Cervical Intramedullary Cavernous Angioma: A Report of 3 Surgical Case

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ABSTRACT

The detection rate of spinal intramedullary cavernous angioma (IMCA) has increased with the increasing use of magnetic resonance imaging (MRI) techniques. The risk of progressive deterioration after serial hemorrhagic events is well recognized, but the indications for surgical resection of spinal IMCA are controversial. We present three consecutive cases of surgically treated cervical IMCA. Two patients had obvious signs of sensorimotor deficit corresponding to the level of the lesion. The other patient presented with frequent episodes of dizziness and neck pain. T2-weighted MRI showed characteristic hemosiderin deposits in all 3 cases. The cervical IMCAs in this series were located on the dorsal surface of the spinal cord.

Areas of discoloration (hemosiderin deposits) were easily identified on the surface of the spinal cord in all three cases, glial planes were identified between the lesions and spinal cord. Total resection of the IMCAs was achieved by careful microsurgical dissection under intra-operative monitoring. The Valsalva maneuver was used to check for residual vascular lesions.

When dorsal cervical IMCAs are identified by MRI, surgical treatment should be considered to prevent catastrophic deterioration due to recurrent hemorrhage.

Key words: spinal cavernous angioma, cervical cord, intramedullary
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Introduction

Cavernous angiomas of the central nervous system occur most commonly in the cerebral hemispheres or brain stem, but the spine may also be affected [2,8,9,11,13,14,18]. With the increasing use of MRI, the detection rate of intramedullary cavernous angioma (IMCA) has increased in the spine as well as in the brain [3,4,6,13]. Spinal IMCAs are easily diagnosed by the presence of characteristic hemosiderin deposits that cause low signal intensity on both T1- and T2-weighted MR images [2,11].

Although several authors have reported the risk of progressive deterioration after recurrent hemorrhagic events, the indications for surgical resection of spinal IMCA are controversial, especially in cases with minor symptoms [9,13,17]. Aggressive surgical treatment has been controversial due to the uncertain of natural history of untreated IMCA and the risk of manipulation of the cervical intramedullary cord.

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Table 1: Summary of 3 patients with cervical intramedullary cavernous angioma

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Spinal Level</th>
<th>Clinical Presentation</th>
<th>MRI findings</th>
<th>Gd effect</th>
<th>Follow up Period</th>
<th>Surgery</th>
<th>Outcomes</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>37</td>
<td>M</td>
<td>C5</td>
<td>3 yrs of painful dysesthesia in his right hand. Twice episodes of severe neck pain after coughing.</td>
<td>Iso-intensity</td>
<td>High-intensity with low-intensity rim</td>
<td>No enhancement</td>
<td>63 mos</td>
<td>Total resection</td>
</tr>
<tr>
<td>2</td>
<td>57</td>
<td>F</td>
<td>C1-C2</td>
<td>Acute onset of moderate hemiparesis and severe deep sensory impairment.</td>
<td>High-intensity (moderately high)</td>
<td>High-intensity with low-intensity rim</td>
<td>No enhancement</td>
<td>75 mos</td>
<td>Total resection</td>
</tr>
<tr>
<td>3</td>
<td>70</td>
<td>M</td>
<td>C4</td>
<td>Repeated dizziness attack. No neurological deficits.</td>
<td>Low-intensity with high-intensity rim</td>
<td>High-intensity with low-intensity rim</td>
<td>Slightly enhanced</td>
<td>27 mos</td>
<td>Total resection</td>
</tr>
</tbody>
</table>

We present our experience with three consecutive cases of cervical IMCA treated surgically, with discussion of the timing and indications for surgery.

Cases

**< Case 1 >**

A 37-year-old male presented with a 3-year history of painful dysesthesia in his right hand and an episode of neck pain in the previous year. He was referred to our hospital with severe neck pain triggered by coughing. Neurological examination revealed moderate deep sensory loss in his right arm and intractable painful dysesthesia in his lower arm and hand. MRI showed a typical cervical IMCA at C5. We considered that recurrent minor hemorrhages had occurred and decided upon surgical treatment. After an osteoplastic laminoplasty of C3-5 and opening the dura mater, an area of discoloration was easily identified on the dorsal surface of the spinal cord. Midline myelotomy was performed and a berry shaped hemangioma was exposed. With careful dissection along the glial plane, the intramedullary cavernoma was totally removed. We confirmed complete resection by means of the Valsalva maneuver. MEP and SEP monitoring were stable during surgery. His neurological condition did not deteriorate after surgery, and the painful dysesthesia in his right lower arm had gradually improved. He was able to return to his former job.

**< Case 2 >**

A 57-year-old female had an acute episode of motor weakness in her right arm and leg. Neurological examination on admission revealed right hemiparesis (arm MMT 4/5, leg MMT 3/5) and severe hemi-sensory impairment. MRI demonstrated a round exophytic non-enhanced mass and a moderately compressed spinal cord at C2 level. Vertebral angiography did not detect any vascular lesions. We suspected that the bleeding site was a spinal arteriovenous malformation or a vascular neoplasm. To prevent recurrent bleeding, surgical exploration was performed 2 weeks after this hemorrhagic episode. The vascular lesion was easily accessed by C2 hemilaminectomy and was identified through the thickened arachnoid membrane. A red berry-like tumor protruded from the dorsal surface of
Fig. 1: Case 1. Magnetic resonance images of intramedullary cavernoma located at C5. Sagittal (A) and axial (C) T1-weighted images showing iso-intensity mass. Sagittal (B) and axial (D) T2-weighted images showing small high-intensity mass with low-intensity rim. Gd-DTPA image (E) showing no enhancement. Intraoperative photograph (F). The spinal cord was minimally widened and the discoloration on the dorsal surface of the cord was apparent. After small myelotomy, berry-like shaped hemangioma was exposed.

Fig. 2: Case 2. Magnetic resonance images of intramedullary cavernoma located at C2. The large part of lesion was dorsally exophytic. Sagittal (A) and axial (C) T1-weighted images showing moderate-high-intensity mass. Sagittal (B) and axial (D) T2-weighted images showing high-intensity mass with localized low-intensity rim. Gd-DTPA image (E) showing no enhancement. Intraoperative photograph (F). The discoloration on the dorsal surface of the spinal cord with thickened arachnoid membrane. Red colored berry-like tumor was protruded from dorsal surface.
the spinal cord. A cavernous angioma was seen beneath the C2 dorsal root entry zone. The margin was clear and the cleavage plane was obvious. The lesion was completely excised from the spinal cord by dissection in the surrounding plane of hemosiderin-stained gliotic tissue. Her hemiparesis was unchanged in the immediate postoperative period. Her neurological symptoms then gradually improved. She was able to walk with a cane and her sensory impairment was much reduced.

< Case 3 >

A 70-year old male experienced several episodes of sudden dizziness during the past year. His dizziness was accompanied by a sensation of occipital head heaviness. Neurological examination did not show any objective sensorimotor signs. MRI revealed an intramedullary cavernous angioma with a hemosiderin rim at C4 level. We could not determine whether or not his dizziness was related to his cervical tumor. He gave informed consent to surgical treatment to prevent bleeding from his angioma. He underwent a C3-5 laminoplasty. After the dura and thickened arachnoid membrane were opened, a blue mulberry-shaped angioma was seen on the dorsal aspect of the spinal cord. A dilated posterior median vein ran over the posterior surface of the cavernoma in the midline. Taking care not to sacrifice this vein, sharp dissection was confined to the thin plane of gliotic, hemosiderin-stained tissue surrounding the angiomas. The lesion was removed in an inside-out fashion until gliotic white matter was identified. The Valsalva maneuver was performed to check that the cavernous angioma had been removed. The residual lesion was projected from the deep tissues, and the lesion was thoroughly resected.

Ultimately the spinal cord was decompressed but no definite malformation or other lesion was identified with the Valsalva maneuver repeated three times. The patient’s occipital headache and dizziness were relieved postoperatively. Two years later he was neurologically intact.

Discussion

Although the timing and indication of surgical treatment for cervical IMCAs is controversial [1,4,13,16], some patients who refused surgical treatment have later progressively deteriorated following multiple hemorrhagic episodes, resulting in major neurological deficits. [6,9,13]. Neurosurgeons tend to procrastinate about surgery because of the trivial symptoms that occur in some patients with an episode of minor bleeding from a cervical IMCA. In the past, patients with an asymptomatic cervical IMCA found on MRI were managed conservatively, and followed up with serial examinations. [1]. Patients with spontaneously resolving symptoms after an initial bleed from an IMCA should be treated surgically to avoid further hemorrhage [3,5,7,13,17]. Recurrence of hemorrhage can diminish the chances of recovery because of worsening preoperative neurological condition.

Ogilvy et al [13], classified spinal IMCAs into four types based on their clinical presentation: 1) a discrete episode of neurological deterioration with a varying degree of recovery between episodes, 2) slowly progressive neurological decline, 3) acute onset of symptoms with rapid decline, and 4) acute onset of mild symptoms with subsequent gradual decline lasting weeks and months. We believe that the latter two groups should be surgically treated as soon as possible. The former two groups should be considered for surgery if they suffer from a progressive or step-wise deterioration of spinal cord function. The recommended timing of surgery is as follows: cases with progressive deterioration should be immediately treated by surgery, and cases with stable symptoms should be operated upon within two or three weeks of an initial hemorrhage as is the case with intracranial vascular malformation.

Koyama [10] analyzed a large series of spinal tumors, and reported that the most of spinal intramedullary hemorrhages were due to cavernous angiomas which are unlikely to cause massive bleeding, because of their pathological structure. He also concluded that identification of an angioma was not difficult during surgical procedures, and that vascular lesions were easily removed after evacuation of an old hematoma. McCormick et al. [11] found that an area of discoloration can be easily identified on the surface of the spinal cord. If a lesion is not
Fig. 3: Case 3. Magnetic resonance images of intramedullary cavernoma located at C4. Sagittal (A) and axial (C) T1-weighted images showing low-intensity mass with high-intensity rim. Sagittal (B) and axial (D) T2-weighted images showing hyperintensity mass with low-intensity rim. Gd-DTPA image (E) showing slight enhancement.

Fig. 4: Intraoperative photograph in case 3. Blue color mulberry shaped angiomas was seen on the dorsal aspect of the spinal cord (A). Assumed total removal of angioma (B). However, residual angioma was protruded from resective bed after the Valsalva maneuver (C).
visible on the surface of the cord, a myelotomy can be made in the midline or over the dorsal root entry zone. Once the lesion is found, a glial plane can usually be identified between the lesion and spinal cord, with obvious hemosiderin deposits. We must perform a careful sharp dissection around the surface of the lesion in the surrounding plane of gliotic tissue to avoid injury to normal spinal cord tissue.

As with a cavernous angiomas in the brain stem, it may be difficult to assess the completeness of removal of an IMCA [6,17]. Occasionally, a small remnant of an angioma or even satellite lesions are discovered in the walls of the resection cavity after a presumably complete removal. Several authors state that incomplete resection of angiomas encourages subsequent hemorrhage [6,7,17]. Mishima and Sasaki et al [12] reported the intentional increasing venous pressure with the Valsalva maneuver revealed hidden angiomatosus components after they assumed multilobular cavernous angioma of the brain stem had been removed completely. They recommended increasing venous pressure with the Valsalva maneuver to confirm complete extirpation of multilobular angiomas. This procedure can be applied to spinal intramedullary cavernomas. We also emphasize the importance of checking for residual vascular lesions using the Valsalva maneuver during surgery for IMCA. If an angiomomas remnant persists, this maneuver will result in protrusion of a deeply hidden lesion, as in our Case 3. We recommend that the maneuver should be repeated several times at intervals of several minutes, until the surgeon can confirm complete resection of IMCA.

To avoid surgical complications, intra-operative monitoring is essential [6,7,16]. We have routinely adopted MEP for all cases with spinal surgery involved cervical, thoracic and lumbar lesions. In cases with lesions located on the dorsal surface of the cervical spinal cord, surgery was performed using both MEP and SEP monitoring systems. Fortunately, the three cases in this study did not show significant changes in MEP and SEP during surgical procedures. We believe that surgical procedures should be terminated if MEP or SEP show a reduction to half amplitude.

The ICAMs in our series, as well as most of cases reported by other authors, were located on the dorsal surface of the spinal cord. ICAMs may occur on the ventral surface or in the deep core of the cervical cord – these lesions require extremely skilled surgical technique [6,7,13,17]. Surgery should therefore be undertaken with caution in patients with trivial neurological signs whose lesions are ventral or in the core of the spinal cord.

We concluded that resection of a cervical IMCA is safe and has a low risk of permanent neurological complications with a careful microsurgical technique, intra-operative monitoring by SEP and MEP, and confirmation of total removal by the Valsalva maneuver.

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Reviewer's comment : Masakazu Takayasu, M.D., Ph.D.
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The authors reported 3 cases of cervical intramedullary cavernous angiomas, which were successfully treated. They suggested that surgical treatment was indicated in symptomatic cases with dorsally located angiomas. However, indication for surgical treatment should be more cautious and surgical techniques might be more complicated in ventrally located angiomas. They recommended Valsalva maneuver to check for residual angiomas. This maneuver seemed to be useful. However, it had to be evaluated in larger number of cases.

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The authors reported three cases of intramedullary cervical cord cavernous malformations with successful surgical removal.

The natural history of intramedullary spinal cord (IMSC) cavernous malformations and their potential risk of hemorrhage remain unclear. Sandalcıoğlu et al. observed 17 hemorrhagic episodes in 375 patient-years of life, so that the calculated annual retrospective hemorrhage rate for symptomatic IMSC cavernous malformations was 4.5% per patient/year (Neurosur Rev 2003). The authors proposed that patient with spontaneously resolving symptoms after bleeding should be treated to avoid further hemorrhage. However, in reality, it is not easy to encourage patients to undergo operation when he/she had been completely asymptomatic over an extended period of time after the first bleeding. Considering the risk of transient mild neurological deterioration after surgery, it seems also tough to recommend surgical treatment for the elderly patient when he/she is nearly asymptomatic such as case 3 of this paper. The authors should be congratulated for their excellent surgical results. Intraoperative neurophysiological monitoring is essential. In my personal experience, intraoperative ultrasound is useful to visualize the location and extent of the intramedullary lesion so as to determine the site of myelotomy. Sandalcıoğlu et al. observed that all three patients with previously incomplete resected cavernous malformations experienced new neurological deficits due to significant rebleeding, indicating the potential risk of subsequent hemorrhage for patients with residual lesions (Neurosur Rev 2003). It is crucial to avoid partial removal of the malformations. The authors recommended increasing venous pressure with the Valsalva’s maneuver to identify the residual vascular lesions. This procedure is unique and seems useful.