Tenosynovial Giant Cell Tumor of the Clinoid: Rare Condition

Aaron Gelinne, Erinc Akture, Bruce Tranmer

Key words
- Clinoid
- Giant cell
- Intracranial
- Soft tissue
- Tumor

Abbreviations and Acronyms
D-TGCT: Diffuse tenosynovial giant cell tumor
MRI: Magnetic resonance imaging
MSC: Mesenchymal stem cell
TGCT: Tenosynovial giant cell tumor
TMJ: Temporomandibular joint

Department of Neurological Surgery, University of Vermont Medical Center, Burlington, Vermont, USA

To whom correspondence should be addressed: Aaron Gelinne, B.S. [E-mail: agelinne@med.uvm.edu]

Citation: World Neurosurg. (2018) 118:168-171. https://doi.org/10.1016/j.wneu.2018.07.066

Journal homepage: www.WORLDNEUROSURGERY.org

Available online: www.sciencedirect.com

BACKGROUND: **A tenosynovial giant cell tumor (TGCT) is a rare type of tumor that primarily arises from the tendon sheath, synovium, and bursae. In rare cases, these tumors can affect joints of the head and neck such as the temporomandibular joint. This is the only case to our knowledge of an intracranial TGCT tumor of the clinoid.**

CASE DESCRIPTION: **We present the case of a 25-year-old female with a 2-year history of progressively blurred vision in her left eye without visual field defects. She denied any headaches or symptoms referable to the left eye region. Past medical history was significant for meningitis at 10 months of age. Family history was noncontributory with no history of brain tumors.**

CONCLUSIONS: **A tumor originating from the left anterior clinoid was found intraoperatively and confirmed by histology to be a TGCT.**

BACKGROUND AND IMPORTANCE

Tenosynovial giant cell tumor (TGCT) is a subtype of soft tissue tumors that typically arise from the synovium, tendon sheath, and bursae. Classically, these tumors are divided into 2 discrete categories: localized and diffuse (D-TGCT). Localized tenosynovial giant cell tumors present in articular joints with a benign course and have a favorable prognosis with local excision. D-TGCT, while benign, is locally aggressive and can affect both intra-articular and extraarticular sites, usually around the knee and shoulders. Rarely, D-TGCT tumors have been described in the head and neck, specifically the temporomandibular joint (TMJ).[1-4] To date, no cases have been published that describe the occurrence of TGCT in other extra-articular structures of the skull base. The current case is the only report to our knowledge that describes an isolated TGCT tumor at an intracranial location beyond the boundaries of the temporal bone. The patient has given her consent for the publication of this case.

CLINICAL PRESENTATION

History

A 25-year-old female presented to the author's clinic with a 2-year history of progressively blurred vision in her left eye without visual field defects. She received glasses in the fourth grade and had regular optometric examinations. At her last ophthalmic examination 1 year before our encounter, her left optic nerve appeared pale. She denied any headaches or symptoms referable to the left eye region. Past medical history was significant for meningitis at 10 months of age. Family history was noncontributory with no history of brain tumors.

Examination

On physical examination, she had an afferent pupillary defect on the left side with full extraocular movements and normal accommodation with no strabismus or nystagmus. Visual acuities with correction were 20/20+ in the right eye, 20/60+ in the left eye without pinhole improvement, and further refractive refinement to 20/50. Color vision showed no dyschromatopsia in the right eye, 5/14 correct pseudoisochromatic plates with the left eye. Amsler grid showed no metamorphopsia. Slit lamp examination revealed normal anterior segments, with no vascular tortuosity or arterialization of any vessels. Dilated stereoscopic funduscopy revealed significant pallor of the left optic nerve, which involved the entire nerve head with a cup-to-disc ratio of 0.3–0.4. No other neurologic deficits were noted. Magnetic resonance imaging (MRI) showed a 2-cm hypointense lesion originating from the left clinoid process that was compressing the left internal carotid artery and left optic nerve. A computed tomography scan showed bony hypertrophy in the region of the tumor with calcification (Figure 1).

Operation

At surgery, a left pterional craniotomy was performed. The tumor was found to be originating from the anterior clinoid with destruction of the clinoid by the tumor. The left carotid artery and left optic nerve were distorted and compressed by the tumor. The tumor had minimal vascularity with calcifications in some regions, which peeled off of the carotid artery and optic nerve well. The optic nerve was severely compressed, distorted, and thinned out medially. The optic canal was opened up widely, and the tumor was removed in that region. The ophthalmic artery was seen anteriorly and had been pushed anteriorly by the capsule of the tumor. Gross total removal of the tumor was obtained, and the specimen was sent to pathology for frozen section.

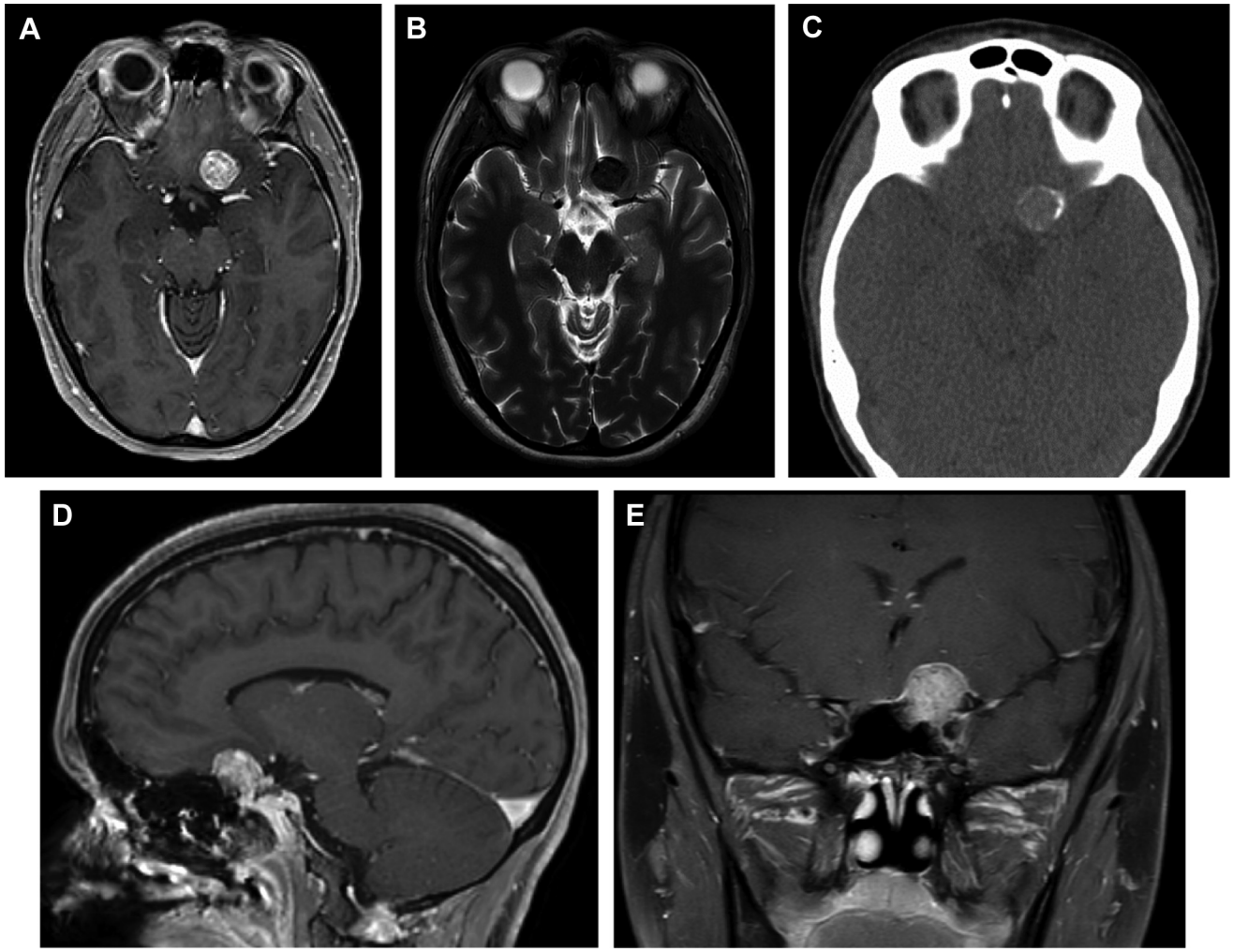

Figure 1. Preoperative magnetic resonance imaging (MRI) and computed tomography imaging. (**A**) T1 MRI axial, (**B**) T2 MRI axial, (**D**) T1 MRI sagittal post contrast, and (**E**) T1 MRI coronal post contrast showing heterogeneously hypointense extra-axial mass centered at left anterior clinoid process with compression of the adjacent left optic nerve and internal carotid artery. (**C**) CT noncontrast showing extra-axial hyperdense mass, partially calcified, centered at the level of the clinoid process on the right.

Pathologic Findings

Histologic examination of the tumor showed an admixture of mononuclear cells and giant cells, negative for S-100 and positive for clusterin and desmin, consistent with diffuse TGCT (Figure 2, clusterin and desmin staining not shown).

Postoperative Course

Postoperatively, the patient's vision was relatively stable. Visual acuity in the right eye was 20/20 with correction and Jaeger 1+ on the near card. In the left eye, acuity was 20/200+ and Jaeger 16. Color vision (Ishihara) showed no dyschromatopsia in the right eye and 1/12 correct pseudoisochromatic plates in the left eye. Follow-up MRI showed a small amount of abnormal tissue from possible post-surgical scarring or residual tumor. Unfortunately, the patient was lost to follow-up due to relocation, but no adjuvant therapies were administered at our facility.

DISCUSSION

Although exceedingly rare, extraarticular TGCT of the head and neck can affect the TMJ with invasion to local structures of the internal ear.[4] However, outside the confines of the temporal bone, isolated TGCT inside the cranium has not been documented. TGCT is rarely life threatening but can cause significant morbidity in a patient due to invasions and impingement on adjacent structures.

Etiology

The etiology of TGCT is largely unknown, but the 2 predominant theories that have prevailed suggest a neoplastic or reactive origin of mononuclear and inflammatory cells.[5,6] Studies that are more recent support a neoplastic origin with clonal proliferation as a cause for TGCT.[7] A chromosomal translocation of 1p13 leads to the overexpression of CSF1 on a small percentage of tumor cells.