Case Report

Schwannoma of the upper cervical spine—a case report

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Abstract

Upper cervical schwannoma is rare, and belongs to benign tumors that is usually asymptomatic. It accounted for only ten percent of schwannomas cases and mostly occurs in 40–50 years old patients. Aggressive and total resection is the treatment of choice for this tumor. Preoperative diagnosis is difficult, relying on clinical suspicion, and confirmed by surgical pathology. We report a 54-year-old male patient with chief complain of progressive weakness and numbness of his right arm for four months. He had a history of lymph node tumor in 2007 and excised in 2011. Neurological decrease was found on the right arm. The radiographic examination showed lytic lesion on the second, third, and fourth cervical spine. Computed tomography scan showed destruction extending to the first cervical vertebra. Capsulated extradural and extramedullary mass and compression to the spinal cord was found from the magnetic resonance imaging. Two-stage operation was performed. The first stage was posterior decompression with occipitocervical fusion and instrumentation, while the second was anterior decompression and iliac strut graft. Both operations performed using the surgical ultrasonic dissector. During three months’ follow-up, it showed neurological improving. Neurological deficit appears when there is compression on the spinal cord. Total resection is the treatment of choice for these tumors. Preoperative imaging should be performed to exclude malignant tumor and found tumor extension.

Introduction

Spinal schwannomas are rare, locate at the intradural spinal cord in adults of 40–50 years old, with the incidence around 0.3–0.4 cases/100,000 persons per year, involving both male and female. It is a benign nerve sheath tumors from schwann cells. The incidence in upper cervical spine accounted for 0.1% of all schwannomas. Intraosseous schwannoma accounted for less than 0.2% of primary bone tumors. Non-intraosseous spinal schwannoma is most commonly found in lumbar and thoracic regions, while cervical part is the least. Usually the patient showed no symptoms. Radiating pain, sensory deficit, motor weakness, and paresis are the signs of spinal cord compression. The tumour originated from nerve root or foramen.

In 2001, Sridhar declared a classification system of benign spinal schwannoma. In this classification, only type V showed the intraosseous schwannoma. Intraspinal tumor with extraspinal component >2.5 cm is classified as giant schwannoma. Difficulty in resection is related to the tumour location, such as in extradural schwannoma and giant invasive schwannoma. Instability is frequently found following the removal of tumour from the vertebral body.

Macroscopically, schwannoma is shown as an encapsulated, solid or cystic spindle cell mesenchymal tumors, while microscopically it is composed of two cellular zones: Anthony type A, which arranges with spindle-shaped schwann cell densely and surrounding it, such as muscle, nerve, and artery.

The symptoms may be apparent in 1–84 months. The most common sign is radicular pain, followed by motor weakness, sensory deficit, and paresis.

Case report

A 54-years-old right-handed male, working as an electronic technician, was admitted in August 2017. He complained of difficulties in writing and fine coordination when performing his job.
Neurological examination discovered at below level C5 and numbness on level C4 bilaterally. The patient first noticed his symptom 6 months ago and has been worsening slowly. Four years ago, he had a history of slow-growing anterior neck mass. A general surgeon performed an excision procedure, which histologically was found as lymph node tumor. After removal of the tumor, he had no other symptom.

A plain radiograph showed lytic lesion on C2–C4 with solid margin and bone destruction on C3 and C4, with scalloping of the third cervical vertebra (Fig. 1A). Contrast-enhanced CT scan of the neck showed destruction on the first to the fourth cervical vertebra with feeding artery originating from the vertebral artery (Fig. 1B and 1C).

MRI showed an extradural extramedullary mass on C2–C5, 39.5 × 30.9 × 45.7 (mm) extend to intracanal region with scalloping destruction on neural foramina and extend to soft tissue posteriorly and anteriorly. The tumor mass also invaded nerve root and vertebrae with compression of the spinal cord to the left side and caused cord edema on C4–C5 (Fig. 1D and 1E).

Embolization and angiography were performed 4 days before operation to reduce blood flow and found that feeding artery diameter was 0.3 mm with stenosis of right vertebral artery on the third cervical vertebra and micro catheter was unable to pass through. Angiography discovered that the tumour compressed the vertebral artery on the third cervical vertebra with the smallest diameter of 0.42 mm, which was only 33% of the normal vertebral artery. Feeding artery found on level C2–C3 with the biggest diameter of 0.34 mm as shown in Fig. 1F.

We performed two-stage procedure, the first on December 2017. We performed posterior decompression and stabilization with instrumentation and fusion. A soft capsulated greyish mass lied on C2–C5, which was removed using the ultrasonic surgical aspirator. Cavitrone ultrasonic surgical aspirator (CUSA) was used to destroy tissue tumor by heat and cavitation. Macroscopically,

Fig. 1. (A) Plain radiograph showed lytic lesion on C2 and C4. Lateral view showed collapsed vertebra of C2–C3. (B) Enhanced CT showed destruction of the first to fourth cervical vertebra. Lateral view showed destruction of C1 and C4. (C) AP view showed destruction of right side of C2–C4. (D) Sagital MRI showed a solid encapsulated mass from C2–C4 on spinal canal and extended to the bone. (E) Coronal MRI showed mass compressing the spinal canal. (F) Angiograph showed feeding artery (short arrow) and basilar artery compressed by the mass and sized only one third of normal diameter (long arrow).
tumor tissue invading posterior side has been removed. Spinal canal was not intruded by the tumors and so no need to sacrifice any nerve. Vertebral artery was compressed by the tumor, but did not invade into the artery. After removal of the tumor, we fused the occipital to the seventh cervical vertebra using pedicle screws (Fig. 2).

During operation, we also took a mass for pathologic examination. Histopathology confirmed as a benign schwannoma with proliferation of schwann cell, spindle core, thin chromatin. Anthony A and Anthony B cells were also found (Fig. 3).

Three weeks after the first operation, we performed the anterior approach procedure. Initially, we planned mandible osteotomy to reach second and third cervical vertebra. Carotid pulse was palpated and the superior recurrent laryngeal nerve was identified and preserved. The prevertebral fascia was visible using a fine needle. The position was checked using image intensifier (Fig. 4A and 4B). The incision extended to submandibular region without mandible osteotomy. The tumor tissue located at anterior part was removed using the ultrasonic surgical aspirator. Iliac graft was taken as buttress on C2–C5 using anterior cervical plate (Fig. 4C and 4D). Anterior plate was used for buttress. The body of C2 and C3 was destroyed and could not hold tight with screws.

Postoperatively, the patient was put in Minerva cast for two months to provide cervical fusion. Postoperative evaluation showed an increase on the motoric and sensoric nerve system from 3/5 on the right side to 5/5 within 2 months. Plain radiology on the 2nd month evaluation found stable fixation without any loosening on screw (Fig. 5). The patient also started to work again in the third month.

MRI evaluation performed one year postoperatively to seek any recurrence of tumor. Patient already gained full motoric and sensoric nerve system and already back to work without any sequel. We performed brain CT and found normal (Fig. 6A). MRI found the mass on the paravertebral level of C2–C3 entering the foraminal canal (Fig. 6B).

Discussion

The case reported is a male patient with schwannoma in upper cervical region, categorized as giant schwannoma with the size of...
39.5 × 30.9 × 45.7 (mm) and extend to intracanal region. Difficulty in resection occurred due to its location at the extradural region and is a giant invaded schwannoma. The tumour already destroyed the vertebral body and extended from C1 to C4. Clinically neurological deficit was found on the right side of the trunk. Motor weakness was not obvious until the late stage. We performed plain radiograph, MRI and CT scan to evaluate the extension of destruction on the vertebral body. We found the tumor already compressing the spinal cord especially on C4–C5 and destruction of the vertebral body. The tumor extruded anteriorly and extended to C3. The tumour was found extended to extradural and extramedullary regions, while in the literature 70%–80% of spinal schwannomas are reported to be intradural. Preoperatively we planned on embolizing the tumor; unfortunately it was unsuccessful due to small caliber of the feeding arteries. Instead we directly performed posterior stabilization, decompression, and occipitocervical fusion. In order to get complete surgical removal of the mass, we performed two-stage operation. Dissection of the tumor was performed using cavitron ultrasound surgical aspirator.

Since instability frequently occurs following removal of the tumor invaded to the vertebra, we performed second stage operation to resect the tumor anteriorly. Two-stage operation was performed to prevent postoperative instability and recurrence.

Seven months after operation, the patient has already gained motor function and was capable of daily activity. Fusion on C2–C4 was found radiologically.

Fig. 4. The level was checked using image intensifier and the tumor was decompressed using cavitron ultrasonic aspirator. (A) Second-stage operation performed via anterior approach using CUSA®; (B) Reaching C2 using CUSA®; (C) Iliac graft; (D) Plate placement.

Fig. 5. Radiological evaluation of lateral view. (A) Two months evaluation, (B) Seven months evaluation, (C) one year evaluation.

Fig. 6. (A) No signs of metastatic tumor were found in brain on CT. (B) MRI of cervical vertebra showed the mass on the right paravertebral C2–C3.
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Nil.

Ethical statement

The patient has already given consent for the case reporting.

Conflicts of interest

The authors declare no conflicts of interest in this study.

References

1. Celli P, Trillo G, Ferrante L. Spinal extradural schwannoma. J Neurosurg Spine. 2005;2:447–456. https://doi.org/10.3171/spi.2005.2.4.0447.
2. Seppälä MT, Haltia MJ, Sankila RJ, et al. Long-term outcome after removal of spinal schwannoma: a clinicopathological study of 187 cases. J Neurosurg. 1995;83:621–626. https://doi.org/10.3171/jns.1995.83.4.0621.
3. Fawcett KJ, Dahlin DC. Neurilemmoma of bone. Am J Clin Pathol. 1967;47:759–766. https://doi.org/10.1093/ajcp/47.6.759.
4. Sridhar K, Ramamurthi R, Vasudevan MC, et al. Giant invasive spinal schwannomas: definition and surgical management. J Neurosurg. 2001;94:210–215.
5. Park SC, Chung SK, Choe G, et al. Spinal intraosseous schwannoma: a case report and review. J Korean Neurosurg Soc. 2009;46:403–408. https://doi.org/10.3340/jkns.2009.46.4.403.