Intramedullary spinal cavernoma: clinical presentation, microsurgical approach, and long-term outcome in a cohort of 48 patients

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OBJECT Intramedullary spinal cavernoma (ISC) is a rare entity and accounts for approximately 5%–12% of all spinal vascular pathologies. The purpose of the present study was to examine the influence of clinical presentation, localization, and different surgical approaches on long-term outcome in patients treated for ISC.

METHODS The authors performed a retrospective single-center study of 48 cases of ISC treated microsurgically over the past 28 years. Analyzed factors included preoperative clinical history, microsurgical strategies, neurological outcome (American Spinal Injury Association [ASIA] grade, Epstein and Cooper grade), and the occurrence of postoperative spinal ataxia. Univariate analysis was performed to identify factors influencing long-term outcome.

RESULTS Preoperatively, 18.8% of all patients experienced a slow, progressive decline in neurological function and 33.3% suffered repetitive episodes of acute neurological deterioration over a time frame of months to years. Moreover, 16.7% noted the sudden onset of a severe neurological deficit, whereas 25% experienced the sudden onset of symptoms with a subsequent gradually progressive decline in neurological function. On long-term follow-up after treatment (mean ± SD, 79.3 ± 35.2 months), 70.8% of patients showed no change in neurological function, 6.3% suffered from a decline, and 22.9% improved neurologically. Thoracolumbar localization (p = 0.043), low preoperative Epstein and Cooper grade for the lower extremities (p < 0.001), and a low preoperative ASIA grade (p < 0.001) were identified as factors associated with an unfavorable outcome (ASIA Grade A–C). The rate of spinal ataxia related to surgical approach was 16.7%.

CONCLUSIONS Postoperative neurological function in ISC patients is determined by the preoperative neurological status. On long-term follow-up after microsurgical treatment, 93.7% of patients presented with a stable or improved condition (ASIA grade); thus, definite microsurgical treatment should be considered as long as patients present with only mild symptoms after the diagnosis of symptomatic ISC.

http://thejns.org/doi/abs/10.3171/2015.5.FOCUS15153

KEY WORDS spinal cavernoma; microsurgery; spinal cord; vascular malformation

Intramedullary spinal cavernoma (ISC) is a rare vascular disease and accounts for 5%–12% of all spinal vascular pathologies. The incidence of cavernoma located in the CNS, including the brain and spinal cord, is about 1.9 cases/100,000 persons/year with 3%–5% of lesions located in the spinal cord. An ISC may become clinically apparent with the acute onset of neurological deficits and a wide range of symptoms due to either an acute macrohemorrhage forming a space-occupying lesion, possibly accompanied by edema of the spinal cord, or a worsening of preexisting symptoms as the result of recurrent hemorrhage. However, repetitive intrallesional microhemorrhages can lead to a more slowly progressive decline in neurological function. The sudden onset of pain is rarely seen but is most likely the result of subarachnoid hemorrhage.
A number of published case reports and case series of ISC have documented surgical treatment options as well as the natural history of the disease.\(^6,14,22,35,38\) For the purpose of diagnosis, MRI still represents the gold standard, showing a typical hyperintense lesion on native T2-weighted sequences, usually surrounded by a hypointense hemosiderin rim after hemorrhage (Fig. 1). With respect to causative treatment options, complete microsurgical removal of the pathology is recognized as the therapy of choice whenever possible.\(^2,4,11,23,25,27,37\) Approaches to the spinal cord are performed either directly to the pathology in cases of ISCs reaching the pial surface or via the central dorsal sulcus (midline myelotomy) for deep-seated lesions. Spinal cord access via the dorsolateral entry zone is especially suitable for superficial lateral pathologies by opening the dentate ligament and gently rotating the spinal cord.\(^36\) Custom-tailored approaches are mandatory to minimize perioperative complications, achieve complete resections, and optimize postoperative outcome.\(^3,5\) Indications for microsurgical removal are present when an ISC becomes clinically apparent and presents as surgically accessible. The annual rate for a first hemorrhage is up to 4.5% in retrospective series, assuming lesions were present since birth. For recurrent hemorrhage, the annual rate is up to 66% per patient per year, possibly leading to further, rapid, permanent neurological deterioration.\(^1,4\)

Little data correlate clinical, anatomical, and surgical aspects of ISCs with the long-term neurological outcome of surgically treated patients. The present study was conducted to evaluate long-term outcome in a cohort of 48 microsurgically treated patients with ISC and to identify factors influencing neurological status after treatment to facilitate decision making for clinicians confronted with ISC.

Methods

We performed a retrospective analysis of the medical records between 1986 and 2014, identifying all patients with the diagnosis of an ISC treated microsurgically at our neurovascular center. Other vascular pathologies of the spinal cord such as spinal dural arteriovenous fistula or spinal arteriovenous malformations were excluded. Patient histories were assessed using the 4-type scale introduced by Ogilvy et al., which discriminates between acute and chronic progression as well as phases of neurological recovery (Table 1).\(^3,31\) Long-term functional outcome was assessed using the American Spinal Injury Association (ASIA) impairment scale, which was applied pre- and postoperatively and on long-term follow-up. As an additional clinical parameter, the Epstein and Cooper (EC) grading system for intramedullary tumors was applied for all patients preoperatively and immediately postoperatively.\(^10,21\) Favorable outcome on long-term follow-up was defined as an ASIA grade of D or E. For the assessment of dorsal column function after dorsal myelotomy, patients were additionally seen in the outpatient clinic and neurologically examined (Romberg’s sign, Unterberger’s test, tandem gait assessment, heel-shin test).

Using a 3-pin Mayfield skull clamp if the lesion was located in the cervical spine or in the cervicothoracic region, we performed surgery with the patient in prone position.

We utilized various approaches to the spinal cord (for example, laminotomy, laminectomies, hemilaminectomies), depending on the extent and location of the pathology (Fig. 2). To improve targeting of the approach and minimize surgical trauma, ultrasonography was used before opening the dura mater.\(^3,32\) Myelotomy was performed via the posterior median sulcus (dorsal midline entry zone), lateral through the dorsolateral entry zone, or directly into the pathology (Fig. 3). Microsurgical dissection was performed using the surrounding gliotic layer as a preparation margin. In some cases, a CO₂ surgical laser system (SLS-25, Surgilase) was used to dissect without traction. Throughout the preparation, all efforts were made to protect the surrounding spinal cord vessels (Figs. 4 and 5). Intraoperative electrophysiological monitoring (IOM) was applied in all cases treated after 1996. In these cases, somatosensory evoked potentials along with motor evoked potentials (MEPs) and morphological D-wave changes were analyzed. After complete microsurgical removal of the ISC, the arachnoid layer was sutured (7.0 Ethilon, Ethicon), the dura was closed, and the vertebral arches were fastened in place (in cases of laminotomy).

Neuropathologists routinely analyzed the resected specimen. Staining procedures utilized hematoxylin and eosin, van Gieson’s elastic stain, Masson’s trichrome stain, and Turnbull’s stain for siderin, all following standard laboratory procedures. Histologically, the most frequent differential diagnosis was melanocytoma.

Descriptive statistics were used to characterize the study population. The Student t-test was used to compare means of metric variables and the likelihood-ratio chi-square test was used for categorical variables whenever appropriate. Univariate logistic regression analysis was used to identify potential factors influencing long-term clinical outcome (ASIA grade, EC grade). A p < 0.05 was considered statistically significant. All statistics were performed using SPSS (IBM SPSS Statistics for Windows, version 22.0, IBM Corp.). Values are expressed as the mean ± standard deviation, unless indicated otherwise.

Results

Out of a series of 397 patients with intramedullary spinal cord lesions treated at our institution during the study period, we identified 48 patients with ISC (12.1%). Twenty-
three females (47.9%) and 25 males (52.1%), with a mean age of 41.3 ± 15.6 years, had ISC's located in the thoracic (56.3%), cervical (39.5%), and thoracolumbar (4.2%) regions.

Preoperatively, 18.8% of patients reported a slow, progressive decline in neurological function, 33.3% showed discrete and acute episodes of neurological deterioration with various degrees of recovery in between, and 41.7% of patients reported an acute onset of symptoms followed by either a rapid (16.7%) or gradually progressive (25.0%) decline of neurological function; the remaining 6.2% presented with incidental findings of ISC. Neurological symptoms were defined as sensory disorders (for example, dysesthesias, impaired proprioception), motor disabilities (for example, paresis), or autonomic dysfunction (for example, disturbances in bladder or bowel control).

The time span from the onset of symptoms to the diagnosis of ISC varied between hours in cases of acute and severe neurological impairment, and 96 months in cases of mild neurological impairment (mean 16.1 ± 20.3 months).

The treatment of choice was complete microsurgical removal, which was performed in all 48 cases. Concerning the operative approach to the spinal cord, various degrees of bony resection were performed. For small and lateral ISC's reaching the surface, hemilaminectomies (1-level: 8.3%, n = 4; 2-level: 2.1%, n = 1) were found suitable and supplied a sufficient intraoperative overview. On the other hand, deep or large ISC's required more extensive approaches (1- or 2-level laminotomies: 18.8%, n = 9; 1- or 2-level laminectomies: 54.2%, n = 26) allowing for an overview of the complete spinal cord anatomy (Fig. 2). Other, much more infrequently performed approaches constituted ventral approaches (2.1%, n = 1) and, for example, combined suboccipital and cervical dorsal laminectomies or interlaminal fenestrations (14.6%, n = 7).

Microsurgical myelotomy was performed either via the posterior median sulcus (37.5%) or directly in cases in which the lesion had contact with the surface (54.2%). In 4 patients (8.3%) a lateral approach (lateral entry zone) with division of the dentate ligament on one side was performed (Fig. 3B). Other custom-tailored options included a ventral approach to the cervical spine when the ISC was located in the ventral spinal cord, extending to the medullary surface, which was performed once (Fig. 3C).

Functional outcome assessment using the ASIA impairment scale immediately postoperatively before discharge demonstrated that 54.2% of the patients were unchanged neurologically, 33.3% had worsened, and 12.5% had improved compared with the preoperative state.

Worsening or improvement was defined as a change by at least 1 grade on the scale. On long-term follow-up (mean 79.3 ± 35.2 months) 70.8% of the patients demonstrated no change in neurological function compared with their preoperative status, 6.3% had worsened, and 22.9% had improved (Fig. 6). Overall, 83% of patients presented with a good ASIA grade (D or E) on long-term follow-up.

Factors possibly influencing the ASIA grade on long-term follow-up were identified by univariate logistic regression analysis. Three factors were associated with significant effects on the long-term postoperative ASIA grade: thoracolumbar localization, a low preoperative EC grade for the lower extremities, and a low preoperative ASIA grade (Table 2).

Regarding ISC localization, pathologies located in the thoracolumbar spinal cord (n = 2), as opposed to the cervical or thoracic, did show a greater risk for a postoperative ASIA grade of A–C. In our series, 2 patients with thoracolumbar ISC's were identified, and both of them had an ASIA grade of C on long-term follow-up. Forty-six patients with either cervical or thoracic ISC had a higher probability of presenting with an ASIA Grade E or D on long-term follow-up (p = 0.043; Figs. 6 and 7).

The preoperative clinical status as reflected by the ASIA grade for the lower extremities, and a low preoperative ASIA grade (Table 2).

Of the 48 patients included in our study, 18 (37.5%) were also seen on an outpatient basis in the Department of Neurology and underwent physical examination with a focus on the clinical aspects of spinal ataxia. Because of the long follow-up period, patients were often not available for a physical examination. All 18 of these patients had been treated within the past decade. Seven patients (38.9%) experienced moderate or severe neurological impairments prior to the operation (for example, paraparesis or previously existing spinal ataxia). Postoperatively, 3 patients (16.7%) showed signs of newly occurring spinal ataxia. In all 3 of these cases, midline myelotomies were performed to access the pathology (approach-related ataxia, median follow-up for this group: 48 months).

One patient suffered from a recurrence of an initially

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**TABLE 1. Ogilvy classification of preoperative clinical history**

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>Pattern of discrete and acute episodes of neurological deterioration over mos to yrs separated by various degrees of recovery</td>
</tr>
<tr>
<td>2</td>
<td>Slow, progressive decline in neurological function</td>
</tr>
<tr>
<td>3</td>
<td>Acute onset of symptoms followed by rapid neurological decline</td>
</tr>
<tr>
<td>4</td>
<td>Acute onset of mild symptoms followed by a gradually progressive decline in neurological function over wks to mos</td>
</tr>
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</table>

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**FIG. 2.** Approaches to the spinal column varied depending on the extent and localization of the ISC. The most frequent approaches included 1- or 2-level laminotomies or laminectomies. In some cases of lateral ISC, hemilaminectomies were sufficient for accessing the spinal cord. We also used combined approaches to the cervical spinal cord, making decompression of the foramen magnum and laminotomies of C-1 necessary.
completely microsurgically resected thoracic ISC. She underwent reoperation with a straightforward approach and has not suffered another recurrence in 11 years of additional follow-up. Regarding perioperative complications, 1 patient suffered from an acute epidural hematoma, which was immediately evacuated; this patient had no postoperative neurological deterioration on follow-up. In the presented cohort, no CSF leakages or wound infections leading to a second surgical intervention occurred.

One patient required cervical spondylodesis 4 months after extirpation of a cervical ISC because of segmental instability leading to a secondary kyphosis. In this patient, the initial cervical approach had been performed via a 2-level laminotomy.

Discussion

In this study, factors influencing long-term clinical outcome and microsurgical strategies are presented along with the preoperative clinical histories of 48 microsurgi-
intramedullary spinal cavernoma
cally treated ISC patients. Factors significantly associated with an unfavorable outcome (defined as ASIA Grades A–C) include a thoracolumbar localization, a low preoperative EC grade for the lower extremities, and a low preoperative ASIA grade. On long-term follow-up, 93% percent of surgically treated patients presented with an improved or unchanged neurological status compared with their preoperative status, and 83% of patients had an ASIA Grade D or E.

The major goal of surgical treatment is the complete and immediate resection of the ISC without permanent neurological deterioration. Currently, the feasibility of microsurgical treatment along with patterns of clinical manifestations has been recognized, and the microsurgical removal of ISC is recommended when the pathology becomes clinically apparent and appears accessible.17,36

If symptomatic patients present with an ISC showing signs of hemorrhage on MRI, microsurgical removal is suggested within 2 and up to 6 weeks after the disorder becomes clinically apparent. Results from previous studies of cavernoma surgery, for example, in the brainstem, suggest early microsurgical treatment if anatomically reasonable. If surgery is delayed for several weeks, patients are at greater risk for neurological deterioration.4 This finding is most likely attributable to scarring around the cavernoma so that dissection becomes more invasive and increases the risk for neurological deficits. If surgery is performed too early, edema and vulnerability of the spinal cord due to hemorrhage may lead to postoperative deterioration. If surgery is performed too late, gliosis and limited space may contribute to aggravated surgical removal with possibly negative effects on clinical status after surgery.

Observation, on the other hand, is recommended for both asymptomatic lesions not reaching the pial layer and symptomatic deep-seated lesions with minimal or transient symptoms.17 In a recent series focusing on the conservative treatment of ISC, patients displaying sensory and/or motor deficits were shown to profit from surgical treatment.20 Only asymptomatic patients or patients with pain as the only symptom were shown to justify a “wait and see” strategy.

In our cohort, 33.3% of patients had a worse ASIA grade in the immediate postoperative phase compared with their preoperative status. This immediate deterioration was most likely attributable to intraoperative intramedullary dissection, and the rate fell to 6.3% of patients with permanent worsening on long-term follow-up after surgery. In the literature, transient neurological worsening after surgery occurs in up to 50% of cases, but on long-term follow-up after surgery permanent worsening decreases to 0%–20%.1,13,18,37

Regarding the assessment of spinal ataxia, especially after myelotomy, the ASIA grade does not accurately reflect the true degree of impairment since it mostly addresses motor deficits due to muscle strength. Three (16.7%) of 18 patients, who on follow-up were neurologically examined with an emphasis on ataxia, were found to have newly diagnosed spinal ataxia postoperatively. All of these patients had been treated with midline myelotomy to access the pathology. The implications of these findings are hard to determine since preoperative neurological deficits may distort the true incidence of newly occurring spinal ataxia postoperatively, making an interpretation of the findings (preoperative deficit versus complication versus operative morbidity due to myelotomy) even more difficult.

Intraoperatively, the lack of substantial deterioration in IOM with a special focus on D-wave changes confirms the surgical strategy and helps to avoid a worsening neurological outcome postoperatively.12,24,33

A major prognostic factor determining neurological outcome was the preoperative neurological status (ASIA grade: p < 0.001 and EC grade for the lower extremities: p < 0.001), which matches the results from another previous series.36 Both scores focus on motor function of the lower extremities and can be interpreted together.

FIG. 5. Microsurgical removal of an exophytically growing spinal cavernoma (A) in the cervical spinal cord (C-4). Preparation of the cavernoma using bipolar forceps with coagulation of the nourishing vessels (B), leaving the cavernoma bed after complete extirpation of the pathology (C).

FIG. 6. On long-term follow-up, 83% of patients presented with an ASIA grade of D or E. Directly after the operation, 33.3% of patients experienced temporary deterioration of their neurological status, as measured using the ASIA scale. Permanent neurological deterioration according to this scale was seen in 6.3% of patients.
However, 16.7% of patients suffered preoperatively from the acute onset of symptoms associated with severe neurological deficits. Given the risk for severe permanent neurological deficits and rapid neurological decline combined with the potential for a good postoperative outcome, early microsurgical therapy is strongly advocated whenever the lesion is accessible and patients suffer from only mild symptoms. From our point of view, symptomatic patients with deep-seated lesions and progressive mild symptoms can also be safely treated without the risk of postoperative deterioration, since the annual initial hemorrhage risk is estimated to be around 4.5% and higher, especially if one considers that rapid neurological deteriorations can occur. At what time point mild symptoms become progressive may be very subjective; therefore, neurological symptoms must be closely monitored in the case of deep-seated lesions managed with a conservative observation strategy.

As a second factor influencing long-term outcome, a thoracolumbar localization was shown to be associated with a less favorable neurological status on follow-up. The implications of these findings in our cohort are difficult to evaluate reliably since only 2 patients had lesions in the thoracolumbar spine; thus, statistical significance in the univariate analysis must be interpreted carefully. Both patients with thoracolumbar ISC presented with an ASIA Grade C on long-term follow-up, which reflected the preoperative status (ASIA Grade C).

In 58.3% of cases, a CO2 laser was used. Given the institutional experience of our center, especially for myelotomy and intramedullary preparation, the use of a laser enabled the surgeon to dissect without any traction to the surrounding nervous tissue. In clinical practice, once a surgeon becomes familiar with the device, the CO2 laser proves to be an effective and safe operative tool that ensures as gentle preparation as possible. A microsurgical alternative to the CO2 laser is the cavitational ultrasonic surgical aspirator (CUSA) in spinal cord tumor surgery.9,26 Regarding the bony approach to the spinal cord, various strategies ranging from keyhole approaches (for example, hemilaminectomies) to more extensive multiple-level laminectomies in large ISC were applied in our cohort. Recent approaches focus on nondestructive, minimally invasive strategies if possible to obtain an adequate anatomical overview. If multilevel laminectomies in the cervical spine are necessary (due to the need for gently rotating the spinal cord to reach the lateral lesion or the extent of the cavernoma), they are routinely combined with dorsal stabilization techniques to avoid kyphotic malalignment.19 To access ventrally located cervical cavernomas reaching the pial surface, a custom-tailored approach via partial corpectomy has been shown to be feasible. In these rare cases, the broad dorsal exposures of the spinal cord can be avoided by rotating the spinal cord to reach the pathology. On the one hand, however, this approach requires extensive experience evaluating IOM data, in particular D-wave monitoring data (in this case, the stimulating electrode was placed ventrally); on the other hand, it requires an excellent understanding of the vascular supply of the spinal cord. The routine use of IOM was established in 1997, whereas the use of the CO2 laser was established much earlier in our department, that is, from the beginning of the presented series (mid-1980s). Intraoperative electrophysi-

### TABLE 2. Univariate ordinal logistic regression analysis of variables tested possibly affecting postoperative ASIA long-term outcome

<table>
<thead>
<tr>
<th>Variable</th>
<th>p Value</th>
</tr>
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<tbody>
<tr>
<td>Age</td>
<td>0.316</td>
</tr>
<tr>
<td>Sex</td>
<td>0.657</td>
</tr>
<tr>
<td>Localization</td>
<td>0.043</td>
</tr>
<tr>
<td>Onset of symptoms until time of treatment</td>
<td>0.417</td>
</tr>
<tr>
<td>Spinal approach</td>
<td>0.110</td>
</tr>
<tr>
<td>Myelotomy</td>
<td>0.068</td>
</tr>
<tr>
<td>Laser use</td>
<td>0.711</td>
</tr>
<tr>
<td>Preop clinical course</td>
<td>0.761</td>
</tr>
<tr>
<td>Preop EC grade UE</td>
<td>0.881</td>
</tr>
<tr>
<td>Preop EC grade LE</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Preop ASIA grade</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

LE = lower extremity; UE = upper extremity.
* Localization, preoperative EC grade as well as preoperative ASIA grade significantly influence long-term postoperative ASIA grade.
ological monitoring has proven to be an especially helpful surgical tool capable of influencing the intraoperative strategy. However, sometimes electrophysiological deteriorations seem to be related to a surgical maneuver but are eventually proven to be false negative. In these cases, the intraoperative situation together with the electrophysiological findings and the experience of the operating surgeon influence the decision of whether to modify the surgical strategy or to continue with the operation as planned.

Additionally, D-wave alterations were recently shown to be a consistent parameter predicting the postoperative neurological status with higher reliability than MEPs. As clinical improvements often take years to develop, some of the recently treated patients (5 within the last 2 years) might undergo additional improvement in the future, possibly improving the general long-term outcome of the cohort presented.

A limitation of this study was its design as a retrospective analysis.

**Conclusions**

Microsurgical treatment of ISC displays an unrivaled safe and efficient causative treatment option and should be considered when patients present with symptoms. On long-term follow-up, more than 93% of patients present with an unchanged or improved neurological status after complete removal of the pathology, as compared with their preoperative status. Risk factors for an unfavorable outcome (ASIA Grades A–C) were a low preoperative ASIA grade and a pathology located in the thoracolumbar region. Given the rarity of the ISC and the necessity to be reserved for spinal neurovascular centers offering the complete spectrum of microsurgical therapies including IOM.

**Acknowledgments**

We thank Dr. D. Winkler for his kind support in data collection and Nicklas von Spreckelsen for his support creating graphs.

**References**


Author Contributions
Conception and design: Reitz, Raimund, Fritzsche, Westphal, Eicker. Acquisition of data: Reitz, Burkhardt, Raimund, Fritzsche. Analysis and interpretation of data: Reitz, Vettorazzi, Raimund, Fritzsche, Regelsberger, Eicker. Drafting the article: Reitz, Raimund, Regelsberger, Westphal, Eicker. Critically revising the article: Reitz, Burkhardt, Vettorazzi, Schmidt, Westphal. Reviewed submitted version of manuscript: Reitz. Approved the final version of the manuscript on behalf of all authors: Reitz. Statistical analysis: Reitz. Administrative/technical/material support: Reitz. Study supervision: Reitz, Eicker.

Supplemental Information
Previous Presentation
Portions of this work were presented as proceedings at the 9th Annual Meeting of the German Spine Society held in Leipzig, Germany, on December 12, 2014.

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