant cell tumors, which have a recurrence rate as high as 40% to 60% and can potentially (though uncommonly) metastasize.

PVNS involving the temporal bone is rare, with less than 15 cases described in the literature (3, 17). PVNS is a benign, proliferative tumor involving either the synovium or tendon sheath. The hip and knee are the most common sites of PVNS. It can occur in two forms: a diffuse form involving the entire synovium of the joint or a localized form presenting as a solid nodule. PVNS involving the TMJ is usually the diffuse form (18). Imaging studies typically show erosion of bone and cartilage, as well as cystic (expansile) changes of the adjacent bone (19). Histologically, there is proliferation of large round and spindle-shaped cells, plump epithelioid histiocytic cells, foam cells, and multinucleated osteoclastic giant cells with abundant hemosiderin deposition (20).

In the case presented, a giant cell tumor was treated with preoperative embolization, which caused tumor necrosis, hemorrhage, and hemosiderin deposition. The combination of giant cells with hemosiderin deposition in a lesion near the TMJ prompted an initial diagnosis of PVNS. However, the lack of erosive changes involving the mandibular head on CT scans and

at surgery, and the lack of synovial involvement of the TMJ at surgery, made the diagnosis unlikely. The pathologic and imaging findings were reexamined together, and this led to the final diagnosis of giant cell tumor. Previous reports have tended to emphasize the difficulty in differentiating giant cell tumor from giant cell reparative granuloma. However, the radiologist also needs to be aware of the histologic similarities between giant cell tumor and PVNS involving the temporal bone, and how preoperative interventional radiologic techniques can cause a giant cell tumor to simulate the histologic appearance of PVNS.

References

1. Hirschl S, Katz A. Giant cell reparative granuloma outside the jaw bone. *Hum Pathol* 1974;5:171–181
2. Colclasure JB, Shea MC Jr, Graham MA. Giant cell lesions of the temporal bone. *Am J Otol* 1981;2:188–192
3. Eisig S, Dorfman HD, Cusamano RJ, Kantrowitz AB. Pigmented villonodular synovitis of the temporomandibular joint. *Oral Surg Oral Med Oral Pathol* 1992;73:328–333
4. Rickert RR. Pigmented villonodular synovitis of the temporomandibular joint. *Otolaryngol Head Neck Surg* 1982;90:668–670
5. Lapayowker MS, Miller WT, Levy WM, Harwick RD. Pigmented villonodular synovitis of the temporomandibular joint. *Radiology* 1973;108:313–316
6. Rosen MP. Giant-cell tumor of the temporal bone. *AJR Am J Roentgenol* 1991;156:1290–1292
7. Saleh EA, Taibah AK, Naguib M, et al. Giant cell tumor of the lateral skull base: a case report. *Otolaryngol Head Neck Surg* 1994;111:314–318
8. Rock JP, Mahmood A, Cramer HB. Giant cell tumor of the skull base. *Am J Otol* 1994;15:268–272
9. Findlay JM, Chiasson D, Hudson AR, Chui M. Giant cell tumor of the middle cranial fossa. *J Neurosurg* 1987;66:924–928
10. Cook HF, Miller R, Yamada R. Giant cell tumor of the infratemporal fossa: report of a case. *J Oral Maxillofac Surg* 1986;44:651–656
11. Bertoni F, Unni KK, Beabout JW, Ebersold MJ. Giant cell tumor of the skull. *Cancer* 1992;70:1124–1132
12. Oda Y, Tsuneyoshi M, Shinohara N. Solid variant of aneurysmal bone cyst (extragnathic giant cell reparative granuloma) in the axial skeleton and long bones. *Cancer* 1992;70:2642–2649
13. Felsberg GJ, Tien RD, McLendon RE. Frontoethmoidal giant cell reparative granuloma *AJNR Am J Neuroradiol* 1995;16:1551–1554
14. Jaffe HL. Giant cell reparative granuloma, traumatic bone cyst, and fibrous (fibro-osseous) dysplasia of the jaw bones. *Oral Surg* 1953;6:159–175
15. Fechner RE, Slaughter FH, Pope TH. Extraordinary growth of giant cell reparative granuloma during pregnancy. *Arch Otolaryngol* 1984;110:116–119
16. Kaugers GE, Niamtu J, Svirsky JA. Cherubism: diagnosis, treatment, and comparison with giant cell granulomas and giant cell tumors. *Oral Surg Oral Med Oral Pathol* 1992;73:369–374
17. Franchi A, Frosini P, Santoro R. Pigmented villonodular synovitis of the temporomandibular joint: report of a case. *J Laryngol Otol* 1994;108:166–167
18. Curtin HD, Williams R, Gallia L, Meyers EN. Pigmented villonodular synovitis of the temporomandibular joint. *Comput Radiol* 1983;7:257–260
19. O'Sullivan TJ, Alport EC, Whiston HG. Pigmented villonodular synovitis of the temporomandibular joint. *J Otolaryngol* 1984;13:123–126
20. Miyamoto Y, Tani T, Hamaya K. Pigmented villonodular synovitis of the temporomandibular joint. *Plast Reconstr Surg* 1977;59:283–286