

A Case of Hypoglossal Neurilemmoma Resected Via Burr-hole Craniectomy

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Hypoglossal neurilemmoma is extremely rare. Intracranial hypoglossal neurilemmoma has been reported to the present most commonly as a space-occupying lesion with symptoms of raised intracranial pressure. A 68-year-old woman presented with deviation of the tongue to the left on protrusion. Preoperative radiological images revealed an extra-axial mass in and around the hypoglossal canal. The tumor was totally resected via retrosigmoid suboccipital approach with burrhole craniectomy. Histopathological examination verified a neurilemmoma. She had no neurologic abnormality except hypoglossal palsy which recovered completely in six months. Retrosigmoid suboccipital approach with burrhole craniectomy can be an useful approach in intracranial hypoglossal neurilemmoma without extracranial extension or with minimal extracranial extension into the hypoglossal canal.

KEY WORDS : Hypoglossal nerve · Neurilemmoma · Suboccipital approach.

Introduction

Hypoglossal neurilemmomas are very rare^{1,2,6,12,14,19-21}. These tumors usually grow intracranially causing an enlargement and erosion of the hypoglossal canal, however sometimes those can extend into the extracranial part^{1,5,14,20}. The retrosigmoid suboccipital approach traditionally provides excellent exposure and access to the neurovascular structures of the cerebellopontine angle(CPA). Many authors advocate it as the procedure of choice during vestibular nerve section for intractable peripheral vestibular disorders, microvascular decompression(MVD) for hyperactive cranial nerve syndromes, and hearing preservation during surgery for vestibular schwannomas^{3,4,11}. Here we briefly summarize a case of the hypoglossal neurilemmoma, presenting as hypoglossal palsy. In addition, we describe technical aspects of the retrosigmoid suboccipital approach with one burrhole craniectomy for hypoglossal neurilemmoma.

Case Report

A 68-year-old female patient visited with a complaint of slowly progressive deviation of the tongue to the left on

protrusion and nuchal pain, which developed 5 months ago. The patient was alert and well oriented. She showed full range of sensory-motor function. Physical examination revealed mild atrophy of the left tongue and no cutaneous lesion. No neurological finding other than hypoglossal palsy was present. She had neither relevant family nor past medical history. Initial magnetic resonance(MR) images demonstrated the intracranial part of a high signal tumor between the medulla and the medial part of the left cerebellum (Fig. 1), that was exhibited as extra-

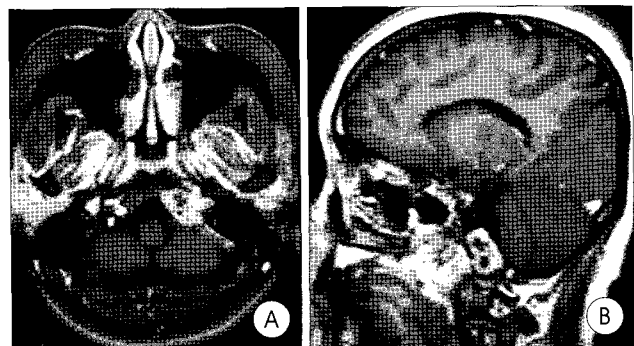


Fig. 1. Preoperative T1-weighted gadolinium-enhanced magnetic resonance images in 68-year-old female presented with hypoglossal palsy. Axial (A) and sagittal (B) images show intensely enhancing mass with cysts that filled and extended into the hypoglossal canal.

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Fig. 2. Intraoperative photograph shows a tumor (white asterisk), a branch of the posterior inferior cerebellar artery (black arrow), and the cranial root of accessory nerve (X).

axial mass in the posterior fossa. The size of the tumor was measured $25 \times 20 \times 15$ mm. It showed low signal on T1-weighted images and high signal on T2-weighted images with marginal enhancement. Although there were no displacement of the carotid and vertebral arteries and the medulla, the mass extended extracranially along the left hypoglossal canal through the eroded bone defect. When considering slow growth of the tumor and advanced patient's age, we selected rather minimal approach. Preoperatively, we discussed the possibility of radiosurgery for the remnant tumor with the patient and her family members. Under the general anesthesia, the tumor was removed via the retrosigmoid suboccipital approach with one burrhole craniectomy. The whitish-yellow tumor was well encapsulated (Fig. 2). Several branches of the hypoglossal nerve adhered to the upper surface of the tumor, and the tumor extended into the enlarged hypoglossal canal. The nerve fibers were carefully dissected off the surface of the tumor preserving their integrity. The tumor was resected totally after a part of the hypoglossal canal was drilled out. Histopathological ex-

amination of the resected tumor confirmed as a neurilemmoma (Fig. 3A). The cell proliferation consisted of spindle cells with often wavy nuclei. S-100 immunostaining showed intense positivity of the spindle cells (Fig. 3B). Postoperatively, the computed tomography(CT) with bone window showed no residual tumor (Fig. 4). She had no postoperative neurologic deficits except for the left hypoglossal palsy which was recovered on follow up study 6 months later.

Technical aspects of the retrosigmoid suboccipital approach with burrhole craniectomy

The patient was placed in the lateral position under the endotracheal general anesthesia. Following insertion of stimulus and response leads for electrophysiologic monitoring of cranial nerve VII~XI, the head is fixed with Yasargil head clamp. A 5cm retroauricular skin incision was made overlying the asterion. A 2cm diameter craniectomy extending inferiorly from the asterion was then made using air speed drill and Kerrison punch. After the sigmoid and transverse sinus margin was identified, triangular dural opening was made with the intact dura base along the medial edge of the sigmoid sinus. In an attempt to minimize contamination of the subarachnoid space with bone dust, drilling stopped when the dura was incised and the posterior fossa was entered. Draining of 30~50cc of cerebrospinal fluid(CSF) made it easy to expose the CPA with minimal or no cerebellar retraction. If once the CPA was exposed, lower cranial nerves and tumor could be easily identified on the microscope. A basket-like rootlet adherent to the surface of the tumor was found running over the tumor and extending towards the hypoglossal canal. The tumor covered by arachnoidal membranes was exposed. Its capsule was incised and the lesion was internally debulked. Dissection from the surrounding cerebellum, brain stem, and lower cranial nerves was performed without much efforts, except for the hypoglossal nerve. Bleeding control was done with oxycellulose

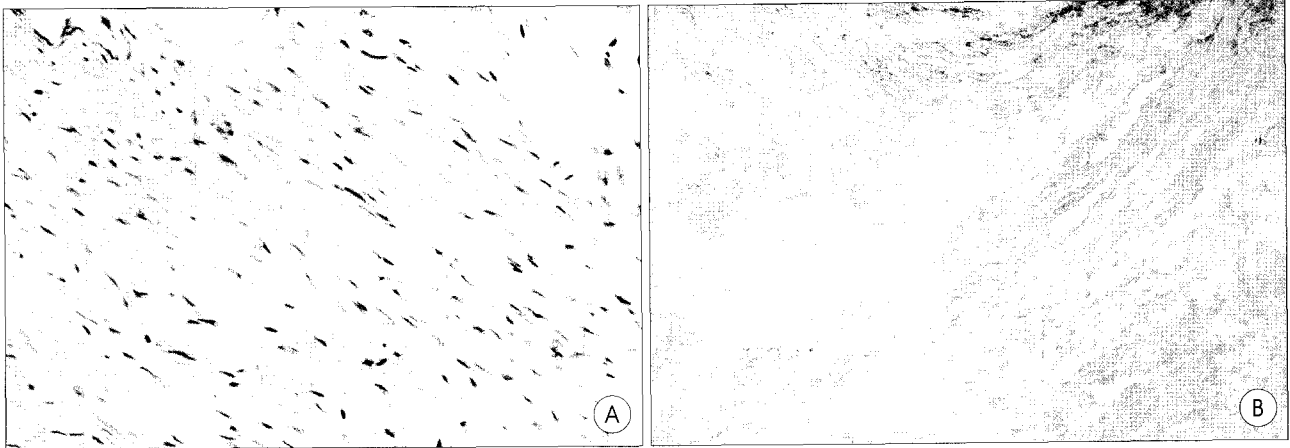


Fig. 3. A : Histopathological examination shows arranged and wavy tumor cells within long fascicles (H&E, $\times 200$). B : Immunohistochemical staining for S-100. The tumor cells are positive for S-100 immunohistochemical staining (IHC, $\times 200$).

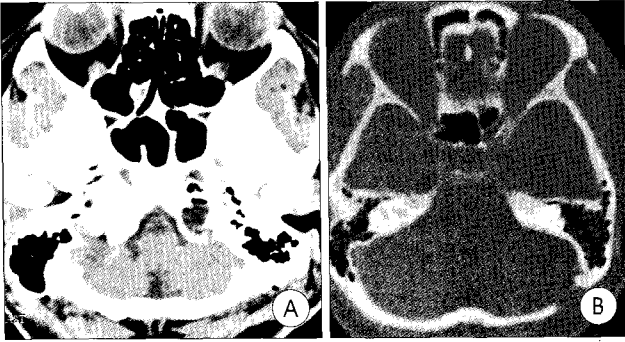


Fig. 4. A : Axial computed tomography(CT) with enhancement shows total removal status of the tumor in the cistern and hypoglossal canal. B : CT scan with bone window shows a small bony defect (surgical corridor).

and cottonoid. When the intracranial portion of the procedure was completed, the dura was sufficiently reapproximated. After surgical glue application, the site of burrhole craniectomy was filled with bone dust and fragments wrapped in a large oxycellulose.

Discussion

Hypoglossal neurilemmoma is quite rare and only less than 100 cases have been reported previously¹⁹. It accounts for only 5% of all non-acoustic intracranial neurilemmomas⁹. It is mostly intracranial tumor and usually arise intracranially, causing an enlargement and erosion of the hypoglossal canal. Hypoglossal neurilemmoma, on rare occasion, can extend into the extracranial compartment towards the jugular foramen, carotid canal, or infratemporal fossa^{1,5,14,20}. Most intracranial neurilemmomas arise from sensory divisions of the cranial nerves, including acoustic, vestibular, and trigeminal nerves. On the other hand, the most common sign is hypoglossal nerve palsy with ipsilateral hemiatrophy and weakness of the tongue in hypoglossal neurilemmoma²⁰. Hypoglossal nerve palsy has particular diagnostic value, since more than 90% of patients presented with this finding^{10,21}.

High-resolution CT scan and MR images are most crucial radiological tools, which together provide a complete evaluation of the tumor type and extension, extent of bone destruction and involvement of the surrounding structure. High-resolution CT scan with bone window is indispensable for detecting bone involvement : an enlargement or an erosion of the hypoglossal canal has a significant clue on the differential diagnosis compared with jugular foramen tumors^{2,5,6,17}. Gadolinium-enhanced MR displays the morphological features of the tumor mass and defines its localization and relationships with the dura, vertebral artery, jugular bulb, the lower cranial nerves, and the brain stem^{14,16,17}. Besides, angio-MR images provides a further complete pre-operative evaluation, showing the involvement and/or displacement of such major vessels as the vertebral artery,

the sigmoid sinus and the jugular bulb^{7,12,17}.

Since neurilemmomas are benign tumors, the mainstay of the treatment is complete surgical removal. But, surgical approaches to the hypoglossal canal and the jugular foramen are often complex and require time consuming procedures associated with significant morbidity. Their locations in a complex region of the skull base can make complete removal difficult owing to the complicate anatomical relationship to neurovascular and brainstem structures. The choice of the best surgical approach is determined by the type of tumor extension, bone destruction and involvement of the surrounding structures. According to the current literatures, the dorsolateral suboccipital transcondylar approach is indicated in the removal of intradural tumors located in the lower clivus and anterolaterally to the cervicomedullary junction^{7,8,13,16}. In dumbbell-shaped hypoglossal neurilemmoma, far lateral approach with partial resection of the condyle to open the hypoglossal canal is the recent preferred surgical approach^{12,18}. If size is small and confirmed into the hypoglossal canal, only the necessary parts of the canal should be drilled so as not to destroy the condyle and induce craniovertebral instability¹².

On the other hand, radiosurgery also can be a good alternative for an elderly patient. However, if the tumor is crushing the brain stem, radiosurgery is not safe for all occasions, and a cystic tumor is less likely to respond to the radiosurgery^{12,15}.

Conclusion

Hypoglossal neurilemmoma is an unexceptionally rare tumor. High-resolution CT, MR images and angio-MR are useful radiological tools for early diagnosis and preoperative surgical planning. Suboccipital approach with burrhole craniectomy can be a useful approach when operating intracranial hypoglossal neurilemmoma without extracranial extension or with minimal extracranial extension into the hypoglossal canal.

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