Malignant Transformation of an Osteochondroma into a Chondrosarcoma in the Skull Base: A Rare Case Report

Kirby Quinn[1]* MD, Khushi Saigal[2] BS, Virginia Fernandez DO[3], Gaurav Saigal[1] MD

[1]*Department of Diagnostic Radiology, Division of Neuroradiology, University of Miami Miller School of Medicine/Jackson Health Systems, Miami FL, USA.*

[2]*University of Florida College of Medicine, Gainesville FL, USA.*

[3]*Department of Pathology, University of Miami Miller School of Medicine/Jackson Health Systems, Miami FL, USA.*

***Correspondence:** Kirby Quinn, Department of Diagnostic Radiology, Division of Neuroradiology, University of Miami Miller School of Medicine/Jackson Health Systems, Miami FL, USA.

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ABSTRACT

Osteochondromas are benign tumors of skeletal bones that are not commonly seen in the skull. Primary chondrosarcomas of the skull base, on the other hand, are more frequently seen and tend to arise from the sphenoethmoidal regions and petroclival synchondrosis. Chondrosarcoma can also develop secondarily from malignant transformation. However, only one other known reported case of malignant transformation of an osteochondroma into a chondrosarcoma of the skull base has been described in the literature. We present the case of a 29 year old male who presented with right periorbital pain and was found to have an extra-axial lobulated skull base mass with classical features of chondrosarcoma; which ended up being concordant with pathology. However, the petroclival synchondrosis was found to remain intact and not involving the lesion, thus precluding a primary chondrosarcoma that would typically arise from the synchondrosis. Instead, an osseous protrusion arising off the body of the right sphenoid bone was found originating from the lesion, and thus felt to represent a malignant transformation of a primary osteochondroma of the skull base into a chondrosarcoma.

CASE REPORT

INTRODUCTION:

Although osteochondromas are the most prevalent benign tumors in skeletal bones, they are extremely infrequent in the skull. Even rarer is malignant transformation into a chondrosarcoma. We present a rare case of a skull base chondrosarcoma caused by malignant transformation of an underlying osteochondroma.

CASE PRESENTATION:

We present a case of a 29-year-old male with no relevant past medical history who presented with a several month history of intermittent episodes of right-sided periorbital pain with extension along the right temporal bone with mild associated right eye pain and erythema. Initial magnetic resonance imaging (MRI) of the brain revealed a T1 isointense/T2 hyperintense heterogeneously enhancing extra-axial lobulated mass centered at the right cavernous sinus/Meckel's cave (Figure 1). Lobulated portions of the mass extended into the right perimesencephalic and prepontine cisterns. The lesion approximated the right anterior clinoid process anteriorly, and was seen along the lateral aspect of the dorsum sella and along the posterolateral clivus to the petroclival fissure, which was seen intact. The

mass appeared to encase the right cavernous carotid, causing narrowing of the right cavernous carotid with flow preserved distally. There were underlying foci of susceptibility artifact within the mass, corresponding to calcifications. A computed tomography (CT) scan of the skull base appeared to demonstrate an osseus protrusion from the body of the right sphenoid, with an associated soft tissue mass demonstrating an underlying 'rings and arcs' pattern of calcification (Figure 2). While the CT and MRI findings were characteristic of a chondrosarcoma, the lack of involvement of the petroclival synchondrosis was confusing. Based on the associated osseus protrusion from the body of the sphenoid, we hypothesized that the mass was likely a chondrosarcoma secondary to malignant transformation of an underlying osteochondroma.

The patient underwent a right orbital approach craniotomy with gross total resection of the mass within the cavernous sinus, along the floor of the middle cranial fossa and in the infratemporal fossa. Pathology revealed conventional chondrosarcoma, myxoid and hyaline types, intermediate grade 2/3 and with approximately 80% of the tumor demonstrating necrosis (Figure 4). The patient's postoperative course was uneventful, and the patient was discharged on postoperative day three. Repeat MRI of the skull base was obtained approximately 3 months postoperatively, showing the resection bed with expected postoperative changes. There was, however, a suspicious area of heterogenous enhancement at the posterior aspect of the petro-clival junction, which was felt to reflect residual tumor versus treatment changes (Figure 3).

DISCUSSION

Osteochondromas are the most common benign tumors of the skeletal bones, located mostly at the metaphysis [1]. Osteochondromas may be solitary or multiple, as in the case of the autosomal dominant Hereditary Multiple Exostoses (HME) [1], but in our case the patient presented with a solitary lesion and has no other known osteochondromas. They are exceedingly rare in the skull, with intracranial osteochondromas representing only 0.1-0.2% of all intracranial tumors and only 29 of these being reported in literature [1-3]. Of those total reported cases, the majority were skull base (46.4%), and of those in the skull base, 5 were centered in the posterior clinoid process, followed by 4 in the paraseller-middle cranial fossa region, 2 in the sella turcica, 2 in the foramen magnum and 1 in the petrous bone [3]. Chondrosarcomas are most commonly found in the metaphysis of long bones as well, with a predilection for the lower extremities [4,5]. Merely 2% of all chondrosarcoma cases occur at the skull base, indicating a prevalence of 0.1-0.2% [6]. Most of the reported cases of chondrosarcomas in the skull base (90%) are sporadic and arise de novo [7]. Predisposing conditions have been reported in rare instances, such as previous skull base fractures, Olliers disease and Mafucci syndrome [8]. In rare instances, metaplasia of the dura or arachnoid mater and the choroid plexus has been suggested as well [9]. Secondary chondrosarcomas, which are far less common than primary chondrosarcomas, can arise centrally from an enchondroma or peripherally from an osteochondroma [5]. To our knowledge, presentation of a chondrosarcoma in the skull base due to malignant transformation of an underlying osteochondroma has only been reported once before in literature [10]. However, we do face the limitation in not having pathological confirmation of primary osteochondroma, but postulate this to be the most likely pathophysiological explanation for this chondrosarcoma location with a preserved petroclival synchondrosis.

The most common locations of skull base chondrosarcomas are the petroclival synchondrosis or the sphenoethmoidal region. Clinical presentation is due to mass effect on the adjacent cranial nerves and brain structures, with the most common symptoms being headaches and diplopia [6,11]. In our case, presentation was mainly right-sided orbital and temporal pain, likely secondary to involvement of trigeminal nerve in the involved Meckel's cave. In the case described, there was a mass seen in the right parasellar region with the CT and MRI findings suggestive of a chondrosarcoma. These included the off-midline location, the characteristic 'rings and arcs' calcification seen on CT, the heterogenous destructive appearance on both CT and MRI and the T2 hyperintensity of the mass. However, the petroclival synchondrosis appeared intact and did not appear to be the epicenter of the lesion. The primary other differential which was considered was a chordoma, which was excluded because of the off-midline location. The patient had no known history of a primary, thus making metastatic etiology unlikely as well. A meningioma, particularly an aggressive type, was considered as the most likely alterative etiology, even though the pattern of calcification of the matrix was not typical for a meningioma. On reviewing the images again, we realized there was a bony protrusion arising from the body of the right sphenoid bone, which had the appearance of an osteochondroma. We concluded that this was likely a secondary chondrosarcoma due to malignant transformation of an osteochondroma. Pathology confirmed the lesion to be a chondrosarcoma.

CONCLUSION

We describe a rare case of suspected malignant transformation of an osteochondroma of the skull base into a chondrosarcoma. Knowledge of this occurrence is important since chondrosarcomas can occur outside of their typical primary locations when secondary in etiology.

TEACHING POINT

Primary and secondary chondrosarcomas have similar imaging features, however, primary chondrosarcomas in the clivus typically involve the petroclival synchondrosis and in the case of a clival chondrosarcoma with preserved petroclival synchondrosis, consideration of a secondary chondrosarcoma should be made.

QUESTIONS

1. What is the typical CT description of a chondrosarcoma?
   a. Bone in bone.

b. Rings and arcs.
c. Expansile.
d. Lytic.

Answer: (b) The typical CT description of a chondrosarcoma is of the 'rings and arcs' due to the characteristic appearance of the chondroid matrix. This is true of whether the chondrosarcoma is primary or secondary in etiology.

2. Chondrosarcomas of the skull base typically (not in this case) arise from the . . . ?

a. Clivus.
b. Sphenoid bone.
c. Petrous apex.
d. Petroclival synchondrosis.

Answer: (d) Usually, skull base chondrosarcomas arise from the petroclival synchondrosis or can also be seen in the sphenoethmoidal region, arising from chondroid tissue. In our case, there was no evidence that the tumor arose from the petroclival synchondrosis because it remained intact.

3. What is the usual mechanism of pathogenesis of the majority of chondrosarcomas?

a. As part of a hereditary syndrome, predisposing to tumor formation.
b. Degeneration from a primary benign bone lesion.
c. De-novo.
d. Post-traumatic.

Answer: (c): Most (90%) of chondrosarcomas are reported to arise de novo and are considered primary chondrosarcomas. Secondary chondrosarcomas, which are far less common than primary chondrosarcomas, can arise centrally from an enchondroma or peripherally from an osteochondroma, which is what we postulate in this case report.

REFERENCES

[1] Caulkins RK, Atkins K, Washmuth NB, Wei S, Wang D. Osteochondroma of the Skull: A Case Report. *Int J Anat Var*. 2021. 14(6): 99-100.

[2] Hongo H, Oya S, Abe A, Mtsui T. Solitary Osteochondroma of the Skull Base: A Case Report and Literature Review. *J Neurol Surg Rep*. 2015; 76(1): e13-e17. PMID: 26251790.

[3] Thapa BR, Katwal S. Intracranial osteochondroma arising from the posterior clinoid process: a rare case report with diagnostic challenges and comprehensive literature review. *Ann Med Surg (Lond).* 2024; 86(4): 2352-2356. PMID: 38576951.

[4] Damron TA, Ward WG, Stewart A. Osteosarcoma, Chondrosarcoma, and Ewing's Sarcoma. *Clin Orthop Relat Res*. 2007: 459: 40-47. PMID: 17414166.

[5] Gazendum A, Popovic S, Parasu N, Ghert M. Chondrosarcoma: A Clinical Review. *J Clin Med*. 2023; 12(7): 2506. PMID: 37048590.

[6] Konovalov A, Shekhtman O, Shekhtman AP, Bezborodova T. Chondrosarcoma of the Skull Base: A Case Study and Literature Review. *Cureus*. 2020; 12(12): e12412. PMID: 33659104.

[7] Kim JH, Lee SK. Classification of Chondrosarcoma: From Characteristic to Challenging Imaging Findings. *Cancers (Basel)*. 2023; 15(6): 1703. PMID: 36980590.

[8] Horvai A, Unni KK. Premalignant conditions of bone. *J Orthop Sci*. 2006; 11(4): 412-423. PMID: 16897210.

[9] Battal B, Zamora C. Imaging of Skull Base Tumors. *Tomography*. 2023; 9(4): 1196-1235.

[10] Alpers BJ. Cerebral Osteochondroma of Dural Origin. *Ann Surg.* 1935; 101(1): 27-37. PMID: 17856448.

[11] Carlson ML, O'Connell BP, Breen JT, et al. Petroclival Chondrosarcoma: A Multicenter Review of 55 Cases and New Staging System. *Oto Neurotol*. 2016; 37(7): 940-950. PMID: 27273403.

FIGURES

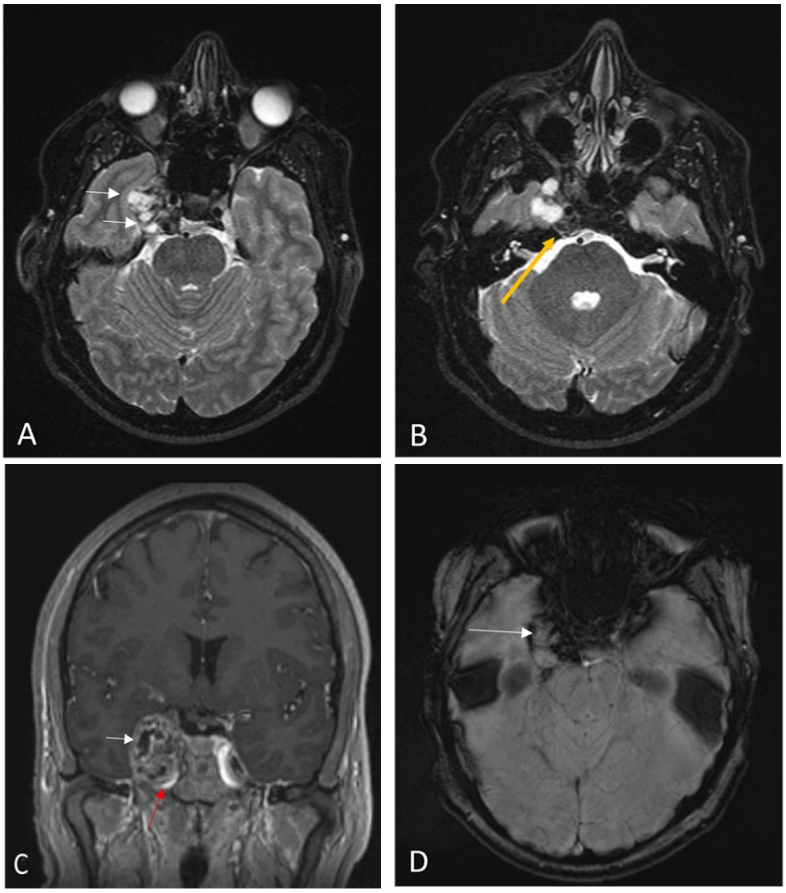

Figure 1: Preoperative MRI of the skull base. Axial T2 weighted (A, B). Coronal MPRAGE post contrast T1 (C) and Axial SWI (D) images demonstrate a mixed signal, predominantly a hyperintense T2 heterogeneously enhancing mass (white arrows) in the right parasellar region involving the cavernous sinus and the Meckel's cave (white arrows). The cavernous carotid artery is narrowed (red arrow). Mass extends posteriorly to the petroclival synchondrosis (yellow arrow), which is intact. SWI sequence demonstrates underlying foci of susceptibility consistent with calcification (long white arrow).

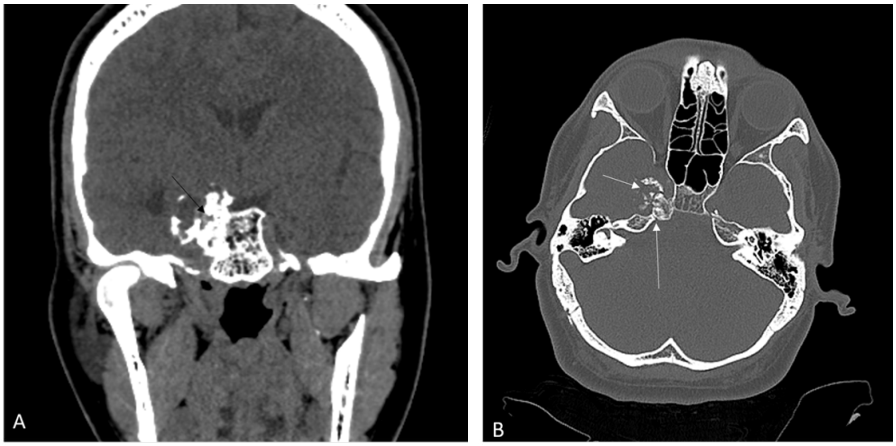

Figure 2: Coronal bone window (A) and axial soft tissue window (B) images from non-contrast CT of the brain demonstrate a large mass protruding from the body of the sphenoid (black arrow). Underlying calcifications in the form of 'rings and arcs' are noted (short white arrow). Note the petroclival synchondrosis which appears intact (long white arrow).

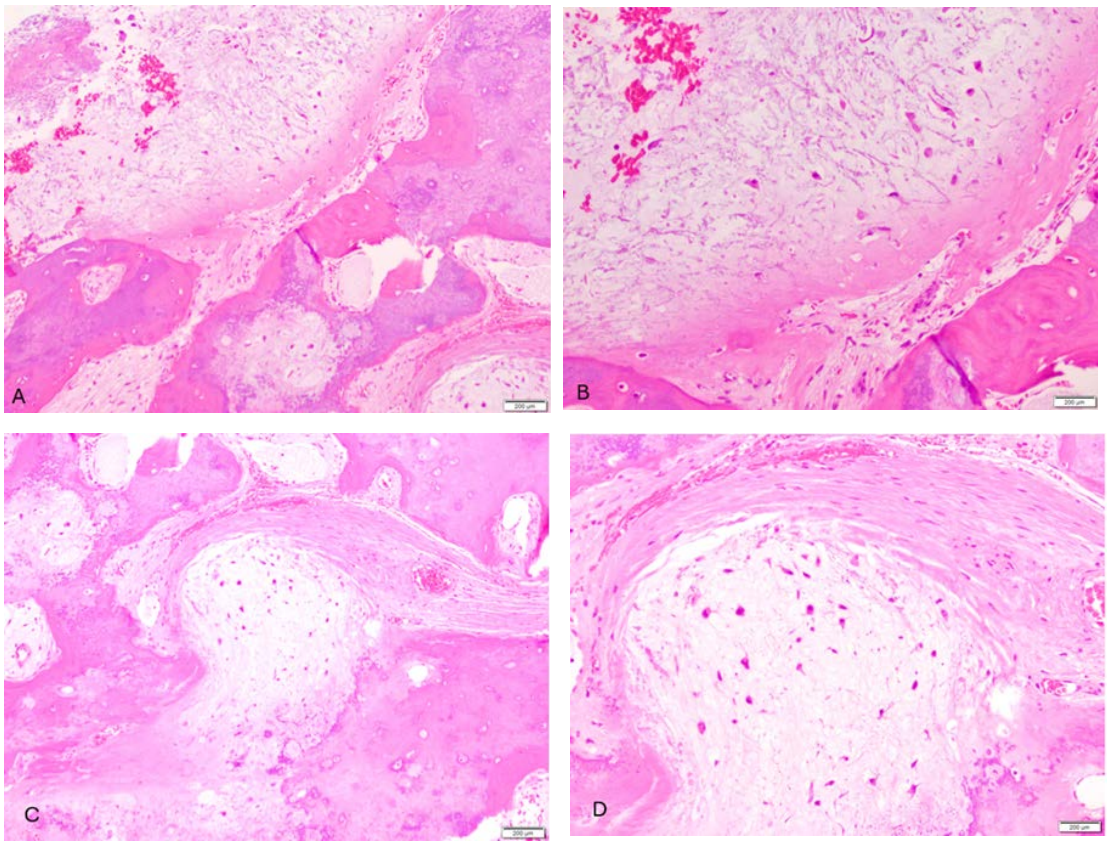

Figure 3: Hematoxylin-eosin stains illustrating the morphology of conventional chondrosarcoma with areas of necrosis with 10x magnification (A, C) and 20x magnification (B, D).

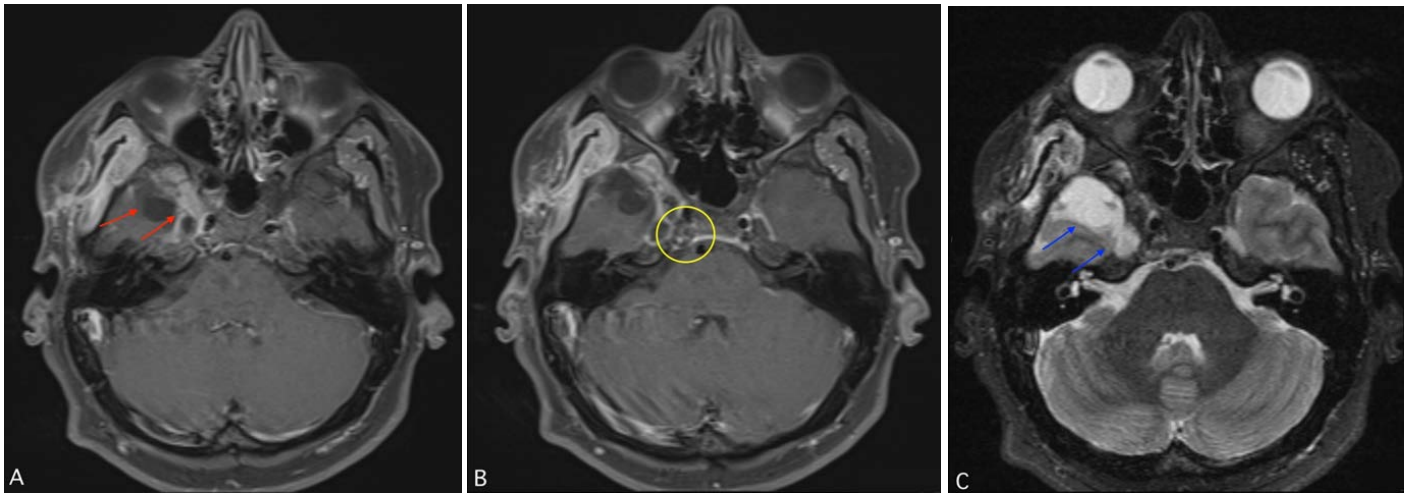

Figure 4: Postoperative MRI of the skull base. Axial T1 postcontrast (A, B) and axial T2 (C) images demonstrate postoperative changes of interval right pterional craniotomy for resection of the skull base chondrosarcoma. Lobulated area of homogenous enhancement within the medial floor of the right middle cranial fossa along the previously resected right sphenoid wing felt to represent postoperative changes (A) with corresponding T2 hyperintensity (blue arrows). Within the posterior aspect of the right petroclival junction (yellow circle), there is a focus of heterogenous enhancement, which could reflect residual tumor versus treatment changes.

KEYWORDS

Chondrosarcoma, osteochondroma, petroclival synchondrosis.

ABBREVIATIONS

MRI = Magnetic Resonance Imaging
CT = Computed Tomography
HME = Hereditary Multiple Exostoses