Outcomes following resection of intramedullary spinal cord cavernous malformations: a 25-year experience

Clinical article


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Object. The management of intramedullary spinal cord cavernous malformations (CMs) is controversial. At Barrow Neurological Institute, the authors selectively offer surgical treatment for symptomatic spinal cord CMs. The purpose of this paper is to review the clinical outcomes in patients after resection of these lesions based on a single-center experience over a 25-year period.

Methods. The records of 80 patients who underwent resection of pathologically confirmed spinal cord CMs from January 1985 to May 2010 were analyzed retrospectively. Preoperative clinical status and imaging findings were evaluated as well as immediate and long-term postoperative outcomes.

Results. Compared with their preoperative Frankel grade, 11% of patients were worse, 83% were the same, and 6% improved immediately after surgery. At a mean follow-up interval of 5 years, 10% of patients were worse, 68% were the same, and 23% were improved compared with their preoperative status. Five percent of patients underwent reoperation for resection of a symptomatic residual or recurrent lesion. Immediate complications were encountered in 6% of patients, including CSF leakage and deep venous thrombosis. Long-term complications were encountered in 14% of patients and included kyphotic deformity, stenosis, and spinal cord tethering. A significant correlation was found between long-term outcome and anteroposterior length of the lesion (p = 0.01).

Conclusions. The resection of intramedullary spinal cord CMs can be achieved with good long-term outcomes and an acceptable risk of immediate or delayed complications. (DOI: 10.3171/2011.1.SPINE10454)

Key Words • cavernous malformation • outcome • intramedullary spinal cord • resection

AVENOUS malformations can be found throughout the CNS, but only rarely occur within the spinal cord. Intramedullary spinal cord CMs account for only 5% of all CMs of the CNS and for 5%–12% of all spinal cord vascular lesions. They can manifest with the sudden onset of neurologic deficit, with a gradually progressive decline in neurologic function or, rarely, with the sudden onset of pain caused by subarachnoid hemorrhage. Resection is the only definitive treatment for symptomatic intramedullary spinal cord CMs, but surgery in this highly eloquent tissue poses a significant risk of morbidity. Given the variability in reported outcomes and the uncertain natural history of CMs, resection of these lesions remains controversial.

At our institution, we offer surgery to patients with symptomatic CMs of the spinal cord whose general medical health and lesion location are favorable for resection. Our operative approach is typically through 1 of 3 surgical corridors: a midline myelotomy, a posterolateral myelotomy through the DREZ and substantia gelatinosa, or a lateral myelotomy just ventral to the dentate ligaments. Cavernous malformations are rarely resected via an anterior approach because of the tenuous blood supply in this region.

We previously published a study involving a series of 17 patients with spinal cord CMs who underwent operations between 1985 and 1995. In this report, we update our series of patients with spinal cord CMs by including the outcomes of all patients who underwent resection of a spinal cord CM from January 1995 to May 2010. Our cumulative experience of 80 patients over 25 years represents the largest single series of resected spinal cord CMs reported to date.

Methods

Institutional Review Board approval for this study was obtained. Subsequently, the hospital records of 80 patients who underwent resection of intramedullary spinal cord CMs were reviewed. The authors report their outcomes and complications.

Abbreviations used in this paper: CM = cavernous malformation; DREZ = dorsal root entry zone.
cortical CMs at our institution by the senior author (R.F.S.) between January 1985 and May 2010 were studied retrospectively. Data were acquired from clinic notes, operative reports, hospital charts, and pre- and postoperative MR images of the spine and brain.

**Patient Demographics**

Patient demographics are summarized in Table 1. There were 42 females and 38 males, with a mean age of 39.9 years (range 14–77 years). Six patients had a family history of CMs. One patient had undergone radiation therapy for a malignant rhabdomyosarcoma of the left sinonasal cavities 15 years before presentation. Clinical presentations were classified based on the 5 types of intramedullary spinal cord CMs as described by Ogilvy et al. 

<table>
<thead>
<tr>
<th>Variable</th>
<th>Value</th>
</tr>
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<tbody>
<tr>
<td>mean age ± SD (yrs)</td>
<td>39.9 ± 13.3</td>
</tr>
<tr>
<td>age range (yrs)</td>
<td>14–77</td>
</tr>
<tr>
<td>M/F (%)</td>
<td>48:52</td>
</tr>
<tr>
<td>clinical presentation (%)</td>
<td>discrete, acute episodes 43 (54), slow, progressive decline 30 (37), acute onset, rapid decline 3 (4), acute onset, gradually progressive decline 4 (5)</td>
</tr>
<tr>
<td>lesion location (%)</td>
<td>cervical spine 46 (55), thoracic spine 28 (35), conus medullaris 6 (7)</td>
</tr>
<tr>
<td>lesion dimensions (mean in mm ± SD)</td>
<td>craniocaudal 11.5 ± 5.2, anteroposterior 6.7 ± 2.8, transverse 6.8 ± 2.9</td>
</tr>
</tbody>
</table>

Operative Technique

The operative technique used for resection of intramedullary spinal cord CMs by the senior author has been published previously. Briefly, all patients underwent a posterior midline incision followed by laminectomy (28 patients [35%]) or laminoplasty (52 patients [65%]). Opening of the dura in the midline, or slightly off midline, was followed either by a midline myelotomy (24 patients), posterolateral myelotomy over the DREZ (30 patients), or lateral myelotomy (8 patients). When the CM was visible through the pia without intervening spinal cord tissue, myelotomy was performed directly over the lesion itself (18 patients), which was paramedian in the posterior aspect of the spinal cord in 15 patients and was located anterolaterally in 3 patients. Microdissection instruments were used to resect the CM. In a few cases, a carbon dioxide laser was used. Veins were associated
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with the CMs in all cases, and every attempt was made to preserve these vessels during dissection and removal of the lesion. Only patients with histologically confirmed CMs were included in this study.

**Follow-Up**

Follow-up information was obtained through notes from the last postoperative clinic visit. Because our institution services a large referral base, data were alternatively obtained through notes from outside hospitals or clinics if the patient was discharged to another center for follow-up. The patient’s clinical status, resolution of pain, and any additional and related surgical procedures that the patient may have undergone by the time of follow-up were noted. Immediate clinical outcome was documented for all 80 patients. The mean postoperative follow-up duration for all patients in whom more than 1 month of fol-

<table>
<thead>
<tr>
<th>Frankel Grade</th>
<th>Preop (%)</th>
<th>Immediate Postop (%)</th>
<th>Long-Term Follow-up (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>1 (1.3)</td>
<td>1 (1.3)</td>
<td>1 (1.6)</td>
</tr>
<tr>
<td>B</td>
<td>1 (1.3)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>C</td>
<td>9 (11.3)</td>
<td>14 (17.5)</td>
<td>4 (6.4)</td>
</tr>
<tr>
<td>D</td>
<td>47 (58.8)</td>
<td>44 (55)</td>
<td>37 (59.7)</td>
</tr>
<tr>
<td>E</td>
<td>22 (27.5)</td>
<td>21 (26.3)</td>
<td>20 (32.3)</td>
</tr>
<tr>
<td>total</td>
<td>80 (100)</td>
<td>80 (100)</td>
<td>62 (100)</td>
</tr>
</tbody>
</table>

* The mean follow-up duration was 5 years.

![Fig. 1](image_url)  
**Fig. 1.** Imaging of a representative patient with an intramedullary spinal cord CM.  
A: Sagittal T1-weighted MR image demonstrating a mixed intensity lesion behind the C-7 vertebral body consistent with an intramedullary spinal cord CM.  
B: Sagittal T2-weighted MR image of the same lesion.  
C: Axial T2-weighted MR image showing the lesion located laterally within the spinal cord. This lesion was resected through a myelotomy at the DREZ, and the lesion was extirpated with the use of a carbon dioxide laser.  
D: Postoperative sagittal T1-weighted MR image shows no residual lesion.
Complications and Outcomes

Immediate and delayed complications related to the surgical procedure or general anesthetic were recorded from the time of surgery at our institution until the date of last follow-up. Unintended incomplete resection of a spinal cord CM manifesting with recurrent symptomatology on a delayed basis was also included. Immediate postoperative clinical outcome (in all 80 patients) and neurological status at the time of long-term follow-up (in 62 patients) was also assessed using the Frankel scale. Comparisons between preoperative and immediate postoperative clinical status as well as with neurological outcome at the time of last follow-up were made.

Statistical Analysis

Statistical analysis was performed using GraphPad InStat statistical software (GraphPad, Inc.). Categorical variables were analyzed using Fisher exact test for dichotomous categories or the chi-square test for independence when more than 2 categories were analyzed. Continuous variables were analyzed using Student t-test when comparing 2 groups, or ANOVA between groups when comparing results from more than 2 groups. All probability values < 0.05 were considered statistically significant.

Results

Immediate Radiographic Outcome

A possible residual lesion was noted on early postoperative MR imaging in 3 patients. In 1 of these patients, no additional symptoms were noted at the time of last follow-up 7 years later. Another of these patients experienced 2 subsequent episodes of mildly worsening symptoms for which no additional surgery was performed as of follow-up 1 year later. The third patient underwent re-resection for recurrent symptoms 6 years after the initial operation.

Immediate Surgical Complications

Five patients (6%) suffered complications from surgery or general anesthesia (Table 3). In 1 patient, a serous epidural fluid collection causing significant mass effect on the thecal sac required decompression on postoperative Day 11. Postoperatively, 2 patients experienced CSF leaks, one of which required placement of a lumbar drain. Another 2 patients were diagnosed with deep venous thrombosis in the early postoperative period.

Immediate Clinical Outcome

Immediately after surgery, 1 patient (1%) was classified as Frankel Grade A, none were Grade B, 4 (6%) were Grade C, 37 (60%) were Grade D, and 20 (32%) were Grade E (Table 2). Compared with their preoperative neurological status, the Frankel grades of 6 patients (10%) were worse. The Frankel grade improved in 14 patients (23%), and the neurological status of 42 patients (68%) remained the same (Fig. 2). When compared with their immediate postoperative Frankel grade, 4 patients (6%) worsened, 16 (26%) improved, and 42 (68%) remained the same over the follow-up period. Of the 45 patients who presented with pain and in whom long-term follow-up was available, 25 (56%) had experienced improvement in their pain at long-term follow-up.

The correlation between lesion size ≤ 10 mm in greatest dimension and improved long-term outcome compared with preoperative clinical status (dichotomized as better versus same/worse) showed a trend toward statistical significance (p = 0.054, Fisher exact test). An ANOVA (nonparametric, 2-tailed) test showed no correlation between outcome (better, same, or worse) and continuous variables including craniocaudal dimension, transverse dimension, age, or length of follow-up. However, the difference in anteroposterior dimensions was statistically significant (p = 0.0139) when compared between the 3 groups: better (mean 14.2 mm), same (mean 28.4 mm), and worse (mean 38.6 mm). A multiple comparisons test within this ANOVA showed that statistical significance was maintained when comparing better versus same (p < 0.05) and better versus worse (p < 0.05) but not same versus worse (not significant).

Using the chi-square test for independence, the correlation between long-term outcome (classified as better, same, or worse) with location of cavernoma (cervical, thoracic, or conus), approach (DREZ, lateral, anterolateral, paramedian, or midline), or preoperative Frankel grades (A, B, C, D, or E) did not reach significance. The correlation between pain relief and an approach to the lesion through a DREZ myelotomy versus the other approaches was also not significant (p = 1.0, Fisher exact test).

Of the patients presenting to our institution without prior resection, 3 patients underwent reoperation for resection of a symptomatic residual/recurrent CM. Only 1
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of these patients was described as having a residual lesion noted on early postoperative imaging. The other 2 patients demonstrated symptomatic worsening 2 and 4 years after their initial resection, respectively, and imaging at the time of re-presentation demonstrated a recurrent hemorrhage despite no obvious residual lesion on earlier scans. All 3 of these patients underwent reoperation and resection of histologically confirmed lesions.

Two additional patients developed recurrent symptoms over the course of their follow-up (at 2 and 5 years) that were reminiscent of their initial presentation. Follow-up imaging in these patients, however, revealed stable postoperative changes without evidence of residual or recurrent lesions.

Delayed Surgical Complications

Of the 62 patients in whom long-term follow-up was available, 9 (14%) experienced delayed complications related to the surgical procedure (Table 3). Four patients (6%) who underwent resection for cervical CMs developed symptoms related to postaminectomy (1 patient) or laminoplasty (3 patients) kyphosis involving the same levels of prior surgery. There was no statistically significant difference in the occurrence of kyphosis between each type of procedure (p = 1.0, Fisher exact test). All 4 patients underwent anterior cervical stabilization and fusion procedures at a mean interval of 6 years after their initial surgical procedure. Two patients (3%) developed symptomatic cervical stenosis involving the surgical site and required posterior decompression procedures at a mean of 4 years after their initial resection. Another 3 patients (5%) developed new symptoms related to spinal cord tethering at the surgical site a mean of 7 years after their initial resection. One of these patients required a single detethering procedure at 10 years; one required 2 procedures at 8 and 20 years, including a suboccipital decompression for symptomatic cerebellar tonsillar herniation; and one required 3 detethering procedures at 3, 6, and 8 years after the initial resection.

Discussion

Ogilvy et al.10 described 5 different patterns of clinical presentation in patients with spinal cord CMs. In their report, the most common presentation, which occurred in 36% of their patients, involved discrete, acute episodes of neurological deterioration over months to years. The next most common presentation—slow, progressive decline in neurological function—occurred in 22%. In our series, 54% of patients presented with discrete, acute episodes, and 37% presented with a slow, progressive decline in neurological function. Similar to the series of Ogilvy et al.10 none of our patients presented with subarachnoid hemorrhage from a lesion on the surface of the spinal cord.

Patients with familial CMs, defined as 1 or more first-degree relatives with a suspected CM of the brain or spine, comprised only 8% of our series. Patients with a history of familial CMs were more likely to have coexistent brain CMs (p = 0.01, Fisher exact test). Furthermore, age at presentation was not statistically different between patients with and without a family history of CMs (37.2 vs 40.1 years, respectively; p = 0.6, Student t-test). In our subset of patients with a spinal cord CM who also underwent brain imaging, 34% had at least 1 brain lesion. Although potentially influenced by selection bias, this figure is slightly lower than our previously published data based on 17 patients12 and similar to the experience of Cohen-Gadol et al.3 who reported a coexistence rate of 40% in their 33 patients.

Outcomes after resection of intramedullary spinal cord CMs have varied. Prior to this report, the largest surgical outcome series was published by Labaigue et al.9 in 2008. In their multicenter retrospective study of 53 patients, long-term follow-up data (mean 7.3 years) were available for 37 patients and demonstrated improvement in 54%, no change in 16%, and clinical worsening in 30%.

In another series of 26 patients reported by Jallo et al.5 in 2006, 50% worsened immediately after surgery. At a mean follow-up of 4.5 years, only 8% were worse, 46% were stable, and 46% reported improvement compared with their preoperative functional status.5 In their study, however, long-term follow-up was subjective and described without the use of an established neurological or functional grading system. A more recent clinical series of 16 patients showed that 75% improved and 25% were stable based on Frankel grade during the mean follow-up period of 23.1 months.2 In a systematic review of 186 patients with spinal cord CMs, resection resulted in a functional outcome that was stable in 61%, improved in 31%, and worse in 8% of patients.2

In our series, immediately after surgery 9 (11%) of the 80 patients were worse based on Frankel grade. All patients who were clinically worse immediately after surgery had deteriorated by 1 Frankel grade (E to D in 5 cases and D to C in 4 cases). Over the follow-up period, however, 10% were worse, 23% were better, and 68% were
the same compared with their preoperative grades. Of the patients who were worse (6 of 62), 5 were worse by 1 Frankel grade and 1 was worse by 2 Frankel grades. The latter patient was clinically the same immediately after surgery compared with his preoperative grade of E, but 5 years after his initial resection he suddenly deteriorated to a Frankel grade of C. Magnetic resonance imaging at the time of his deterioration showed stable postoperative changes with no evidence of a residual or recurrent lesion.

The significant association between long-term clinical outcome and anteroposterior diameter in this series is interesting, because size of lesion has not previously been associated with outcome after resection of spinal cord CMs.\textsuperscript{9,11} Although the exact reasons for the association in our series are unknown, it is possible that smaller lesions are less likely to produce permanent damage to the surrounding spinal cord tissue and that the resection of such lesions results in less overall trauma to the surrounding spinal cord. Because outcome was not associated with craniocaudal or transverse length and because almost all of the lesions were located posteriorly within the spinal cord, it is interesting to speculate whether the worsened clinical outcome is due to encroachment of the lesion more anteriorly toward the motor tracts.

In this review, 51 (64\%) of 80 patients had related symptoms of pain at the time of their presentation. In most cases, this pain was described as a burning dysesthesia, but numerous cases of radicular and axial pain were also reported. Of the patients for whom such data were available at follow-up, 56\% experienced an improvement in their preoperative pain. We previously addressed the result of resection of intramedullary spinal cord CMs on pain resolution.\textsuperscript{7} In that report, 52\% of patients showed improvement in their preoperative pain symptoms at a mean follow-up of 44.5 months, which is similar to our current results. Furthermore, although it may appear plausible that a myelotomy through the DREZ could result in improvement in pain symptoms, no such association was found in this study.

The delayed surgical complications in our series included postlaminectomy or laminoplasty kyphosis in 6\% of patients, and there was no statistically significant difference in the occurrence of kyphosis between each type of procedure. Due to the number of patients in this study, however, and due to the relatively low incidence of cervical kyphosis after both laminectomy and laminoplasty, little can be inferred about the long-term benefit of replacing the lamina.

In 2 of 3 patients with an unexpected residual lesion noted on postoperative imaging, recurrent symptoms were experienced several years later. However, postoperative imaging of spinal CMs is often difficult to interpret, more so than that of CMs resected in other noneloquent locations because the adjacent hemosiderin-stained tissue is not resected. The predictive value of a “possible residual lesion” therefore is often regarded as questionable, especially given a gross-total resection as presumed intraoperatively, and a watch-and-wait approach is generally pursued. Perhaps as MR imaging improves, it will be possible to more clearly differentiate a true residual lesion from a small amount of blood or fluid in the resection cavity. Given the potential long-term benefits, such an advance could help facilitate the surgical goal of a complete resection, even if it implies an early return to the operating room for reexploration.

The retrospective nature of our study and absence of a control group preclude a direct comparison of the surgical outcomes with the natural history of symptomatic spinal CMs. In a retrospective review of spinal cord CMs managed conservatively by Kharkar et al.,\textsuperscript{9} however, 9 of 10 patients remained the same or improved at a mean follow-up of 80 months. Comparing our results with their group of conservatively managed patients is difficult because of the potential for significant selection bias in their series.

In our experience, on the other hand, symptomatic lesions of the spinal cord behave fairly aggressively, similar to CMs of the brainstem. The results of this study, therefore, support the complete resection of symptomatic intramedullary spinal cord CMs to prevent further neurological decline.

Conclusions

This single-center, single-surgeon experience demonstrates that spinal cord CMs can be resected with good long-term outcomes and with an acceptable risk of complications. Clinical outcomes correlated with the anteroposterior diameter of the lesion, which is a novel finding, although further studies are required to more clearly define the reasons for this association.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Spetzler, Mitha, Vishteh. Acquisition of data: Mitha, Turner, Abla, Vishteh. Analysis and interpretation of data: Spetzler, Mitha, Turner, Abla. Drafting the article: Mitha, Turner, Abla. Critically revising the article: all authors. Reviewed final version of the manuscript and approved it for submission: all authors. Statistical analysis: Mitha, Abla. Administrative/technical/material support: Mitha, Turner. Study supervision: Spetzler, Vishteh.

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