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Case Report
Clinical Pathology

Treatments of tenosynovial giant cell tumours of the temperomandibular joint: a report of three cases and a review of literature

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Abstract. Tenosynovial giant cell tumours (TGCTs) are benign lesions affecting synovial joints. The classified subtypes are localized and diffuse. They seldom occur in the temporomandibular joint (TMJ). The aim of this study is to report on three new cases and to review the literature.

One patient had surgical debulking with adjuvant external beam radiation therapy (EBRT). After 1 year of follow-up, no evidence of disease was presented. The second patient was misdiagnosed and treated with denosumab. Debulking with adjuvant EBRT followed. Ten months postoperatively, no disease progression was seen. The third patient received systemic nilotinib and remained stable for over 5 years.

The literature review included 106 cases of which 95 had diffuse subtype. Most patients, had surgical excision. Thirteen (14%) patients received adjuvant EBRT. Eleven (14%) recurrences were identified. After 1-, 5- and 10 years of follow-up, an overall progression-free survival (PFS) of 99% (95% confidence interval (CI) 0.96–1), 80% (95% CI 0.68–0.94), 67% (95% CI 0.51–0.90) was calculated, respectively. Treatments for diffuse-TGCT-TMJ should be individualized depending on age, severity of symptoms, extent of disease and progression, expected mutilation of surgical interference, and current systemic treatment options. In stable disease a 'wait and see' policy, is a viable option. Additional treatments should be reserved for symptomatic, irresectable tumours or residual disease after surgical treatment with persistent complaints.

Key words: tenosynovial giant cell tumours; temporomandibular joint; pre-auricular swelling; pigmented villonodular synovitis; treatments.

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Tenosynovial giant cell tumours (TGCTs)[2], formerly known as pigmented villonodular synovitis (PVNS), are benign lesions that can affect any synovial joint[3]. It is most common in large weight-bearing joints such as the knee, hip and ankle[1]. However rare, it does occur in the temporomandibular joint (TMJ) where it is even less common compared to malignancies such as sarcomas or metastases[4].

TGCTs can be classified into localized and diffuse subtypes, which differ in clinical features and behavior[2]. Since 1973[5], over 100 TGCT cases with TMJ involvement have been reported[6,7]. Patients initially present with an indolent course of a painless pre-auricular swelling, that may become symptomatic with limited mouth opening or trismus as the tumour increases in size[8]. The duration of symptoms prior to diagnosis is on average, 11.5 months. Age at presentation is most common in the late 30 s to 40s[9].

On magnetic resonance (MR) imaging, an enlarged mass extending away from the joint with hemosiderin deposition, is typical for TGCTs[10]. This hemosiderin deposition can be depicted as 'blooming' on gradient echo or prominent hypointens on T1- and T2-weighted sequences.

Computed tomography (CT) can reveal areas of lytic bone erosion and sclerosis. Furthermore, it clearly defines the extent of the tumour, which is the focal areas of hyperdensity within the soft-tissue mass.

Both types of TGCTs are microscopically identical with a heterogeneous accumulation of both, neoplastic and non-neoplastic cells. The neoplastic cells overexpress colony stimulating factor 1 (CSF1) as a result of a translocation fusing CSF1 (at 1p11-13) to COL6A3 (at 2q35) that probably attracts the secondary non-neoplastic population of mainly histiocytes. This reactive component is known as the 'paracrine landscape effect'[11,12].

Most reports of TMJ involvement describe the diffuse form, which can be locally aggressive with bone destruction and invasion of contiguous structures. In large joints, high recurrence rates over 50% are shown, depending on time followed[13]. In contrast, for TMJ recurrence rates of 15% with at least 1 year of follow-up were reported. However, in a limited cohort of seven cases, the rate of recurrence at 5 years was 29%[7].

Management of TGCTs entails a complete synovectomy to remove all pathologic tissue. When complete removal is considered mutilating, additional treatments might be an alternative; external beam radiation therapy (EBRT), radiation synovectomy, cryosurgery, total joint arthroplasty and immune or targeted therapy. However, the effect of these treatments is unknown, caused by rarity of the disease and heterogeneity of the patients[14].

The aim of this study is to report on our experience of three TGCT-TMJ cases and review the relevant literature.

Case reports

Patient 1

A 20-year-old female patient had a progressive, pre-auricular, painful swelling and hearing loss, on the left side. She had complaints of headaches and nausea. Examination confirmed a painful pre-auricular solid-elastic, partially fluctuating swelling of 5 cm in diameter. On computed tomography (CT) and MR imaging (Fig. 1A), a large solid multilobular mass in the infratemporal fossa was detected with extension into the temporal bone showing destructive and lytic growth. The solid components were remarkably low in signal intensity on T1- and T2-weighted MR images, without bone matrix. An increased pressure on the temporal lobe was seen. No malignant characteristics were shown.

A CT wired biopsy was performed from the solid osseous expansile temporal part of the tumour. It showed a benign mesenchymal lesion with giant cells. Differential diagnostic, a chondroblastoma with secondary aneurysmal bone cyst, or a diffuse type of TGCT were considered.

Three months after initial visit, the lesion extending from the TMJ involving the skull base and intracranial extension (extradural), was surgically removed. The tumour showed a multilobular yellow, rust-brown aspect with pigmentation. The adjacent dura remained intact. The skull base, the lateral os zygomaticus and the medial part of the temporal bone were eroded. A titanium mesh was implanted in the temporal bone defect. The musculus temporalis was positioned in the TMJ.

Because of the adjacent internal carotid artery, residual tumour and visible pigmentation remained. Definitive pathology report confirmed an irradical resection of a diffuse type of TGCT. The patient received adjuvant external beam

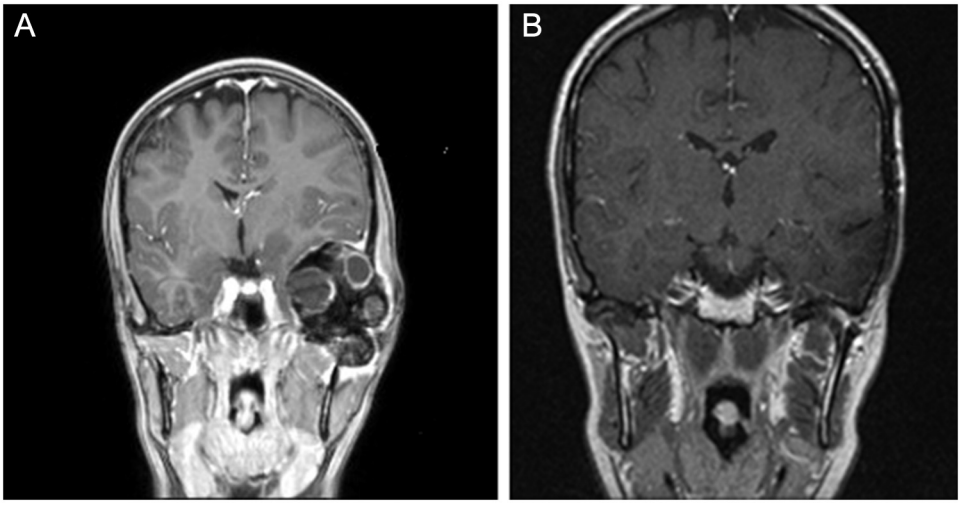

Fig. 1. (A) MR images of a 20-year-old female with TGCT-TMJ on the left (patient 1). Preoperative MR images demonstrating a large solid multilobular mass in the infratemporal fossa with extension into the left temporal bone. An increased pressure on the temporal lobe was seen. No malignant characteristics were shown. (B) MR images of a 20-year-old female with TGCT-TMJ left sided, 1year postoperatively (patient 1) after TGCT-TMJ resection on the left side. There is minimal residual coloring around the TMJ, indicating scar tissue or reactive, without visible hemosiderine deposits.

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