

Extraskelatal myxoid chondrosarcoma involving the jugular foramen and cerebellopontine angle: a case report

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Abstract

We report a rare case of extraskelatal myxoid chondrosarcoma (EMC) involving the jugular foramen and cerebellopontine angle. A 39-year-old male patient presented with initial complaints of headaches, unilateral hearing loss, and loss of balance. Cranial magnetic resonance imaging revealed an enhancing multilobulated right jugular foramen mass with medial upward extension into the cerebellopontine angle and lateral downward extension into the carotid-jugular space. The patient was preoperatively diagnosed with having a paraganglioma solely due to the tumor localization and magnetic resonance imaging finding of strong enhancement patterns. However, upon surgical resection of the intracranial component of the tumor via right lateral suboccipital craniectomy, cytologic and subsequent histopathologic examination helped delineate the diagnosis of EMC. Given the rarity of tumors involving both jugular foramen and cerebellopontine angle, we discuss histopathologic differentiating features and diagnostic pitfalls between EMC and other possible mimickers.

Keywords: Jugular foramen; extraskelatal myxoid chondrosarcoma; histopathology; immunohistochemistry; magnetic resonance imaging.

Introduction

Lesions extending to or developing from the jugular foramen are uncommon. The most frequent tumor of the jugular foramen – paraganglioma, or glomus tumor, has a reported incidence of 1 in 1.3 million [1]. Similarly, extraskelatal myxoid chondrosarcoma (EMC) is also a rare entity. Accounting for <3% of soft tissue sarcomas, EMC is a malignancy of mesenchymal tissue commonly presenting in the proximal lower extremity and limb-girdle, with the thigh being the most common site [2]. We report an exceptional case of intracranial EMC involving the jugular foramen and cerebellopontine angle, with a detailed analysis of histopathologic and radiologic distinguishing features. To our knowledge, EMC of the jugular foramen with extension into the cerebellopontine angle has only been reported once in the literature [3].

Clinical Summary

A 39-year-old male patient visited an otolaryngologist with complaints of right-sided hearing loss, headache, and loss of balance. His consciousness level was clear and neurologi-

cal examination revealed no further abnormalities. He was diagnosed with Meniere's disease and treated accordingly. After persistent worsening of his complaints, a cranial magnetic resonance imaging (MRI) was performed and revealed a multilobulated right jugular foramen mass with intracranial extension into the cerebellopontine angle, resulting in compression of the cerebellum and fourth ventricle. The lesion also had caudal extension into the right carotid-jugular space, obstructing the jugular vein. The mass was approximately 5.5 x 5 x 4.5 cm in size. It was homogeneously hyperintense on T2-weighted images (**Figure 1 a-c**), and homogeneously hypointense on T1-weighted images (**Figure 1d**); inhomogeneous enhancement was evident after contrast medium administration (**Figure 1e**). Diffusion-weighted imaging did not show any restricted diffusion. No hemorrhagic foci or calcifications were observed on susceptibility-weighted imaging. Combined with the clinical features, these imaging findings led to the preliminary diagnosis of paraganglioma. Due to the mass effect of the intracranial cerebellopontine component of the tumor, the patient underwent subtotal tumor resection via right lateral suboccipital craniectomy, followed by gamma knife radiosur-

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gery. The tumor was gray-tan with an invaded capsule (**Figure 1f**). Postoperative recovery was complicated with right-sided facial paralysis but was otherwise unremarkable. The patient's initial symptoms resolved after the operation and he remains in remission 15 months after the operation.

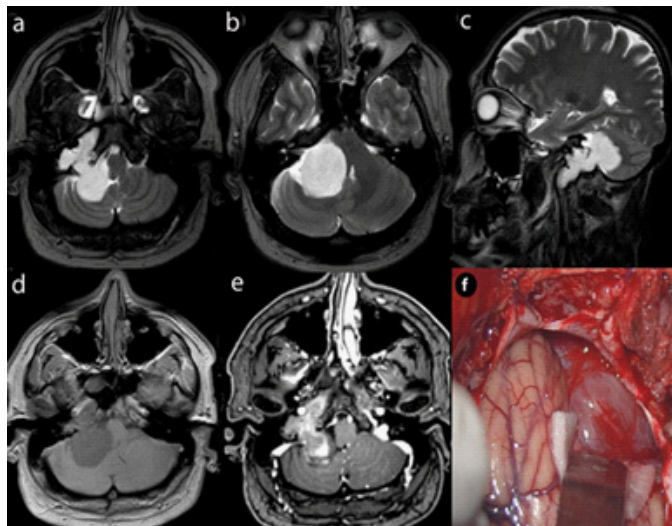


Figure 1: Brain magnetic resonance imaging (MRI) findings and intraoperative specimen of the tumor. Brain MRI. Axial (a,b) and sagittal (c) T2-weighted images show brightly homogeneous hyperintense, multilobulated right jugular foramen mass with intracranial extension to the cerebellopontine angle. (d) The tumor is hypointense on the axial T1-weighted image. (e) There is an intense enhancement on axial contrast-enhanced T1-weighted image. (f) An intraoperative view demonstrates a gray-tan tumor with an invaded capsule.

Pathological Findings

Initial cytologic examination revealed uniform tumor cells with round nuclei and eosinophilic cytoplasm (**Figure 2A**). Evaluation of the histopathologic specimen demonstrated neoplastic cells arranged in anastomosed chords within a myxoid matrix. Moreover, the neoplastic cells demonstrated cytoplasmic vacuoles, nuclear atypia, and a lack of mitotic activity (Figure 2B). Immunohistochemical staining of the specimen demonstrated positivity for periodic acid Schiff (PAS) confined to the myxoid background (**Figure 2C**). Diffuse positivity for S100 was present (**Figure 2D**). Ki-67 labeling index was estimated at 2% (**Figure 2E**). The tumor cells also exhibited positive expression for epithelial membrane antigen (EMA) (**Figure 2F**). Negative staining was noted for glial fibrillary acidic protein (GFAP), pan-cytokeratin (pan CK), brachyury (T), synaptophysin, and progesterone receptor expression. Finally, taking all of these findings into account, the tumor was diagnosed as low-grade myxoid chondrosarcoma.

Discussion

In 1972, EMC was defined as a rare neoplasm with poorly demarcated borders made up of chords or strands of cells in the bed of mucoïd stroma [6]. While EMC has been reported to arise in widespread unpredictable locations [7], the jugular foramen is a rare site of presentation that may lead to diagnostic difficulties without proper histopathologic analysis.

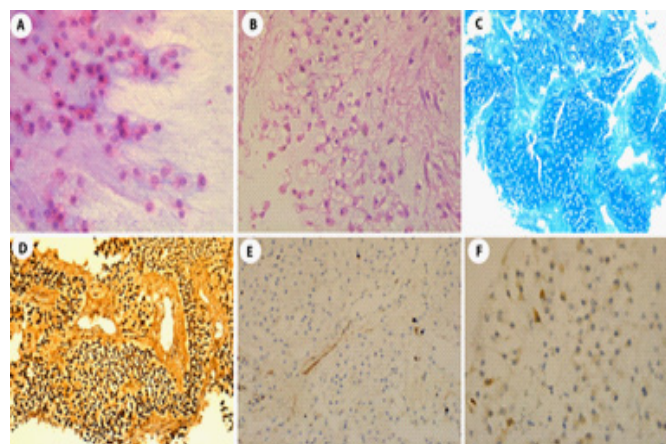


Figure 2: Microphotographs of the tumor sections stained with hematoxylin and eosin (A, B), PAS (C), S100 (D), Ki-67 (E), and EMA (F). Cytologic intraoperative specimen demonstrates uniform cells with round nuclei and eosinophilic cytoplasm, within a myxoid stroma (A). Tumor cells with prominent cytoplasmic vacuoles are observed (B). Tumor cells are arranged in lobules within a background of alcianophilic mucin (C). There is nuclear and cytoplasmic reactivity of tumor cells for S-100 protein (D). Scarce reactivity of tumor cells' nuclei, quantified at 2% (E). Uniform round tumor cells showing scattered positivity (F).

Furthermore, given the density of neurovascular structures traversing the jugular foramen, tumors extending within this area can be challenging for surgical management, further elucidating the importance of proper diagnosis that guides management.

The most commonly identified lesions extending from jugular foramen tumors are paragangliomas, also called glomus tumors, followed by nerve sheath tumors (e.g. schwannomas) and meningiomas, respectively [8]. Initial signs of tumors of this site are pulsatile tinnitus, hearing loss, and headache [3], with the latter two presenting in our patient. Ramina et al proposed that paragangliomas predominantly present with tinnitus and hearing loss, whereas schwannomas and meningiomas are more likely to cause lower nerve deficits [9]. Although the presenting symptoms and localization of the tumor were highly suggestive of a paraganglioma, pathological examination of the material precluded the provisional diagnosis.

Histopathological analysis revealed an abundance of vacuoles in the cytoplasm of the neoplastic cells and cord-like arrangement within a myxoid matrix, a common feature of chordoid tumors. Chordoid tumor is an umbrella term encompassing chordoid meningioma, chordoid glioma, chordoma, and myxoid chondrosarcoma [10]. Therefore, they were included in the differential diagnosis. Since paraganglioma has been reported to rarely display myxoid components, it was also considered [11]. Immunohistochemical staining for each of EMA, GFAP, pan CK, S-100, T, synaptophysin, and progesterone was crucial to draw a conclusion amongst the subtypes of chordoid lesions. Since chordoid glioma typically arises in the third ventricle, and characteristically shows diffuse positivity for GFAP, it was ruled out since our specimen stained negative for GFAP. Furthermore, while chordoid meningioma typically shows reactivity to S100, the staining pattern is focal, not diffuse, and is often accompanied by progesterone receptor

reactivity, a feature not present in our specimen. Given some studies report myxoid chondrosarcoma mimicking chordoma in histopathology [12-14], a careful distinction was necessary. Brachyury (T) is a protein denoting notochordal development, making it a sensitive and specific marker of chordoma. T is putatively positive in all chordomas [14]. Despite the cytoplasmic vacuoles being somewhat compatible with the characteristic physaliphorous cells of chordomas, negative labeling for T favored the diagnosis of myxoid chondrosarcoma over chordoma. Finally, the absence of Zellballen pattern, or nest-shaped cellular orientation, along with negative staining for synaptophysin, helped factor out the possibility of the lesion being a paraganglioma with myxoid features. Thus, based on the histopathologic finding of cells with vacuolated cytoplasm, arranged in chordoid morphology in a myxoid matrix, as well as positive immunohistochemical staining for EMA & S-100, the specimen was evaluated as a low-grade extraskeletal myxoid chondrosarcoma. While the cytogenetic analysis was not conducted in our institution, it should be noted that it provides important diagnostic utility. EMC is characterized by a unique chromosomal translocation, commonly involving gene fusion of EWSR1 to NR4A3 within t(9;22) (q22; q11.2), a feature that can further elucidate EMC from potential mimickers [15-17].

In conclusion, this case demonstrates the paramount importance of pathology in assessing lesions in atypical locations. We describe a rare case with initially ambiguous features, as evidenced by the obscure radiological findings. Immunohistochemical distinction, mainly through EMA, S100, and Brachyury, was key to ruling out mimicking alternative diagnoses that may have changed the postoperative management approach, as a low-grade tumor such as EMC differs in management compared to a more aggressive mimicker, like chordomas.

Ethical statement

The patient gave informed signed consent for the writing and publication of this article.

Disclosure of interest

The authors declare that they have no conflict of interest.

References

- Moffat DA, Hardy DG. Surgical management of large glomus jugulare tumours: infra- and trans-temporal approach. *J Laryngol Otol.* 1989;103(12):1167-1180. [DOI:10.1017/s0022215100111284].
- Lucas DR, Stenman G. Extraskeletal myxoid chondrosarcoma. In: Fletcher CD, Bridge JA, Hogendoorn PCW, Mertens F, editors. *World Health Organization classification of tumour, pathology, and genetics of tumours of soft tissue and bone.* Lyon: International Agency for Research on Cancer; 2013;223-4.
- Cummings TJ, Bridge JA, Fukushima T. Extraskeletal myxoid chondrosarcoma of the jugular foramen. *Clin Neuropathol.* 2004;23(5):232-237.
- Slaba S, Haddad A, Zafatayeff S, et al. Imaging of an exceptional tumor: myxoid chondrosarcoma of the jugular foramen. *J Med Liban.* 2001;49(4):231-233.
- Harvey SA, Wiet RJ, Kazan R. Chondrosarcoma of the jugular foramen. *Am J Otol.* 1994;15(2):257-263.
- Enzinger FM, Shiraki M. Extraskeletal myxoid chondrosarcoma. An analysis of 34 cases. *Hum Pathol.* 1972;3(3):421-435. [DOI:10.1016/s0046-8177(72)80042-x].
- Oliveira AM, Sebo TJ, McGrory JE, et al. Extraskeletal myxoid chondrosarcoma: a clinicopathologic, immunohistochemical, and ploidy analysis of 23 cases. *Mod Pathol.* 2000;13(8):900-908. [DOI:10.1038/modpathol].
- Vogl TJ, Bisdas S. Differential diagnosis of jugular foramen lesions. *Skull Base.* 2009;19(1):3-16. [DOI:10.1055/s-0028-1103121].
- Ramina R, Maniglia JJ, Fernandes YB, et al. Jugular foramen tumors: diagnosis and treatment. *Neurosurg Focus.* 2004;17(2):E5. Published 2004 Aug 15. [DOI:10.3171/foc.2004.17.2.5].
- Takei H, Powell SZ, Rivera A. Immunohistochemistry and brain tumors. In: *Encyclopedia of the Neurological Sciences.* 2nd ed. Oxford, UK: Academic Press; 2014:679-683
- Laforga JB, Vaquero M, Juanpere N. Paragastric paraganglioma: a case report with unusual alveolar pattern and myxoid component. *Diagn Cytopathol.* 2012;40(9):815-819. [DOI:10.1002/dc.21665].
- Doyle LA, Hornick JL, Chapter 4 Immunohistology of Neoplasms of Soft Tissue and Bone, *Diagnostic Immunohistochemistry,* Publisher: Elsevier, 2019.
- Sangoi AR, Dulai MS, Beck AH, Brat DJ, Vogel H. Distinguishing chordoid meningiomas from their histologic mimics: an immunohistochemical evaluation. *Am J Surg Pathol.* 2009;33(5):669-681. [DOI:10.1097/PAS.0b013e318194c566].
- Vujovic S, Henderson S, Presneau N, et al. Brachyury, a crucial regulator of notochordal development, is a novel biomarker for chordomas. *J Pathol.* 2006;209(2):157-165. [DOI:10.1002/path.1969].
- Hirabayashi Y, Ishida T, Yoshida MA, et al. Translocation (9;22)(q22;q12). A recurrent chromosome abnormality in extraskeletal myxoid chondrosarcoma. *Cancer Genet Cytogenet.* 1995;81(1):33-37. [DOI:10.1016/0165-4608(94)00201-0].
- Sandberg AA. Genetics of chondrosarcoma and related tumors. *Curr Opin Oncol.* 2004;16(4):342-354. [DOI:10.1097/01.cco.0000129678.72521.e5].
- Stenman G, Andersson H, Mandahl N, Meis-Kindblom JM, Kindblom LG. Translocation t(9;22)(q22;q12) is a primary cytogenetic abnormality in extraskeletal myxoid chondrosarcoma. *Int J Cancer.* 1995;62(4):398-402. [DOI:10.1002/ijc.2910620407].