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<u>Chondroid Tenosynovial Giant Cell Tumour – A rarity in the TMJ</u>

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Abstract

Tenosynovial giant cell tumours (TGCT) of the temporomandibular joint (TMJ) are extremely rare, particularly the subset of chondroid tumours. TGCT can be broadly divided into localised and diffuse types (1), of which there have only been 116 reported cases in the TMJ. Rarer still are the subset of chondroid tenosynovial giant cell tumours (CTGCT), of which only 30 cases affecting the TMJ have been reported (2). We present a case of CTGCT involving the TMJ including discussion of presentation and management.

Case Report

A 67-year-old lady presented to the local oral and maxillofacial surgery department with a 7-8 years history of pain involving her left TMJ that restricted daily function. The pain was reasonably controlled with analgesia and a soft diet. She also reported a clicking noise on the affected side, as well as a significant deterioration in her hearing. There were no other associated symptoms or history. The patient was known to have medication-controlled hypertension and her social history was unremarkable.

Examination revealed a non-tender swelling in the left preauricular region, with associated mandibular deviation to the left side. Mild weakness of the buccal branch of her facial nerve was noted. Otoscopy demonstrated an obstructed ear canal.

Computerised tomography (CT) and Magnetic Resonance Imaging (MRI) scans identified a large, destructive bony lesion involving the left mastoid, TMJ and skull base regions (Figure 1). There was also evidence of abnormal soft tissue extending in to the left ear and a biopsy was recommended.

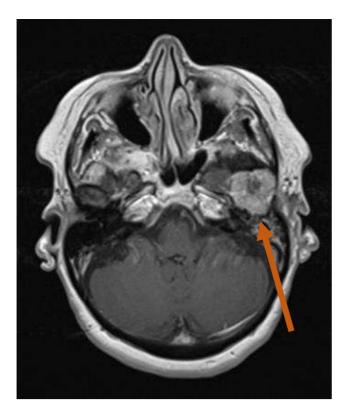


Figure 1 – Axial view of T1 weighted MRI with tumour evident in the left TMJ (arrow)

An open biopsy of the left TMJ was performed. Histopathology demonstrated eosinophilic ovoid and spindle cells with bland nuclei, with focal accumulations of multinucleated giant cells. There were also areas of lobular myxoid structure as well as chondroid tissue in keeping with a CTGCT (Figure 2). There was no evidence of malignancy.

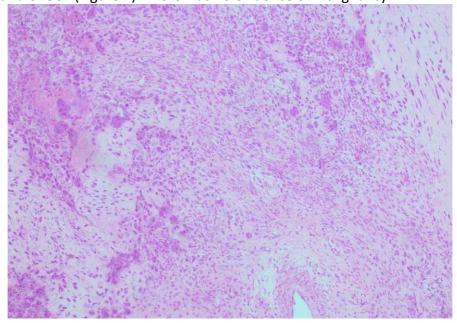


Figure 2 - Haematoxylin and eosin section with evidence of multinucleated giant cells, spindle cells and chondroid areas (original magnification x200).

The patient subsequently underwent a left condylectomy with left temporalis flap interposition (Figure 3). Histopathology confirmed the initial diagnosis though excision margins could not be determined due to the fragmented nature of the specimen.

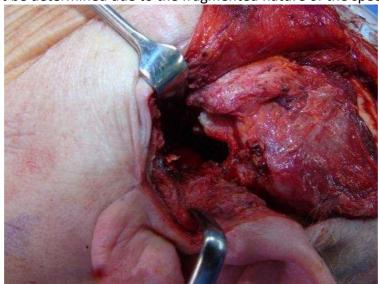


Figure 3 - Intra-operative view of tumour location

The patient's recovery has been uneventful with no evidence of recurrence at 15 months and excellent recovery of function.

Discussion

TGCT in the TMJ was first described by Lapayowker et al in 1973 (3). Though most often benign, 30 malignant cases of TGCT have been reported with only one affecting the TMJ. Benign forms may still prove to be locally destructive and may erode into the skull base (4). Typically, a TGCT presents with a preauricular mass, trismus/stiffness and clicking of the TMJ (5). TGCTs are often misdiagnosed – particularly as temporomandibular joint dysfunction (TMD). A review identified duration to diagnosis for these patients ranging from two months to 15 years, averaging at 30 months. Our patient had symptoms ongoing for eight years. There also appears to be no sex predilection (1). This highlights the importance of further investigations if conservative management is proving ineffective.

Panoramic radiographs are the first form of investigation. CT scans are useful in identifying the bony extent of the lesion, especially any skull base erosion. However, MRI scans are the imaging of choice for diagnosis and management. The presence of haemosiderin in these lesions causes a low signal intensity in the MRI resulting in a 'blooming effect' (6).

In the literature, various differential diagnoses have been considered, however, preauricular swellings are often benign but in many instances they can be malignant, including adenoid cystic carcinomas, mucoepidermoid carcinomas, primary and metastatic squamous cell carcinomas – hence requiring thorough consideration (7). Additionally, due to presence of chondroid metaplasia in CTGCTs, other lesions such as chondroblastomas and chondrosarcomas should also be considered (2). Histologically, CTGCTs appear red-brown in colour with masses of villi, synovial folds and often chondroid tissue typically containing multinucleated giant cells (5).

Due to the destructive potential of CTGCTs, complete surgical resection is the ideal treatment, as seen in this case. In some cases, particularly in severe cases or incomplete resections, adjuvant treatment including radiotherapy may need to be considered (2,4). Though there was a lack of clarity of the resection margins in our case, close clinical follow-up was recommended. A variable recurrence rate (7-11%) of these lesions has been reported (1). This is comparatively lower than recurrence in other parts of the body. Close follow-up is necessary over a period of several years due to this risk.

CTGCTs, though rare, should be considered when dealing with preauricular swellings where common diagnoses have been disproven. A full work-up is required to assess the nature of the lesion, particularly any invasive growth. Complete surgical resection is the gold standard, with close clinical follow-up.

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