



# Recurrent Temporal Bone Tenosynovial Giant Cell Tumor with Chondroid Metaplasia: the Use of Imaging to Assess Recurrence

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**Key words:** tenosynovial giant cell tumor, temporal bone, CT, MR, PET

**SUMMARY** – *Tenosynovial giant cell tumor (TGCT) is a benign proliferative lesion of unclear etiology. It is predominantly monoarticular and involves the synovium of the joint, tendon sheath, and bursa. TGCT of the temporomandibular joint (TMJ) is rare and aggressive resulting in destruction of surrounding structures. The diagnosis may be suggested by imaging, mainly by the MR features and PET/CT, and confirmed by histopathology. We describe the case of a 50-year-old man who presented with right-sided hearing loss, tinnitus and TMJ pain. Pathology revealed tenosynovial giant cell tumor with chondroid metaplasia. Six years later he developed a recurrence, which was documented to our knowledge for the first time with CT, MR and FDG PET/CT imaging.*

## Introduction

Tenosynovial giant cell tumor (TGCT) is a benign proliferative lesion of unclear etiology, which is predominantly monoarticular and involves the synovium of the joint, tendon sheath, and bursa. TGCT of the temporomandibular joint (TMJ) is rare with only 58 cases having been reported. It may be clinically misinterpreted as a parotid mass, may involve the adjacent skull base, and may have variable extension to the temporal bone.

We describe a case of TGCT arising from the temporomandibular articular capsule and extending into the temporal bone, with recurrence after six years. Histologically, the mass presented areas of classic TGCT with foci of chondroid metaplasia. The mass was completely documented with CT, MR and FDG PET/CT imaging, which to our knowledge is the first such case to be documented with all of these imaging modalities.

## Case Report

A 50-year-old man with a five-year history of right-sided hearing loss and TMJ pain presented with constant, non-pulsatile tinnitus. The otologic examination at presentation was unremarkable. Imaging studies revealed a destructive lesion in the right infratemporal fossa and glenoid fossa, extending into the overlying temporal bone. An ill-defined lytic mass within the right temporal bone was documented on CT (Figure 1A), with slightly enhancing soft tissue components extending over the roof of the glenoid fossa without gross abnormality of the mandibular condyle. The margins of the lesion had a scalloped appearance. There was extension to the external auditory canal and erosion along both the anterior and posterior margins of the canal. This mass showed increased F18-FDG uptake in the temporal bone (max SUV of 6.4) and mildly increased FDG uptake in the soft tissue of the external auditory canal (max

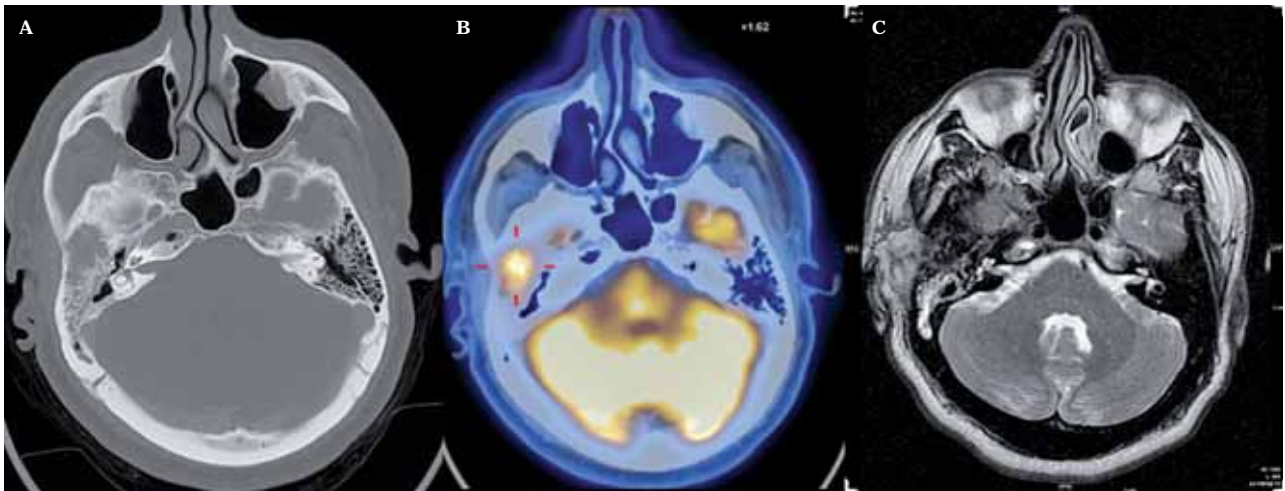


Figure 1 A) Preoperative axial CT scan in bone algorithm shows an ill-defined lytic mass within the right temporal bone. B) Preoperative PET-CT with corresponding area of increased avidity for F18-FDG. C) Preoperative axial T2-weighted imaging reveals predominantly hypointense right temporal mass.

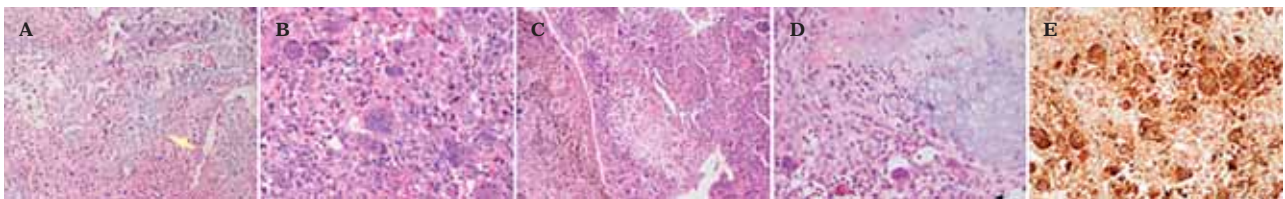


Figure 2 A) Initial biopsy showing chondroid matrix with “chicken wire” calcification (arrow) resembling chondroblastoma. H&E, original magnification,  $\times 20$ . B) Resection specimen showing conventional tenosynovial giant cell tumor with mononuclear histiocyte-like cells, scattered osteoclast-type giant cells and hemosiderin pigment. H&E, original magnification,  $\times 20$ . C) Tenosynovial giant cell tumor with chondroid metaplasia showing abundant hemosiderin pigment on the left side and chondroid area in the center. H&E, original magnification,  $\times 10$ . D) Closer view of the interface between classic tenosynovial giant cell tumor and area of chondroid metaplasia. H&E, original magnification,  $\times 20$ . E) Multinucleated giant cells and a subpopulation of the mononuclear cells are positive for CD68. CD68 immunostain, original magnification,  $\times 20$ .

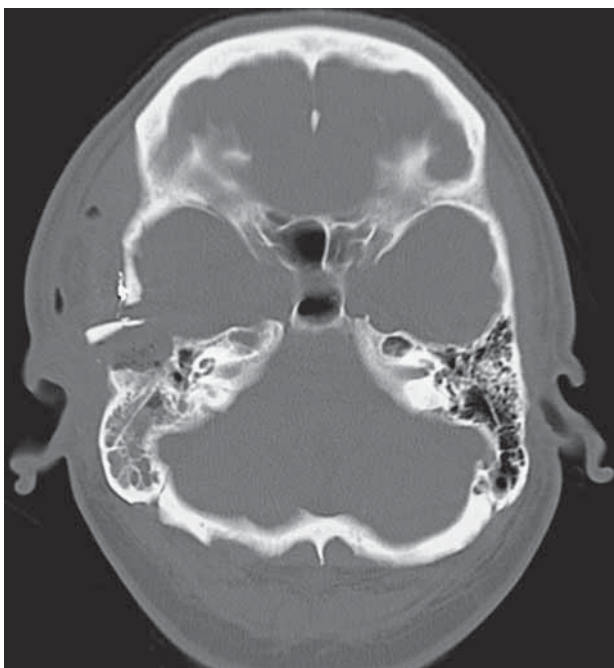


Figure 3 Immediate postoperative axial CT scan in bone algorithm reveals a surgical bed with smooth bony margins.

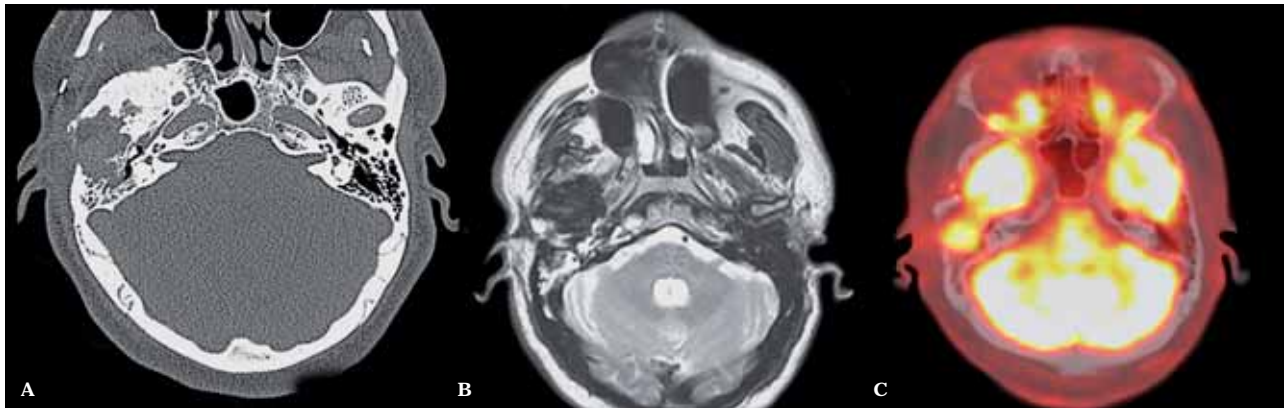


Figure 4 A) Axial CT scan in bone algorithm, of 6 years of follow-up, shows a lytic lesion with irregular, ill-defined bony margins. B) Follow-up MR with axial T2-weighted imaging hypointensity, not specific requiring correlation with PET avidity for F18-FDG. C) PET-CT shows increased F18-FDG avidity in the temporal portion of the surgical bed, reflecting recurrence.

SUV of 3.5) (Figure 1B). On MR imaging the soft tissue mass presented a predominantly hypointense signal on T1- and T2-weighted images (Figure 1C). There was no significant adenopathy in the neck region.

A biopsy was initially performed through a right preauricular incision, which exposed tumor invading the cartilaginous portion of the external auditory canal and the superior and medial aspects of the glenoid fossa. The capsule of the condyle was resected, where there was obvious destruction and tumor, as well as the nidus of the origin of the tumor. A temporoparietal approach then allowed resection of the remaining temporal bone tumor. The initial biopsy revealed histological features similar to a chondroblastoma of bone, with “chicken wire” calcifications within a chondroid matrix (Figure 2A). Pathological examination of the resection specimen showed tumor involving synovium and bone with classic features of TGCT (Figure 2B), including mononuclear histiocyte-like cells, osteoclast-type giant cells, extensive hemosiderin deposition and areas of chondroid metaplasia (Figure 2C and/or 2D), focally resembling chondroblastoma. The tumor cells were diffusely positive for CD68 and focally positive for S100 protein. The final diagnosis was tenosynovial giant cell tumor with chondroid metaplasia.

Postoperative CT and MR studies showed soft tissue material throughout the operative bed representing a flap and inflammatory operative changes. Of note were smooth post surgical bony margins (Figure 3). No suspected residual tumor was observed at that time. How-

ever, MR and CT imaging performed six years after the surgery again showed a lytic lesion with scalloped bony margins extending into the greater wing of the sphenoid bone associated with mildly enhancing soft tissue extending back into the mastoid and middle ear (Figure 4A,B). These features were suspicious for recurrent tumor, confirmed on a new PET/CT, which showed a mass with increased 18-FDG uptake in the operative bed of the right temporal bone (Figure 4C). Additionally there was a diffuse sclerotic reaction in the greater sphenoid wing and right mandibular condyle, and there was an enhanced soft tissue thickening around the lateral aspect of external auditory canal, compatible with postoperative changes. The suspected recurrence was biopsy proven and wide surgical removal was performed.

## Discussion

Tenosynovial giant cell tumor (TGCT) is defined by the World Health Organization as a locally aggressive neoplasm composed of synovial-like mononuclear cells admixed with multinucleate giant cells, foam cells, siderophages and inflammatory cells. It affects most commonly the knee and hip in young adults<sup>1</sup>. Involvement of TMJ is rare. In the current literature there are 58 cases described with one third of them having intracranial involvement<sup>2</sup>. Some authors advocate that the majority of tumors affecting the TMJ are extraarticular, more infiltrative, extend to the temporal bone, and have higher recurrence rates<sup>1,5</sup>. By defi-



nition, the extraarticular form of TGCT consists of an infiltrative soft tissue mass, with or without involvement of the adjacent joint<sup>1</sup>. The macroscopic appearance is usually multinodular, instead of presenting the typical villous patterns seen in the intraarticular forms<sup>1</sup>.

Skull base involvement and intracranial extension have been previously described<sup>3-6</sup> as has a case presenting as a middle cranial fossa mass<sup>7</sup>. The rare malignant form of TGCT with metastatic lesions in the lungs and lymph nodes has also been documented<sup>8</sup>.

Recurrence is common, may be multiple, with 33-50% developing as the extraarticular form<sup>1</sup>. As recurrences are associated with positive surgical margins, wide excision should be the treatment of choice<sup>1,9</sup>. Cai et al. reported a recurrence rate of 11% for TGCT of the TMJ<sup>6</sup>. Genetic studies have revealed a translocation involving the genes CSF-1 on chromosome 1 and COL6a3 on chromosome 2 in primary lesions, as well as recurrences and metastases<sup>1,10</sup>.

Clinically, patients usually have symptoms for many years and the most common complaints are preauricular swelling, pain or limitation of motion<sup>1,2,9,11</sup>. Imaging features of TGCT of the TMJ consist of a soft tissue mass frequently associated with bone erosion with scalloped margins and cyst formation as better depicted with CT and areas of hyperattenuation due to iron deposition<sup>9</sup>. On MR, the nodular masses usually exhibit low signal intensity on T1- and T2-weighted images, reflecting characteristic hemosiderin deposition<sup>2,9,11</sup>. However the MR appearance may vary depending on the proportions of lipid, hemosiderin, fibrous stroma, pannus, fluid, and cellular elements<sup>6</sup>. MR is sensitive and specific for the diagnosis of TGCT and also helps in surgical planning. As in our case, avidity on FDG PET/CT has been described<sup>12</sup>.

Histopathological features include synovial-lined spaces, blood-filled pseudoalveolar spaces in 10% of the cases, mitotic activity, variable cellularity, with small or large histiocyte-like cells, osteoclast-like multinucleated giant cells and hemosiderin deposits<sup>1,2,13</sup>. Giant cells are positive for CD68 and CD45, and mononuclear cells are positive for CD681. Interestingly O'Connell et al. document that 67% of TGCT contain dendritic cells expressing S100 protein<sup>14</sup>. A minority of cells are neoplastic and overexpress CFS<sup>12,13</sup>. Cartilaginous and osseous metaplasia are rare<sup>13</sup>.

In our case there was a mixed immunophe-

notype pattern with histiocyte-like and osteoclast-like giant cells positive for CD68, and also mononuclear cells in chondroid areas positive for S100 protein. This mixed histopathology was previously reported by Oda et al. who documented two cases and by Hoch et al.<sup>13</sup> who documented five cases of TGCT with chondroid metaplasia of the TMJ containing hyaline chondroid nodules and dystrophic calcification in chondroid areas, histologically mimicking chondroblastoma. Although TGCT of the TMJ is rare, interestingly the TMJ and temporal bone are the most common sites of skull chondroblastoma<sup>13</sup>.

Chondroblastoma should be in the differential diagnosis, as imaging features may overlap, and the histopathological distinction from TGCT may be hard to establish<sup>13</sup>. The potential relationship to chondroblastoma remains unclear<sup>13</sup>. The differential diagnosis of TGCT on MRI also includes synovial chondromatosis, tumoral calcium pyrophosphate dihydrate crystal deposition disease, rheumatoid arthritis, synovial sarcoma, hemophilia, and synovial hemangioma<sup>2,3,9,13</sup>. The occurrence of TGCT with synovial chondromatosis is also reported in the literature<sup>15</sup>, but chondromatosis is characterized by the presence of loose bodies (hyaline cartilage) with minimal calcification.

Finally, complete excision with wide margins is the treatment of choice<sup>4</sup>. Radiation therapy may be considered when vital structures are involved and to prevent recurrence<sup>4,8</sup>. We suggest that MR and PET/CT, when available, should be strongly considered in the follow-up to monitor tumor recurrence<sup>7</sup>.

## Conclusion

TGCT of the TMJ with chondroid metaplasia is benign but locally destructive. Although the definite diagnosis is histological, CT, MR and now FDG PET/CT imaging play an important role in the preoperative setting, excluding other important etiologies and suggesting the diagnosis of TGCT if hemosiderin deposition is documented. Our case reinforces evidence that TGCT with chondroid metaplasia should be considered in the differential diagnosis of TGCT of the TMJ. The gold standard treatment is complete resection with good long-term outcome. Our case suggests that the best imaging to assess a recurrence may be MR and PET/CT.

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