

Imaging features for diffuse-type tenosynovial giant cell tumor of the temporomandibular joint

A case report

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Abstract

Rationale: The tenosynovial giant cell tumor (TGCT) is a benign but locally aggressive tumor that arises from the synovial membrane of joints, tendon sheaths, and bursae. Although any joint can be affected, involvement of the temporomandibular joint (TMJ) was reported very rarely, and there is no relevant report on ¹⁸F-FDG PET/computerized tomography (CT).

Patient concerns and diagnoses: We present here a rare case of diffuse-type of TGCT (D-TGCT) arising from the right TMJ in a 74-year-old woman. The patient was discovered a mass of the right temporal fossa during a head CT scan. However, she did not receive any treatment and was discharged from the hospital. She visited our institution again after 4 years with worsening headache and swelling of the right preauricular area. An enhanced CT demonstrated a 6.0 × 3.4 × 5.0 cm mass of mixed density involving the right TMJ, with evident enhancement and extensive erosion of adjacent bones. Magnetic resonance imaging (MRI) showed hypointensity in the solid part of the mass but high signal intensity in the cystic part or necrosis on T2-weighted images (T2WI). In ¹⁸F-FDG PET/CT images, the solid portion of the mass had increased FDG uptake with a SUVmax of 19.8. It was then diagnosed as D-TGCT by postoperative pathology.

Lessons: The case report shows the imaging features of the TGCT, including CT, MRI, and ¹⁸F-FDG PET/CT, especially the typical hypointensity on T2WI. Careful preoperative examination and complete resection are the factors that lead to the optimal treatment of the TGCT.

Abbreviations: CT = computerized tomography, D-TGCT = diffuse type of TGCT, MRI = magnetic resonance imaging, T2WI = T2-weighted image, TGCT = tenosynovial giant cell tumor, TMJ = temporomandibular joint.

Keywords: CT, FDG PET/CT, MRI, temporomandibular joint, tenosynovial giant cell tumor

1. Introduction

The tenosynovial giant cell tumor (TGCT) is a benign but locally aggressive tumor that arises from the synovial membrane of joints, tendon sheaths, and bursae. The term of TGCT was introduced by Jaffe et al^[1]. It is considered as fibrohistiocytic tumor by the World Health Organization classification of bone and soft tissue tumors.^[2] Broadly, TGCT is classified according to growth pattern, including localized and diffuse forms. The localized form of TGCT is more prevalent, frequently affecting the small joints of the hands and feet. In contrast, diffuse-type of TGCT (D-TGCT) is less common, which is also referred to as the synonym of pigmented villonodular synovitis.^[3] It typically affects the knee, hip, or shoulder, and rarely presents in the temporomandibular joint (TMJ).^[4–6] Imaging features of the

TGCT is specific, especially the typical hypointensity on T2-weighted images (T2WI) of the magnetic resonance imaging (MRI). Here, we present a rare case of a female patient diagnosed as D-TGCT of the right TMJ with the aim to share the imaging features of the TGCT including computerized tomography (CT), MRI and ¹⁸F-FDG PET/CT.

2. Case report

A 74-year-old woman had a mild and enduring headache occasionally 4 years ago. Her families insisted on taking her to the hospital for a head CT examination, and a 5.5 × 3.0 × 4.5 cm mass was found on the right temporal fossa (Fig. 1). Because the quality of her life had not been affected by the mild headache, she did not receive any treatment and was discharged from the hospital. She visited our institution again after 4 years with worsening headache and swelling of the right preauricular area. Axial and coronal head enhanced CT and MRI scans were subsequently obtained.

Head enhanced CT scan demonstrated a 6.0 × 3.4 × 5.0 cm mass with mixed density involving the right TMJ, increased than before in size, with evident enhancement and extensive adjacent bony erosion, including right temporal bone, zygomatic bone, and the right greater sphenoid wing. Coronal images demonstrated that the tumor did not infiltrate the brain parenchyma with the complete dura mater (Fig. 2). MRI showed a large lobulated mass involving the right TMJ, which was mixed appearance with hypo-, hyper-, and isointensity on coronal T1-weighted images and hypointensity in the solid part but high signal intensity in the cystic part or necrosis on axial T2WI (Fig. 3). An increased FDG uptake for the mass in the right TMJ

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Figure 1. Axial head computerized tomography demonstrating a cystic-solid mixed mass involving the right temporomandibular joint, with scattered calcification (4 years ago).

region was found in the maximum intensity projection image (Fig. 4) and axial and coronal images of ^{18}F -FDG PET/CT, with a SUVmax of 19.6 (Fig. 5). No other abnormal focally increased activity was detected in whole-body ^{18}F -FDG PET/CT images.

Intraoperative view showed that the tumor located above the right zygomatic bone, and the hernian adjacent the tumor had been widely damaged involving the right TMJ. The tumor was tightly adhered to adjacent tissue. With the complete dura mater, the tumor did not infiltrate the medial cranial fossa. Photomicrograph for the pathologic specimen showed that compact hyperplastic monocytes, with the potential to differentiate into foam cells and fibroblast. There were also a number of scattered hemosiderin-laden macrophages, multinucleated giant cells, and pale blue calcification in the fibrous stroma. Immunohistochemistry showed that the tumor was strongly positive for Vimentin and CD68, focally positive for Ki-67, CD31, CD56, and CD34, and negative for AE1/AE3(PCK), EMA, CK7, CK20, S-100, NSE, GFAP, PR, HMB-45, and Desmin (Fig. 6). Combining the histological information with the clinical presentation yielded a diagnosis of D-TGCT of the right TMJ.

The symptoms of headache and swelling of the right preauricular area have disappeared after the operation. Up to

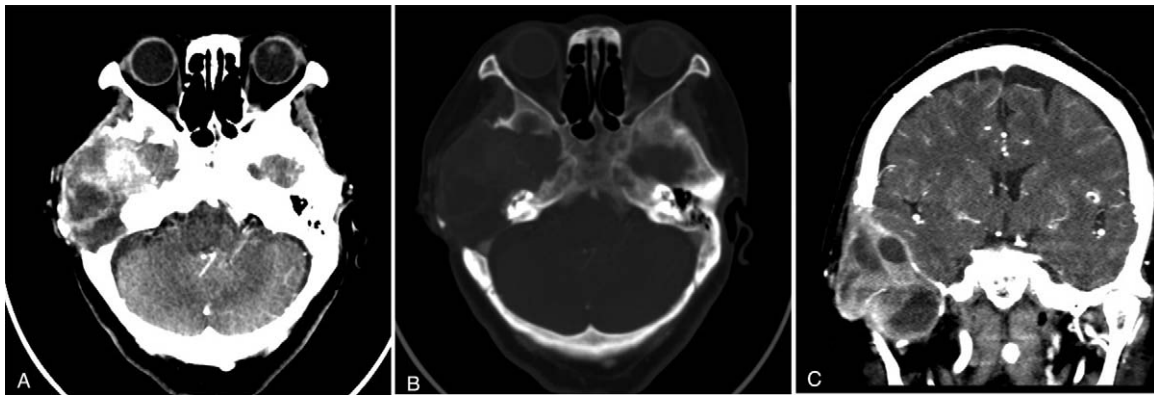


Figure 2. Contrast-enhanced computerized tomography (CT) images of the head demonstrated a mass with mixed density involving the right temporomandibular joint, with evident enhancement in the solid part (A, axial). The bone window showed the large mass causing extensive erosion of right temporal bone, zygomatic bone, and the right greater sphenoid wing (B, axial). Coronal CT images demonstrated that the tumor did not infiltrate the brain parenchyma with the complete dura mater (C, soft tissue window).

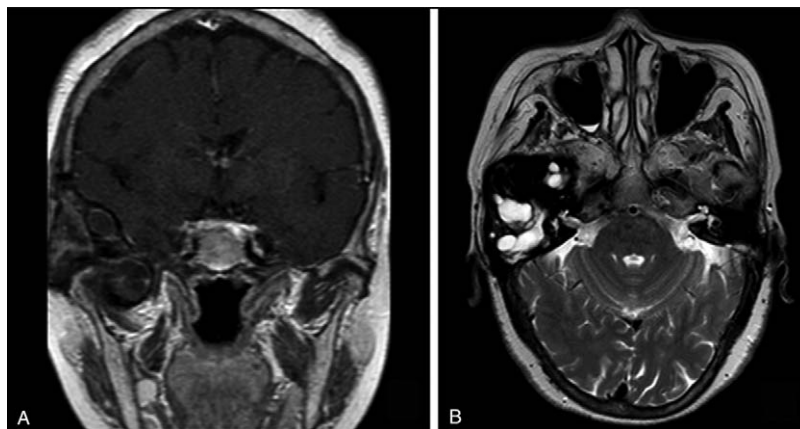


Figure 3. Magnetic resonance imaging showed a large lobulated mass involving the right temporomandibular joint, which was mixed appearance with hypo-, hyper-, and isointensity on coronal T1-weighted images (A) and hypointensity in the solid part but high signal intensity in the cystic part or necrosis on axial T2-weighted images (B).

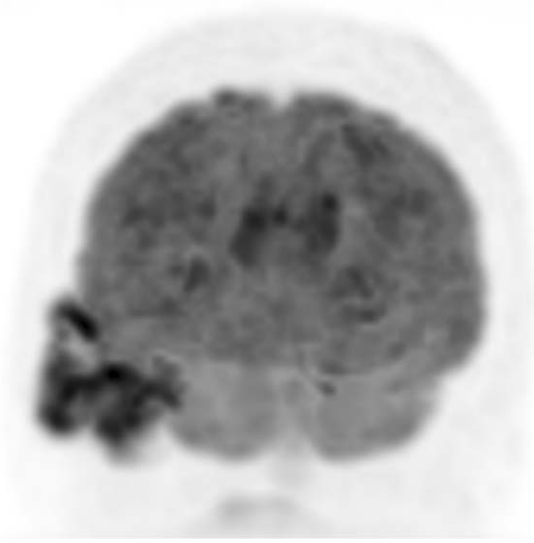


Figure 4. An increased FDG uptake located in the right temporomandibular joint region was showed in the maximum intensity projection image of ¹⁸F-FDG PET/computerized tomography.

the nearest follow-up (a half year after operation), there was no discomfort and recurrent imaging findings in radiology.

3. Discussion

The TGCT is a benign but locally aggressive tumor that arises from the synovial membrane of joints, tendon sheaths, and bursae. Although any joint can be affected, involvement of the TMJ was reported very rarely. Clinically, the TGCT usually occurs in all decades of life with predominance in the third or fourth decade.^[7] The etiology of TGCT is controversial, someone has suggested that recurrent trauma and hemarthrosis may be the contributing factor.^[8] Microscopically, the tumor is composed of

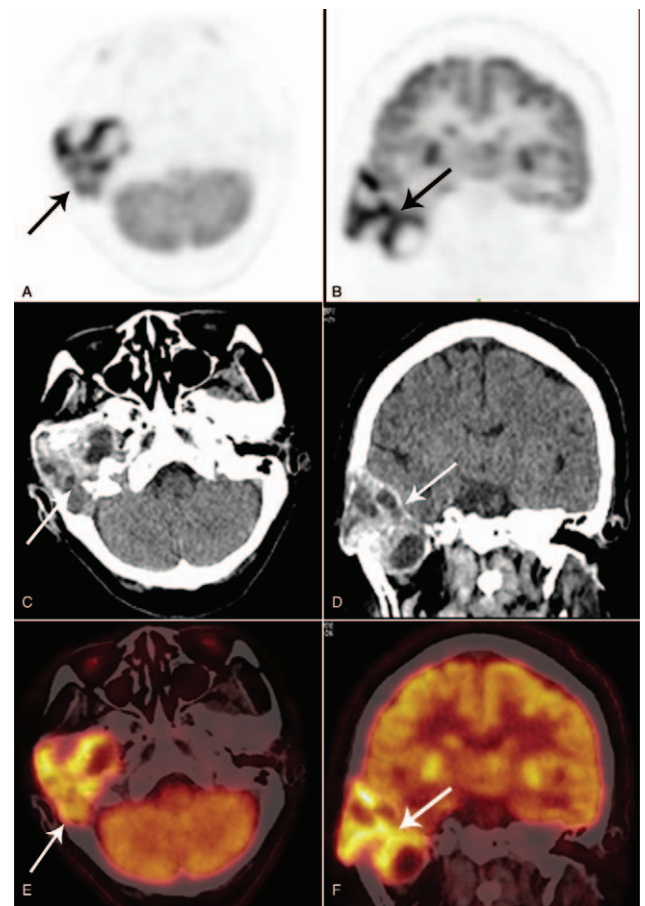


Figure 5. A soft-tissue mass with increased FDG uptake located in the right temporomandibular joint was showed in ¹⁸F-FDG PET/computerized tomography (CT) images. An increased FDG uptake for the mass was found in PET images (A, axial; B, coronal). In corresponding PET/CT fusion images, the solid portion of the mass had increased FDG uptake (arrows) with a SUVmax of 19.8 (C and D—CT; E and F—fusion).

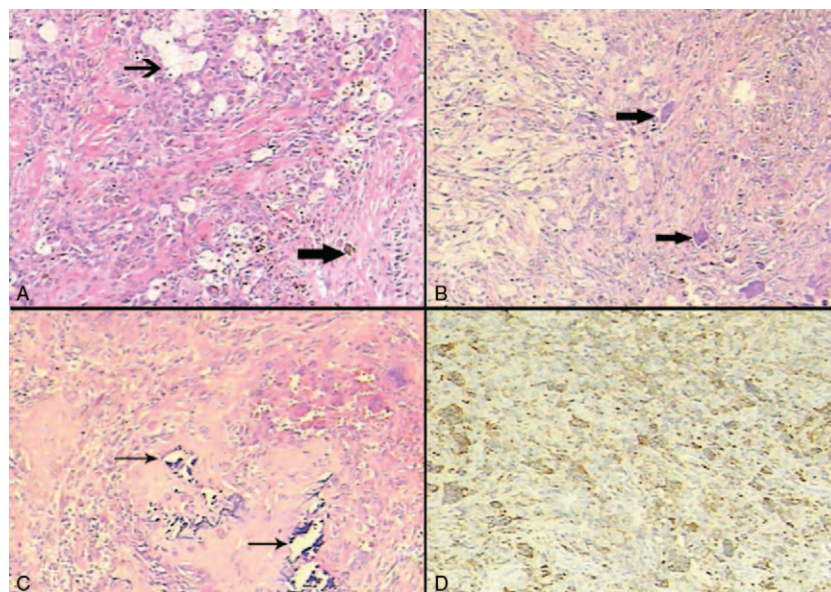


Figure 6. Photomicrograph (A–C, HE, original magnification, ×100; D, immunohistochemistry, ×100) showed compact hyperplastic monocytes, foam cells (A, small arrow), and hemosiderin-laden macrophages (A, large arrow). There were also a number of scattered multinucleated giant cells (B, arrows), and pale blue calcification (C, arrows) in the fibrous stroma. The monocytes and macrophages with a positive CD68 were showed in the immunohistochemistry (D).

monocyte, multinucleated giant cells and foam cells distributing in a fibrous stroma, presenting hemosiderin deposition.^[9]

CT and MRI were proved to be very helpful in showing the extension of the lesion as well as bone destruction. In particular, with the deposition of hemosiderin, the images invariably showed hypointensity throughout the lesion on T2WI. However, the appearance of TGCT on MRI can be variable, depending on the relative proportion of the lipid, hemosiderin, fibrous stroma, and cellular elements.^[10] PET findings were often positive because of the abnormal focus of increased activity, which was a potential pitfall to consider it as the malignant lesion.^[11,12]

The standard treatment for the TGCT is surgery, with partial or complete excision of the lesion and involved bones. With the property of locally aggressive, the TGCT is often not completely removed, especially diffuse type. Some scholars believe that incomplete excision is the root of the recurrence.^[13]

4. Conclusion

In this case, we demonstrate a rare disease of TGCT involving the TMJ, which has specific manifestation in MRI and ¹⁸F-FDG PET/CT. Clinicians and oral pathologists should keep in mind the possibility of the TGCT when aggressive lesions involve the TMJ, with irregular low signal on T2WI, abnormally intense radioisotope uptake on ¹⁸F-FDG PET/CT and adjacent bone destruction. Careful preoperative examination and complete resection are the factors that lead to the optimal treatment of TGCT.

5. Consent

Informed consent was signed by the patient for the publication of this report and related images.

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