



Tenosynovial Giant Cell Tumor of the Temporomandibular Joint with Skull Base Involvement: Multidisciplinary Management of a Rare Case and Review of the Literature

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Abstract

Objectives: This article presents a case of a rare tenosynovial giant cell tumor (TGCT) involving the right temporomandibular joint (TMJ) and skull base requiring complex multidisciplinary ablation and reconstruction.

Design: Case report of a TGCT affecting the TMJ with skull base involvement.

Setting: Academic medical center.

Main Outcome Measures: Feasibility of approach, return to normal dental occlusion and diet after treatment.

Results: A 24 year old dentate patient presented with a . She underwent a right temporal craniotomy with middle fossa approach and extradural excision, mandibular condylectomy, infratemporal fossa resection, resection of the bony ear canal, adipofascial free flap reconstruction, and maxillomandibular fixation. Her post-operative course was uneventful, and she returned to a regular diet with normo-occlusion post-operatively. She was treated with adjuvant radiotherapy and maintained a regular diet at her 3-month post-operative visit.

Conclusion: TGCT's with skull base involvement require treatment with a multidisciplinary surgical approach. Soft tissue reconstruction of the glenoid fossa is feasible and can allow for return to a normal diet in dentate patients.

Introduction

Tenosynovial giant cell tumors (TGCT) encompass a family of tumors that typically originate from the synovial membranes, most commonly the joints, tendon sheaths, and tendons. These tumors manifest into either a localized form which is usually indolent and most commonly presents in the fingers and hands, or the diffuse form, which is less common, but presents in larger joints such as the knees and shoulders.¹ Given the unusual origin and spread, TGCT involving the temporomandibular joint (TMJ) with are exceedingly rare, with fewer than 60 cases reported within current literature.² Even more rare are tumors with reported destruction of the glenoid fossa of the TMJ. Given the scarcity of such cases and unique challenge of concurrent skull base reconstruction and dental rehabilitation, devising an optimal management strategy presents a distinct challenge for clinicians. This case report focuses on the multidisciplinary management of a TGCT of the TMJ with skull base involvement in a dentate 24-year-old female and aims to provide future guidance for similar cases.

Case Report

The patient presented initially to clinic with purulent right sided otorrhea, episodic otalgia, dysgeusia, and headaches for 1 year. Physical examination revealed right-sided facial edema and swelling. Diagnostic imaging included contrasted computed tomography (CT) and magnetic resonance imaging. These revealed a T1 isointense, T2-heterogeneously intense expansile lesion measuring 4.1 x 4.4 x 4.6 cm within the right temporomandibular joint. The lesion's destructive nature, with central necrosis and adjacent dural enhancement, suggested a more aggressive pathology (Figure 1A-F).

Core needle biopsy and pathologic immunohistochemical studies revealed tumor cells with chondroid metaplasia and dystrophic calcification. These cells stained positive for clusterin and negative for desmin and H3K36M. Findings were consistent with tenosynovial giant cell tumor.

To address both the intracranial and extracranial tumor components as well as occlusal rehabilitation, multidisciplinary planning with head and neck surgery, neurosurgery, otology, and oral and maxillofacial surgery was required. It was recommended that treatment include a right temporal craniotomy with middle fossa approach and extradural excision, mandibular condylectomy, infratemporal fossa resection, resection of the bony ear canal, free tissue transfer, and maxillomandibular fixation. Subsequent reconstruction of the right ear canal utilized split calvarial bone harvested from the craniotomy site. Comprehensive soft tissue reconstruction of the resection defect site was performed using an adipofascial left radial forearm free flap. The patient was then placed in guiding elastics for maintenance of occlusion in the early postoperative period. The postoperative period was uneventful without complications, and the patient was ultimately discharged on post-operative day 5. After multidisciplinary tumor board discussion, adjuvant radiotherapy was recommended due to clinically aggressive nature of the tumor and positive surgical margins. Following adjuvant radiotherapy, the patient reported mild right-sided otalgia, bilateral pain with mastication, but maintained a full regular diet without weight loss. She was last seen in post-treatment follow up 3 months after radiotherapy and without complaint.

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Figures



Figure 1A

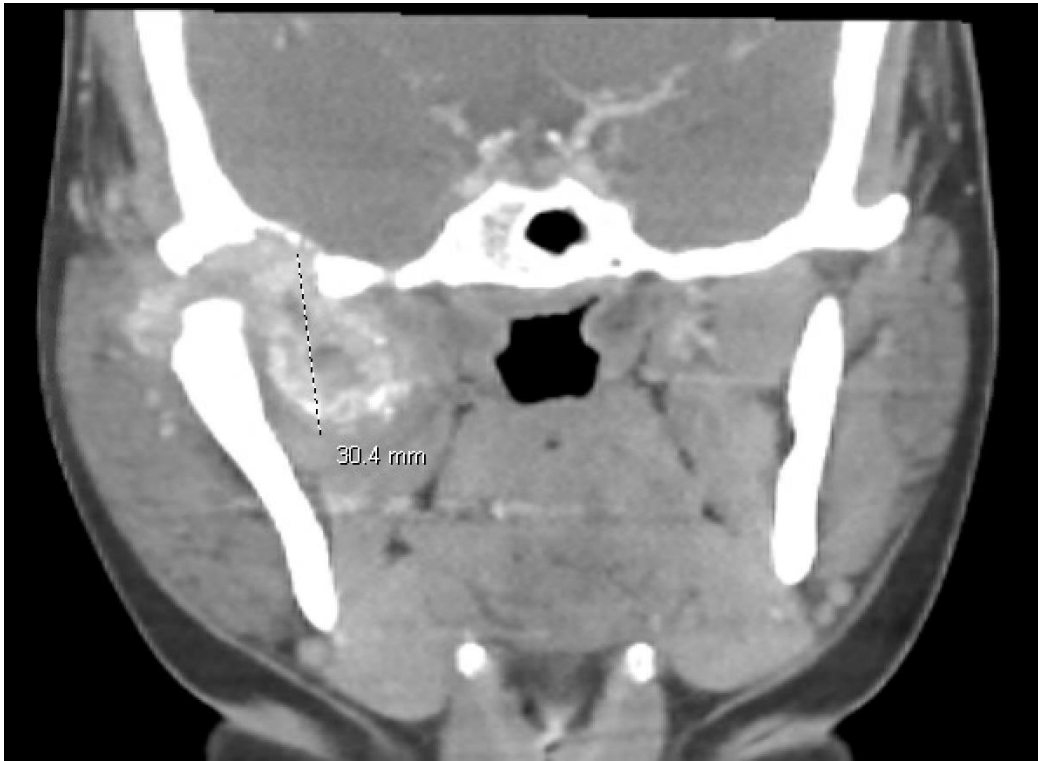


Figure 1B

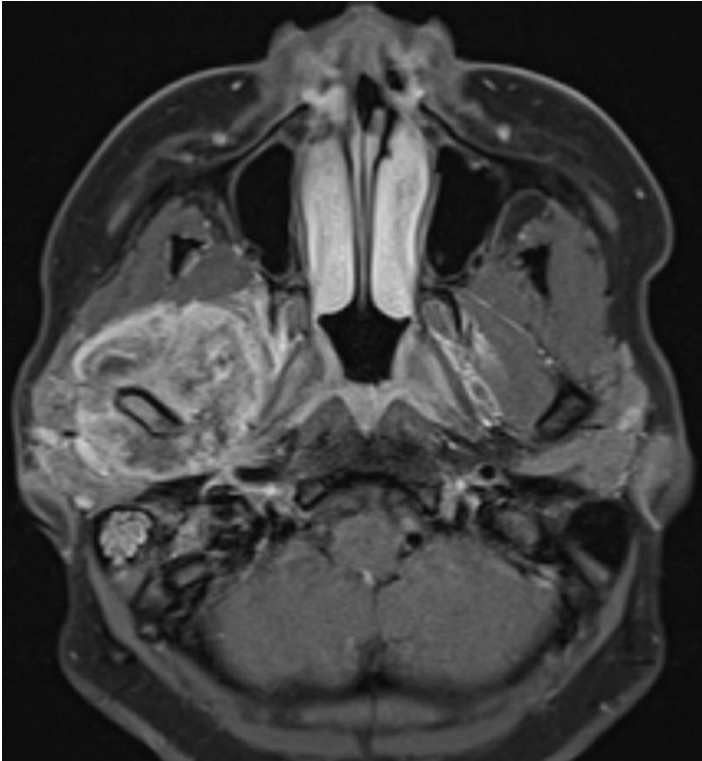


Figure 1C

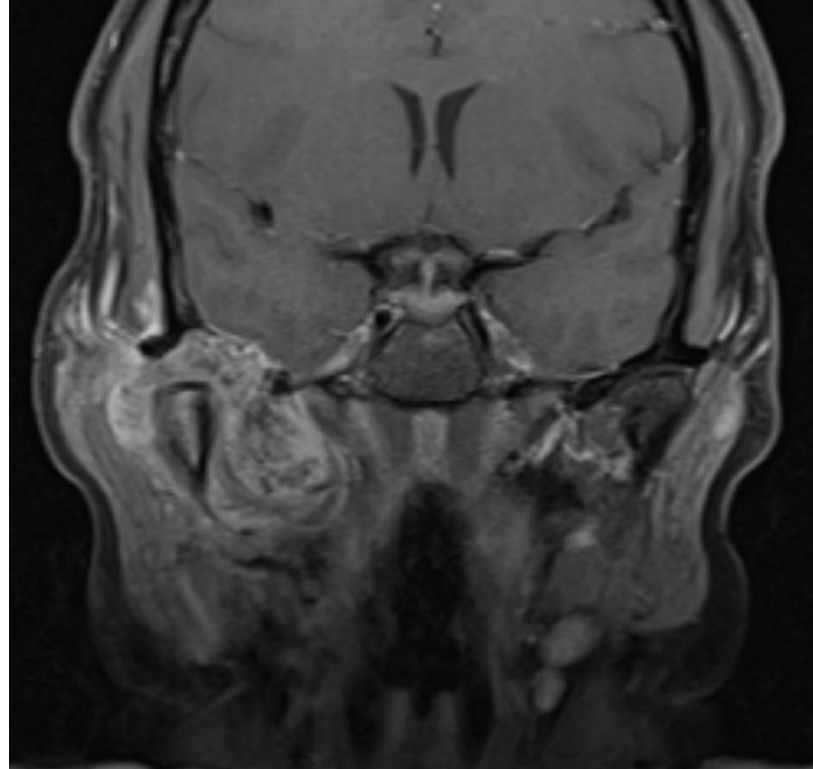


Figure 1D



Figure 1E

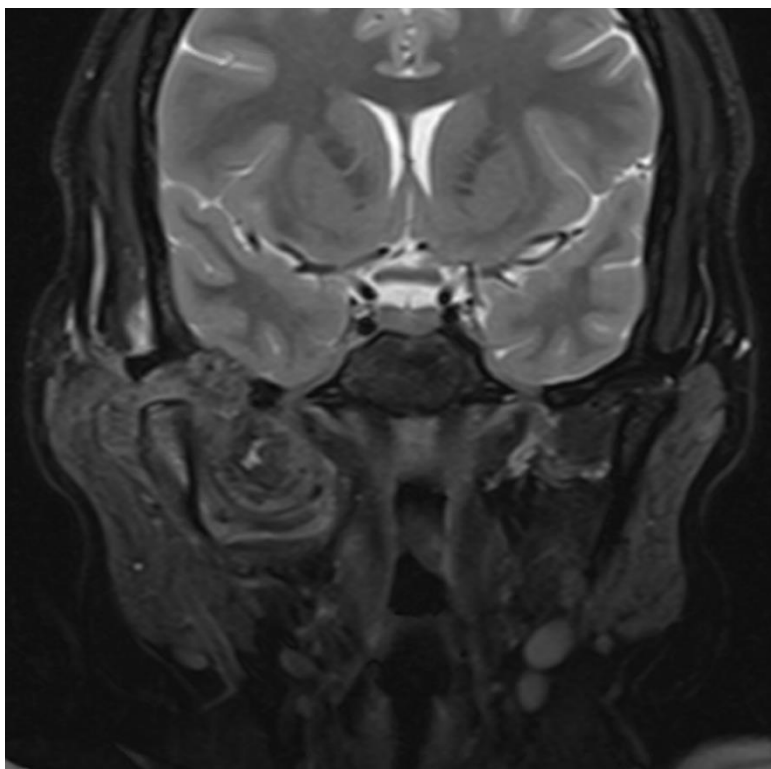


Figure 1F

Figure 1. A) Axial computed tomography (CT) view of the tenosynovial giant cell tumor (TGCT) of the temporomandibular joint (TMJ) showing central necrosis, B) Coronal CT view of the TGCT of the TMJ demonstrating skull base erosion, C) Post-contrast T1-weighted axial magnetic resonance imaging (MRI) demonstrating an isointense TGCT of the TMJ, D) Post-contrast T1-weighted coronal MRI demonstrating the isointense TGCT of the TMJ with middle cranial fossa (MCF) extent and adjacent dural enhancement, E) Post-contrast T2-weighted axial

Discussion

TGCT's of the TMJ with skull base erosion are exceedingly rare. In this case, multidisciplinary management was required due to tumor extent necessitating total resection of the TMJ, including the mandibular condyle, EAC, and glenoid fossa. Reconstruction of total TMJ defects remains poorly defined in the existing literature. In cases where the glenoid fossa remains intact, reconstruction of the TMJ with alloplastic material, cadaveric allograft, costochondral autograft, or osseous free flap is technically feasible.³⁻⁵ However, given the concomitant skull base defect in total TMJ resection, reconstruction of the mandibular condyle risks displacement of the neocondyle into the middle cranial fossa. Vascularized soft tissue reconstruction has, therefore, been investigated as a reconstructive option in patients with total TMJ defects. In Lee et al.'s series of six patients undergoing total TMJ reconstruction with free adipofascial flaps, patients were shown to have excellent occlusal and functional outcomes, though individuals requiring adjuvant radiotherapy did have worse inter-incisor distance and higher mandibular functional impairment questionnaire (MFIQ) scores.⁶ In our patient's case, soft tissue reconstruction was selected given her defect, and in accordance with Lee et al's findings, had excellent post-treatment occlusal and functional outcomes given her ability to return to a regular diet.

The complexity of our case highlights the importance of multidisciplinary approach, combining the efforts of head and neck surgery, otology, neurosurgery, and oral and maxillofacial surgery. Furthermore, the utilization of soft tissue alone for reconstruction was demonstrated to be effective in providing adequate occlusion and a return to normal diet.

Acknowledgments:

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