Chondroid Tenosynovial Giant Cell Tumor: A Rare Case Involving the Infratemporal Fossa

Abdullah Al Bakri*, Abdulaziz Bakathir, Ahmed Hashmi, Noor Al Saadi, Salma Al Shibani, Faisal Al Kalbani, Hunaina Al Kindi

Oman Medical Specialty Board, Muscat, Oman

Received: 20 November 2019

Accepted: 1 February 2020

*Corresponding author: albakri12@hotmail.com

DOI. 10.5001/omj.2021.11

ABSTRACT

Tenosynovial Giant Cell Tumor is a benign soft-tissue neoplasm that rarely occur in the craniofacial region. We report hear a case of a 27-year-old male patient presented to our unit in September 2017 with severe temporomandibular joint (TMJ) pain and progressive limitation in mouth opening. Based on clinical and imaging examinations, a well-defined soft tissue lesion was identified within the right infratemporal fossa causing pressure on the TMJ and the surrounding structures. The lesion was surgically excised through trans-mandibular and endoscopic approaches. Histopathology diagnosis revealed a rare chondroid subset of tenosynovial giant cell tumor. At 18 months follow-up review, the patient showed resolution of the jaw pain, good functional and esthetic outcomes, and no evidence of recurrence.

Keywords: Skull base; Temporomandibular Joint; Giant Cell Tumor of Tendon Sheath; Case Report; Oman.
Tenosynovial Giant Cell Tumor (TGCT) is a rare benign soft-tissue lesion that was first described by Chassaignac in 1852. Jafee and his co-workers further defined the lesion to include synovial tissue, tendon sheath, bursa, and joint structure.\textsuperscript{1} TGCT usually affect patients in their third to fifth decades of life with roughly equal sex distribution.\textsuperscript{2,3} TGCT have been reported to mainly involve the finger joints and tendon sheaths of the foot, ankle and knee. The occurrence of TCGT in head and neck region is exceedingly rare and relatively uncommon in the area of temporomandibular joint (TMJ) and skull base.\textsuperscript{1-5} The TCGT lesion is broadly classified into localized and diffuse type with literature reporting the localized form accounting for over 75\%.\textsuperscript{3-6} Recently published work by Ehrenstein \textit{et al}, reported that the prevalence per 100,000 persons for localized TCGT was 44.3 and 11.5 for the diffuse form.\textsuperscript{4} The pathogenesis of TGCT is unknown, but studies have proposed that the neoplasm could be related to bleeding following trauma and lipid metabolism diseases.\textsuperscript{2,4}

Several treatment modalities have been proposed for management of TGCT. While total surgical excision remains the main form of treatment, adjunctive therapies including radiotherapy and/or chemotherapy are being advocated in cases of incomplete excision and recurrence.\textsuperscript{3-5}

This short report describes an unusual and unique presentation of a craniofacial TGCT in a 27-year-old male and highlights the clinical presentation, investigations performed, and the surgical management that have led to a successful outcome with an 18-month follow-up.

\textbf{CASE REPORT}
A 27-year-old male was referred to our department in September 2017 with a chief complaint of a 6-month progressive trismus and jaw pain. His past medical history was unremarkable and there was no report of recent trauma or infection. Clinical assessment showed limited mouth opening of 25 mm and pain affecting the right side of the TMJ area. There was no associated clicking or deviation on mouth opening.

An orthopantomograph did not reveal any significant abnormality of the jaw and non-contrast Computed Tomography (CT) showed a well-defined round mass in the right infratemporal fossa with no cranial or TMJ bony extensions [Fig. 1]. Magnetic Resonance Imaging (MRI) revealed a defined round soft tissue mass measuring 3.2x3.1x2.8 cm, medial to right TMJ head with minimal extension into the soft tissue component of the TMJ [Fig. 2]. Both CT and MRI excluded cranial extension and vascular nature of the lesion but highlighted a close proximity to the internal carotid and internal maxillary arteries.

**Figure 1:** None-contrast CT (A) sagittal and (B) coronal views of the patient showing around soft tissue lesion with no extensions to the surrounding bones.
Figure 2: Magnetic resonance imaging of the patient (A) coronal T1-weighted view, (B) axial T1-weighted view and (C) sagittal T1-weighted view showing round soft tissue lesion with no bony involvement of the skull base and TMJ, Minimal extension into the soft tissue component of the TMJ.

Based on the clinical and imaging modalities, a differential diagnosis of neurofibroma, trigeminal schwannoma and giant cell tumor was proposed. As the lesion was located at a relatively inaccessible position and being close to critical anatomical structures, the preoperative diagnostic biopsy was neither feasible nor advisable, therefore, the patient underwent total surgical excision of the lesion under general anesthesia via trans-oral anterior lateral mandibulotomy approach for direct access to the infratemporal fossa. The approach involved a midline lip split that was extended to submandibular neck crease. The mandibular symphysis was then exposed, marked and fixed with two mini bone plates prior to being osteotomies. Subsequently, the mandible was laterally rotated, the lingual and inferior alveolar nerves were identified and protected, and the lateral pterygoid muscle was excised to closely approach the lesion, which was then visualized via endoscope and freed from all attachments ensuring total
and complete excision. The maxillary artery branches were found circulating the mass and they were identified and clipped endoscopically [Fig. 3].

Figure 3: Surgical procedure stages; (A) marking of the skin incision, (B) marking of the mandibular osteotomy and osteosynthesis fixation with two bone plates, (C) lateral mandibulotomy facilitating direct access to the infratemporal fossa, and (D) endoscopic view of the lesion.

The histopathology result showed fibrohistiocytic cells with hemosiderophages, foamy macrophages and osteoclast-type multinucleated giant cells on a background of fibrocollagenous tissue with chondroid islands and osteoid formation. The cells appeared to form a lining giving a partly cystic appearance. There were large areas of coagulated necrosis, cholesterol clefts, hemorrhage and scattered dystrophic calcifications [Fig. 4]. The histopathological features were consistent with chondroid TGCT, a rare subset of localized TGCT that was completely excised.

Figure 4: Histopathology images of the tenosynovial giant cell lesion in haematoxylin and eosin stain (A) x200 magnification showing cholesterol clefts (arrows) and hemosiderin, (B) x200
magnification showing multinucleated giant cells (arrows) in a background of fibrocollagenous tissue with chondroid islands, hemosiderophages and foamy macrophages, (C) x100 magnification showing areas of tissue necrosis (arrow).

At 18-month post-operative review, the patient showed remarkable recovery with improvement in mouth opening to 35 mm and resolution of the associated jaw pain. In addition, the 18-month post-operative MRI showed no evidence of recurrence [Fig. 5].

Figure 5: 18-month post-operative magnetic resonance imaging of the patient (A) coronal T1-weighted view, (B) axial T1-weighted view and (C) sagittal T1-weighted view showing no recurrence or residual soft tissue mass.

DISCUSSION

TGCT is a benign soft-tissue neoplasm that could be seen in head and neck region.\(^2,3\) The neoplasm can be broadly classified according to clinical appearance and biological conduct into localized or diffused form. The localized form that is also referred to as giant cell tumor of tendon sheath appears as isolated nodule of tendon sheath. The diffuse, commonly referred as pigmented villonodular synovitis, is an aggressive form that classically infiltrate into the joint, often recur, and may exhibit features of malignant transformation.\(^2,3,6\) Scientific literature states that both forms are rare with most reported case studies focusing on the diffused form.\(^2,5,8\) Our
presented case fall under the category of localized TGCT and have been diagnosed relatively early compared to published cases.⁵,⁶,⁸

The clinical presentation of TGCT over the TMJ and skull base region is considered as a rare phenomenon, with only limited cases being reported in the literature.⁵,⁶,⁹,¹⁰ A recently published population-based study covering a period of 16 years in Denmark failed to show any evidence of TGCT involving the face thus, highlighting the relative rare occurrence of TGCT in the facial region.⁴ The age of the population affected ranges from 25 - 60 years and with some reports showing a slight female predominance especially in the localized form of TGCT.⁴,⁵,⁸-¹⁰ Freeman et al and Tel et al, reported that the average age at presentation was 33.7 years with roughly 1:1 male to female ratio.³,¹⁰ Our presented case falls into this age range and in line with published reports.²-⁶,⁸-¹²

Clinically, most of the patients with TGCT presents with a very much general and non-specific symptoms including vague jaw pain and gradually progressive trismus with or without an associated facial swelling.⁵,⁶ Such incipient presentation of symptoms may lead to delayed diagnosis and significant expansion of the lesion with risk of involvement of the surrounding vital anatomical structures.¹⁰,¹¹,¹³ Published study by Zhang et al, has highlighted that the time to diagnosis of TGCT involving the TMJ and the surrounding tissues was of a range from 2 months to 15 years with an average duration of symptoms of 30 months.¹² Our presented case had an early CT and MRI as the protocol in our department facilitate that for patients with TMJ symptoms, thus, early diagnosis and treatment.
Total surgical resection of TGCT is the widely recommended treatment modality.\textsuperscript{3,5-10} The surgical approach depends mainly on the location of the lesion, hence, may include different surgical approaches.\textsuperscript{14,15} We have utilized the trans-oral lateral mandibulotomy approach combined with the endoscopic assistance. This resulted in providing us with a wide surgical access and minimal morbidity to the patient.

The literature reports an overall TGCT recurrence rate of 7-32.3\% post surgical excision.\textsuperscript{2-8} Published work has highlighted that many factors could influence the potential risk of recurrence and these includes; the destructive biological behaviour of the tumor, the degree of surgical resection (total or subtotal), and post-resection adjunctive radiotherapy usage.\textsuperscript{3,5,16} Published meta-analysis research by Freeman \textit{et al}, highlighted that gross total resection of the localised TGCT is an optimal and effective treatment modality. Futhermore, they have observed that in cases of subtotal resection, the use of adjunct radiotherapy has significantly reduced the rate of recurrence from 70\% to 14.3\%, and accordingly, suggested that when sub-total resection is encountered, adjunct radiotherapy is highly recommended as an effective treatment for controlling the disease.\textsuperscript{3} Due to the potential risk of recurrence, patients with TGCT are recommended to be placed on long-term follow-up.\textsuperscript{3,6} At 18-month follow-up, our presented patient showed an overall satisfactory recovery with no evidence of recurrence and he remains under our regular annual long-term review.

\textbf{CONCLUSION}

TGCT is a benign soft-tissue tumor, which is rarely observed in the craniofacial region. TGCT should be included in the armamentarium of clinical diagnosis for any lesions arising within or in
close proximity to the TMJ. Total resection of the lesion offers good treatment outcome and low recurrence rate.

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