

Clinical Article

Tailored Surgical Approaches for Benign Craniovertebral Junction Tumors

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Objective : We report our surgical experience in the treatment of 16 consecutive patients with benign craniovertebral junction (CVJ) tumor, observed from 2003 to 2008 at our department.

Methods : We had treated 6 foramen magnum meningiomas, 6 cervicomedullary hemangioblastomas, 1 accessory nerve schwannoma, 1 hypoglossal nerve schwannoma, 1 C2 root schwannoma, and 1 cavernous hemangioma. Clinical results were evaluated by Karnofsky Performance Scale (KPS) and all patients underwent preoperative neuroradiological evaluation with computed tomography (CT) and magnetic resonance image (MRI). Angiography was performed in 15 patients and preoperative embolization was done in 2 patients.

Results : Five far-lateral, 1 supracondylar and 10 midline suboccipital approaches were performed. Gross total removal was achieved in 15 cases (94%) and subtotal removal in 1 patient (6%). None of the patients required occipitocervical fusion. Radiological follow-up showed no recurrence in cases totally removed. Postoperative decrease of KPS scores was recorded in only 1 patient. The treatment of cervicomedullary solid hemangioblastoma presented particular issues : by preoperative embolization, we removed tumor totally without an excessive bleeding or brainstem injury. In one of foramen magnum meningioma, we carried out subtotal removal due to hard tumor consistency and encasement of neurovascular structures.

Conclusion : The choice of surgical approaches and the extent of bone resection should be defined according to the location and size of individual tumors. Moreover, we emphasize that preoperative neuroradiological evaluations on presumptive tumor type could be helpful to the surgeon in tailoring the technique and providing the required exposure for different lesions, without unnecessary surgical steps.

KEY WORDS : Benign tumor · Complication · Craniovertebral junction · Neuroradiology · Surgical resection.

INTRODUCTION

Surgical management of craniovertebral junction (CVJ) tumor is challenging for the neurosurgeon. Nevertheless, several surgical approaches to CVJ tumors have been recently developed, with the aim of reducing operative mortality and morbidity¹⁸⁾. According to the previous literatures, the most frequent neoplastic lesions of the CVJ are meningiomas, neurinomas, chordomas, paragangliomas, epidermoids, dermoids and chondrosarcomas²⁰⁾. There were so many factors influencing treatment of CVJ lesions including etiology, the

location of the lesion, and the growth pattern of the individual¹⁸⁾. There were also various routes to access the CVJ : anterior, anterolateral, lateral, posterolateral, posterior approaches¹⁸⁾. Because these lesions have a different bony destruction and neurovascular structure involvement such as the vertebralbasilar system and lower cranial nerves, tailored surgical approaches and preoperative neuroradiologic evaluation with computed tomography (CT) scan, magnetic resonance imaging (MRI), digital subtraction angiography (DSA) are necessary^{7,8,18)}. We report our surgical experience for benign CVJ tumors in a view of histopathological diagnosis, clinicoradiologic findings, and adequate surgical techniques.

MATERIALS AND METHODS

Patients and populations

Between January 2003 and March 2008, 1,341 patients with intracranial tumors were treated using surgical resection in

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our hospital. Among these patients, with the exception of jugular foramen tumors, only 16 cases were diagnosed with benign CVJ tumors by means of radiological studies and their surgical records (the incidence rate : 1.2%). The medical record and operative videos of these 16 patients with benign CVJ tumors were retrospectively reviewed and further analysis was undertaken of their neuroradiological and histopathological findings.

Evaluation of data

To define the clinical characteristics of the patients with benign CVJ tumors, clinical data such as the age, sex, presenting symptoms and neurological signs, symptom duration, and the Karnofsky Performance Scale (KPS) score were evaluated. We also reviewed the preoperative MRI and CT scans, focused on the exact location and neurovascular encasement by the tumors. We performed DSA for 5 foramen meningioma, 6 hemangioblastoma, 1 accessory nerve schwannoma, 1 hypoglossal nerve schwannoma, and 1 C2 root schwannoma, 1 cavernous hemangioma. In these cases, we evaluated more detailed information about supplying and drainage vascular system, and the relationship between the tumor and major artery or its branches.

Various surgical approaches were attempted, based on the preoperative diagnosis, exact location and size, and neurovascular encasement by the lesion. Through the review of the

surgical and operative videos, we evaluated the operative findings, such as the adherence of the tumor to the peritumoral structures, the accidental neurovascular injury. We also tried to find out the surgical pitfall to improve the access to the lesion or reduce unexpected injury of the important structures. The extent of tumor removal was classified as gross total removal, subtotal removal, and partial removal. Total removal was defined as complete resection of the tumor mass. Subtotal removal was defined as resection of the tumor mass with small tumor remnants left behind at important structures, such as the vertebrobasilar system and its perforating arteries, cranial nerves or brain stem. Partial resection was defined as incomplete resection of the tumor mass.

Operation related complications and the KPS score at discharge and the last follow-up visit were assessed. Each patient underwent enhanced CT scan immediately after operation and gadolinium-enhanced MRI was checked 3 and 12 months postoperatively. After then, gadolinium-enhanced MRI follow-up was achieved annually. Using the postoperative imaging studies, we confirmed the extent of removal and recurrence of treated lesion.

RESULTS

The information of the individual patients is shown in Table 1. Six patients presented with a foramen magnum meningioma,

Table 1. Demographic data of the patients

| No | Age/Sex | Preop KPS | Diagnosis | Approach | Resection | Postop KPS | Cx. |
|----|---------|-----------|----------------------|--|------------|------------|--------------------|
| 1 | 55/F | 80 | FM meningioma | Far lateral SOC | Simpson II | 100 | - |
| 2 | 70/F | 100 | FM meningioma | Far lateral SOC | Simpson II | 100 | - |
| 3 | 65/F | 90 | FM meningioma | Far lateral SOC | Simpson IV | 80 | Hemiparesis (mild) |
| 4 | 64/F | 90 | FM meningioma | Far lateral SOC | Simpson II | 90 | - |
| 5 | 61/F | 60 | FM meningioma | Midline SOC + C1 laminectomy | Simpson II | 70 | - |
| 6 | 72/F | 90 | FM meningioma | Midline SOC + C1 laminectomy | Simpson I | 100 | - |
| 7 | 70/F | 100 | XI schwannoma | Far lateral SOC | GTR | 100 | - |
| 8 | 30/M | 90 | XII schwannoma | Supracondylar | GTR | 90 | - |
| 9 | 24/F | 50 | Hemangioblastoma | Midline SOC + C1 laminectomy | GTR | 100 | - |
| 10 | 55/M | 100 | Hemangioblastoma | Midline SOC + C1 laminectomy | GTR | 100 | - |
| 11 | 19/M | 100 | Hemangioblastoma | Midline SOC | GTR | 100 | - |
| 12 | 21/F | 100 | Hemangioblastoma | Midline SOC | GTR | 100 | - |
| 13 | 35/F | 100 | Hemangioblastoma VHL | Midline SOC | GTR | 100 | Pneumothorax |
| 14 | 37/F | 50 | Hemangioblastoma VHL | Midline SOC (postop ETV) (preop EVD) | GTR | 50 | Paraplegia |
| 15 | 21/F | 70 | C2 root neurofibroma | Midline SOC + C1, 2 laminectomy | GTR | 100 | - |
| 16 | 28/M | 80 | Medullary cavernoma | Midline SOC | GTR | 100 | CSF leakage |

Cx. : Complication, EVD : extraventricular drainage, ETV : endoscopic 3rd ventriculostomy, FM : Foramen magnum, GTR : gross total resection, KPS : Karnofsky Performance Scale score, SOC : suboccipital craniotomy, VHL : Von Hippel-Lindau syndrome

6 with a cervicomedullary hemangioblastoma, 2 with a lower cranial nerve schwannoma (XI, XII), and each one case with a C2 root neurofibroma, medullary cavernoma respectively. The mean age was 45.4 years (range, 19-70 years). Female sex predominated by a factor of 2 : 1 (11 female and 5 male patients). The mean follow-up was 27.2 months. At admission, headache or posterior neck pain was observed in 9 patients (56%), motor and sensory change in 6 (38%), lower cranial nerve related symptoms in 3 (19%), deterioration of mental status in 1 (6%), intractable hiccup in 1 (6%), and one patient was detected incidentally on MRI for concomitant cerebral infarction. Out of 16 patients, preoperative embolization was done in 2 patients.

Patients have been operated through the following approaches : 10 midline suboccipital craniotomy (SOC), 5 far-lateral SOC, 1 supracondylar approach. Midline SOC was performed in all 6 hemangioblastoma, 2 posterior-type foramen magnum meningioma, C2 root neurofibroma, and medullary cavernoma cases. The laminectomy of upper cervical spine was added to conventional midline SOC in 5 cases, with definitive downward extension of mass into the upper cervical region on preoperative MRI. In 6 foramen meningioma patients, far lateral SOC was performed in 4 ventral type cases (Fig. 1) and midline SOC with C1 laminectomy in 2 posterior type cases (Fig. 2). Drilling of posterior one-third of the occipital condyle was done with 4 far-

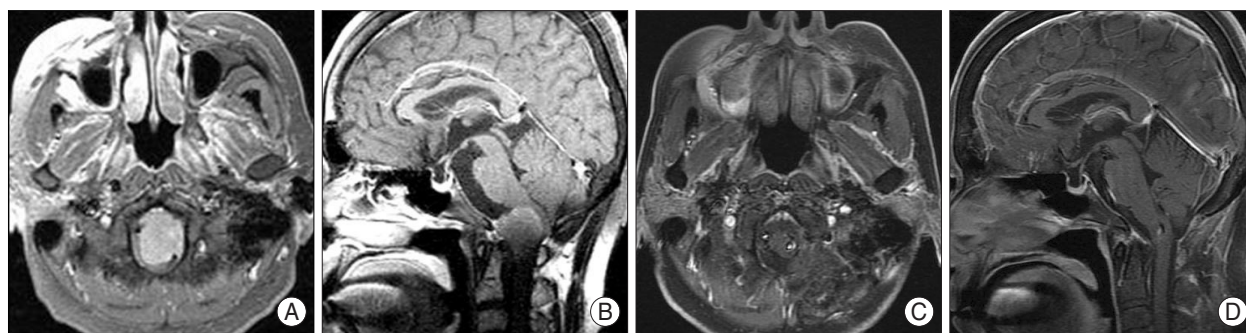


Fig. 1. Pre- and postoperative radiological findings in ventral type foramen magnum meningioma. Preoperative MRI showed a 2.5×2 cm sized extra-axial enhancing mass in cervicomedullary junction with displacement of adjacent brainstem (A and B). There was no recurrence during 4 years after surgery (C and D).

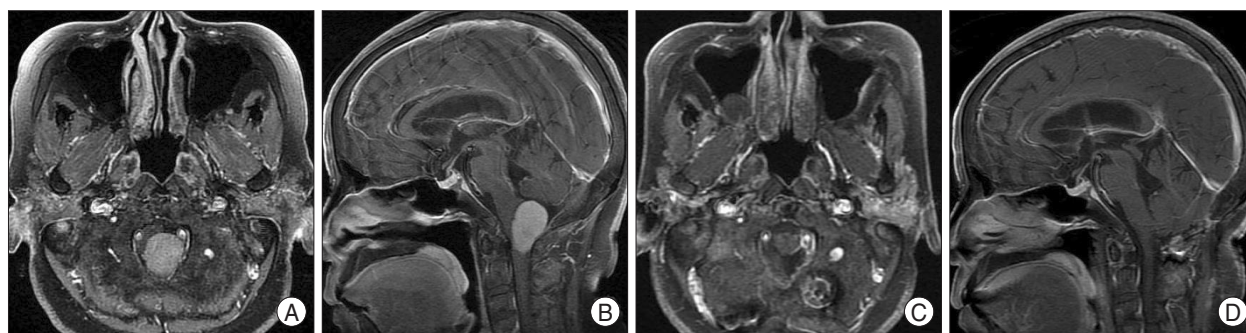


Fig. 2. Pre- and postoperative radiological findings in posterior type foramen magnum meningioma. Preoperative MRI showed a 2×3 cm sized extra-axial homogenous enhancing mass in posterolateral portion of foramen magnum with displacement of cervical spinal cord, medulla anteriorly (A and B). There was no recurrence during 2 years after surgery (C and D).

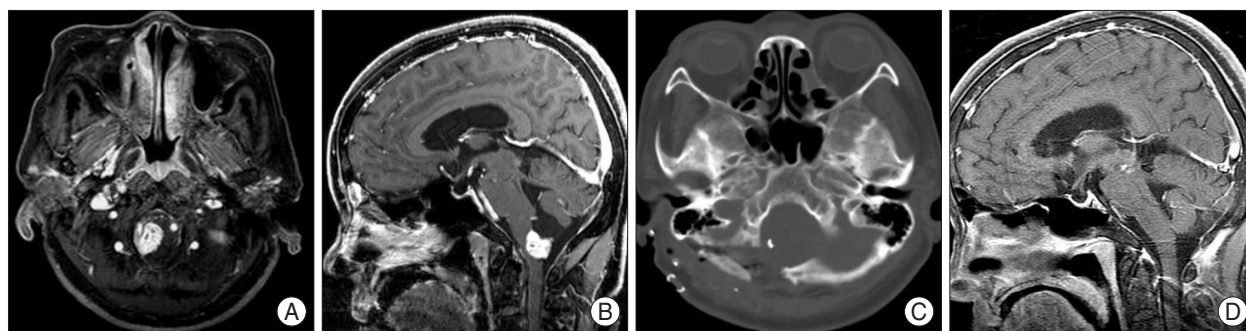


Fig. 3. Pre- and postoperative radiological findings in accessory nerve schwannoma. Preoperative MRI showed a 2.5×3 cm sized heterogenous enhancing mass in cervicomedullary junction (A and B). Postoperative CT scan showed evidence of right suboccipital craniotomy (C). There was no recurrence during 3 years after surgery (D).

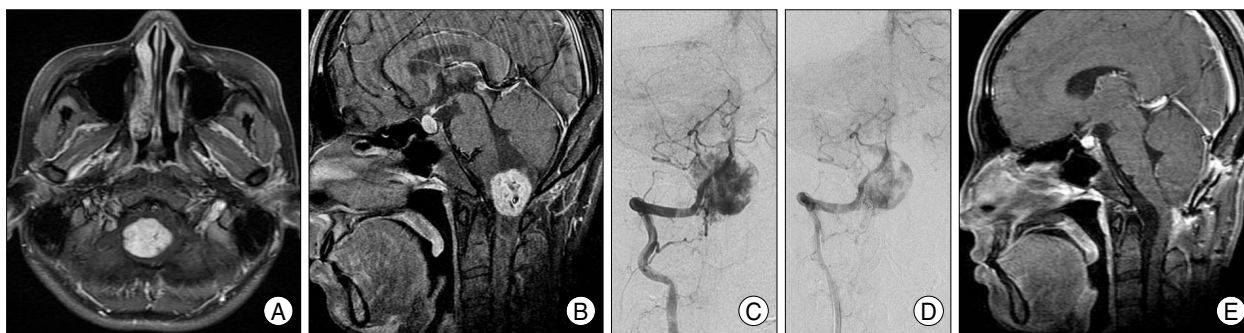


Fig. 4. Pre- and postoperative radiological findings in hemangioblastoma. Preoperative MRI showed a 2.5×3.5 cm sized extra-axial heterogenous enhancing mass in cervicomedullary junction (A and B). Preoperative embolization of main feeding artery by using gelfoam, and microcoils was achieved. The vascularity was reduced markedly. (C and D). There was no recurrence during 3 years after surgery (E).

lateral SOC. One hypoglossal nerve schwannoma patient underwent supracondylar approach, and 1 accessory nerve schwannoma underwent far lateral approach (Fig. 3). The others (6 hemangioblastoma, C2 root neurofibroma, and medullary cavernoma) underwent midline SOC with/ without C1 laminectomy (Fig. 4). Selection of either midline SOC with/without C1 laminectomy or far lateral SOC depended on the extent of the tumors in CVJ.

Gross total removal was achieved in all patients except one. In one of foramen magnum meningioma, we carried out subtotal removal due to hard tumor consistency and encasement of neurovascular structures. We did not need any occipitocervical fusion. Postoperative decrease of KPS scores was recorded in only one patient. During follow up period, radiological exams found no sign of recurrence in cases with total removal of tumor. For the case with remnant, radiological exams showed no increasement of size.

DISCUSSION

CVJ contained several important neurovascular structures : medulla, vertebrasilar system, low cranial nerves (IX, X, XI and XII)⁸. Therefore, tumors involving these structures have been considered hard to achieve complete resection and minimize postoperative morbidity²⁰. Standard preoperative work-up such as CT scan, MRI, and/or angiography is needed. On MRI, gadolinium-enhanced sequences help to precisely delimit the dural attachment zone, the tumor, and its relation to neural and vascular structures. On T2-weighted images, the presence of an arachnoid plane between the tumor and the neuraxis is sometimes visible. Bone windows CT scan is helpful in case of extradural extension to investigate bone erosion and to schedule preoperatively the need for fusion⁴. Conventional angiography is not necessary in all patients but useful in follows, embolization of highly vascularized tumor, a balloon occlusion test in case of vertebral artery (VA) encasement⁴. Intraoperative neurophysiological monitorings also

have been used for preventing injury of the neural structures including somatosensory evoked potentials, brainstem auditory evoked potentials, and electromyographic monitorings of low cranial nerves by recordings through an endotracheal tube (CN X), and with a needle in the sternomastoid muscle (CN XI) and tongue (CN XII)³.

Meningiomas locating in the region of the foramen magnum poses particular problems, because they originate anterior to the brain stem or demonstrate significant anterior extension involving several vital structures. A number of surgical approaches have been used in the past to expose and remove completely such tumors. A transoral approach has been used for ventral type meningiomas, but the surgical field may be contaminated and limited laterally^{18,19}. Because of these limitations of anterior or anterolateral approaches, lateral approaches are most widely used to remove meningiomas anterior to the brain stem. Hammon and Heros proposed a lateral SOC that was used as an initial attempt to perform a more lateral suboccipital exposure for vertebrasilar aneurysms^{12,13}. Since then, the extreme lateral craniocervical approach has been progressively developed as a more and more lateral approach to provide access to lesions in the midline on the anterior aspect of the foramen magnum^{1,2,9,23}. The anatomical structures limiting the surgeon laterally are the occipital condyle, the jugular bulb, the sigmoid sinus, and the vertebral artery²². The surgical windows for the surgeon are sufficient once a standard lateral suboccipital craniotomy is combined with drilling the posterior third of the occipital condyle^{2,13,15,18}. In our study, we also performed far lateral suboccipital craniotomy with four ventral type meningioma patients. One meningioma case with encasement of vertebral artery was likely to be resected incompletely. Many authors described encasement of the vertebral artery as a limiting factor for removal of craniocervical meningiomas^{10,11,24}.

Hemangioblastomas can occur throughout CNS system that originate primarily in the cerebellum (83-95%), spinal cord (3.2-13%), and medulla oblongata (2.1%)⁶. Hemang-

ioloblastomas are benign slow-growing neoplasms that carry a significant operative risk²⁵. Surgeons have been encountered severe problems during surgery because of the risk of massive bleeding¹⁶. Some authors described that coagulation of the dominant and the other feeders first followed by shrinking the tumor and finally coagulating the drain veins was a very important surgical strategy for successful and safe removal of these highly vascularized intramedullary tumors²⁵. So most of the authors suggested that digital subtraction angiography (DSA) prior to the operation was necessary to understand the relationship between the feeding arteries and drain veins associated with the hemangioblastomas¹⁶. In our series, we performed DSA in all hemangioblastoma patients including spinal angiography in patients with VHL syndrome. Two hemangioblastoma patients were also treated with embolization of feeding arteries to prevent the massive bleeding during the surgery. A midline SOC with/without removal of the posterior arch of C1 was undertaken. After the dural opening, we could perform exact midline myelotomy with confirming the obex displaced laterally by hemangioblastoma compression. The mass was removed en bloc after isolating the nidus. T1- and T2-weighted MRI was performed in all patients for the preoperative planning, screening and follow-up of the patients. There were no surgery related complications including Ondine's curse so called central sleep apnea and neurogenic hypertension induced by the pathologic affecting the nucleus tractus solitarius in the brainstem²⁵.

Schwannomas are benign tumors that constitute approximately 8% of all primary intracranial tumors²¹. Of these, schwannomas arising from low cranial nerves are extremely rare¹⁴. Recently, Bulsara et al.⁵ proposed three tumor types according to the imaging result as follows : Type A, intradural tumor, Type B, dumbbell-shaped tumor; and Type C, dumbbell-shaped tumor with high cervical extension. They also recommended different types of surgical approaches considering extent of the tumors⁵. In our series, we performed far-lateral SOC in accessory nerve schwannoma, and supracondylar approach in hypoglossal nerve schwannoma. We achieved gross total resection but, hypoglossal nerve palsy was remained in the latter patient. Especially, in the tumors originating from the cranial nerves, appropriate surgical strategies and careful precautions were needed for the prevention of the injury of the adjacent cranial nerves and vascular structures.

Cavernomas placed in deep nuclei and in the brainstem were considered inoperable diseases. However, recently many surgical indications have been accepted in cavernomas which the location was near the pial surface, and has a mass effect or significant hemorrhagic evidences¹⁷. In this study, 30 year-old male suffered from intractable hiccup and left hemiparesis was admitted. MRI revealed cavernous angioma with he-

morrhage in left side of cervicomedullary junction. We performed midline SOC and achieved gross total resection of cavernoma without any surgical related complications except transient CSF leakage treated by lumbar drainage. The patient got fully recovered motor activity and was free from intractable hiccup a few days after surgery. Removal of the lesion was achieved by making small parenchymal opening and initial reduction of the lesion by evacuation of the hematoma, shrinking it by coagulation and limited decompression to permit removal of the lesion in one or several pieces¹⁷.

CONCLUSION

The selection of surgical approaches and the extent of bone resection should be defined according to the location and size of individual tumors. We found the approaches, such as a lateral SOC or midline SOC combined with appropriate laminectomies, to be sufficient for tumor removal in most CVJ benign tumors. For ventrally located tumors, drilling the posterior third of the condyle provides sufficient additional space to access. As removing the tumor, a "surgical space" was created, allowing feasible access to more extensive area. Moreover, we emphasize that preoperative neuroradiological evaluations on presumptive tumor type could be helpful to the surgeon in tailoring the technique and providing the required exposure for different lesions, without unnecessary surgical procedures.

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