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Glomus Jugulare Tumor Presenting with Intracerebellar Hemorrhage

ABSTRACT—What is believed to be the first case of a glomus jugulare tumor presenting with intracerebellar hemorrhage is described. A 25-year-old normotensive man suddenly suffered from severe headache, nausea, vomiting, vertigo, and ataxia due to an intracerebellar hemorrhage. Magnetic resonance imaging and angiography revealed a highly vascular jugulare foramen tumor extending into the intracranial space adjacent to the hematoma. Total removal of the tumor was performed successfully via the combined pre- and retrosigmoid approach, and the histologic diagnosis was a glomus jugulare tumor. We concluded that one of the numerous draining veins on the surface of intracranial tumor, which were observed during the operation, was the origin of the intracerebellar hemorrhage.

CASE REPORT

The clinical features of glomus jugulare tumors (GJTs) can vary widely due to their biologic nature and complicated developmental patterns. The origin of this tumor is a glomus body, which is a part of the chemoreceptor system.^{1,2} Although the incidence is low, labile hypertension due to catecholamine secretion is one of the initial symptoms of GJTs.³⁻⁷ GJTs commonly occur in the middle ear and the temporal bone and may injure the auditory apparatus and cranial nerves in the temporal bone and jugulare foramen.⁸⁻¹³ Therefore, common presenting symptoms of GJTs are tinnitus, hearing loss, otalgia, vertigo, facial nerve palsy, dysphagia, and other related symptoms. GJTs may also extend into the intracranial space in the posterior fossa, resulting in cerebellar ataxia and intracranial hypertension.^{8,12-16} Al-

though GJTs are extremely hypervascular, spontaneous hemorrhage in these tumors is rare.^{9,17} Moreover, no direct evidence of intracranial hemorrhage in GJTs has been reported. Here we describe the first case of GJT presenting with intracerebellar hemorrhage.

A 25-year-old man was admitted to a local hospital complaining of sudden and severe headache, vomiting, and vertigo on September 11, 1998. Computed tomography revealed an intracerebellar hematoma with extension into the fourth, third, and lateral ventricles and a cerebellopontine angle (CPA) tumor (Fig. 1). At the admission examination, the patient was revealed to be normotensive and alert but had cerebellar ataxia and Vth and VIIth-Xth cranial nerve palsy. Although he never had any medical consultation, he had been suffering from right hearing disturbance and dysphagia for 2 and 3 years, respectively. He was treated with an intravenous injection of mannitol and glyceol to reduce in-

Figure 1. Plain computed tomography demonstrates an intracerebellar hemorrhage extending into the IVth ventricle (asterisk) and high-density mass (arrow) in the cerebellopontine angle adjacent to the hematoma.

tracranial pressure and then referred to our hospital for further examination and treatment on September 17, 1998.

Magnetic resonance imaging (MRI) clearly demonstrated a jugulare foramen tumor extending into the CPA adjacent to the intracerebellar hematoma (Fig. 2). Angiography demonstrated that the tumor was highly vascularized from the external carotid artery system and had numerous enlarged draining veins (Fig. 3). The right transverse-sigmoid sinus could not be visualized by four vessel angiography, indicating that the right jugular bulb was occluded by the tumor. The tumor was diagnosed as a GJT, based on the MRI and angiographic findings, and surgical removal assisted by preoperative feeder embolization was scheduled.

Preoperative Embolization

To reduce the amount of bleeding during the tumor resection, selective feeder embolization was performed a day before the operation. The right posterior auricular artery and ascending pharyngeal artery, which supply the tumor, were embolized with polyvinyl alcohol particles, polyvinyl acetate, and braided occlusion device coils (Target Therapeutics). The tumor stain was significantly reduced after the procedure (Fig. 4).

Figure 2. Sagittal (A) and coronal (B) sections of MRI after Gd-DTPA administration revealed that the tumor (arrow) adjacent to the intracerebellar hematoma (arrowhead) presents a dumbbell-shaped developmental pattern from the jugulare foramen.

Figure 3. Right external carotid angiogram shows a significant tumor stain supplied by the retroauricular and ascending pharyngeal arteries (A) and numerous enlarged draining veins (B).

Operation

The operation was performed on October 6, 1998. The tumor was totally removed by the combined pre- and postsigmoid approach. The tumor was found to relate to the Xth cranial nerve and to be extending into the subdural space via the jugulare foramen. The tumor was well devascularized by the preoperative embolization, and the amount of intraoperative bleeding was small. Numerous enlarged veins were observed on the surface of the tumor, which extended into the subdural space.

The histologic diagnosis was a GJT. The patient's postoperative course was uneventful. Although right IXth, Xth, and XIth cranial nerves were intentionally sacrificed during the tumor removal, the patient's neu-

rologic condition was not aggravated after surgery. He moved to a rehabilitation hospital on November 11, 1998.

DISCUSSION

Common causes of intracerebellar hemorrhage are hypertension, intracerebellar tumors, and vascular anomalies such as arteriovenous malformation, cavernous angioma, and so on. Extramedullary tumors are unlikely to induce intracerebellar hemorrhage. GJTs are characterized by their extremely high vascularity, and the control of bleeding is the major problem during surgical removal.^{8,10,12,13,15,18,19} However, spontaneous

Figure 4. Right external carotid angiogram before (A) and after (B) embolization. The tumor stain was significantly reduced after feeder embolization.

bleeding in GJTs is rare. A few authors described bleeding from the ear when the tumor extends into the external auditory canal.^{9,17} Moddy et al.²⁰ reported a case of extensive calcification in a GJT and described that "the patient probably was afflicted with this process for many years and presented with a subarachnoid hemorrhage." Although the evidence of subarachnoid hemorrhage was not confirmed in their case, we believe that this is the only report in which the intracranial hemorrhage due to GJT is described. The intracerebellar hemorrhage in the present case is undoubtedly due to a GJT. Hypertension is the major cause of intracerebellar hemorrhage. Moreover, GJTs may induce extensive hypertension by secreting catecholamines.³⁻⁷ However, no case with hypertensive intracerebral hemorrhage due to catecholamine-secreting GJTs has been reported. Moreover, our patient was normotensive, and there was no evidence of catecholamine production. Based on the angiographic and intraoperative findings, we concluded that one of the enlarged draining veins on the surface of the intradural tumor was the origin of the intracerebellar hemorrhage. Bleeding from GJTs is likely to occur because of their high vascularity. The actual incidence of intracranial hemorrhage is, however, extremely low. The developmental patterns of GJTs vary widely, and Fisch¹⁵ classified these tumors into four categories: classes A–D. Fisch class D tumors demonstrate intracranial invasion. Although precise data are not available, some class D tumors may be covered by the dura mater and may not extend into the subdural space. This might be the reason for the low incidence of intracranial hemorrhage from GJTs in spite of their high vascularity.

Surgical removal of GJTs is complicated, especially when they extend into the intracranial space, by virtue of their location and vascular nature.^{12,21–24} Surgical morbidity is usually related to deficits of the lower cranial nerves, which might be injured at the jugulare foramen.^{8,21} GJTs are radiosensitive, and many authors have demonstrated the efficacy of radiation therapy, indicating that the first choice of treatment for GJTs without lower cranial nerve palsy might be radiation.^{9,11,13} However, the ideal treatment of these biologically benign tumors should be surgical removal. Although their extremely high vascularity is the major problem during the surgical removal, the preoperative embolization of feeding arteries can significantly reduce the amount of bleeding, making the removal of GJTs easy and safe.^{19,25,26} However, the problem of postoperative lower cranial nerve palsy still remains. Fortunately, our case had a long history of dysphagia, indicating that the function of the affected lower cranial nerves was compensated at the time of operation. Thus, we decided to remove this tumor surgically after preoperative embolization. Tumors located in the temporal bone and the extracranial space can be removed by the infratemporal fossa approach¹⁸ or the combined approach through the neck and temporal bone.¹⁰ The strategy for removal of

GJTs with intracranial extension has been described by several authors.^{12,15,23,24} Because of the small size of the extracranial tumor and the massive intracranial extension, we used the combined pre- and retrosigmoid approach without the transcervical (or infratemporal fossa) approach. After the lateral suboccipital craniectomy, the posterior part of the mastoid process and pyramis was resected to open the posterior wall of the jugulare foramen. The posterior fossa dura was opened, and the right sigmoid sinus was ligated and cut at the proximal portion. Thus, the entire tumor was exposed and removed safely and completely.

GJTs are rare, and their clinical features and treatment strategy are complicated due to their location and biologic nature. Here we described an unusual but possible presentation of GJTs and a strategy for removal of GJTs extending into the subdural space with the assistance of preoperative embolization.

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