CASE REPORT

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SCHWANNOMA ORIGINATING FROM LOWER CRANIAL NERVES: REPORT OF 4 CASES

HIROFUMI OYAMA, AKIRA KITO, HIDEKI MAKI, KENICHI HATTORI, TOMOYUKI NODA and KENTARO WADA

Department of Neurosurgery, Ogaki Municipal Hospital, Ogaki, Japan

ABSTRACT

Four cases of schwannoma originating from the lower cranial nerves are presented. Case 1 is a schwannoma of the vagus nerve in the parapharyngeal space. The operation was performed by the transcervical approach. Although the tumor capsule was not dissected from the vagus nerve, hoarseness and dysphagia happened transiently after the operation. Case 2 is a schwannoma in the jugular foramen. The operation was performed by the infralabyrinthine approach. Although only the intracapsular tumor was enucleated, facial palsy, hoarseness, dysphagia and paresis of the deltoid muscle occurred transiently after the operation. The patient's hearing had also slightly deteriorated. Case 3 is a dumbbell-typed schwannoma originating from the hypoglossal nerve. The hypoglossal canal was markedly enlarged by the tumor. As the hypoglossal nerves were embedded in the tumor, the tumor around the hypoglossal nerves was not resected. The tumor was significantly enlarged for a while after stereotactic irradiation. Case 4 is an intracranial cystic schwannoma originating from the IXth or Xth cranial nerves. The tumor was resected through the cerebello-medullary fissure. The tumor capsule attached to the brain stem was not removed. Hoarseness and dysphagia happened transiently after the operation. Cranial nerve palsy readily occurs after the removal of the schwannoma originating from the lower cranial nerves. Mechanical injury caused by retraction, extension and compression of the nerve and heat injury during the drilling of the petrous bone should be cautiously avoided.

Key Words: Schwannoma, Hypoglossal nerve, Vagus nerve, Parapharyngeal tumor, Infralabyrinthine approach

INTRODUCTION

The development of lower cranial (IXth–XIIth) nerves-derived schwannoma in the cervical region, jugular foramen, and hypoglossal canal is comparatively rare. As for the cervical region, the nerves of origin were identified in 62% and they were the brachial plexus, facial nerve, trigeminal nerve, vagus nerve and sympathetic chain.[1)] Sixty-nine percent of the patients presented with symptoms secondary to mass effect or nerve deficit and the main symptoms were neck mass (27%), dyspnea/dysphonia (15%), nasal obstruction/epistaxis (12%), odynophagia/dysphagia (12%) and shoulder/upper extremity pain (8%).[1)] The parapharyngeal space is the most common site of tumor origin (31%), followed by the neck (23%), skull base (19%), sinonasal cavity (15%), middle ear (8%) and posterior pharynx (4%).[1)]

Corresponding Author: Hirofumi Oyama, M.D.

Department of Neurosurgery, Ogaki Municipal Hospital, Minamikawamachi 4-86, Ogaki, Gifu 503-8502, Japan

Phone: 0584-81-3341, Fax: 0584-75-5715, E-mail: oya3776@arrow.ocn.ne.jp

It was reported that the origin of the jugular foramen schwannoma was the glossopharyngeal (IXth) nerve in 40%, vagus (Xth) nerve in 25% and spinal accessory (XIth) nerve in 35% of cases.[2] As for symptoms, the presentation of the patients reportedly varied according to the tumor growth pattern.[3] Deafness, vertigo, and ataxia were present in all patients with a major intracranial component.[3] By contrast, lower cranial nerve involvement, including hoarseness and weakness of the trapezius and sternocleidomastoid muscles, occurred in patients in whom the tumor was primarily within the bone or extracranial.[3] Patients with the major component of the schwannoma within the bone also experienced deafness.[3]

Hypoglossal schwannoma is considered to be comparatively rare. The locations of these tumors were reported to be intracranial (31.5%), both intra- and extracranial (50%) and extracranial (18.5%).[4] They mainly presented with cerebellar dysfunction (45.3%) and pareses of the facial (VIIth) (17.6%), IXth (57.4%), Xth (40.3%), XIth (20.9%), and hypoglossal (XIIth) (85.7%) cranial nerves.[4]

We describe 4 cases of schwannomas in those areas and discuss the operative complications and methods to avoid them.

CASE REPORT

Case 1 (Vagal nerve-derived schwannoma in the parapharyngeal space)

A 31-year-old female presented with difficulty in swallowing. She showed no other neurological abnormalities. A palpable tumor was observed under the right mandible. Computed tomography (CT) and magnetic resonance imaging (MRI) showed a schwannoma in the right parapharyngeal space (Fig. 1A). A 10-cm-long transverse incision was placed 2 cm below the

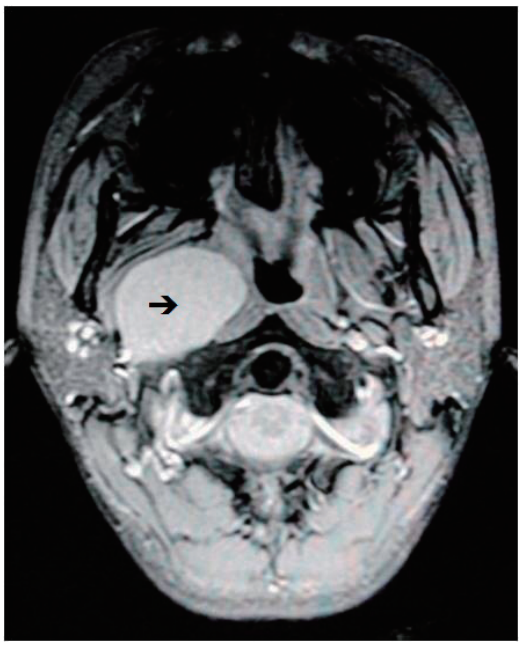

Fig. 1A Preoperative gadolinium-enhanced magnetic resonance imaging, T1-weighted image, axial view. Tumor (arrow) is found in right parapharyngeal space.

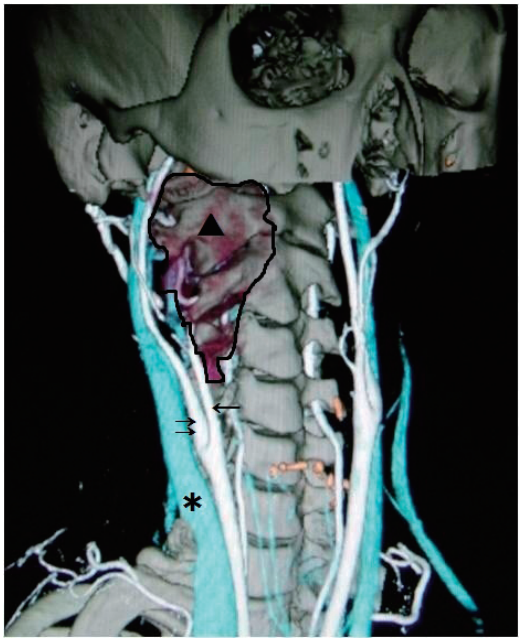

Fig. 1B Preoperative enhanced 3-dimensional computed tomography scan. Tumor (purple-colored, enclosed with black lines, arrowhead) is located behind external carotid artery (white-colored, arrow). Internal carotid artery (white-colored, double arrow) and internal jugular vein (blue-colored, asterisk) are pushed backward by tumor.

right lower mandible to exfoliate and elevate the caudal portion of the submaxillary gland. The digastric muscle and facial artery/vein were cut to allow the external carotid artery to be moved out of place. Although the internal carotid artery was thin and adhered to the dorsal side of the tumor capsule, the artery could be exfoliated (Fig. 1B). It was impossible to exfoliate the vagal nerve because it was completely integrated with the tumor capsule on the dorsal side of the tumor. Although the cranial portion of the tumor was not exfoliated, and an extremely thin tumor capsule remained, the entire tumor was removed completely. Pathological examination showed a palisade arrangement of spindle cells, and thus the tumor was diagnosed to be schwannoma. The patient showed a mild swallowing disorder and moderate dysphonia following surgery, but both were resolved within 3 months, and there was no recurrence as of 11 months after surgery.

Case 2 (Jugular foramen schwannoma)

A 56-year-old female patient presented with dizziness, hearing loss of 58.8 dB in her left ear, hoarseness, and a swallowing disorder. CT and MRI showed that an extradural tumor had damaged the left petrous bone (Fig. 2A). Cerebral angiography showed that the left sigmoid sinus was obstructed, and the tumor was fed by the left ascending pharyngeal artery. The intracapsular tumor was completely removed using a left infralabyrinthine approach (Fig. 2B). The tumor had mainly located in the pars nervosa containing the IXth, Xth and XIth cranial nerves, but the nerve of the tumor origin was not identified. Pathological examination showed that there were cells with spindle- and elliptically-shaped nuclei with infiltration of inflammatory cells. There were also some cells with large heterotypic nuclei. In addition, there were many blood vessels with hemorrhage. Immunostaining showed that the tumor was positive for S-100 protein

and negative for epithelial membrane antigen, and it was diagnosed to be schwannoma. There was some loss of hearing, in addition to transient hoarseness, swallowing disorder, accessory nerve paralysis (difficulty in elevating left shoulder), and left facial nerve paralysis following the resection. These symptoms resolved almost completely and there was no recurrence as of 18 months after surgery.

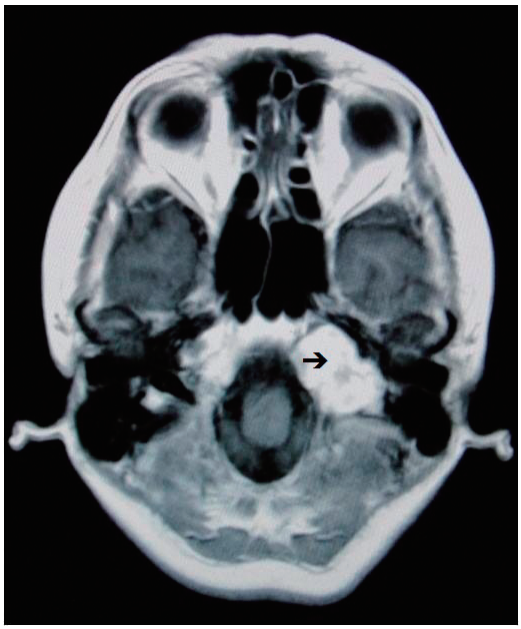

Fig. 2A Preoperative gadolinium-enhanced magnetic resonance imaging, T1-weighted image, axial view. Tumor (arrow) is found in left anterior petrous bone.

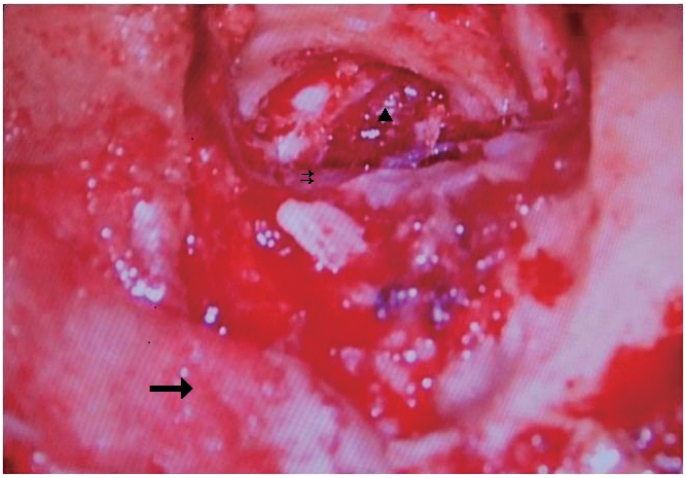

Fig. 2B Operative view, infralabyrinthine approach. Left sigmoid sinus (arrow) is dissected from petrous bone. Red-colored tumor (arrow head) is found beyond compressed blue-colored jugular bulb (double arrow).

Case 3 (Dumbbell-shaped hypoglossal schwannoma)

A 25-year-old male patient presented with headache and nausea. The right side of the tongue showed paralysis and atrophy. CT and MRI demonstrated a dumbbell-shaped tumor in the right hypoglossal canal, extending in and out of the skull (Fig. 3A). There was a large multilocular cyst in the right cerebellopontine angle, strongly compressing the brain. In addition, the right

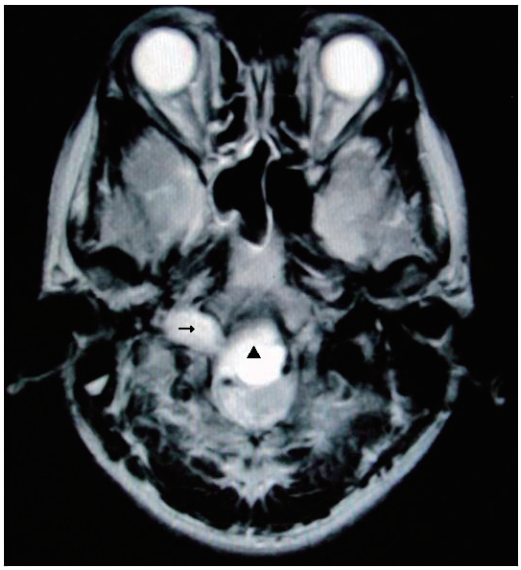

Fig. 3A Preoperative magnetic resonance imaging, T2-weighted image, axial view. Tumor is found in right hypoglossal canal (arrow) and posterior fossa (arrowhead).

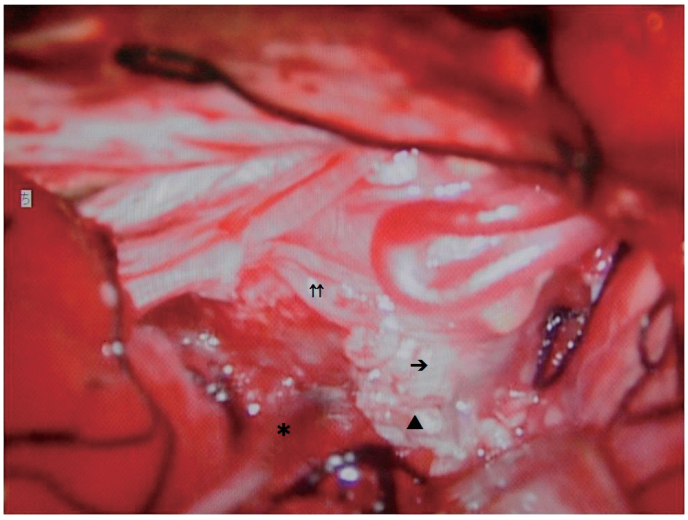

Fig. 3B Operative view, right retromastoid suboccipital approach. Tumor remnant (arrowhead) is found around hypoglossal nerves (arrow). Accessory nerves (double arrow) and right posterior inferior cerebellar artery (asterisk) are also shown.

hypoglossal canal was enlarged by the tumor. Most of the tumor was surgically resected, except for a portion surrounding the hypoglossal nerve (Fig. 3B). Pathological examination clearly confirmed the disorganized bundles of spindle-shaped cells with marked cyst formation, and the tumor was diagnosed to be schwannoma with severe degeneration. Vomiting continued for a while following surgery. The tumor grew during the course in which it developed a cyst formation after stereotactic irradiation. Temporal headache and nausea developed in addition to difficulty in oral intake. However, these symptoms improved and the tumor decreased over time. Now, the tongue atrophy remains and the tumor has increased slightly 37 months after surgery.

Case 4 (Lower cranial nerve-derived cystic schwannoma in the posterior cranial fossa)

A 47-year-old male patient presented with vomiting and right hearing loss. A cystic tumor was found in the right posterior cranial fossa. Subtotal excision was performed through the cerebello-medullary fissure by the first author in another hospital. The tumor developed from the IXth and Xth cranial nerves and parts of these nerves were sacrificed. A part of the tumor was strongly adhesive to the brain stem, and was not resected. Pathological examination of the cyst wall revealed that there were spindle-shaped cells, and the tumor was diagnosed to be schwannoma. The patient experienced transient hoarseness and swallowing disorder after the surgery, which improved later. No recurrence was observed over a 14-year follow-up.

DISCUSSION

Tumor in the parapharyngeal space

Various tumors develop in the parapharyngeal space including parotid gland-derived tumor and metastasis of malignant tumors, such as malignant lymphoma, nasopharyngeal cancer, oral cancer, and thyroid cancer. It was reported that numerous cases of nerve cell-origin tumor were vagus nerve- and sympathetic nerve-derived tumors.[5,6] Forty-two percent of the patients had schwannomas from the cervical sympathetic chain and 58% had schwannomas of the cervical vagus nerve.[6] The carotid artery and internal jugular vein are often separated in cases with vagus nerve-derived tumors, while the internal carotid artery and external carotid artery are often separated in cases with sympathetic nerve-derived tumors.[6] In the current case 1, the carotid artery and internal jugular vein were not separated, but the internal carotid artery and external carotid artery were separated. A sympathetic nerve-derived tumor was suspected from the radiological findings. However, the vagal nerve was discovered to be completely integrated with the tumor capsule during the operation and the tumor was conclusively diagnosed as vagal nerve-derived schwannoma.

The proper treatment for this type of tumor is surgical removal with preservation of the nerve, if possible, and reanastomosis and/or nerve grafting if that is not possible.[7] Surgical methods used to resect schwannomas include a transcervical approach, a cervical-transparotid approach, and a cervical-transpharyngeal approach, but a transcervical approach can be used in most cases like the current case 1. The cranial and sympathetic nerves were not sacrificed in Case 1.

Eighty-eight percent of the patients reportedly had complete surgical excision, and 12% had subtotal resection.[1] Adhesion of the tumor to nerves requires tumor enucleation, or cutting of the adhesive nerve with successive neuroanastomosis.[8,9] Postoperative nerve injury occurred in 62% of the patients with a resolution in 44%.[1] Hoarseness, choking and coughing while eating were reported to happen after operation, and horner syndrome was also a consequence of sympathetic chain resection.[5,7] In the current case 1, the vagal nerve was completely integrated with the tumor capsule during the operation, so intracapsular resection was performed. However, transient lower

cranial nerve palsy manifested after the operation. The injury was thought to have occurred when the vagus nerve was extended or pulled during the process of the tumor resection.

Jugular foramen schwannoma

Jugular foramen schwannomas can be classified into type A: a tumor in the posterior cranial fossa; type B: a tumor in the jugular foramen; type C: a tumor in the extracranial portion; or type D: a dumbbell tumor extending in and out of the skull.[10] During surgery for type A tumor, the resection is usually performed with a retrosigmoid approach. An infralabyrinthine approach is used for many type B and C tumors. Type D tumors are resected using a combined approach. Reconstruction of the extradural dead space with a pedicled flap is important.

Lower cranial nerve injury was reported to occur in 15–50% of cases after surgery.[2,11,12,13] New postoperative deficits in previously normal cases were rarely seen, however, worsening of preoperative deficits was frequently noted.[2] The deficits were reported as follows: N. VII (transient 11.3%, permanent 3.8%), N. VIII (transient 7.5%, permanent 7.5%), N. IX & X (transient 30.1%, permanent 26.4%), N. XI (transient 17%, permanent 11.3%), N. XII (transient 17%, permanent 13.2%), and ataxia (17%).[2] In the current Case 2, transient deficits of the lower cranial nerves happened, although intracapsular resection was performed. This might be due to the fact that the lower cranial nerves in the pars nervosa were pushed to the wall during curettage. Since the lower cranial nerves were already weak due to compression by the tumor, concomitant use of endoscopy is preferable.

It was reported that 11.3–11.9% of surgical patients had disturbance of facial function.[2,11] Temporal facial nerve paralysis in Case 2 may have been caused by heat generated during drilling of the petrous bone, because no exfoliation was performed between the mastoid process and digastric muscle. Cooling with intermittent watering was insufficient, and continuous cooling with cold water would have been preferable.

Hypoglossal schwannoma

This tumor is characterized by an enlarged hypoglossal canal. It is best to open the hypoglossal canal using a transcondylar approach for complete resection. It was reported that 42.9% of the patients showed improvement in the function of the lower cranial nerves after the operation.[14] Electromyography monitoring might be useful for removing the tumor while maintaining the function of the hypoglossal nerve.[15] In Case 3, complete resection was avoided since the tumor strongly adhered to the lower cranial nerves.[16]

The tumor may temporarily increase in size after stereotactic irradiation of the residual tumor, promoting aggravation of symptoms in the current Case 3. Pre-existing tongue atrophy reportedly persisted even after removal of the tumor.[14] As atrophy was already observed on the right side of the tongue in Case 3, there was the option to perform complete resection with sacrifice of the right hypoglossal nerve. Some studies have reported a favorable prognosis in cases that underwent neurorrhaphy with sural nerve grafting after cutting the hypoglossal nerve.[17]

CONCLUSION

Intracapsular enucleation is the most effective and safest procedure for tumor removal. Even if only the thin capsule remains, recurrence is unlikely after the operation. The surrounding or original cranial nerves adhere to the tumor capsule, making anatomical separation difficult. However, mechanical damage can be imposed on the surrounding or original cranial nerves when the tumor capsule is retracted or compressed. This is due to the thin and weakened cranial nerves

adhered to the tumor capsule being retracted or compressed with the tumor capsule. In Case 1, the deficits were thought to happen mainly due to the retraction of the tumor capsule. In Case 2, the deficits were suggested to be caused by the compression from inside the tumor capsule.

Gentle operative manipulation is mandatory to avoid cranial nerve damage. At first, the tumor capsule should be enucleated as much as possible. As the capsule becomes thinner, the search for the adhered cranial nerves on the back side becomes easier. Next, so as to avoid compression of the cranial nerves, the tumor capsule should not be compressed from inside during enucleation and hemostasis. Then, the tumor capsule should be removed piece by piece at the previous portion. Retraction of the tumor capsule should be avoided. Temporary or permanent nervous injury may occur easily, even though the continuity of the lower cranial nerve can be maintained. It is important to avoid injuries caused by traction, extension, compression, and heating of nerves during surgery for lower cranial nerve-derived schwannoma.

REFERENCES

1) Malone JP, Lee WJ, Levin RJ. Clinical characteristics and treatment outcome for nonvestibular schwannomas of the head and neck. *Am J Otolaryngol,* 2005; 26: 108–112.
2) Bulsara KR, Sameshima T, Friedman AH, Fukushima T. Microsurgical management of 53 jugular foramen schwannomas: lessons learned incorporated into a modified grading system. *J Neurosurg,* 2008; 109: 794–803.
3) Kaye AH, Hahn JF, Kinney SE, Hardy RW, Jr; Bay JW. Jugular foramen schwannomas. *J Neurosurg,* 1984; 60: 1045–1053.
4) Hoshi M, Yoshida K, Ogawa K, Kawase T. Hypoglossal neurinoma--two case reports. *Neurol Med Chir (Tokyo),* 2000; 40: 489–493.
5) Guerrissi JO. Solitary benign schwannomas in major nerve systems of the head and neck. *J Craniofac Surg,* 2009; 20: 957–961.
6) Saito DM, Glastonbury CM, El-Sayed IH, Eisele DW. Parapharyngeal space schwannomas: preoperative imaging determination of the nerve of origin. *Arch Otolaryngol Head Neck Surg,* 2007; 133: 662–667.
7) Chang SC, Schi YM. Neurilemmoma of the vagus nerve. A case report and brief literature review. *Laryngoscope,* 1984; 94: 946–949.
8) Gilmer-Hill HS, Kline DG. Neurogenic tumors of the cervical vagus nerve: report of four cases and review of the literature. *Neurosurgery,* 2000; 46: 1498–1503.
9) Langner E, Del Negro A, Akashi HK, Araújo PP, Tincani AJ, Martins AS. Schwannomas in the head and neck: retrospective analysis of 21 patients and review of the literature. *Sao Paulo Med J,* 2007; 125: 220–222.
10) Samii M, Babu RP, Tatagiba M, Sepehrnia A. Surgical treatment of jugular foramen schwannomas. *J Neurosurg,* 1995; 82: 924–932.
11) Fayad JN, Keles B, Brackmann DE. Jugular foramen tumors: clinical characteristics and treatment outcomes. *Otol Neurotol,* 2010; 31: 299–305.
12) Sanna M, Bacciu A, Falcioni M, Taibah A. Surgical management of jugular foramen schwannomas with hearing and facial nerve function preservation: a series of 23 cases and review of the literature. *Laryngoscope,* 2006; 116: 2191–2204.
13) Wilson MA, Hillman TA, Wiggins RH, Shelton C. Jugular foramen schwannomas: diagnosis, management, and outcomes. *Laryngoscope,* 2005; 115: 1486–1492.
14) Ichimura S, Yoshida K, Kawase T. Surgical approach for hypoglossal schwannomas to prevent deformity of the atlanto-occipital joint. *Acta Neurochir (Wien),* 2009; 151: 575–579. Epub 2009 Apr 1.
15) Ishikawa M, Kusaka G, Takashima K, Kamochi H, Shinoda S. Intraoperative monitoring during surgery for hypoglossal schwannoma. *J Clin Neurosci,* 2010; 17: 1053–1056. Epub 2010 May 21.
16) Yoshida K. Surgical strategy for hypoglossal schwannoma. *No Shinkei Geka,* 2010; 38: 17–23.
17) Mathiesen T, Svensson M, Lundgren J, Kihlström L, Parisotto R, Bagger-Sjöbäck D. Hypoglossal schwannoma-successful reinnervation and functional recovery of the tongue following tumour removal and nerve grafting. *Acta Neurochir (Wien),* 2009; 151: 837–841. Epub 2009 Mar 17.