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Case Report

Jugular foramen schwannoma mimicking paraganglioma: case report and review of imaging findings

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ARTICLE INFO

Article history:

Received 15 September 2015

Accepted 5 December 2015

Available online 18 January 2016

Keywords:

Schwannoma

Paraganglioma

Jugular foramen

Flow voids

MRI

Imaging

ABSTRACT

We present the case of a 30-year-old woman who presented with an 11-year history of chronic occipital headaches and a 12-month history of worsening difficulty speaking and/or swallowing, facial spasms, hearing loss, and dizziness. A large lytic mass was found centered in the left jugular foramen (JF) on computed tomography examination; follow-up magnetic resonance imaging showed an avidly enhancing mass with prominent central flow voids. Histopathologic examination after surgical resection revealed the mass to be a schwannoma. Prominent central vascularity is an unusual presentation for JF schwannomas. Our report provides a review of magnetic resonance imaging features of intrinsic JF lesions relevant to our case.

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Introduction

Schwannomas are benign tumors of nerve sheath origin that most often arise from the superior vestibular branch of the eighth cranial nerve in the cerebellopontine angle [1]. However, schwannomas may rarely originate from the ninth or tenth cranial nerves within the jugular foramen (JF) [2]. The contents of the JF guide a unique differential in the consideration of an intrinsic JF lesion. Besides schwannomas, paragangliomas may arise from scattered collections of paraganglial tissue, and meningiomas may form from an intraforaminal connective tissue layer. Additional

considerations include metastatic tumor, primitive neuroectodermal tumor, and jugular vein pseudomass [3,4].

The imaging characteristics of these lesions are often similar. However, paragangliomas are classically associated with a “salt & pepper” appearance on magnetic resonance imaging (MRI) because of multiple areas of signal void interspersed with hyperintense foci due to slow flow or hemorrhage. Accordingly, paragangliomas are highly vascular on angiography [5]. In contrast, schwannomas are typically described as avascular or hypovascular on angiography, without central flow voids on MRI [2,4,5]. We describe an unusual case of a JF schwannoma with prominent central

Competing Interests: The authors have declared that no competing interests exist.

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<http://dx.doi.org/10.1016/j.radcr.2015.12.002>

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flow voids on MRI and present an overview of the relevant imaging findings.

Case report

A 30-year-old woman with a family history of cancer and an 11-year history of chronic occipital headaches presented with a chief complaint of increasing difficulty speaking and reaching high notes whereas singing over the past year. She also reported difficulty swallowing, left-sided facial spasms, left-sided hearing loss, tinnitus, and dizziness. On physical examination, deficits in the left eighth through twelfth cranial nerves were identified.

A contrast-computed tomography (CT) scan of her head was performed for further evaluation and revealed a scalloped expansion of the JF with a mildly enhancing soft tissue mass measuring 2.6 cm in diameter (Fig. 1A). The mass showed intracranial extension to the cerebellopontine angle and extracranial extension into the carotid space (Figs. 1B and C).

Subsequent MRI examination showed a $3.8 \times 3.5 \times 3.5 \text{ cm}^3$ dumbbell-shaped lesion with a “salt & pepper” appearance on coronal T2W-imaging in the left JF (Fig. 2A), with involvement of the left internal auditory canal, hypoglossal canal, clivus, and cerebellopontine angle. This lesion demonstrated avid enhancement on postcontrast imaging (Fig. 2B). There was associated mass effect on the left medulla and anterior inferior left cerebellum, and obscuration of the left-sided cranial nerves VII–XII. The distal left cervical internal carotid artery just proximal to the horizontal petrous segment was asymmetrically small in caliber, and the left internal jugular vein was not seen.

An elective operative approach was selected. Carotid angiography and tumor embolization were performed before surgery. External carotid angiogram revealed angiographic tumor blush just distal to the origin of the ascending pharyngeal artery anteriorly at the left skull base with vascular supply from the ascending pharyngeal artery (Fig. 3).

Postembolization angiogram showed no identifiable residual tumor blush.

Mastoidectomy and suboccipital craniectomy were performed and both translabyrinthine and retrosigmoid approaches were used to gain access to the tumor. Microsurgical resection of the tumor was performed under intraoperative cranial nerve monitoring, with intentional residual tumor left behind in the inferior pole of the JF and in the internal auditory canal because of indistinct tissue plane between nerve and tumor capsule. Closure was accomplished via abdominal fat graft and titanium plating.

Histopathologic examination showed palisading nuclei and spindle cells, and the sample was positive for S100 protein and negative for neurofibrillary and epithelial stains on immunohistochemistry. The final pathologic diagnosis was schwannoma with no evidence of malignancy. The diagnosis was definitive with no evidence of paraganglioma.

The patient recovered appropriately postoperatively, and was discharged on postoperative day 7. There was minimal improvement from her preoperative clinical symptomatology. The patient returned 3 weeks later complaining of postnasal drip, rhinorrhea, headaches, fever, sore throat, and malaise; a cerebrospinal fluid leak was identified and the patient underwent exploration of the posterior and middle cranial fossae for closure of the defect. The patient was discharged 3 days after the secondary repair, and as of the time of this report is recovering appropriately.

Discussion

JF masses can be classified on the basis of whether they arise from structures within the foramen or external to it, that is, intrinsically or extrinsically [4]. Extrinsic lesions include chordoma, inflammatory lesions, and rhabdomyosarcoma; and intrinsic lesions include paraganglioma, schwannoma, meningioma, metastatic disease, jugular vein pseudomass, and others. Based on the clinical history and imaging

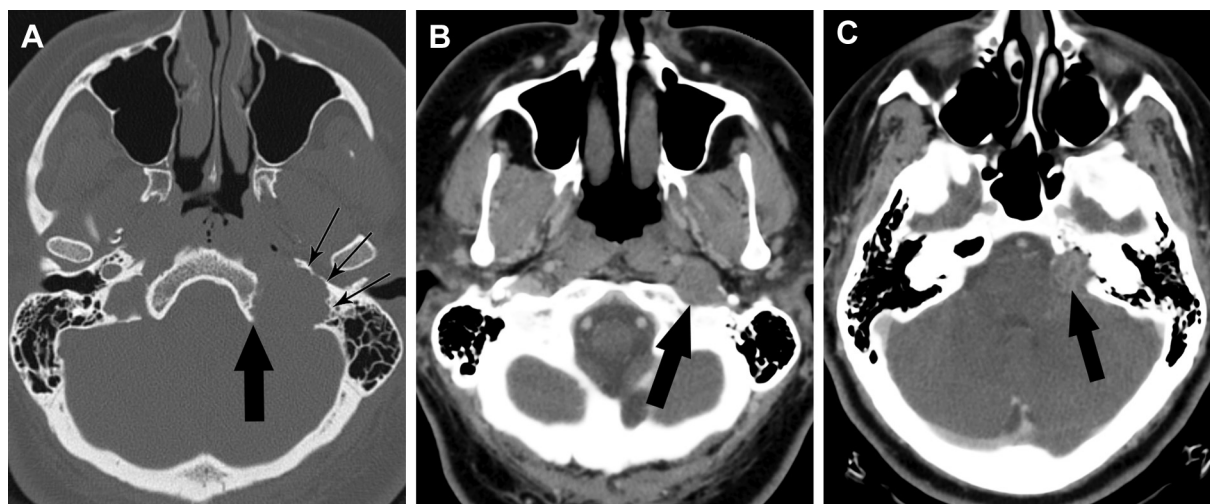


Fig. 1 – Enhanced-CT examination showing a mildly enhancing mass in the L-JF with (A) scalloped expansion of the JF (small arrows) with medial extension (large arrow) on bone windows; (B) extracranial extension in the carotid space; and (C) intracranial extension with involvement of the cerebellopontine angle (arrow).

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