Case report

Intradural extra-arachnoid schwannoma of the upper cervical spine

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A very rare case of intradural extra-arachnoid schwannoma involving the upper cervical spine is described. The tumour most likely originated from the extra-subarachnoid angle region, with its unusual pattern of expansion within the spinal canal resulting from such characteristics of the upper cervical region as a relatively wide spinal canal, an extensive ventral subarachnoid space, and membranous ventromedian and lateral anchoring structures which influenced the direction of tumour enlargement.

Keywords: schwannoma; cervical spine; intradural extra-arachnoid tumour

Introduction

Increasing numbers of patients with dumb-bell-shaped spinal cord tumour have been reported. We treated a patient with an unusual level of spinal involvement and configuration of the tumour on axial images.

Case report

A 57-year-old man presented to Tokai University Hospital with a 4-month history of progressive nuchal and shoulder pain and numbness of the fingertips of both hands. One month prior to admission he had been diagnosed with spinal cord tumour by magnetic resonance (MR) imaging at another institution.

Physical examination on admission revealed a slender individual with severe nuchal pain, mild gait disturbance, and moderate weakness of both upper and lower extremities. Examination of sensation showed incomplete involvement below the occipital area. Deep tendon reflexes were increased. Babinski and Chaddock signs were absent. Ankle clonus was observed bilaterally. He also gave a history consistent with neurogenic bladder, and anal sphincter tone was abnormal.

Radiographs revealed no abnormal widening of the cervical spinal canal. Sagittal MRI showed a large anteriorly located mass producing significant compression of the spinal cord at C1–2. The mass included an isointense area surrounded by low intensity on T1-weighted images; T2-weighted images showed reversal of these intensities. The cervical spinal cord was displaced rightward and posteriorly. Enhancement after gadolinium contrast administration was ob-

served in a wide zone along the margin of the tumour on axial images (Figure 1a, b).

Lumbar puncture at L2–3 yielded clear cerebrospinal fluid. The concentration of protein was 54 mg/dl; the glucose concentration was normal. A Queckenstedt test gave normal findings, as did the cell count. Myelography demonstrated slight compression of the spinal cord by the tumour at C1–2. Computed tomographic myelography also showed an anteriorly located mass (Figure 2). A vertebral angiogram disclosed no abnormal shift of the vertebral artery.

We could not determine from images whether the tumour was located in the epidural space or in the intradural extra-arachnoid space.

After sudden neurologic deterioration, the patient underwent *en bloc* laminectomy from C1 to C2. The left second cervical root was swollen, and a part of the mass was seen to extend toward the epidural space. After we opened the dura mater, we observed the exposed dorsal arachnoid membrane and opened it longitudinally. Right posterolateral displacement of the spinal cord was evident. No tumour was seen within the subarachnoid space (Figure 3a). With the cord medially retracted, the ventral arachnoid membrane was visible without any perforation. We could then detect the ventral intradural extra-arachnoid space contained a large tumour after making an incision of the ventral arachnoid membrane (Figure 3b). A pair of an inlet and outlet of the associated spinal nerve root was cut inside the intradural extra-arachnoid space, after which the tumour could be removed (Figure 3c). The cutting portion of the inlet of the spinal nerve root inside the intradural extra-arachnoid space was attended by vessels, but it seemed to show a normal appearance.

Further evaluation of the proximal portion of the spinal nerve root was performed and we could also observe normal appearance of it inside the intra-

Figure 1 T1-weighted MR images after gadolinium contrast administration. (a) On sagittal and (b) axial views, the mass exhibited marked ringlike enhancement and a regular margin. Axial imaging showed the cervical spinal cord to be displaced posterolaterally to the right by the tumour

arachnoid space just beside the spinal cord. This means that this case is not a straight forward intra-arachnoid extramedullary tumour. Complete resection of the remaining associated left C2 root was meticulously performed around the far lateral portion of the spinal canal. After these procedures, the ventral aspect of the dura mater was clearly visible.

Grossly, the tumour measured $12 \times 7 \times 7$ mm and contained a cystic area, possibly representing an area of old hemorrhage or necrosis. Histopathologic examination showed a schwannoma with predominantly Antoni type A histology, with high cellularity and growth of tumour cells in interlacing bundles with nuclear palisading. Furthermore, it was suspected that the origin of the tumour was from the ventral spinal root, because of no ganglion cells and a lot of myelinated fibres (Figure 4). Postoperatively, muscle strength recovered immediately and examination after 1 year revealed no limitation of neck motion, no sensory deficits and an extremely stable gait.

Enhanced MR images 3 years after the procedure showed no abnormal tumour shadow and the authors confirmed that the associated spinal nerve root in the subarachnoid space was not preoperatively influenced by the tumour tissue and the origin of the tumour was from the intradural extra-arachnoid space (Figure 5).

Discussion

An intradural extra-arachnoid spinal cord tumour such as we present here has not been reported to date. The tumour had a dumb-bell configuration; tumours of such shape are reported most frequently in the cervical spine: 44.8% of cases in one series by Honma,⁷ although Hagiwara⁵ has reported a lower incidence of these tumours in the upper cervical spine (5.6%).

Figure 2 Computed tomographic myelogram after subarachnoid injection of contrast material showed the posterolaterally displaced cervical spinal cord at C1–C2 and contrast medium between the mass and the spinal cord (arrow)

Figure 3 Intraoperative photographs. (a) Just after an incision of the dorsal dura mater and dorsal arachnoid. The subarachnoid space contained no tumour. [Ventral arachnoid (←), Dorsal dura mater and arachnoid (⇐), Spinal cord (*)] (b) After an incision of the ventral arachnoid. The mass was evident within the ventral intradural extra-arachnoid space. [Ventrolateral arachnoid (←), Tumour (⇔), Spinal cord (*)] (c) The tumour, removed *en bloc*

According to the Eden classification,⁶ our patient represents the type 1 category, which is not common in the cervical area. We therefore consider this a rare case as to both level of the spinal involvement and axial distribution of the tumour.

Location of the tumour was of great concern in the initial states of surgery. The operative field presented here, suggested the strong likelihood of occurrence of a tumour originating from the extra-subarachnoid angle region. The dorsal root ganglia in the upper cervical area are located more proximally than at more caudal spinal levels due to such anatomic characteristics as a wider spinal canal and smaller intervertebral foramina, which lead to tumour growth inside the canal. Wide range of the extra-subarachnoid angle

Figure 4 Microscopically, the surgical specimen was a typical schwannoma showing an Antoni A pattern composed of interlacing bundles of spindle cells with forming palisades nuclei. No ganglion cells were observed. (Original magnification, × 50; hematoxylin and eosin)

Figure 5 Postoperative enhanced MRI images showed no abnormal tumourous shadow

Figure 6 (a) Tumour growth pattern. The tumour originated from the extra-subarachnoid angle region. (b) The tumour grew within the spinal canal and a part of the mass extended toward the epidural space. (c) Expansion into the intradural extra-arachnoid space without perforation of the arachnoid. (d) Significant compression and posterolateral displacement of the spinal cord ultimately occurred

region inside the canal was also contributory to the unusual pattern of tumour expansion reflected by these anatomic features (Figure 6a–d).

Ventromedial extension of the tumour was likely because of the large capacity of the ventral subarachnoid space in the upper cervical area. Furthermore, the membranous anchoring structures at the median ventral aspect of the cord, the dentate ligament which serves as a lateral anchor for the spinal nerve roots, and the lateral arachnoid membrane probably directed tumour expansion in an unfamiliar direction. As indicated by Rickenbaucher,⁴ Tokunaga,³ and Nauta,¹ such structures originate from the pia mater, perforate the arachnoid membrane, and attach to the dura mater (Figure 7a, b). Such architecture is characteristic of the upper cervical area, although its functional significance is unclear. On the basis of these references, we concluded that the intradural extra-arachnoid tumour expanded anteriorly from the extra-subarachnoid angle region without perforation of the ventral portion of the arachnoid membrane lying between the anchoring structures.

Figure 7 (a) Coronal and (b) axial diagrammatic views of the upper cervical spinal cord. The upper cervical area shows a characteristic ventromedial membranous anchoring structure originating from the pia mater, perforating the arachnoid, and attaching to the dura²

References

1. Nauta HJW, Dolan E, Yasargil MG. Microsurgical anatomy of spinal subarachnoid space. *Surg Neurol* 1983; **19**: 431–437.
2. Lang J, Emminger A. Über die Textur des Ligamentum denticulatum und der Pia matter spinalis. *Z Anat Entwickl* 1963; **123**: 505–522.
3. Tokunga A. Spinal meninges. *Spine & Spinal Cord* 1989; **2**: 921–923.
4. Rickenbacher J, Landoli AM, Theiler K. Einbau des Rückenmark in den Wirbelkanal. In: *Praktische Anatomie, II/7* (ed by Lanz Tv, Wachsmuth W). Springer-Verlag, Berlin, 1982; 235–254.
5. Hagiwara M. A case of dumb-bell tumor of the cervical spinal cord originated from greater occipital nerve. *Kanto Journal of Orthopedics and Traumatology* 1988; **19**: 683–687.
6. Eden K. The dumb-bell tumors of the spine. *Brit J Surg* 1940; **28**: 549–470.
7. Honma G. Result of surgical treatment of spinal cord dumb-bell tumors. *Journal of Orthopedics and Traumatology* 1989; **32**: 1513–1521.