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Diffusion-Weighted Magnetic Resonance Imaging Findings in a Patient with Trigeminal Ganglioneuroma

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A case of intracranial ganglioneuroma arising from the trigeminal nerve in the pontine and cerebellopontine angle cistern, in a 44-year-old female, is presented with an emphasis on diffusion-weighted imaging findings. We will discuss on how the tumor in the very unusual location should be differentiated particularly focused on diffusion-weighted imaging findings.

Index terms: *Trigeminal nerve; Ganglioneuroma; Diffusion-weighted imaging*

INTRODUCTION

Ganglioneuromas are rare benign tumors of neural crest origin; they arise from peripheral sympathetic ganglia or the adrenal glands. These tumors are most commonly discovered in older pediatric patients or adults; they are discovered incidentally, through thoracic or abdominal imaging studies performed for unrelated conditions. Intracranial involvement of ganglioneuromas is extremely rare (1, 2), and there has been only one report of ganglioneuroma arising from the trigeminal nerve. We report a very rare case of trigeminal ganglioneuroma in the prepontine and cerebellopontine angle cistern, in a 44-year-old female and describe the

magnetic resonance (MR) imaging characteristics, including diffusion-weighted imaging (DWI) findings.

CASE REPORT

A 44-year-old female was admitted to our hospital because of a 2-year history of facial pain and numbness in the left side. The patient complained of an intermittent stabbing pain and numbness over the left zygomatic region. The rest of the neurologic examination was normal. A brain MR imaging study was performed with a 3T unit (Magnetom TrioTim, SIEMENS, Erlangen, Germany), which revealed a 2 x 1.8 x 1.5 cm well-circumscribed mass in the prepontine and cerebellopontine angle cistern on the left side (Fig. 1). The left fifth cranial nerve was displaced superomedially, and the seventh-eighth nerve complex inferiorly by the mass. The mass showed homogeneous iso-signal intensity on a T1-weighted image relative to the brain parenchyma (Fig. 1A) as well as heterogeneous increased signal intensity on a T2-weighted image (Fig. 1B). A gradient echo image showed no evidence of intratumoral hemorrhage or calcification. The mass demonstrated minimal contrast enhancement after intravenous injection of gadolinium (Fig. 1C). With DWI, the lesion was hyperintense compared to adjacent brain

parenchyma (Fig. 1D). The apparent diffusion coefficient (ADC) values were obtained using a circular region of interest with a diameter of 6 mm at 4 nonoverlapping

locations in the tumor (Fig. 1E). The mean ADC values obtained with b values of 0 and 1000 sec/mm² of the tumor were 0.72 x 10⁻³ mm²/s, compared to 3.01 x 10⁻³ mm²/s for
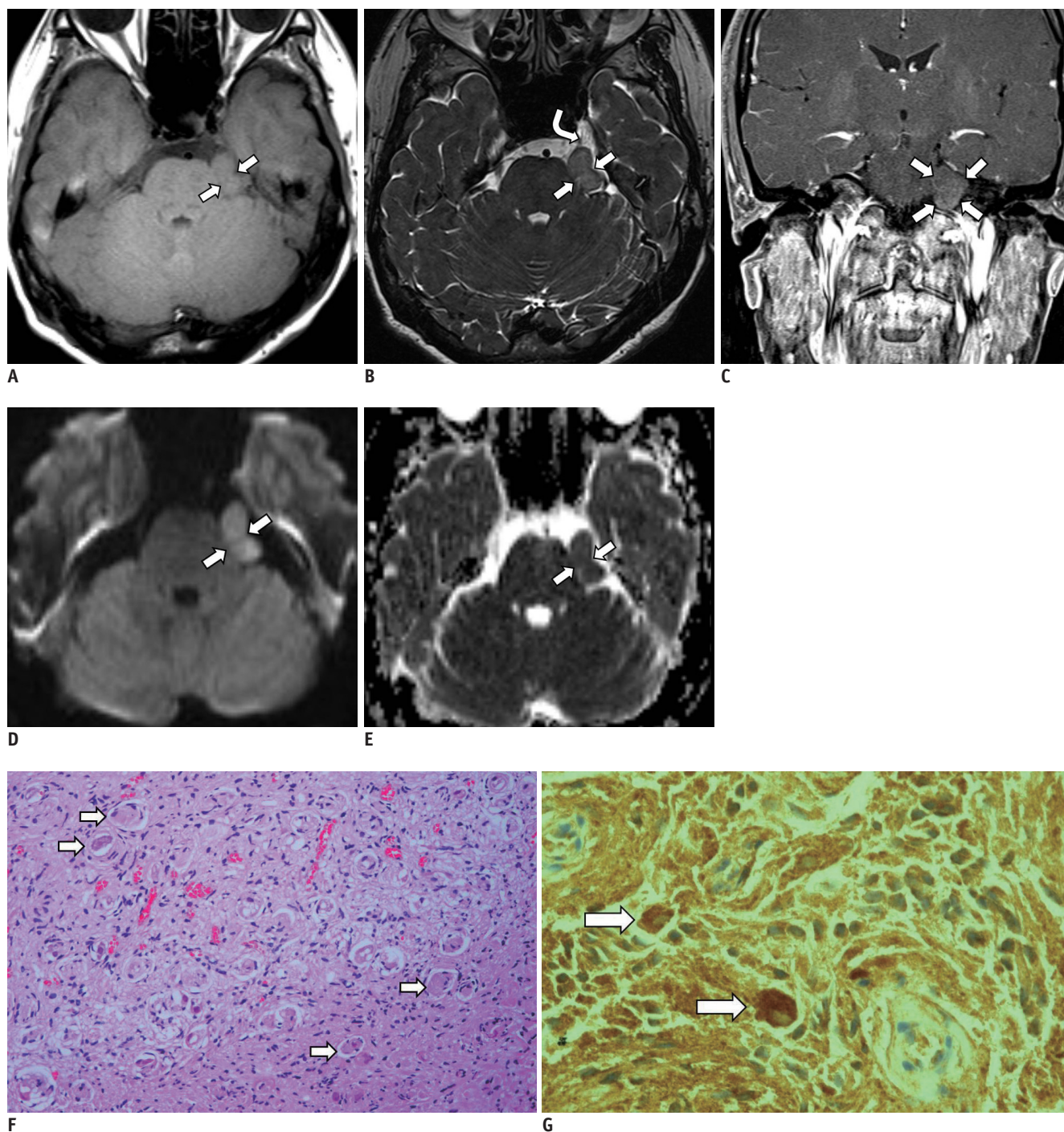

Fig. 1. Trigeminal ganglioneuroma in 44-year-old female.
A. Axial T1-weighted image shows well-defined mass (arrows) in left prepontine and cerebellopontine angle cistern. This has same signal intensity as adjacent brain parenchyma. **B.** Thin section heavily T2-weighted image shows mass (arrows) with abutting and displacement of left trigeminal nerve (curved arrow). **C.** Coronal postcontrast T1-weighted image shows minimal contrast enhancement of tumor (arrows). **D.** Axial echo-planar diffusion-weighed imaging with b = 1000 s/mm² shows tumor with hyperintense lesion relative to adjacent brain parenchyma. **E.** Apparent diffusion coefficient map shows mild hypointensity of tumor (arrows). **F.** Photomicrograph shows that tumor consists of scattered mature ganglion cells (arrows) and spindle-shaped cells with scanty myxoid changes, H&E stain, x 200. **G.** Photomicrograph shows relatively strong S-100 expression of ganglion cells (arrows), x 400.

CSF (measured in the fourth ventricle) and 0.71×10^{-3} mm^2/s for adjacent brain tissue (measured in the pons). The preoperative diagnosis for this poorly enhancing mass with a DWI high signal intensity lesion in the prepontine and cerebellopontine angle cistern was an epidermoid cyst.

The patient underwent a left suboccipital craniotomy and tumor removal surgery. During surgery, a well-circumscribed solid mass was found to be firmly attached to the trigeminal nerve. The trigeminal nerve was split by the tumor and appeared to be thin. The tumor was resected with attached nerve fibers. Histopathologic examination demonstrated a benign tumor composed of mature ganglion cells and spindle-shaped cells in an abundant collagenous stroma (Fig. 1F). Tumor cells were positive for S-100 protein by immunohistochemical staining (Fig. 1G), and the pathologic diagnosis was ganglioneuroma. Postoperatively, facial pain and numbness disappeared, and follow-up MR imaging, obtained 16 months later showed no evidence of tumor recurrence.

DISCUSSION

Ganglioneuromas are typically found within the posterior mediastinum and retroperitoneum, though less common locations include the adrenal medulla, parapharyngeal region, and visceral ganglia. Rarely, ganglioneuromas have been observed in the tongue, prostate, skin, and bone (3). Intracranial ganglioneuroma is extremely rare, and only a few cases have been reported in previous literature (1, 2). Histologically, these tumors are composed of single or clustered mature ganglion cells and mature Schwann cells (3).

Typical radiologic findings of ganglioneuroma arising from the peripheral sympathetic nerve have been described. At CT, ganglioneuromas are well-demarcated hypoattenuated lesions, with calcifications in 20-42% of cases. These tumors are not significantly enhanced after the administration of contrast materials, but delayed CT images can show slight or moderate enhancement (3, 4). At MR, ganglioneuromas are generally hypointense on T1-weighted images and heterogeneously hyperintense on T2-weighted images with respect to adjacent muscle.

Abe et al. (1) reported the first case of trigeminal ganglioneuroma and described conventional MR imaging findings, which are similar to those of thoracoabdominal ganglioneuromas. MR images included in their report demonstrated a well-defined mass with low signal intensity

on a T1-weighted image, high signal intensity on a T2-weighted image, and relatively homogenous enhancement on a post-contrast T1-weighted image. In our patient, the lesion was iso-signal intense to the adjacent pons on the T1-weighted image and showed slightly increased signal intensity on the T2-weighted image. It has previously been reported that ganglioneuromas with markedly high signal intensity on a T2-weighted image histologically consist of a large amount of myxoid stroma and relatively few cellular components and collagen fibers. In contrast, tumors with intermediate to subtle high signal intensities on a T2-weighted image, as in our case, consisted of numerous cellular and fibrous components and few myxoid stroma (4, 5). In our case, the mass showed poor enhancement on a postcontrast T1-weighted image. MR imaging enhancement of ganglioneuromas varies from mild to marked (3). Early enhancement with dynamic MR imaging is not typically seen in ganglioneuromas. Thoracoabdominal ganglioneuromas can demonstrate gradually increasing enhancement with dynamic MR imaging (3-5).

Diffusion-weighed imaging findings and ADC values with regard to intracranial ganglioneuroma have not been published previously. Our case showed a homogeneous hyperintense signal throughout the tumor, on a DWI, and revealed a mean tumor ADC value of 0.72×10^{-3} mm^2/s. Gahr et al. (6) recently reported DWI findings of the ganglioneuromas/ganglioneuroblastomas originating from the thoracoabdominal sympathetic nervous system. These tumors showed relatively high ADC values (mean ADC: 1.60×10^{-3} mm^2/s, range 1.13-1.99×10^{-3} mm^2/s). The differences between Gahr's cases and our case may be due to the different tumor histopathologies. One might postulate that high cellular density and scanty myxoid stroma, revealed through histopathology, may be attributed to relatively low ADC values in our case. Unfortunately, histopathologic analysis was not available in the Gahr's study.

Intratumoral hemorrhages are not typical features of ganglioneuroma (3). Qing et al. (7) reported that focal central hemorrhages were observed in 2 of 17 adrenal ganglioneuromas. Our case showed no intratumoral susceptibility signals on gradient echo images, as well as absence of intratumoral hemorrhage or calcification with histopathology.

An epidermoid cyst should be included in the differential diagnosis of T2-increased signal intensity lesions in the prepontine and cerebellopontine angle cisterns. Intracranial epidermoid cysts are typically hyperintense compared with

brain parenchyma with DWI. The high signal intensity of epidermoid cysts with DWI is caused by the intrinsic T2 shine-through effects of the lesion. Reported ADC values of intracranial epidermoid cysts range from 0.98 x 10^{-3} mm^2/s up to 1.36 x 10^{-3} mm^2/s (8); these are higher than the values of trigeminal ganglioneuroma in our case. Other differential diagnoses of masses in the prepontine and cerebellopontine angle cisterns include schwannoma, meningioma, neurofibroma, and metastasis. The majority of these tumors do not show high signal intensity with DWI and demonstrate relatively strong enhancement on post-contrast T1-weighted images. Other rare tumors that have been reported to occur in the trigeminal nerve include benign triton tumor (neuromuscular hamartoma) and ganglioglioma. Neuromuscular hamartomas were reported to be profoundly hypointense on T2-weighted images and demonstrated mild homogenous contrast enhancement (9). A trigeminal ganglioglioma may be difficult to distinguish from a ganglioneuroma like that in our case. This tumor showed a nonspecific homogenous MR signal intensity indistinguishable from the adjacent brain stem with all imaging sequences and no contrast enhancement (10). DWI findings of neuromuscular hamartoma and ganglioglioma, arising from the trigeminal nerve have not been reported.

Conclusion

Trigeminal ganglioneuroma is a very rare tumor; however, this tumor should be considered in the differential diagnosis of tumors with low ADC values in the prepontine and cerebellopontine angle cistern.

REFERENCES

1. Abe T, Asano T, Manabe T, Matsuura H, Furuta T, Taguchi K. Trigeminal ganglioneuroma. *Brain Tumor Pathol* 1999;16:49-53

2. Arseni C, Horvath L, Carp N, Ciurea V. Intracranial ganglioneuromas in children. *Acta Neurochir (Wien)* 1975;32:279-286

3. Lonergan GJ, Schwab CM, Suarez ES, Carlson CL. Neuroblastoma, ganglioneuroblastoma, and ganglioneuroma: radiologic-pathologic correlation. *Radiographics* 2002;22:911-934

4. Cai J, Zeng Y, Zheng H, Qin Y, T K, Zhao J. Retroperitoneal ganglioneuroma in children: CT and MRI features with histologic correlation. *Eur J Radiol* 2010;75:315-320

5. Zhang Y, Nishimura H, Kato S, Fujimoto K, Ohkuma K, Kojima K, et al. MRI of ganglioneuroma: histologic correlation study. *J Comput Assist Tomogr* 2001;25:617-623

6. Gahr N, Darge K, Hahn G, Kreher BW, von Buiren M, Uhl M. Diffusion-weighted MRI for differentiation of neuroblastoma and ganglioneuroblastoma/ganglioneuroma. *Eur J Radiol* 2011;79:443-446

7. Qing Y, Bin X, Jian W, Li G, Linhui W, Bing L, et al. Adrenal ganglioneuromas: a 10-year experience in a Chinese population. *Surgery* 2010;147:854-860

8. Mermuys K, Wilms G, Demaerel P. Epidermoid cyst of the fourth ventricle: diffusion-weighted and flair MR imaging findings. *JBR-BTR* 2008;91:58-60

9. Castro DE, Raghuram K, Phillips CD. Benign triton tumor of the trigeminal nerve. *AJNR Am J Neuroradiol* 2005;26:967-969

10. Athale S, Hallet KK, Jinkins JR. Ganglioglioma of the trigeminal nerve: MRI. *Neuroradiology* 1999;41:576-578