

INNOVATIVE CLINICAL IMAGE

Hemangioblastoma of the Cerebellopontine Angle Evaluated with Pseudocontinuous Arterial Spin Labeling

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Hemangioblastomas of the cerebellopontine angle (CPA) that emerge extra-axially from the peripheral nervous system are extremely rare. We report a case of hemangioblastoma of the CPA evaluated by pseudocontinuous arterial spin labeling (pCASL). The high rate of tumor blood flow determined using pCASL provided additional useful information for the differential diagnosis of the CPA tumors in this patient.

Keywords: *hemangioblastoma, cerebellopontine angle, pseudocontinuous arterial spin labeling*

Hemangioblastoma of the cerebellopontine angle (CPA) that emerges extra-axially from the peripheral nervous system is extremely rare, having been reported only in four cases: three in the vestibular nerve and one in the trigeminal nerve.¹ Hemangioblastoma of the CPA represents a diagnostic challenge because the CPA is an atypical location for this tumor and the imaging findings are similar to those of vestibular schwannoma and meningioma, which are much more common in this region. Owing to the risk of perioperative bleeding, presurgical diagnosis is of great importance as it allows clinicians to select the optimal management approach for a particular patient.

Arterial spin labeling (ASL) is a method for noninvasively evaluating blood flow, in which the intrinsic spin state of the blood is used as a tracer. This perfusion method has been successfully applied to evaluate brain tumors. ASL sequences have recently involved the use of pseudocontinuous ASL (pCASL) and 3D fast spin-echo acquisition techniques, which exhibit few susceptibility artifacts for evaluating the vascularity of skull base tumors. pCASL techniques reportedly provided useful information that enables clinicians to differentiate between skull base meningioma and schwannoma.²

An 84-year-old man presented with vertigo and deafness. He had a left-sided cerebellar tumor that was resected 34 years ago, but its histologic diagnosis was unknown. His family history was unremarkable. The patient's clinical workup included detailed documentation of the clinical history, ophthalmoscopic

examination, magnetic resonance imaging (MRI) of the brain and spine, and computed tomography of the abdomen. Tests for von Hippel–Lindau disease yielded negative results. Brain MRI performed using a 3T scanner (Ingenia; Philips Healthcare, Best, The Netherlands) revealed a round tumor in the right CPA with a maximum diameter of 18 mm that demonstrated hypointensity on T₁-weighted images and heterogeneous hyperintensity on T₂-weighted images (Figs. 1a and 1b). T₂-weighted images showed several flow voids in and around the mass (Fig. 1b). On diffusion-weighting imaging (DWI), the mass had a low signal (Fig. 1c). Contrast-enhanced T₁-weighted images showed strong enhancement of the mass (Fig. 1d). 3D fast spin-echo sequence with background suppression covering the entire brain was used for pCASL. The parameters for pCASL images were as follows: TR = 6000 ms, TE = 40 ms, FOV = 240 mm, reconstructed matrix size = 80 × 80, number of excitations = 3, labeling duration = 1650 ms, post-labeling delay = 2000 ms, slice thickness = 3 mm, number of slices = 40, acquisition time = 5 min. Tumor blood flow (TBF), evaluated by pCASL MRI, was obtained from a region of interest manually drawn on the quantitative CBF map (Fig. 1e) at the maximal, axial, cross-sectional area of the tumor by referring to the findings on contrast-enhanced T₁-weighted images. TBF was calculated using the following equation³:

$$\text{CBF} = \frac{6000 \cdot \lambda \cdot (\text{SI}_{\text{control}} - \text{SI}_{\text{label}}) \cdot e^{-\frac{\text{PLD}}{T_{1,\text{blood}}}}}{2 \cdot \alpha \cdot T_{1,\text{blood}} \cdot \text{SI}_{\text{PD}} \cdot (1 - e^{-\frac{\tau}{T_{1,\text{blood}}}})} [\text{mL}/100 \text{ g}/\text{min}]$$

where λ is the brain/blood partition coefficient in mL/g, $\text{SI}_{\text{control}}$ and SI_{label} are the time-averaged signal intensities in the control and label images, respectively. $T_{1,\text{blood}}$ is the longitudinal relaxation time of blood in seconds, α is the labeling efficiency, SI_{PD} is the signal intensity of a proton density-weighted image, and τ is the label duration. λ is 0.9 mL/g. $T_{1,\text{blood}}$ at 3T is 1650 ms. α for pCASL is 0.85. The mean rate of TBF was

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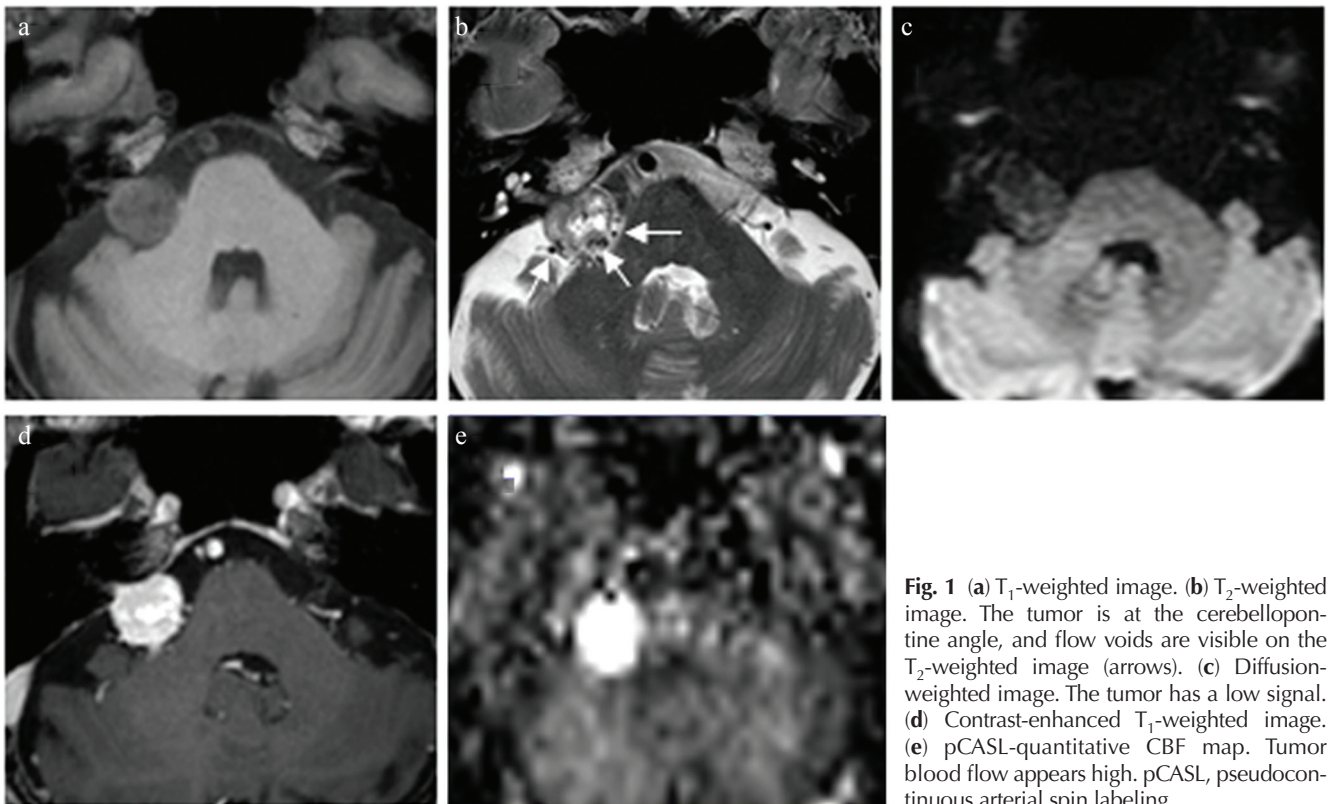


Fig. 1 (a) T₁-weighted image. (b) T₂-weighted image. The tumor is at the cerebellopontine angle, and flow voids are visible on the T₂-weighted image (arrows). (c) Diffusion-weighted image. The tumor has a low signal. (d) Contrast-enhanced T₁-weighted image. (e) pCASL-quantitative CBF map. Tumor blood flow appears high. pCASL, pseudocontinuous arterial spin labeling.

360 mL/100 g/min. Based on the typical MRI findings, the presurgical diagnosis was hemangioblastoma, despite the atypical location; however, the differential diagnoses included schwannoma and meningioma of the CPA. During the surgery, the tumor was found to be extra-axially located at the right CPA and was attached to the dura and right vestibular nerve. The tumor was partially resected, and the pathological findings revealed hemangioblastoma. The final diagnosis was sporadic hemangioblastoma of the CPA.

In this patient, conventional MRI findings were consistent with those of hemangioblastomas; they included flow voids in and around the tumor, low signal on DWI, and strong enhancement after injection of contrast media. ASL has been shown to be useful for differentiating hemangioblastomas from other brain tumors, including metastatic brain tumors, gliomas, meningiomas, and schwannomas, by indicating that hemangioblastoma has the highest rate of TBF.^{4,5} The high rate of TBF that we found with pCASL provided additional useful information for the differential diagnosis of the CPA tumors in this patient.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

References

1. Laviv Y, Thomas A, Kasper EM. Hypervascular lesions of the cerebellopontine angle: the relevance of angiography as a diagnostic and therapeutic tool and the role of stereotactic radiosurgery in management. A comprehensive review. *World Neurosurg* 2017; 100:100–117.
2. Yamamoto T, Takeuchi H, Kinoshita K, Kosaka N, Kimura H. Assessment of tumor blood flow and its correlation with histopathologic features in skull base meningiomas and schwannomas by using pseudo-continuous arterial spin labeling images. *Eur J Radiol* 2014; 83:817–823.
3. Alsop DC, Detre JA, Golay X, et al. Recommended implementation of arterial spin-labeled perfusion MRI for clinical applications: a consensus of the ISMRM perfusion study group and the European consortium for ASL in dementia. *Magn Reson Med* 2015; 73:102–116.
4. Noguchi T, Yoshiura T, Hiwatashi A, et al. Perfusion imaging of brain tumors using arterial spin-labeling: correlation with histopathologic vascular density. *AJNR Am J Neuroradiol* 2008;29:688–693.
5. Yamashita K, Yoshiura T, Hiwatashi A, et al. Arterial spin labeling of hemangioblastoma: differentiation from metastatic brain tumors based on quantitative blood flow measurement. *Neuroradiology* 2012; 54:809–813.