Staged Resection of Large Hypervascular Vestibular Schwannomas in Young Adults

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ABSTRACT

Two young adults underwent resection of large hypervascular vestibular schwannomas (acoustic neuromas) via two-stage surgery. The first patient, a 27-year-old woman with hydrocephalus, had a large hypervascular vestibular tumor in the left cerebellopontine angle (CPA) supplied by the left anterior inferior cerebellar artery (AICA) and posterior inferior cerebellar artery (PICA). The second patient, a 34-year-old woman, had a large AICA-supplied hypervascular vestibular tumor in the left CPA that displaced the brain stem significantly. At the initial stage, only the lateral aspect of the tumor was debulked due to excessive bleeding from the tumor bed. Angiography 1 or 2 months after the initial operation showed that the tumor was hypovascular. At the second stage, the remnant medial aspect of the tumor was relatively avascular and nonadherent to the brain stem. Without blood transfusion during the second stage, the tumor was removed totally in the first patient and subtotally in the second patient. Pathological examination revealed that dilated blood vessels were prominently increased at the first surgery; however, at the second surgery, the number of blood vessels had decreased, showing necrosis and degeneration. Although there are no absolute indications for the staged resection of vestibular schwannomas, this procedure may represent one of the safest options for these difficult lesions in young adults.

KEYWORDS: Acoustic neuroma, hypervascularity, large tumor, staged resection, vestibular schwannoma, young adult

The resection of vestibular schwannomas (acoustic neuromas) remains a great challenge to neurosurgeons and otologists. Complete resection is often curative but may be infeasible if the tumor extends into the internal auditory canal or adheres to the surrounding brain and cranial nerves. Although uncommon, tumor vascularity can interfere with the removal of vestibular schwannomas by complicating adequate hemostasis.¹ Large hypervascular tumors are associated with the highest rates of surgical morbidity and mortality because of their abundant vascularity and arteriovenous (AV) shunting, and the indistinct plane between the pia and tumor.²⁻⁵ Their complete excision with preservation of cranial nerve function and without blood transfusion is not easily accomplished during a single operation. Although numerous reports have described different approaches, few have discussed staged resection of these lesions.²⁻⁵ We report on two young adults with large, highly vascular vestibular schwannomas that were resected via two-stage surgery.

CASE REPORTS

Case 1

A 27-year-old woman experienced a hearing disturbance in the left ear for more than 1 year. The patient also had progressive headache, intermittent difficulty with balance, and impaired visual acuity 3 months before admission. A computed tomographic (CT) scan revealed a left-sided large posterior fossa mass associated with hydrocephalus. A ventriculoperitoneal shunt was placed emergently for the massive hydrocephalus. Magnetic resonance imaging (MRI) demonstrated an enhancing large vestibular tumor (maximum size, about 6 cm) in the left cerebellopontine angle (CPA) that had significantly displaced the brain stem (Fig. 1A). Cerebral angiography showed a hypervascular tumor supplied by the left anterior inferior cerebellar

artery (AICA) and the posterior inferior cerebellar artery (PICA) (Fig. 1B).

A microcatheter system (Tracker 18 catheter) was passed through the guiding catheter, and its tip was placed as close as possible to the tumor-feeding artery. After the meatal loop of the AICA was embolized with polyvinyl alcohol (PVA) foam, the tumor stain was reduced without complications.

Two days after the embolization, the patient underwent a suboccipital craniectomy. The tumor was deeply embedded in the brain stem and the pia mater-tumor interface was indistinct. Resection was extremely difficult due to excessive bleeding from the tumor bed. Only the lateral aspect of the tumor was debulked before the incision was closed. About 2000 ml of blood was transfused. Postoperatively, the patient had a complete facial nerve palsy and hearing loss on the left side.

One month after the initial operation, MRI revealed a medial remnant of the tumor (Fig. 1C). Two months after the initial operation, cerebral angiography showed that the tumor was hypovascular (Fig. 1D). After a staging interval of 2 months, the residual tumor was totally resected via the suboccipital approach. At surgery, the tumor was soft and relatively avascular. The residual medial portion of the tumor had become decompressed and had shifted out of the deep recesses of the brain stem, allowing its resection. The tumor had originated from the vestibular nerve. A blood transfusion was unnecessary in this second-stage operation. Six months after the second operation, the patient underwent a facial-to-hypoglossal anastomosis and had a House-Brackmann Grade 3 facial function⁶ at her most recent follow-up visit. Five years after the operation, follow-up MRI revealed no residual tumor (Fig. 1E).

Case 2

A 34-year-old woman had a 1-year history of headache as well as decreased hearing on the left side. Neurological examination revealed left-sided

Figure 1 Case 1. (A) Postgadolinium T1-weighted coronal MRI showing a large left-sided vestibular tumor with poor central enhancement. (B) Left lateral projection vertebral angiogram showing hypervascularity within the tumor. Most of the blood supply is from the anterior inferior cerebellar artery in the left cerebellopontine angle. (C) Postgadolinium T1-weighted coronal MRI showing a medial remnant of the tumor. (D) Left lateral projection vertebral angiogram showing that the hypervascularity within the tumor has disappeared.

Figure 1 (Continued) (E) Postgadolinium T1-weighted coronal MRI obtained at the 5-year follow-up visit showing no evidence of residual tumor or mass effect. (F) Pathological findings showing diffuse proliferation of spindle-shaped tumor cells in a palisading pattern and hypervascularity within the tumor at the first operation. (G) Necrotic lesions and degeneration within the tumor and reduction of hypervascularity at the second surgery (hematoxylin and eosin x 20).

cerebellar signs and hearing disturbance. MRI demonstrated a large homogeneously enhancing vestibular tumor on the left side (maximum size, about 5 cm) (Fig. 2A). Cerebral angiography showed hypervascularity and AV shunting of the tumor, which was supplied from the left AICA (Fig. 2B).

The patient underwent staged tumor resection via the suboccipital route. At the first operation, excessive bleeding from the tumor bed was stopped with aneurysm clips, and the lateral aspect of the tumor was debulked. The aneurysm clips were left in place. The patient recovered without new deficits. One month after the first operation, MRI demonstrated a defect in the lateral tumor,

with partial decompression of the brain stem and fourth ventricle (Fig. 2C). Cerebral angiography showed the tumor to be hypovascular (Fig. 2D).

After a staging interval of 5 weeks, the residual tumor was found to be relatively avascular and nonadherent to the brain stem. The residual medial portion of the tumor was resected subtotally. The tumor had originated from the vestibular nerve. A blood transfusion was unnecessary during this procedure. The patient had no neurological deficits after surgery except for the left-sided hearing disturbance. Follow-up MRI 1 year after the operation revealed a small residual tumor and no compression of the brain stem or fourth ventricle (Fig. 2E).

Figure 2 Case 2. (A) Postgadolinium T1-weighted axial MRI showing a large vestibular tumor in the left cerebellopontine angle obliterating the fourth ventricle. (B) Left lateral projection vertebral angiogram showing hypervascularity within the tumor. Most of the blood supply is from the anterior inferior cerebellar artery and multiple large-caliber early draining veins. (C) Postgadolinium T1-weighted axial MRI showing a medial tumor remnant and artifact from aneurysm clips in the lateral aspect of the tumor. (D) Left lateral projection vertebral angiogram showing that the hypervascularity within the tumor has been reduced.

Pathological Findings

In Case 1, the tumor was characterized by diffuse proliferation of elongated spindle tumor cells with a palisading pattern. This was diagnosed as a

schwannoma, predominantly Antoni type A (Fig. 1F). In Case 2, the tumor cells with microcystic matrix were arranged haphazardly. It was diagnosed as a schwannoma, predominantly Antoni type B (Fig. 2F). In Cases 1 and 2, dilated blood

Figure 2 (Continued) (E) Postgadolinium T1-weighted axial MRI obtained at the 1-year follow-up visit showing a small tumor residual without compression of the brain stem or fourth ventricle. Pathological findings revealing (F) Antoni B tissue and hypervascularity within the tumor at the first operation, and (G) necrotic lesions and degeneration within the tumor at the second surgery (hematoxylin and eosin x 20).

vessels were increased prominently at the first surgery. At the second surgery, however, the number of blood vessels in both tumors was decreased, showing necrosis and degeneration (Figs. 1G and 2G).

DISCUSSION

The concept of staged resection for vestibular schwannomas has been described.^{2,3,5} In 1925, Dandy reported the staged removal of several ves-

tibular schwannomas in which the residual tumor was typically soft, necrotic, and avascular.³ To date, however, there is no absolute indication for the staged resection of vestibular tumors. Comey et al.² stressed that staged resection should at least be considered for all large tumors, especially those shown to be deeply embedded within the substance of the brain stem and cerebellum on preoperative imaging. Because large tumors distort the brain stem and the cerebellum extensively, the plane between the tumor and pia mater is often indistinct.^{2,3,5} In an attempt to dissect these tumors from the brain stem, the cranial nerves, arteries, and pons may be injured.

The characteristic signal abnormality that predicts a difficult pial dissection, such as a signal abnormality on T2-weighted MRI in the adjacent brain stem in the presence of a large petroclival meningioma, is not seen in the substance of the brain stem on preoperative MRI of vestibular schwannomas.² In our patients, a signal abnormality in the adjacent brain stem on T2-weighted MRIs was not observed, although the interface between the pia mater and tumor was indistinct.

The degree of vascularity may not significantly impair the experienced surgeon's ability to remove hypervascular vestibular schwannomas.¹ Comey et al. noted that another indication for staging included excessive bleeding from the tumor bed.² LeMay et al. reported that a hypervascular tumor may behave similarly to an AV malformation; therefore, similar treatment strategies should be employed.⁴ They stressed that if preoperative embolization cannot be performed and a large vascular component of the tumor remains, radiotherapy or staged operative resections must be considered.^{2,7}

In Case 1, the need for staging was based on the excessive bleeding from the tumor bed during the initial operation. In this patient, preoperative embolization of the feeding artery was ineffective, and it was difficult to control the bleeding. In Case 2, an "intentional" staged operation was planned because of the experience gained from the surgical results of Case 1. Therefore, after the lateral aspect

of the tumor was resected at the initial operation, we stopped the excessive bleeding from the tumor bed with aneurysm clips and left the clips in the operative field. In these two patients, severe engorgement and pulsatility of the surrounding brain and residual tumor were not observed after the debulking.

In their 83 vestibular tumors resected in stages, Comey et al. consistently found that the residual tumor encountered at the subsequent staged resection was relatively avascular.² In our patients, cerebral angiography 1 or 2 months after the initial operation showed that the remnant tumor was hypovascular. The timing of the second operation might depend on the confirmation of reduced hypervascularity of the tumor by cerebral angiography. In the second stage of surgery, we did not encounter excessive bleeding from the tumor bed, nor was a blood transfusion needed. This phenomenon may be related to the tumor's blood supply. In our present cases, pathological analysis revealed that some or all of the blood supply from AICA to the tumor was removed by debulking the lateral aspect of the tumor at the initial surgery, and spontaneous thrombosis occurred within the vasculature of the tumor.

In addition to decreased vascularity, the medial remnant of the tumor usually became significantly decompressed and shifted out of the brain stem, making it more surgically accessible.^{2,3,5} In Case 1, the medial remnant of the tumor was resected totally without excessive bleeding from the tumor at the subsequent stage, despite the patient's complete facial nerve palsy and hearing loss at the initial stage. In Case 2, the tumor was resected subtotally without neurological deficits except for the hearing disturbance from the staged operation.

A vestibular tumor can grow very rapidly with minimal symptomatology in young people.^{8,9} Long-term outcomes of vestibular schwannomas after radiosurgery have been reported, and its efficacy has been confirmed by a Pittsburgh group.¹⁰ Prasad et al. has stated that gamma surgery should be used to treat postoperative residual tumors as

well as tumors in patients with medical conditions that preclude surgery.¹¹ If the remnant tumor regrows, stereotactic radiosurgery may be considered.

Comey et al. reported that a lower incidence of operative morbidity and mortality was associated with the staged resection of their 55 large tumors compared to nonstaged resections in several similar series.² Thus, the staged resection of large hypervascular vestibular tumors in young adults offers an effective, safe method for dealing with these difficult lesions even though additional surgery is required. Considering that the tumor residue is relatively friable and avascular at the second stage of resection, this technique is reasonable. Although there are no absolute indications for the staged resection of vestibular schwannomas, it should be considered an option for difficult cases.

COMMENTS FOR PUBLICATION

Drs. Abe et al. have nicely described their technique for staged resection of large hypervascular vestibular schwannomas in two young adults. Their experience with the first case, in which resection was extremely difficult and associated with excessive bleeding from the tumor bed, led to a staged resection and satisfactory outcome and allowed them to be prepared when a second case presented itself similarly. Lessons learned from the first case allowed the second patient to be treated surgically without the need for a blood transfusion.

The authors appropriately cite the literature on the topic and corroborate the techniques and lessons learned at the University of Pittsburgh, where Dr. Jannetta's team reported 83 such patients in 1995.

Their conclusion that staged resection of vestibular schwannomas be considered for difficult cases is a prudent recommendation supported by both their experience and the experience of other skull base surgeons.

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