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HOW I DO IT - TUMOR - SCHWANNOMA

Transjugular transsigmoid approach for triple dumbbell-shaped jugular foramen schwannomas

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Abstract

Background Jugular foramen tumors, particularly those that are triple dumbbell-shaped with intracranial, intraforaminal, and extracranial extensions, are difficult to access surgically. However, advances in neuroimaging, neuromonitoring, and skull base surgery have enabled their safe resection with lower rates of morbidity and mortality.

Method We share our experience with the surgical technique for the management of triple dumbbell-shaped jugular foramen schwannomas.

Conclusion The infralabyrinthine transjugular transsigmoid approach with high cervical exposure under continuous vagus nerve monitoring enables gross total resection of triple dumbbell-shaped jugular foramen schwannomas, aiming at surgical cure of these benign tumors for appropriately selected patients.

Keywords High cervical exposure · Postauricular transtemporal approach · Posterior fossa · Neuromas · Skull base surgery · Transmastoid retrolabyrinthine approach

Abbreviations

ABR	Auditory brainstem response
CN	Cranial nerve
MEP	Motor evoked potential
MRI	Magnetic resonance image
SEP	Somatosensory evoked potential

Relevant surgical anatomy

Cranial nerve (CN) IX and CNs X and XI all enter the jugular foramen, respectively, separated by the dural septum between CNs IX and X [5]. After entering the foramen, they pass downward on the ventral wall of the jugular bulb and exit the foramen while descending along the internal jugular vein. CN IX descends anteriorly along the lateral side of the carotid artery. CN X descends between the carotid artery and the jugular vein, whereas CN XII joins after traversing the hypoglossal canal. CN XI passes posteriorly across the lateral surface of the jugular vein.

The jugular bulb occupies a large posterolateral compartment of the foramen, so-called the pars venosa or the sigmoid part. It receives venous drainage from the sigmoid sinus, the posterior condylar vein, and the anterior condylar confluence. The anterior condylar confluence comprises the inferior petrosal sinus and adjacent venous connections, and has a main venous channel communicating with the jugular bulb.

Description of the technique

Positioning and skin incision

After placing the lumbar drainage, the patient is placed in the supine-lateral position with slight head retroflexion (Fig. 1a). Using a large C-shaped postauricular incision extending as far as the ventral margin of the sternocleidomastoid muscle (Fig. 1b), the suboccipital muscles are dissected while elevating a pedicled muscular flap for dural closure. The greater auricular

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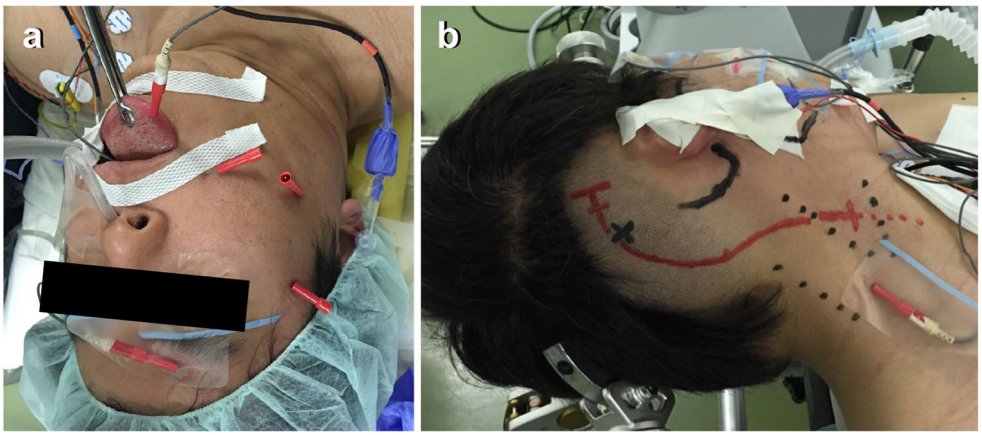

Fig. 1 Operative setting. **a** We routinely monitor ABR, SEP, MEP, and electromyograms of CNs V, VII, X, XI, and XII. **b** A large C-shaped postauricular incision is made on the left side as far as the ventral margin of the sternocleidomastoid muscle (black dotted line)

nerve is carefully preserved, just in case nerve reconstruction is required.

Craniotomy and high cervical exposure

After making a small suboccipital craniotomy (Fig. 2a), posterior mastoidectomy exposes the sigmoid sinus. This mastoidectomy aims to expose the sigmoid-jugular venous complex, and the opening of neither the fallopian nor the semicircular canals is required (Fig. 2c). To avoid any damage to CN VII, facial electromyogram is carefully monitored during the drilling. We prefer to utilize the continuous facial nerve monitoring by placing the stimulating electrode in the aditus. The high cervical exposure reveals the courses of CN XI and of the internal jugular vein (Fig. 2b). Removal of the C1 transverse process is required when exposure of the jugular vein is insufficient.

Venous ligation and intracranial tumor removal with continuous vagus nerve monitoring

The internal jugular vein is ligated, transected, and reflected upward to expose the extracranial tumor located ventral to the vein (Fig. 2e). After ligation of the sigmoid sinus using a Deschamps aneurysm needle (Fig. 2d), dural incision is started from the posterior fossa and extended to cross the sigmoid sinus.

In the cerebellopontine angle, the intact fibers of the lower CNs are carefully separated from the intracranial tumor capsule (Fig. 2f–h). In cases in which the tumor originates from CN IX, which is approximately half of the cases in our experience, intact CNs X and XI are commonly displaced caudally. After identifying the junction of CN X from the brainstem, a ball-type monopolar stimulating electrode is placed on the proximal site of CN X. The evoked vagus electromyogram is monitored with electrical stimulation at a frequency of 1 Hz throughout the procedure, to continuously assess the neural conditions in real-time without interrupting the microsurgical procedure [8].

Intrajugular and extracranial tumor removal

After the removal of the intracranial tumor, the dural incision is extended into the jugular foramen to expose the intrajugular tumor (Fig. 2i). Venous flow from the inferior petrosal sinus or anterior condylar confluence is encountered when completing the removal of the intrajugular tumor, and hence the hemorrhage should be promptly controlled using a fibrin glue-soaked hemostatic material (SURGICEL® cotton ball). The extracranial tumor is also removed while carefully preserving the intact CNs IX–XII and the internal carotid artery (Fig. 2j, k).

Closure

Primary dural closure is performed, and the remaining dural defect is covered by the sternocleidomastoid muscular flap (Fig. 2i). An abdominal fat pad is required when a wide dead space is needed to be filled. Skin closure of the neck requires special cosmetic consideration, as this incision is not within the hairline.

Indications

As schwannomas are typically benign, slow-growing, radiosensitive tumors, patients with schwannomas can be observed or treated by radiosurgery or surgical removal. Partial removal following radiosurgery (e.g., removal of the intracranial tumor following the radiosurgery for the extracranial tumor) may be one option for the treatment of triple dumbbell-shaped schwannomas. The surgical aim should be carefully considered in each case based on the patient's age, symptoms, tumor growth, and tumor size (particularly intracranial tumor size), and extension. Patients aiming at surgical cure by total

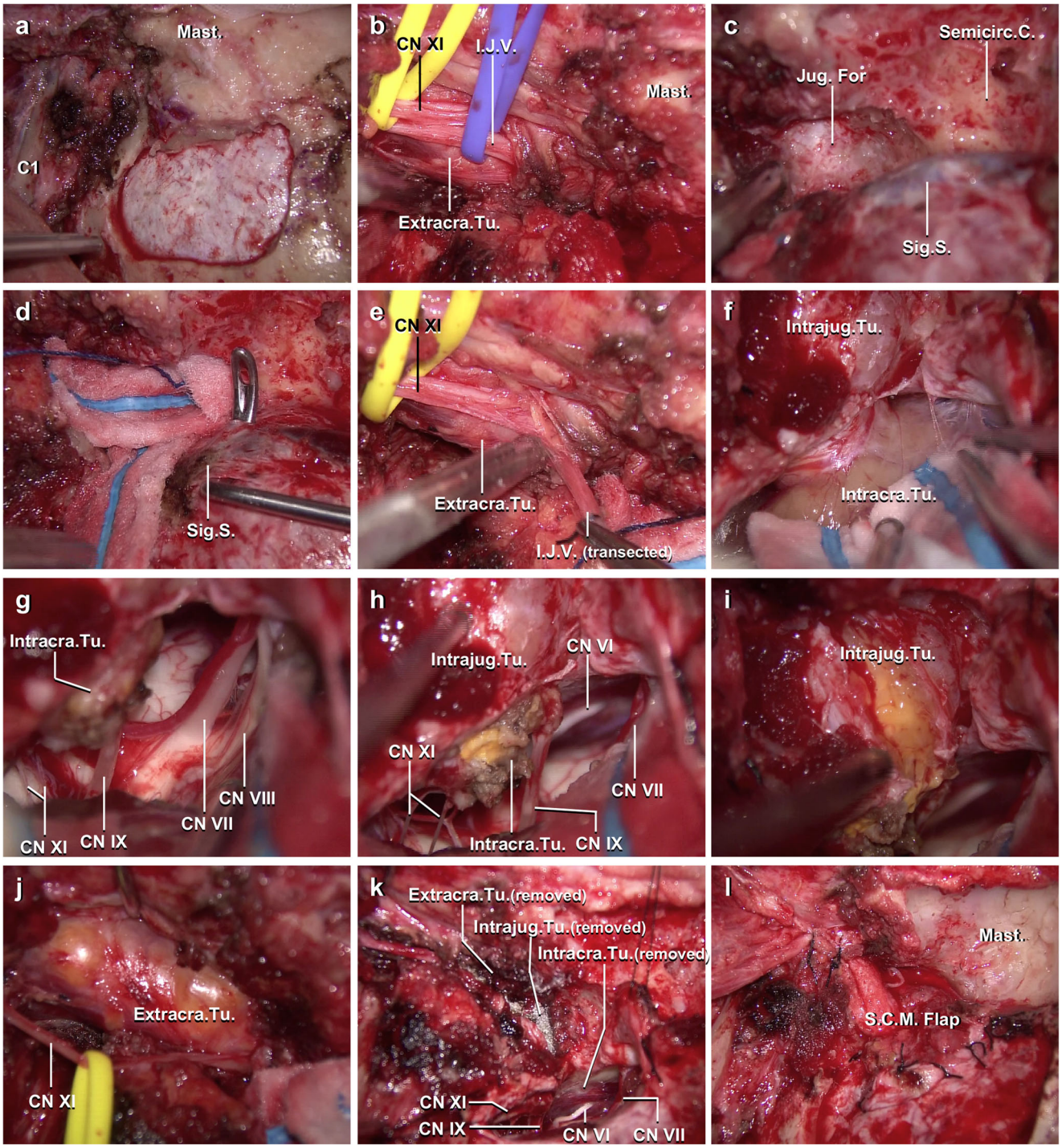

Fig. 2 Intraoperative photographs of a representative case. After small left suboccipital craniotomy (**a**), CN XI and the internal jugular vein were dissected and the extracranial component of the tumor was identified (**b**). Mastoidectomy exposed the entire sigmoid-jugular venous complex around the jugular foramen (**c**). The sigmoid sinus (**d**) and the jugular vein were ligated, and the extracranial tumor was exposed by reflecting upward the transected jugular vein (**e**). Dural incision revealed the intracranial tumor (**f**), and CNs VII and VIII and the intact lower CNs were carefully dissected from the tumor (**g**). The residual tumor in the cranium and that in the jugular foramen were confirmed to be continuous with each other (**h**). The intrajugular (**i**) and extracranial tumors (**j**) were removed while carefully preserving the intact nerves (**k**). The vascularized sternocleidomastoid muscular flap was used for closure (**l**). This tumor originated from CN X, and therefore, continuous vagus nerve monitoring was not performed in this case. CN, cranial nerve; Extracra., extracranial; I.J.V., internal jugular vein; Intracra., intracranial; Intrajug., intrajugular; Jug. For., jugular foramen; Mast., mastoid; S.C.M., sternocleidomastoid muscle; Semicirc. C., semicircular canals; Sig. S., sigmoid sinus; Tu., tumor

removal of a triple dumbbell-shaped tumor should be the only candidates for this surgical procedure (Fig. 3) [1–7, 9, 10]. When the tumor is located only intracranially or when its intrajugular or extracranial extension is limited, the approach preserving the sigmoid-jugular complex, such as retrosigmoid or its suprajugular approach should be selected [8].

Limitations

The transsigmoid approach has been reported as a safe and effective procedure for a surgical treatment of jugular foramen tumors extending intra- and extracranially [1–7, 9, 10], but this surgery is not applicable for patients in whom the sigmoid-jugular complex should not be sacrificed (e.g., patients with a dominant and patent sinus that did not tolerate the test occlusion).

How to avoid complications

Careful application of this technique, detailed intraoperative neuromonitoring, and pre- and postoperative multidisciplinary laryngeal evaluations are mandatory for a successful treatment. We routinely monitor the ABR, SEP, MEP, and electromyograms of CNs VII, X, XI, and XII during this procedure (Fig. 1a), and continuous vagus nerve monitoring is essential to avoid postoperative lower CN deficits [8]. Extubation is usually possible immediately after the surgery, but laryngeal function should be carefully examined with cooperation from otolaryngologists and speech therapists. Laryngoscopy should be performed before restarting oral feeding, and additional videofluorography is considered if dysphagia is suspected. A lumbar drainage is routinely placed for 4–7 days postoperatively, to prevent cerebrospinal fluid leakage.

Specific perioperative considerations

The sigmoid-jugular complex is already occluded preoperatively in many patients (Fig. 3d), but when it is patent, we routinely predict the safeness of its ligation by the balloon occlusion test. Development of adjacent venous structures, including the posterior and mastoid emissary veins (Fig. 3d), displacement of the internal carotid artery, tumor vascularity, bony erosion (Fig. 3c), and craniocervical instability should also be evaluated on the radiological images.

In our 65 surgically treated patients with jugular foramen schwannomas, hearing disturbance was the most common preoperative symptom, which comprised almost 40% of them, and approximately half of them experienced hearing improvement after surgery. Therefore, even if the patient's preoperative hearing is unserviceable, surgical approaches that

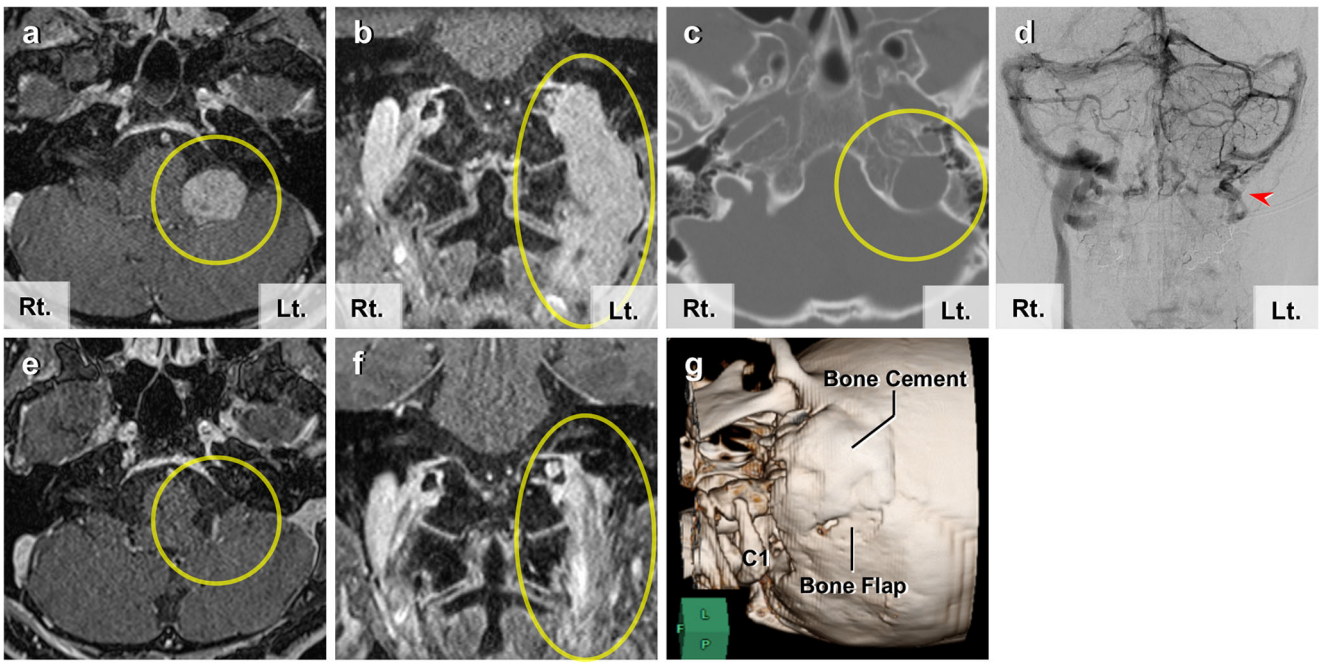

Fig. 3 Radiological images of a representative case. Preoperative MRI displaying a left jugular foramen schwannoma extending intra- (**a**) and extracranially (**b**). The jugular foramen was enlarged (**c**) and the internal jugular vein was occluded (**d**). The posterior condylar vein developed alternatively (**d**, red arrow head). The triple dumbbell-shaped tumor was resected through the transjugular transsigmoid approach with high cervical exposure, and the patient showed no tumor recurrence during the 2 years of follow-up after the operation (**e**, **f**). The bone defect was reconstructed with a bone flap and cement (**g**)

sacrifice or endanger hearing function should not be recommended.

Specific information for the patient

Patients should be informed about alternative treatment choices, general surgical risks, and specific risks as mentioned above, including CN VII and lower CN deficits and cerebrospinal fluid leakage.

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Compliance with ethical standards

Informed consent Informed consent was obtained from all individual participants included in the study.

Conflict of interest The authors declare that they have no competing interests.

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Key points

- Careful patient selection and appropriate setting of the surgical aim for this benign tumor
- Surgical strategy remaining chance of hearing recovery
- Creation of a vascularized flap and preservation of the greater auricular nerve
- Safe exposure of the entire sigmoid-jugular venous complex by mastoidectomy preserving facial and hearing functions
- Precise dissection of intact lower CNs and early beginning of continuous vagus nerve monitoring
- Prompt control of hemorrhage based on detailed understanding of the venous anatomy around the jugular bulb
- Dissection and removal of the intrajugular and extracranial tumor while avoiding any damage to the CNs and internal carotid artery
- Closure using a vascularized flap and postoperative lumbar drainage management to prevent cerebrospinal fluid leakage
- Skin closure considering cosmetic outcome, particularly on the neck outside of the hairline
- Pre- and postoperative multidisciplinary evaluations of laryngeal function with cooperation from otolaryngologists and speech therapists