

Diffuse giant cell tumors of the tendon sheath in temporomandibular joint

Two case reports and review of the literature

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Abstract

Rationale: Diffuse giant cell tumors of the tendon sheath (GCT-TS) in the temporomandibular joint (TMJ) are extremely rare.

Patient concerns: We reported the imaging appearance and the pathological findings of 2 male cases with diffuse GCT-TS in the TMJ (52 years for the case 1 and 50 years for the case 2) who complain of the hearing disorders of left ear when presenting to our hospital.

Diagnosis: Preoperative computerized tomography (CT) scan revealed an irregular mass in the left temporal fossa with the sizes of approximately 5.8 × 3.8 × 4.6 cm for case 1 and 4.2 × 3.2 × 4.3 cm for case 2, respectively. Magnetic resonance imaging (MRI) findings showed a mass of 6.1 × 4.0 × 5.1 cm and 5.3 × 3.6 × 3.3 cm, respectively. Postoperative pathological examination revealed the diffuse multinucleated giant cells accompanied with synovial cells in the diffuse arrangement. Immunohistochemical examination showed CD68 (+), Vim (+), S-100 (-) and 2% of Ki-67 proliferation index. These characteristics are in line with the diagnosis of the diffuse GCT-TS in the TMJ.

Interventions: Both cases underwent the gross total resection followed by radiation therapy.

Outcomes: Two patients experienced no recurrence after follow-up of 1 to 2 year(s).

Lessons: Diffuse GCT-TS in the TMJ is very rare. Clinicians should keep in mind the possibility of the GCT-TS when aggressive lesions involve the TMJ and adjacent bone destruction was found. Careful pre- and postoperative examinations and complete resection are the factors that lead to its optimal treatment, significantly reducing its recurrence rate.

Abbreviations: CT = computerized tomography, GCT-TS = giant cell tumors of the tendon sheath, MRI = magnetic resonance imaging, T2WI = T2-weighted images, TMJ = temporomandibular joint.

Keywords: case report, diagnosis, diffuse giant cell tumors of the tendon sheath, temporomandibular joint, treatment

1. Introduction

Giant cell tumors of the tendon sheath (GCT-TS) is an uncommon benign tumor-like proliferative lesion with an undetermined origin arising from the synovial membranes of joints, bursas, and tendon sheaths.^[1–3] The etiology of GCT-TS remains elusive with possible etiologies of disturbances of lipid metabolism, neoplasm, inflammation, trauma, and hemorrhage.^[4,5] Pathologically, GCT-TS is composed of synovial mononuclear cells and osteoclast-like multinucleated giant cells.^[5] GCT-TS may be intra- or extra-articular, and is classified by clinical presentation and biological behavior as localized or diffuse; the latter is more aggressive, and is also known as villonodular synovitis.^[6] The localized form of GCT-

TS is most prevalent and generally follows an indolent course, frequently affecting the tendon sheath and joints of the hands.^[7] In contrast, diffuse-type GCT-TS are less common and typically affect the knee, hip, or shoulder. The knee is commonly involved in 80% of cases, but any joint may be affected.^[8] Involvement of the temporomandibular joint (TMJ) is uncommon, and about 80 and 30 cases with GTC-TS in the TMJ have been published in the English- and Chinese-language literature, respectively, to our knowledge.^[1,9,10] The predominant symptoms in the TMJ are preauricular mass or swelling, pain, and limitation of mandibular movement.^[11] Because of its destructive nature, immediate treatment upon diagnosis is recommended. This article reports 2 cases of GTC-TS in the TMJ in our hospital. Their clinical, radiological, and pathological features and the treatment were summarized retrospectively, and the related literature was reviewed in an attempt to enhance the understanding about this rare disease.

2. Case presentations

2.1. Case 1

A 52-year-old male patient was admitted to our hospital in May 2015 for the symptoms of “a mass found on the left temporal fossa accompanied by the progressive loss of the left ear hearing for more than 2 years.” The medical history revealed that a hard mass on the right temporal fossa was incidentally found 2 years ago. The physical examination in 1 hospital in Jiuquan city, China, showed that the patient felt no tenderness on the mass and experienced obvious left ear tinnitus, intermittent dizziness without nausea, vomiting, fever, convulsions, consciousness

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HY and FW have contributed equally to this study.

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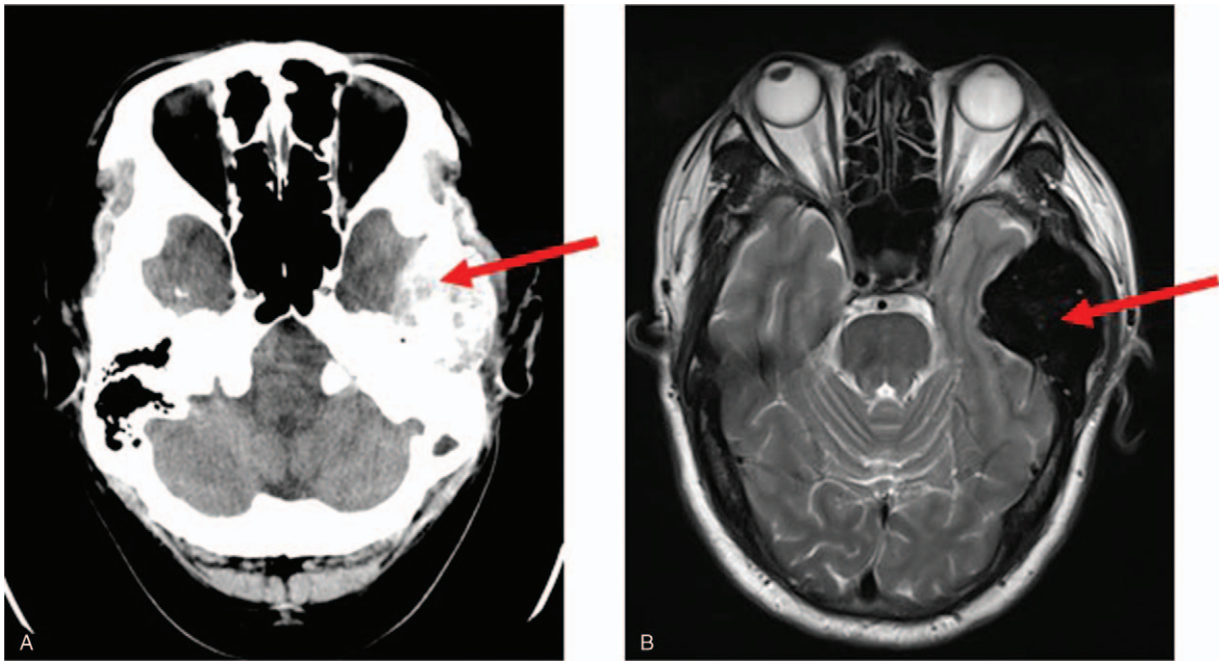


Figure 1. CT scan demonstrates a destructive lesion ($5.8 \times 3.8 \times 4.6$ cm) involving the left temporomandibular joint (TMJ) with significant bony remodeling of the middle fossa floor (A). MRI shows a lesion ($6.1 \times 4.0 \times 5.1$ cm) centered in the TMJ with associated intracranial extension (B). The arrows indicate the lesion in the respective imaging modality (CT or MRI).

disorder, blurred vision, and limb movement disorder. Head computerized tomography (CT) scan demonstrated the intracranial lesion, which was suspected of meningiomas. The surgical treatment was recommended, but the patient refused it. Due to the continuously decreased left ear hearing, intermittent dizziness, the patient visited our hospital to further clarify the diagnosis. After admission, the physical examination showed a mass of approximately 3.0×4.5 cm on the left temporal fossa, which was hard and had no fluctuation. The opening of mouth was normal and the mouth was not skewed. The lower lip was not numb. The function of left side facial nerve was normal and the cervical lymph nodes were not enlarged. No abnormalities in heart, lung, and abdomen were found. CT examination (Fig. 1 A) revealed a mass with size of approximately $5.8 \times 3.8 \times 4.6$ cm and demonstrated that the bone fracture pieces of the left temporal bone were absorbed and involved in the mastoid process. The density of the soft tissue in the adjacent area was visible and the

boundary was still clear. The density of the left middle ear and the mastoid process increased. Magnetic resonance imaging (MRI) showed an irregular mass of approximately $6.1 \times 4.0 \times 5.1$ cm in the left temporal side, which invaded the temporal bone and had no clear boundary with the left mastoid. Osteogenic tumor was suspected because there are multiple abnormal signal lesions in the mastoid in MRI. Chondroblastoma was considered for the multiple calcified nodules. Clinical diagnosis was made to be the space-occupying lesion on the left temporal region, which was suspected of skull osteomas. Intraoperative view showed that the tumor with sizes of approximately $3.0 \times 5.0 \times 3.0$ cm located above the left zygomatic bone, and the skull adjacent the tumor had been widely damaged involving the left TMJ. With the incomplete dura mater, the tumor partly infiltrated the medial cranial fossa. Postoperative pathological examination revealed that the synovial mononuclear cells are pervaded and the cytoplasm is deposited with iron-hemoflavin particles as

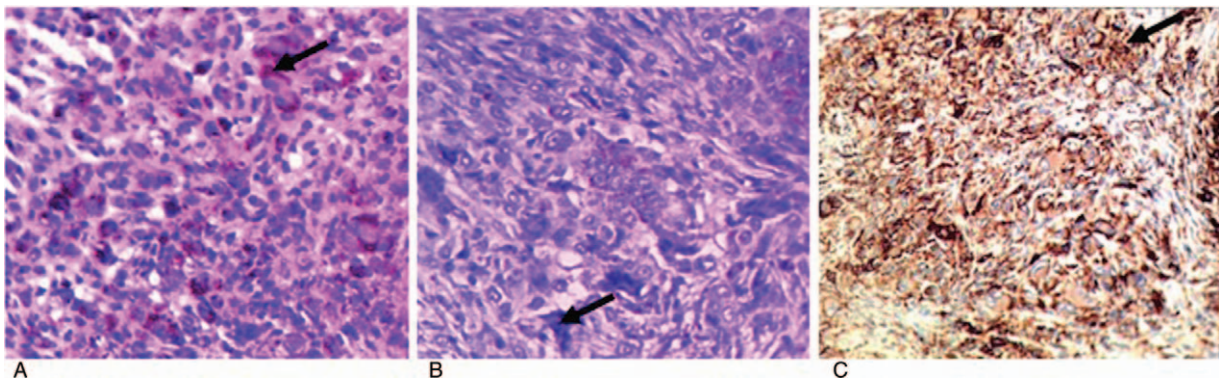


Figure 2. Photomicrograph with hematoxylin-eosin stain showed that the synovial mononuclear cells are pervaded and the cytoplasm is deposited with iron-hemoflavin particles as indicated by arrow (A); Multinucleated giant cells are distributed in the interstitial tissue as indicated by arrow (B); The monocytes and macrophages with a positive CD68 were shown in the immunohistochemistry (C).

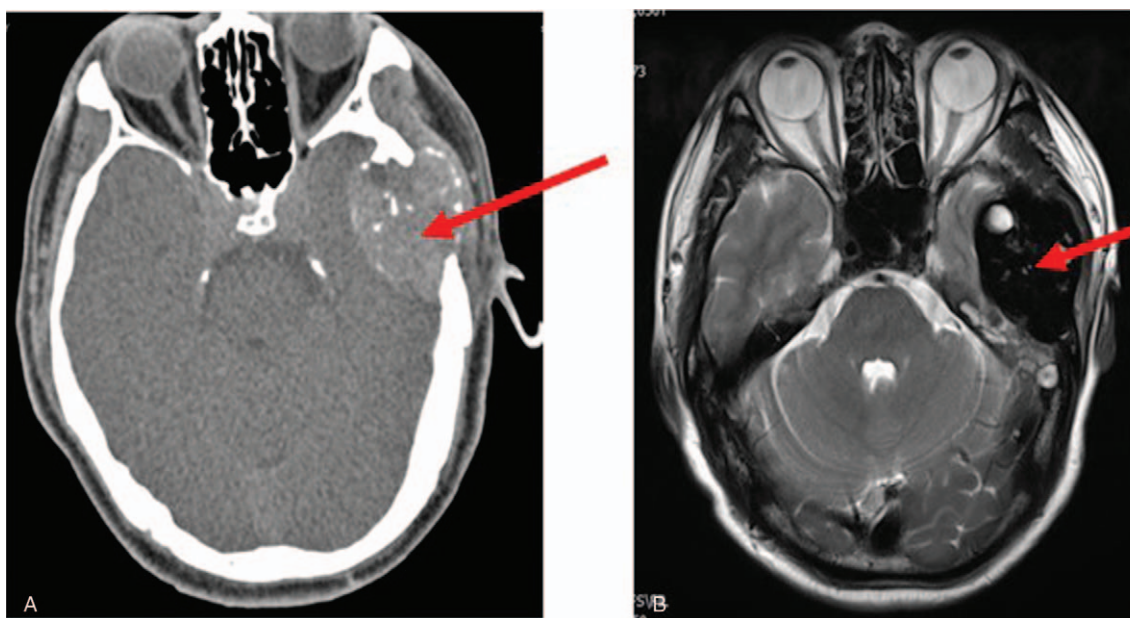


Figure 3. CT scan demonstrates a destructive lesion ($4.2 \times 3.2 \times 4.3$ cm) involving the left temporomandibular joint (TMJ) with significant bony remodeling of the middle fossa floor (A). MRI shows a lesion ($5.3 \times 3.6 \times 3.3$ cm) centered in the TMJ with associated intracranial extension (B). The arrows indicate the lesion in the respective imaging modality (CT or MRI).

indicated by arrow (Fig. 2 A) and that multinucleated giant cells are distributed in the interstitial tissue as indicated by arrow (Fig. 2B). Immunohistochemical examination showed CD68 (+) (Fig. 2C), Vim (+), S-100 (-), and 2% of Ki-67 proliferation index. Combining the histological information with the clinical presentation yielded a diagnosis of diffuse GCT-TS of the left TMJ. The patient underwent radical resection of the tumor followed by radiotherapy in order for preventing the relapse. No recurrence was found after 2 years' follow-up.

2.2. Case 2

A 50-year-old male patient was admitted to our hospital in September 2016 for the symptoms of "left ear tinnitus accompanied with reduced hearing over one year." The medical history revealed that the patient experienced obvious left ear tinnitus accompanied with reduced hearing, however had no headache, dizziness, nausea, vomiting and facial twitching, and numbness for over 1 year. After the symptoms were relieved following the symptomatic treatment in 1 hospital in Longnan city, China, the patient did not receive any further examination. Recently, the patient presented the symptoms of "the left ear tinnitus accompanied with reduced hearing," which were aggravated, especially after he moved his head. The patient came to our hospital for further diagnosis and treatment. The physical examination revealed no gait staggering and standing instability. Neurological checks showed the consciousness, examination cooperation, automatic position, clear speech, normal binocular vision and neural responses, normal bilaterally facial sensation of both pain and temperature, no skew of the jaw and mouth, symmetrical nasolabial sulcus without collapse. CT examination demonstrated the increased internal density of the left middle ear and mastoid without obvious abnormality in the right middle ear, mastoid, cochlea, vestibule, and semicircular canal (Fig. 3 A). The CT findings also showed the destruction of the left temporal bone as well as the round mass shadow with the

size of approximately $4.2 \times 3.2 \times 4.3$ cm. MRI examination showed a large and irregular mass in the left temporal region with sizes of approximately $5.3 \times 3.6 \times 3.3$ cm as well as the temporal bone destruction and mixed signal mass (Fig. 3 B). The mass had unclear border with the left mastoid. Because of the multiple calcified nodules in the mass, the cartilage tumors were considered. Clinical diagnosis was made to be the left temporal space occupying lesion. Intraoperative view, postoperative pathological examination, and immunohistochemical examination were very similar with that showing in case one. The patient also underwent radical resection of the tumor followed by radiotherapy and no recurrence was found after nearly 1-year follow-up.

2.3. Ethic statement

The study was approved by the Ethical Committee of the Second Hospital of Lanzhou University. Written informed consent was obtained from the patients involved in the study.

3. Discussion

GCT-TS is a type of benign soft tissue tumor that was first described by Chassaignac in 1852.^[7,12] GCT-TS is also termed tenosynovial giant cell tumor, pigmented nodular tenosynovitis, xanthogranuloma, benign synovioma, and fibrous xanthoma of synovium.^[2,13,14] The GCT-TS is a benign but locally aggressive tumor that arises from the synovial membrane of joints, tendon sheaths, and bursae.^[14] Although any joint can be affected, involvement of the TMJ was reported very rarely.^[11] The present study added 2 rare cases to the growing body of literature characterizing the clinical features and treatment of GCT-TS in the TMJ already including approximately 80 and 30 cases in the English- and Chinese-language literature, respectively, to our knowledge, enhancing the understanding about this rare disease.^[1,9,10]

Safaei et al^[10] performed a comprehensive literature review by summarizing 58 published cases with GCT-TS in the TMJ. They found that all cases had the mean age of 45 years without predilection of tumor for sex. The patients had a mean duration of 15 months from the symptom onset to diagnosis and the pain was the most common presenting symptom. The great majority of patients underwent surgical resection, and the rate of recurrence was 15% after at least 1 year of follow-up. In another study, Carlson et al^[11] retrospectively reviewed 11 cases with GCT-TS in the TMJ treated at the authors' center and found that the patients had the median age of 49 years, including 8 (73%) men and 7 (64%) cases with right-sided tumor. Five (45%) cases presented with hearing loss, 5 (45%) cases with pain, 4 (36%) cases with aural fullness or pressure, and none with facial nerve paresis. The median from symptom onset to diagnosis was 24 months. The present study showed that the baseline demographics and symptoms of 2 cases are highly similar to that in above-mentioned reviews. Briefly, the mean age at diagnosis was 51 years. The mean duration from symptom onset to diagnosis was 18 months. Both cases presented hearing problems without facial nerve paresis. Exceptionally, both cases are male and had GCT-TS in the left TMJ without postoperative recurrence after follow-up of 1 to 2 year(s). These exceptions might be largely contributed to the small size samples of only 2 cases. In addition, Herman et al^[15] summarized the literature including 26 cases of GCT-TS in the TMJ reported previously and found that 24 (92%) cases presented with a preauricular mass or swelling. All 26 cases (age range: 10–70 years) had the mean duration of 11.4 months from the symptoms onset to diagnosis, having no predilection of GCT-TS for sex and the right or left-sided tumor. In addition, Meng et al^[9] reported 15 patients diagnosed as diffuse GCT-TS in the TMJ in a Chinese-language literature and found the similar clinical data with above-mentioned studies.

The clinical presentations and radiographic findings of patients with possible GCT-TS in the TMJ may originally indicate a soft tissue mass, but the final diagnosis was always made largely depending on histological examination in most of the cases. The possibilities for a differential diagnosis of GCT-TS from inflammatory parotid lesions, pleomorphic adenoma as well as benign and malignant mesenchymal lesions or tumors should be considered. In the report by Carlson et al,^[11] 9 (82%) of the 11 cases were originally diagnosed as chondroblastoma and subsequently reclassified as GCT-TS. The remaining 2 (18%) cases were classified as GCT-TS at initial diagnosis. The results suggested the difficulties in differential diagnosis of GCT-TS with other tumors. The findings of CT and MRI can identify the extension and borders of the lesion and indicate the local bony invasion. The implications of GCT-TS on MRI vary with the specificity of the histological characteristics of the tumor. In addition, CT scans can determine the extension of bony destruction that frequently occurs. Given the rarity of GCT-TS and relatively lack of professional experience on GCT-TS, the clinicians, radiologist, and pathologist should be well cooperated in the diagnosis and strategy determination for treatment. The present 2 cases were misdiagnosed as chondroblastoma by radiograph findings and subsequently reclassified as GCT-TS by histopathological observations.

Despite a benign lesion, radical resection is still the mainstay of treatment.^[11] Early surgical resection upon diagnosis is advisable. This will prevent the destructive proliferation of the tumor resorbing the TMJ, which gradually loses its function, and the deterioration of esthetic and prognosis outcomes. In the report by

Carlson et al,^[11] all 11 patients underwent surgical management with or without radiation therapy. Eight cases undergoing gross total resection, without radiation, showed only 1 recurrence (13%) after follow-up of a median of 116 months. Safaei et al^[10] reported that among 58 patients with at least 1 year, 2 years, and 5 years of follow-up, the recurrence rates were 14%, 15%, and 29%, respectively. Both cases in the present study underwent radical resection of the tumor followed by radiotherapy in order for preventing the relapse. No recurrence was found after 1 to 2 year(s)' follow-up. The discrepancy in recurrence rate between literatures and our study might be explained by the small size samples of only 2 cases, which demonstrated the occasionality and by the utility of radiation for both cases in our study.

4. Conclusion

We demonstrate 2 rare cases with GCT-TS in the TMJ in terms of clinical, radiological, and histopathological feature as well as the surgical treatment management. Clinicians should keep in mind the possibility of the GCT-TS when aggressive lesions involve the TMJ and adjacent bone destruction was found. Careful preoperative examination and complete resection are the factors that lead to the optimal treatment of GCT-TS in the TMJ.

Author contributions

Drafting the manuscript and revising it critically for important intellectual content: YH. Final approval to the version to be published: LC. All authors made substantial contributions to conception and design, or acquisition of data, or analysis/interpretation of data, and read and approved the final manuscript.

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