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 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/mm$^2$ of the tumor were $0.72 \times 10^{-3}$ mm$^2$/s, compared to $3.01 \times 10^{-3}$ mm$^2$/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/mm$^2$ 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 $0.71 \times 10^{-3}$ 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 preoptic 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 $0.72 \times 10^{-3}$ 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: $1.60 \times 10^{-3}$ mm$^2$/s, range 1.13-1.99 $\times 10^{-3}$ 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
DW MR Imaging in Trigeminal Ganglioneuroma

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 $0.98 \times 10^{-3}$ mm$^2$/s up to $1.36 \times 10^{-3}$ mm$^2$/s (8); these are higher than the values of trigeminal ganglioneuroma in our case. Other differential diagnoses of masses in the preoptic 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 preoptic and cerebellopontine angle cistern.

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