Hong Kong J Radiol. 2024;27:Epub. Published 18 November 2024 | https://doi.org/10.12809/hkjr2317772

CASE REPORT

Tenosynovial Giant Cell Tumour of the Temporomandibular Joint with Initial Suspicion of Nodal Metastases: A Case Report

LY Lam[1], WL Wong[1], KWS Ko[1], ZCS Tsang[2], KM Chu[1]

[1]Department of Radiology and Imaging, Queen Elizabeth Hospital, Hong Kong SAR, China
[2]Department of Pathology, Kwong Wah Hospital, Hong Kong SAR, China

CASE PRESENTATION

A 26-year-old female with good past health presented with 1-month history of progressive swelling at the left preauricular region. Physical examination revealed a 4-cm mass over the left preauricular region, not fixed to the underlying structures. Computed tomography (CT) demonstrated a 5-cm hyperdense and heterogeneously enhancing soft tissue mass at the left preauricular region (Figure 1). There was bony erosion of the left mandibular head and articular tubercle of the temporal bone with extension to the base of the middle cranial fossa (Figure 2). No parenchymal invasion of the left temporal lobe was observed. Differential diagnoses included an aggressive parotid tumour with bony extension or an aggressive bone condition arising from the temporal bone such as osteomyelitis or metastasis or primary bone tumours such as aneurysmal bone cyst.

Magnetic resonance imaging (MRI) showed an expansile and lobulated mass with T1- and T2-weighted hypointense signals and moderate contrast enhancement centring at the left temporomandibular joint (TMJ) [Figure 3] and indenting the left middle cranial fossa dura superiorly. There was involvement of the left mandibular condyle and superficial lobe of the left parotid gland. Several nodules showing similar signal characteristics to the index lesion were seen within the left parotid gland. A few lymph nodes were seen along the left jugular chain (Figure 4), with similar signal characteristics. These were suspicious of nodal metastases. Biopsy of the mass revealed mononuclear cells with osteoclast-type giant cells, presence of hemosiderin pigment and expression of clusterin and D2-40 via immunostaining. Features were compatible with diffuse-type tenosynovial giant cell tumour (TGCT).

Wide local excision and cervical lymph node dissection were performed. Final pathology of the specimen confirmed local involvement including the mandibular fossa of the temporal bone (Figure 5), mandibular condyle, zygoma, left parotid gland (Figure 6), and dura at the left temporal fossa. No malignant cells were seen.

Correspondence: *Dr LY Lam, Department of Radiology and Imaging, Queen Elizabeth Hospital, Hong Kong SAR, China*
Email: lly858@ha.org.hk

Submitted: 19 September 2023; Accepted: 10 January 2024. This version may differ from the final version when published in an issue.

Contributors: LYL and WLW designed the study. All authors acquired and analysed the data. LYL and WLW drafted the manuscript. All authors critically revised the manuscript for important intellectual content. All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflicts of Interest: All authors have disclosed no conflicts of interest.

Funding/Support: This study received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Data Availability: All data generated or analysed during the present study are available from the corresponding author on reasonable request.

Ethics Approval: The study was approved by the Kowloon Central Cluster and Kowloon East Cluster Research Ethics Committee of Hospital Authority, Hong Kong (Ref No.: KC/KE-28-0056/ER-1). The requirement for patient consent was waived by the Committee due to loss to follow-up of the patient and the use of de-identified information of the patient in the study.

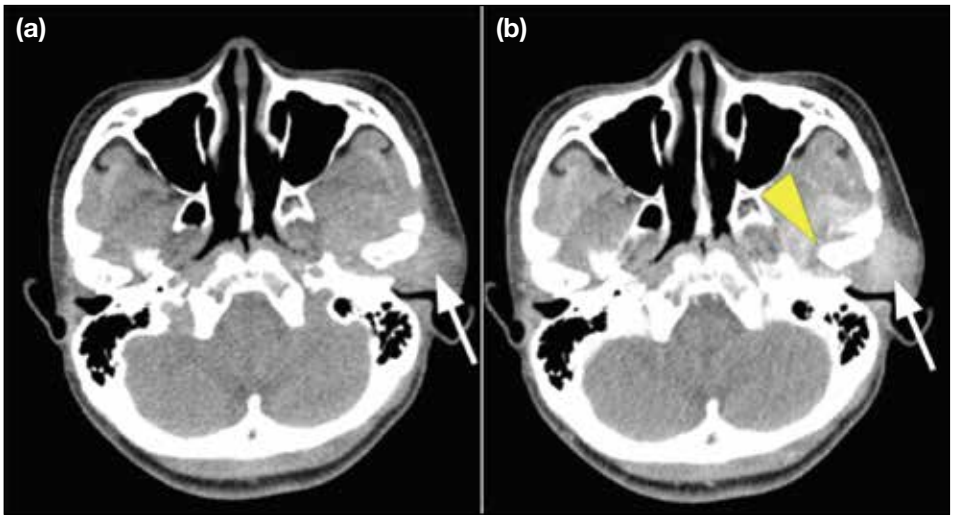

Figure 1. (a) Unenhanced and (b) post-contrast axial computed tomography images in soft tissue window show an enhancing tumour (arrows) at the preauricular region centring around the left temporomandibular joint. A rim of non-enhancing joint effusion is seen (arrowhead).

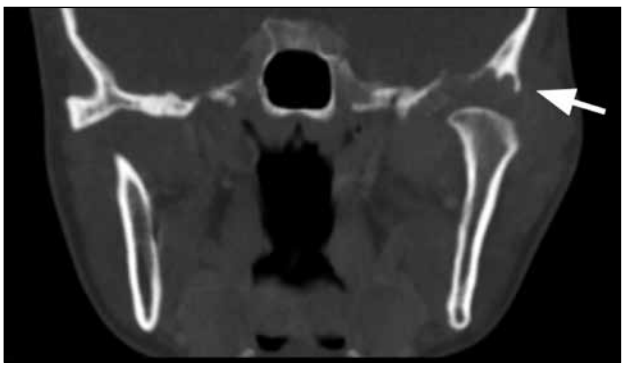

Figure 2. Coronal computed tomography image in bone window shows that the tumour erodes through the temporal bone (arrow).

Cervical lymph nodes from the jugular chain showed hemosiderin laden macrophages without giant cells or malignant cells.

In view of the positive surgical margins, the patient underwent postoperative adjuvant radiotherapy of 40 Gy in 20 fractions. There have been no signs of recurrence on clinical and imaging follow-up by MRI 6 months post-operation.

DISCUSSION

TGCT encompasses a group of neoplasms arising from the synovial membranes of the joints, bursae, or tendons. Previously diffuse TGCTs were well known as pigmented villonodular synovitis. This term is no longer recommended by the World Health Organization[1] as the suffix *-itis* may wrongly imply an inflammatory condition.

Most TGCTs are benign. Malignant TGCT is exceedingly rare, with only 50 reported cases worldwide, typically affecting the lower extremities and most affected adults aged 50 to 60 years.[1]

The aetiology of TGCTs is largely unknown. Some studies attribute the process to repeated intra-articular haemorrhage following trauma, whereas some proposed that it is due to disturbed lipid metabolism.[2] The patient in our case had good past health with no history of facial trauma.

TGCTs are classified according to their location (intra-articular vs. extra-articular) and growth pattern (localised vs. diffuse). There is no sex predilection for intra-articular disease, while there is a slight female predilection for extra-articular disease.[3] The clinical presentation and affected joints differ between localised and diffuse subtypes. Localised TGCTs involve part of the synovium and are commonly found in the fingers.[1] Patients typically present in their 3rd to 5th decade with a female predilection. The second most common location is the hand and wrist regions. They often present as painless and slow growing masses.[4] Diffuse TGCTs are less common with an annual incidence of 4 per million population. These are monoarticular diseases affecting large joints, typically the knees and hips, accounting for 66% to 80% and 4% to 16% of cases, respectively.[1] Other joints such as the ankles, shoulders, and elbows are affected in descending order of frequency.[1,3] Patients are commonly in their early middle age. Patients with diffuse TGCTs present with painful joint swelling with reduced range of movement. Haemarthrosis is commonly

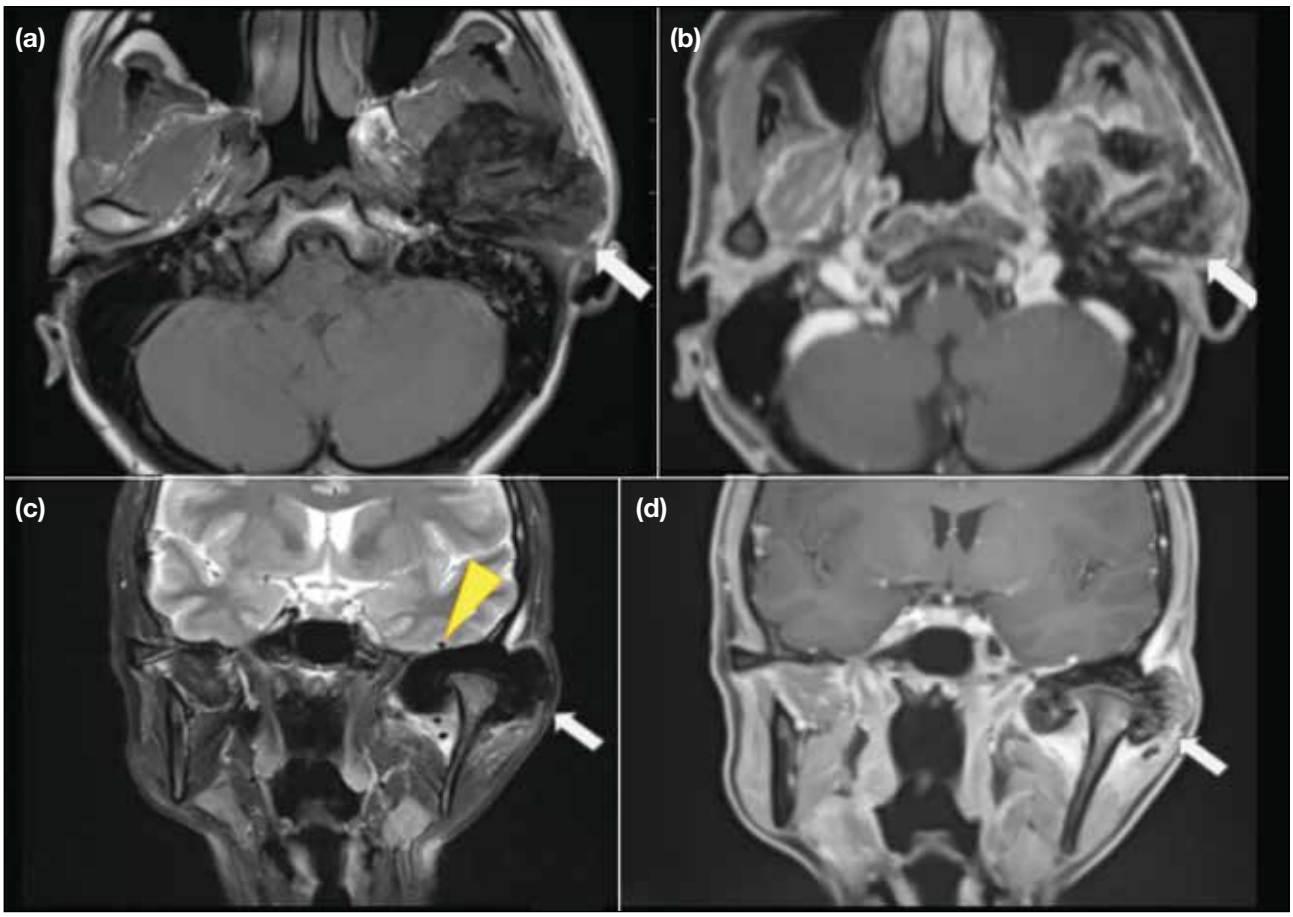

Figure 3. (a) Axial T1-weighted, (b) axial T1-weighted post-gadolinium contrast enhanced, (c) coronal T2-weighted, and (d) coronal T1-weighted post-gadolinium contrast enhanced magnetic resonance images show the tumour (arrows) at the left temporomandibular joint, which is markedly T1- and T2-weighted hypointense with contrast enhancement mainly over the periphery. There is erosion of the base of the temporal fossa with the tumour abutting a cortical vessel (yellow arrowhead in [c]).

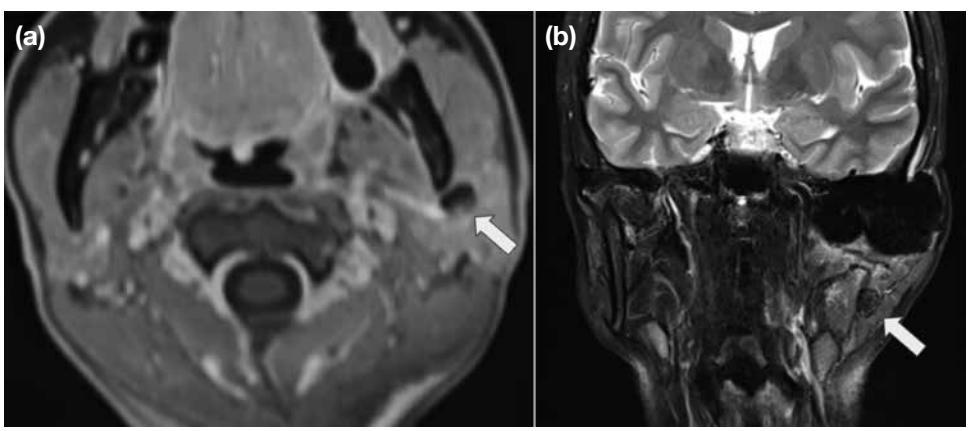

Figure 4. (a) Axial T1-weighted and (b) coronal T2-weighted images show a non-enlarged lymph node (arrows) along the jugular chain with similar T1- and T2-weighted hypointense signal to the primary tumour.

encountered. This is a locally aggressive lesion that tends to have local recurrence following complete excision, with a reported recurrence rate of 35%.[1,3]

TGCT of the TMJ is rare and is typically the diffuse form. The first case of TGCT involving the TMJ was reported in 1973.[5] To date, around 100 cases have been

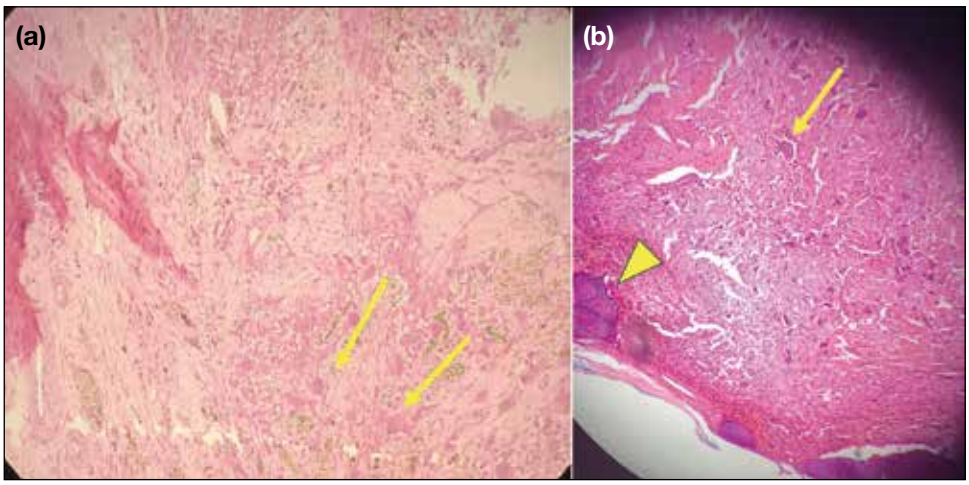

Figure 5. Tissue sections stained with haematoxylin and eosin (×100) show an infiltrative tumour with interspersed osteoclast-like giant cells (arrows in [a] and [b]), in keeping with diffuse-type tenosynovial giant cell tumour. Invasion into the temporal bone (arrowhead) is demonstrated (b).

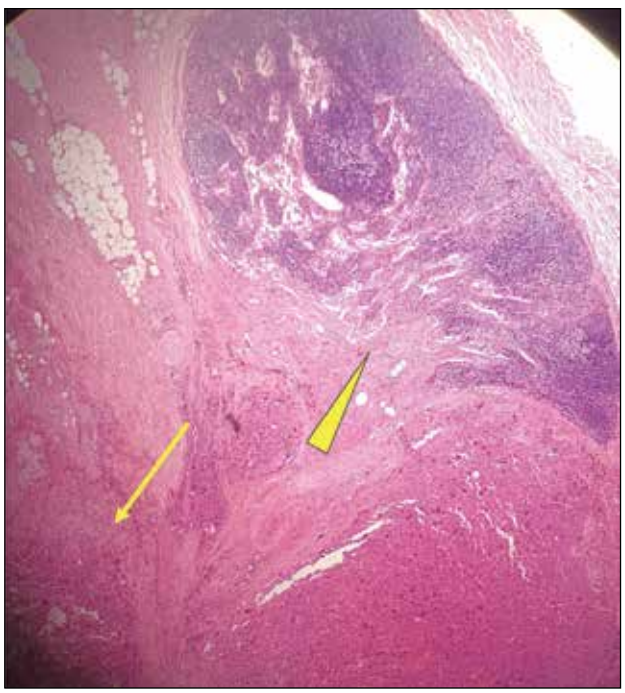

Figure 6. Tissue section of the parotid gland stained with haematoxylin and eosin (×100) shows involvement by the tenosynovial giant cell tumour, with erosion into the blood vessels (arrow) with an intraparotid lymph node (arrowhead) seen.

reported worldwide.[6] The most common presentation is of a preauricular mass. Some cases may present with limited range of movement at the TMJ. Because of their close proximity to the parotid gland, they are often mistaken for parotid lesions.[7]

Microscopically, TGCTs have a variable appearance, consisting of variable proportions of multinucleated giant cells, foamy macrophages, haemosiderin, and stromal collagenisation.

Radiographs of diffuse intra-articular TGCTs commonly demonstrate joint effusion, soft tissue swelling and extrinsic pressure erosion of bone, usually involving both sides of a joint. Radiographs may appear normal in 21% of cases.[3]

On CT and MRI, other than erosion and joint effusion, extensive synovial thickening with villous or nodular projections extending into the joint can be seen. These synovial thickening and masses are hyperdense on CT due to their high iron content. On MRI, they appear hypointense on T1- and T2-weighted sequences due to hemosiderin deposition.[1,7] These were well illustrated in our case. Blooming artefacts are pathognomonic for TGCTs. These tumours show predominantly high signals in short-tau inversion recovery sequence.[7] Contrast enhancement is observed due to their significant vascularity.[3]

Sonographic findings are less specific for the diffuse intra-articular subtype of TGCTs and include complex heterogeneous echogenic masses along the thickened hypoechoic synovium and joint effusion.[3] Doppler imaging commonly reveals increased blood flow.

TGCTs show hypermetabolism on fluorine-18 fluorodeoxyglucose positron emission tomography with an average standard uptake value of 5.9.[3] This may be a potential pitfall for a misdiagnosis of malignant tumour.

In our case, the CT and MRI demonstrated significant

involvement of both sides of the TMJ by the TGCTs, with more erosive effect on the mandibular fossa side than that of the mandibular condyle. This leads to the false impression that the disease is bony in origin. The characteristic magnetic resonance features of hemosiderin deposition were helpful in making the correct diagnosis.

Both malignant TGCTs and TGCTs of the TMJ are very rare conditions; only a few cases of malignant TGCTs of the TMJs have been reported in the literature.[6] By imaging alone, it is difficult to differentiate benign from malignant TGCTs. Features such as rapid growth, aggressive bone destruction, and evidence of distant metastases should raise suspicion of a malignant nature.[5] Common metastatic sites of malignant TGCTs are the regional lymph nodes, lungs, and spine.

In our case, multiple lymph nodes with similar hypointense signals to the index lesion were found, raising an initial suspicion of nodal metastases. Nonetheless the pathology of the index tumour and lymph nodes showed no malignant cells.

A few cases of metastatic spread of benign TGCTs have been described.[8] The presentation of metastases occurred in longer time spans after the initial diagnosis, from years to decades. In our case, the discovery of suspicious lymph nodes was within 1 month of the initial symptoms. The histological findings of the lymph nodes did not meet the criteria for metastases from TGCTs described in the case report by Malik et al.[9] These rendered the possibility of true nodal metastases less likely. The histological findings of haemosiderin laden macrophages and the imaging findings of the lymph nodes can be due to lymphatic drainage of degraded blood products from the haemorrhagic tumour, which has been reported in chronic cases of tumoural haemorrhage.[10] Our findings suggest that imaging alone is not definitive for the diagnosis of nodal metastases and pathology remains the gold standard. Nevertheless clinicians should be vigilant for metastasis in patients with known diffuse TGCTs who present with palpable lymph nodes since benign TGCTs can metastasise and recur after definitive treatment.[8]

The mainstay of treatment for TGCTs is surgical excision with wide margin. Aggressive cases with a high chance of recurrence will benefit from postoperative radiation therapy.[11] In view of the positive surgical margins in the surgical specimen, postoperative radiation and long-term follow-up will be beneficial to our patient to reduce the chance of recurrence and metastases.

CONCLUSION

We report a case of TGCT of the TMJ. Such tumours should be considered in the differential diagnoses of preauricular aggressive swellings. Clinicians should consider nodal metastasis in patients with diffuse TGCTs who present with palpable lymph nodes.

REFERENCES

1. De Saint Aubain Somerhausen N, van de Rijn M; The WHO Classification of Tumours Editorial Board. Tenosynovial giant cell tumour. WHO Classification of Tumours Soft Tissue and Bone Tumours, 5th ed. Lyon: IARC Press; 2020: 133-6.
2. Bemporad JA, Chaloupka JC, Putman CM, Roth TC, Tarro J, Mitra S, et al. Pigmented villonodular synovitis of the temporomandibular joint: diagnostic imaging and endovascular therapeutic embolization of a rare head and neck tumor. AJNR Am J Neuroradiol. 1999;20:159-62.
3. Murphey MD, Rhee JH, Lewis RB, Fanburg-Smith JC, Flemming DJ, Walker EA. Pigmented villonodular synovitis: radiologic-pathologic correlation. Radiographics. 2008;28:1493-518.
4. Peh WC, Shek TW, Ip WY. Growing wrist mass. Ann Rheum Dis. 2001;60:550-3.
5. Yoon HJ, Cho YA, Lee JI, Hong SP, Hong SD. Malignant pigmented villonodular synovitis of the temporomandibular joint with lung metastasis: a case report and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2011;111:e30-6.
6. Verspoor FG, Mastboom MJ, Weijs WL, Koetsveld AC, Schreuder HW, Flucke U. Treatments of tenosynovial giant cell tumours of the temperomandibular joint: a report of three cases and a review of literature. Int J Oral Maxillofac Surg. 2018;47:1288-94.
7. Le WJ, Li MH, Yu Q, Shi HM. Pigmented villonodular synovitis of the temporomandibular joint: CT imaging findings. Clin Imaging. 2014;38:6-10.
8. Chen EL, de Castro CM 4th, Hendzel KD, Iwaz S, Kim MA, Valeshabad AK, et al. Histologically benign metastasizing tenosynovial giant cell tumor mimicking metastatic malignancy: a case report and review of literature. Radiol Case Rep. 2019;14:934-40.
9. Malik K, Raja A, Shirley S. Isolated regional nodal metastasis in giant cell tumor of the bone: case report and review of literature. South Asian J Cancer. 2020;9:58.
10. Elmore SA. Histopathology of the lymph nodes. Toxicol Pathol. 2006;34:425-54.
11. Baniel C, Yoo CH, Jiang A, von Eyben R, Mohler DG, Ganjoo K, et al. Long-term outcomes of diffuse or recurrent tenosynovial giant cell tumor treated with postoperative external beam radiation therapy. Pract Radiat Oncol. 2023;13:e301-7.