



# Metastatic Renal Cell Carcinoma Masquerading as Jugular Foramen Paraganglioma: A Role for Novel Magnetic Resonance Imaging

Andrew J. Thomas, MD<sup>1</sup>; Richard H. Wiggins III, MD, CIIP<sup>1,2</sup>; Richard K. Gurgel, MD<sup>1</sup>

<sup>1</sup>Division of Otolaryngology – Head and Neck Surgery, Department of Surgery; University of Utah, Salt Lake City, UT, USA

<sup>2</sup>Division of Clinical Radiology, Department of Radiology and Imaging Sciences; University of Utah, Salt Lake City, UT, USA

## Abstract

A 55-year-old man presents with what appears clinically and radiographically to be a paraganglioma centered at the jugular foramen (JF). The tumor was surgically resected and surprisingly discovered on post-operative pathology to be metastatic renal cell carcinoma (RCC). Imaging by computed tomography (CT) and magnetic resonance imaging (MRI) was retrospectively compared between three histologically confirmed cases of paraganglioma, and our case of skull base RCC. The JF lesion types were indistinguishable on CT and traditional MRI, however they were clearly distinct in regard to diffusion weighted magnetic resonance imaging (DW-MRI) calculated apparent diffusion coefficient (ADC) values.

## Introduction

Jugular foramen (JF) tumors include jugular paraganglioma (JP), the most common primary tumor of the JF, and less commonly metastatic disease. The most common symptoms of JPs are pulsatile tinnitus and hearing loss, and the most common finding on exam is a pulsatile red mass behind the tympanic membrane. Imaging is the mainstay of diagnosing JF lesions. JPs are generally felt to have characteristic imaging findings: Computed tomography (CT) demonstrates an infiltrative pattern of growth and bony destruction. Magnetic resonance imaging (MRI) reveals the classic “salt-and-pepper” appearance due to heterogeneous hemorrhage/slow flow areas (salt) and high flow vessels creating vascular flow voids (pepper). Contrast enhancement in these lesions is avid. Metastatic disease to the JF is rare, and primary disease is most often of the prostate, breast, kidney, or lung. Renal cell carcinoma (RCC), like JP, is a slow growing, highly vascular tumor. Unlike JP, which is more common in women (3-6:1 women:men), RCC is twice as common in men as in women. Metastatic RCC has rarely been reported in the temporal bone and confused for JP. Radiographically, metastatic RCC may appear similar to JP with a slow-growing pattern of bony permeative destruction on CT, highly vascular with avid contrast enhancement, and similar “salt-and-pepper” appearance as JP on MRI.

## Case

### History:

- 55-year-old man with 2-week history of right sided low frequency pulsatile tinnitus, headaches, and numbness and weakness of the right side of his face.
- No dizziness, ear pain, or otorrhea and had no history of otologic surgery.
- Hypertension.

### Exam and Studies:

- Right hemi-facial hypesthesia as well as slight right facial weakness with facial expression (House-Brackmann 2/6).
- Red, pulsatile, inferiorly based lesion at the floor of the external auditory canal.
- Mild asymmetric right conductive hearing loss.
- Normal 24-hour level of urine metanephrine (87 µg) and normetanephrine (187 µg).

### Imaging:

- Contrast-enhanced computed tomography (CT) of the temporal bones demonstrated a contrast enhancing lesion centered at the right JF eroding surrounding bone of the skull base measuring 3.8 x 3.0 cm (**Figure 1**).
- Magnetic resonance imaging (MRI) of the brain demonstrated avid contrast enhancement, T1 isointense and T2 heterogeneous, with multiple flow voids and cystic degeneration (**Figure 1**).

### Procedures:

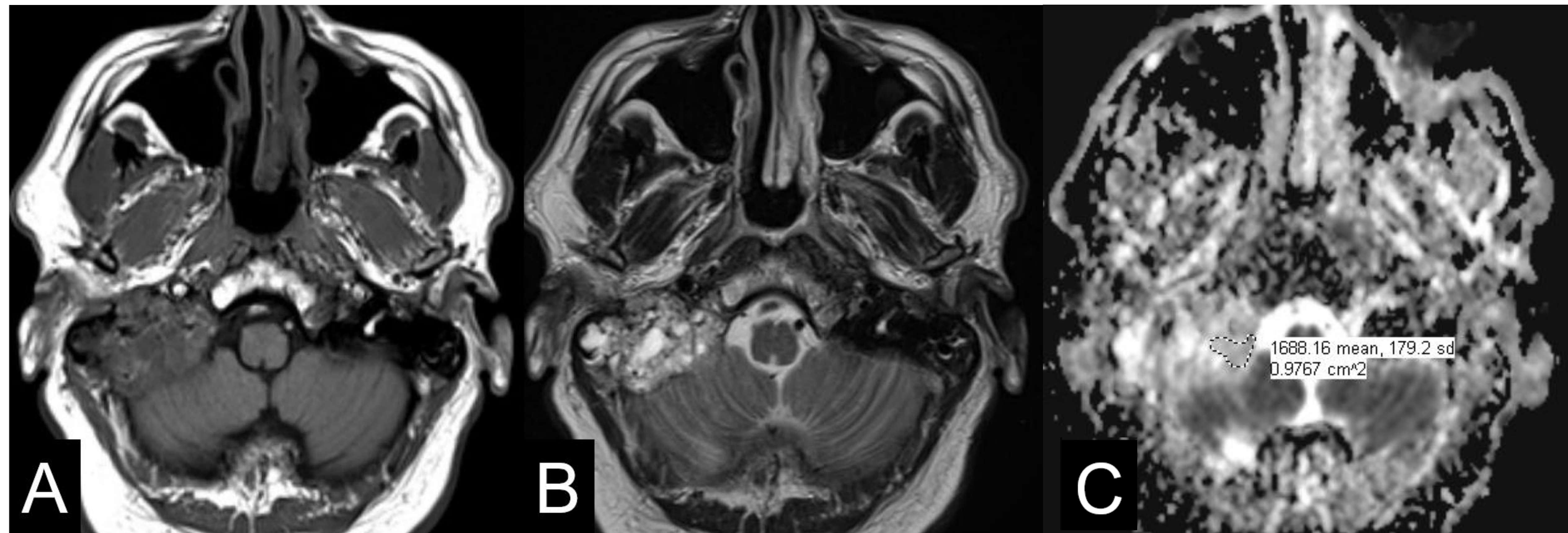
- Angiogram and embolization: Highly vascular tumor
- Transcervical and post-auricular infratemporal approach for removal of the JF lesion (large vascular tumor consistent in appearance with JP).

### Pathology:

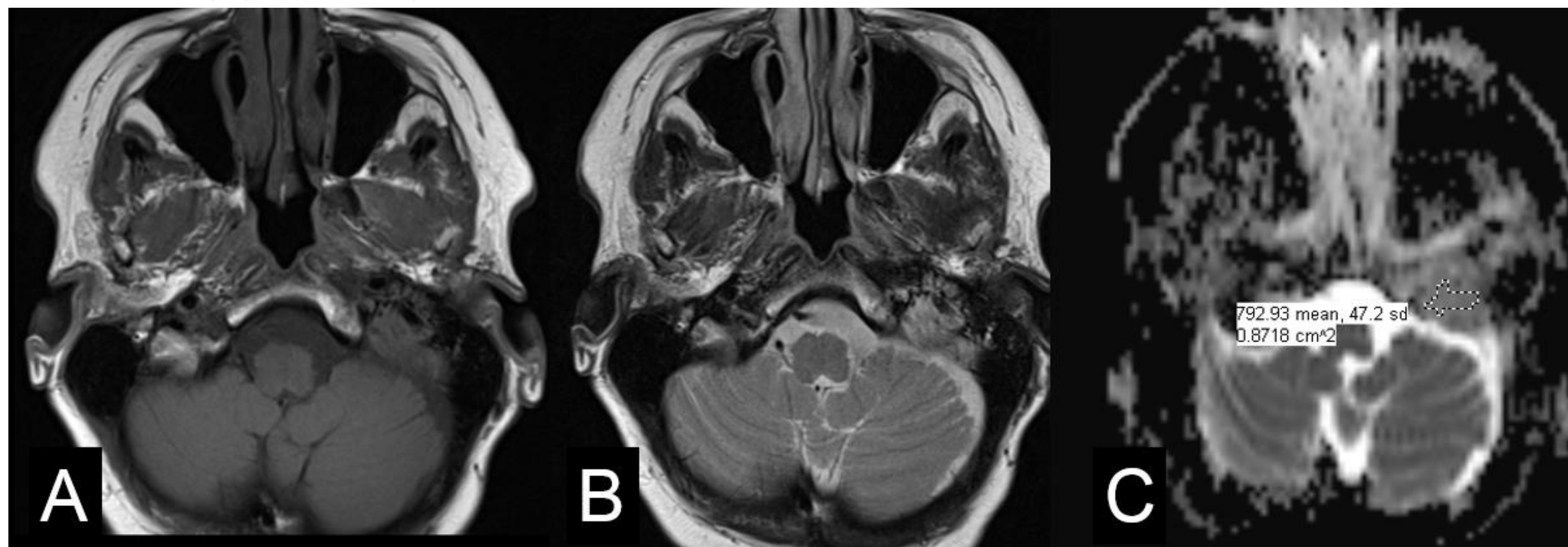
- **Metastatic clear cell tumor** morphologically and immunohistochemically (strongly and diffusely positive for EMA, PAX8, and vimentin) **consistent with renal cell carcinoma (RCC)**. Negative for chromogranin and synaptophysin, which are markers of paraganglioma.

## Contact

Andrew J. Thomas, MD  
Otolaryngology – Head and Neck Surgery  
University of Utah  
Email: andrew.thomas@hsc.utah.edu



**Figure 1:** Jugular foramen metastatic renal cell carcinoma MRI imaging findings. A) T1 low signal B) T2 high signal with heterogeneous flow voids C) DWI imaging with an average (of 3 measurements) calculated ADC value of  $1.685 \times 10^{-3} \text{ mm}^2/\text{s}$  for the tumor.



**Figure 2:** Jugular foramen paraganglioma MRI imaging findings (single case example). A) T1 low signal B) T2 high signal with heterogeneous flow voids C) DWI imaging with an average (of 3 measurements) ADC value of  $0.796 \times 10^{-3} \text{ mm}^2/\text{s}$  for this JP.

JF Tumor Type	CT	T1 MRI	T1 Gad MRI	T2 MRI	Angiogram	Average ADC (SD)
<b>Paraganglioma (JP)</b> (3 measurements x 3 cases reviewed)	Erosive/destructive. Contrast enhancing.	Iso/Low.	Avid contrast enhancement.	High signal. Heterogeneous flow voids.	Highly vascular.	<b>0.695</b> $\times 10^{-3} \text{ mm}^2/\text{s}$ (SD $0.111 \times 10^{-3} \text{ mm}^2/\text{s}$ )
<b>Renal cell carcinoma (RCC)</b> (3 measurements)	Erosive/destructive. Contrast enhancing.	Iso/Low.	Avid contrast enhancement.	High signal. Heterogeneous flow voids.	Highly vascular.	<b>1.685</b> $\times 10^{-3} \text{ mm}^2/\text{s}$ (SD $0.015 \times 10^{-3} \text{ mm}^2/\text{s}$ )
<b>Comparison</b>	Same	Same	Same	Same	Same	<b>P &lt; 0.0001</b>

## Discussion

- Skull base metastatic RCC may be mistaken for JP by clinical and radiographic findings. The “pathognomonic” finding of flow voids in JP are also seen in metastatic RCC.
- Diffusion weighted (DW) MRI measures random movement of water molecules within tissue (Brownian motion). The apparent diffusion coefficient (ADC) quantifies the magnitude of water molecule diffusion, and is inversely related to the cellularity of tissue/tumor.
- We retrospectively calculated ADC values based on routinely obtained DW-MRI for our case of metastatic RCC (**Figure 1**) and three histologically proven cases of JF paraganglioma (**Figure 2**). Conventional MRI findings on T1 and T2 sequences were indistinguishable, as expected, though DW-MRI ADC calculations demonstrated distinct differences.
- These findings are encouraging, for the utility of DW-MRI in distinguishing these entities, but further research is needed to confirm consistent and reliable imaging differences between RCC and JP by DW-MRI.

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