Neurosurg Rev (2003) 26:253–256
DOI 10.1007/s10143-003-0260-2

ORIGINAL ARTICLE

Ibrahim Erol Sandalcioglu · Helmut Wiedemayer · Thomas Gasser · Siamek Asgari · Tobias Engelhorn · Dietmar Stolke

Intramedullary spinal cord cavernous malformations: clinical features and risk of hemorrhage

Received: 21 December 2002 / Accepted: 10 January 2003 / Published online: 1 April 2003

Abstract The aim of this study was to review the natural history of symptomatic intramedullary spinal cord (IMSC) cavernous malformations in order to analyze the underlying mechanisms leading to symptoms and determine the potential risk of lesional hemorrhage. Between January 1990 and June 2001, ten consecutive patients with IMSC cavernous malformations were treated surgically in our institution. Age ranged from 17 to 73 years (mean 34.5). All patients became symptomatic due to one or more hemorrhages leading to neurological deficits of different severity, with a more aggressive course for upper cervical lesions. Pre- and postoperative patient condition was classified according to the Frankel scale. Four patients experienced one hemorrhage, four patients two, one patient three, and another one five repeated hemorrhages. The annual retrospective hemorrhage rate for symptomatic IMSC cavernous malformations was 4.5% per patient/year, with a prospective rehemorrhage risk of 66% per patient/year. The postoperative condition was improved in four patients and unchanged in six, and none grew worse. Detailed analysis of history and clinical course in all patients revealed an acute onset of symptoms with subsequent neurological deterioration after each bleeding episode. Based on the significant risk of rehemorrhage and the gratifying functional results, surgery is indicated for symptomatic IMSC cavernous malformations.

I. E. Sandalcioglu (✉) · H. Wiedemayer · T. Gasser · S. Asgari · D. Stolke
Department of Neurosurgery,
University of Essen,
Hufelandstrasse 55, 45122 Essen, Germany
e-mail: sandalcioglu@uni-essen.de
Tel.: +49-201-7231202
Fax: +49-201-7235909

T. Engelhorn
Department of Neuroradiology,
University of Essen,
Essen, Germany

Keywords Cavernous malformation · Clinical presentation · Hemorrhage · Spinal cord · Vascular malformation

Introduction

Cavernous malformations are uncommon vascular disorders of the central nervous system. Most are intracranial, with a predilection for the supratentorial compartment. There is some evidence that deep-seated intracranial cavernous malformations, such as lesions located in the brainstem, are more likely to be symptomatic due to one or more hemorrhages [14]. As reported recently for brainstem cavernous malformations, symptoms commonly were caused by intra- and/or extralesional hemorrhage [16]. The criteria for hemorrhage were acute onset of symptoms with neurological deficits, neuroradiological signs in MRI, and intraoperative observations. Furthermore, we observed a higher risk of rebleeding when the lesions had already been symptomatic. In contrast, the natural history of intramedullary spinal cord (IMSC) cavernous malformations and their potential risk of hemorrhage remain unclear. In 1992, Ogilvy, analyzing a series of six consecutive patients and 30 from the literature, reported four types of clinical presentation [13]. However, the pathophysiological mechanisms of these types is unexplained. Lesional bleedings or local mass effects caused by progressive growth of the cavernous malformation were mentioned as a possible etiology. The present study reviews ten patients with surgically treated IMSC cavernous malformations to evaluate the clinical features and determine the risk of hemorrhage.

Patients and methods

Between January 1990 and June 2001, a total of 79 patients with IMSC lesions were treated surgically in our institution. Of these, ten harbored IMSC cavernous malformations. There were three male and seven female patients. Age ranged from 17 to 73 years, with a mean of 34.5. Clinical and radiological presentation as well

as intraoperative observations were evaluated. All patients presented with one or more hemorrhage events. Our criteria for a hemorrhage episode were acute onset of symptoms, manifestation of neurological deficits, and corresponding MRI findings demonstrating intramedullary hemorrhage. A hemorrhage event was diagnosed only if all features were present. The patient's pre- and postoperative neurological states were classified according to the Frankel scale [7]. Preoperative radiological investigation included MRI in all cases and spinal angiography in two cases. All patients harbored single symptomatic IMSC cavernous malformations of the neuraxis, and multiple lesions were not detected. The annual retrospective hemorrhage rate was calculated based on the frequency of the hemorrhagic events and patient age in years. The risk of rehemorrhage was evaluated based on the sum of rehemorrhages and the observation interval between bleedings. In all patients, surgical removal was performed under standard microsurgical conditions with intraoperative monitoring of somatosensory evoked potentials. All patients were followed up clinically and by spinal MRI. The average follow-up period was 11.1 months (range 5–32).

Results

Clinical presentation

All patients were symptomatic by one or more hemorrhagic episodes with acute onset of clinical signs and neurological deficits. According to the above mentioned criteria, four patients suffered one hemorrhage, four suffered two, one patient three, and another patient five hemorrhages. In cases with repeated hemorrhage, bleeding led to progressive deterioration of the neurological state. Whereas seven patients showed only mild or moderate neurological deficits (Frankel grades C and D) due to subsequent hemorrhages, the two with cavernous malformations in the upper cervical spinal cord presented with severe tetraparesis and respiratory failure, so that treatment in an intensive care unit with respirator therapy was necessary. We observed 17 hemorrhagic episodes in 375 patient-years of life. Thus, the calculated annual retrospective hemorrhage rate for symptomatic IMSC cavernous malformations was 4.5% per patient/year. According to 16 rehemorrhages in six patients during an observation period of 24.3 years, we calculated a rehemorrhage risk of 66% per patient/year. An overview is given in Table 1.

Surgery

Positioning of the patient was selected according to localization of the IMSC cavernous malformation. Six patients were operated on in a semisitting position and four in a prone position. As a surgical approach, laminectomy was performed in three patients and osteoplastic laminotomy with reconstruction of the posterior spine in seven. Once the dura was opened and the spinal cord exposed, a circumscribed bluish area was visible on the pial surface in seven of the ten patients caused by a subpially located intramedullary hematoma. The spinal cord was markedly swollen due to a hematoma in two of three cases without visible discoloration of the pial surface and in two cases with visible discoloration. In six cases, the lesions were intrinsic, without notable bulging of the spinal cord. Furthermore, abnormal venous vessels

Table 1 Data of patients operated on IMSC cavernous malformation

Case no.	Age (years), sex	Localization	No. of hemorrhages	Duration of symptoms (months)	Preoperative Frankel-grade	Postoperative Frankel-grade	Follow-up (months)
1	48, F	T1–2	2	4	D	D	32
2	47, F	C4–5	2	72	D	D	5
3	28, M	C0–2	2	1	A	D	6
4	24, F	C7	2	3	D	D	8
5	37, M	T3–4	1	5	D	D	6
6	35, F	T1–2	3	168	D	D	12
7	73, M	T12	1	0.5	C	D	12
8	34, F	C2	1	0.5	B	D	17
9	32, F	C6–7	5	36	C	D	7
10	17, F	T7–8	1	0.5	D	D	6

Table 2 Intraoperative findings after dura opening

Case no.	Discoloration	Spinal cord bulging	Venous anomalies
1	+	–	–
2	+	+	–
3	–	+	+
4	+	+	+
5	+	–	–
6	+	–	+
7	+	–	–
8	–	+	–
9	+	–	–
10	–	–	–

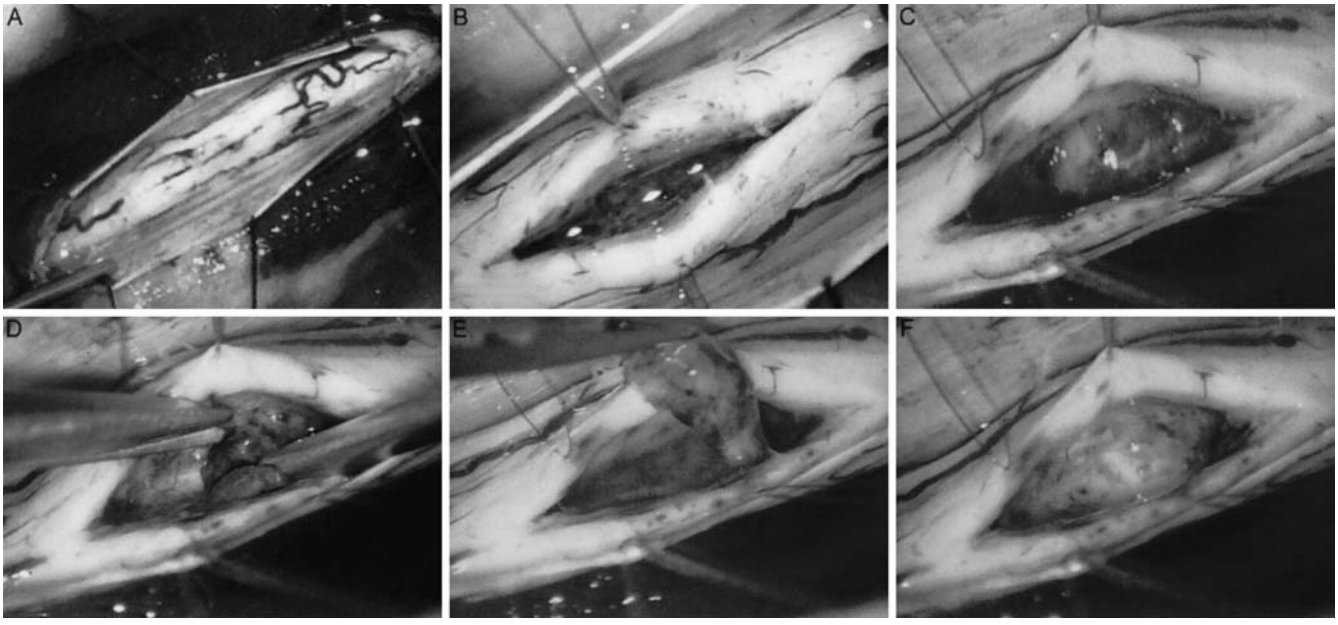

Fig. 1A–F Case 10. Intraoperative photographs of a 17-year-old female presenting with acute onset of back pain and left leg numbness 2 weeks prior to operation. **A** Operative site after T6–8 laminotomy and dura opening show no visible abnormalities on the spinal cord surface. **B–E** After myelotomy, the cavernous malformation was exposed and removed completely following evacuation of the surrounding hematoma. **F** Note preservation of the hemosiderin-stained gliotic tissue

were seen on the spinal cord surface in three cases. Thus, in nine cases, some abnormalities on the operating site were visible (Table 2). Only one patient with a deep-seated, ventrolaterally located lesion showed no abnormalities after exposure of the spinal cord. Although we generally use a midline myelotomy to access intramedullary lesions, in nine cases the entry point to approach the cavernous malformation was guided by these intraoperative observations. Intraoperative ultrasound was used to visualize the extent of the intramedullary lesion and to determine the entry point in the case with no abnormalities on the exposed medullary surface (Fig. 1). In all cases, the cavernous malformation was removed completely after evacuation of the intramedullary hematoma and diagnosis was confirmed histopathologically.

Postoperative course and outcome

Postoperative course was uneventful in all cases, and we observed no complications related to surgical procedure. Transient mild neurological deterioration after surgery occurred in five cases. In one patient, the neurological state was unchanged. Four patients experienced significant improvement of the neurological state in the early postoperative course. We observed an inverse correlation between preoperative neurological state and immediate postoperative course: the worse the preoperative condition, the better the immediate postoperative improvement. At the last follow-up, the neurological state had improved over the preoperative condition in four patients, was unchanged in six, and had worsened in none.

Discussion

Numerous reports comment on the natural history of intracranial cavernous malformations. In contrast, the number of reported cases with IMSC cavernous malformations is small. Mostly, isolated cases or smaller series were reported [1, 2, 3, 4, 8, 9, 10, 11, 12, 13, 17, 18, 21]. Few authors presented series of more than ten cases [5, 6, 19, 20].

In his analysis of 36 patients with symptomatic IMSC cavernous malformations, including six of his own patients, Ogilvy described four types of clinical presentation for symptomatic IMSC cavernous malformation:

- Type 1, with discrete episodes of neurological deterioration
- Type 2, with slow neurological decline
- Type 3, with acute onset of symptoms and rapid decline
- Type 4, with acute onset of mild symptoms and subsequent gradual decline

Except for type 2, the clinical presentation appears to be caused by some kind of acute event leading to neurological deficits of different severity and course. Consequently, it seems reasonable to distinguish two subgroups. One is characterized by slowly progressive myelopathy. These symptoms are probably caused by several minor bleedings or progressive growth of the cavernous malformation leading to local mass effects. The other subgroup presents with major hemorrhage, which causes a sudden onset of symptoms and neurolog-

ical deficits due to bleeding into the cavernous malformation or adjacent neural tissue. The extent of bleeding and its localization determine the clinical presentation in this group according to Ogilvy types 1, 3, and 4.

Whereas the clinical and neurological course of IMSC cavernous malformations is well documented in the literature, surprisingly, detailed analysis of the mechanisms leading to these symptoms and of the risk of hemorrhage for those lesions is lacking. In our series, thorough analysis of history and clinical course revealed that all patients became symptomatic by one or more hemorrhagic events leading to an acute onset of symptoms with neurological deficits. Bleeding was confirmed in all cases by both radiological findings in MRI and intraoperative observations.

Although our number of cases is small, these data shed light on the clinical course of these lesions, the relative risk of hemorrhage, and its effect on neurological condition. With a calculated retrospective annual hemorrhage rate of 4.5% per person/year for symptomatic IMSC cavernous malformations, we observed a risk of hemorrhage similar to that reported for cavernous malformations in the brainstem [15, 16]. With 16 rehemorrhages in six patients, we calculated a rehemorrhage risk of 66% per patient/year. These figures clearly argue for increased risk of neurological deterioration for cases in which the IMSC cavernous malformation had already been symptomatic. An increasing risk for rehemorrhage with neurological deterioration was also observed by other authors [6, 11, 21], although figures determining the risk of hemorrhage and rehemorrhage are lacking.

As reported by Vishteh [20], ten of 17 patients presented with myelopathy, six with radiculopathy, and one with a conus medullaris syndrome. Interestingly, 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.

Two cases treated surgically in our institution and included in the present series were already published in 1991 [12]. One of these patients presented with five hemorrhage episodes during an observation period of 3 years until she was referred to our institution.

Although the decision to operate on patients with only mild symptoms with no significant functional impairment is often difficult, some important arguments favor early surgical intervention. Considering the high risk of rebleeding, the significant morbidity associated with cavernous malformations of the cervical spinal cord, and the gratifying surgical results, in our opinion surgery is indicated for symptomatic IMSC cavernous malformations, although some transient deterioration of the preexisting neurological deficits is to be expected in the early postoperative period.

References

1. Anson JA, Spetzler RF (1993) Surgical resection of intramedullary spinal cord cavernous malformations. J Neurosurg 78:446–451
2. Barnwell SL, Dowd CF, Davis RL, Edwards MSB, Gutin PH, Wilson CB (1990) Cryptic vascular malformations of the spinal cord: diagnosis by magnetic resonance imaging and outcome of surgery. J Neurosurg 72:403–407
3. Canavero S, Pagni CA, Duca S, Bradac GB (1994) Spinal intramedullary cavernous angiomas: a literature meta-analysis. Surg Neurol 41:381–388
4. Cantore G, Delfini R, Cervoni L, Innocenzi G, Orlando ER (1995) Intramedullary cavernous angiomas of the spinal cord: report of six cases. Surg Neurol 43:448–452
5. Crisante L, Herrmann HD (1998) Radical excision of intramedullary cavernous angiomas. Neurosurgery 43:424–430
6. Deutsch H, Jallo GI, Faktorovich A, Epstein F (2000) Spinal intramedullary cavernoma: clinical presentation and surgical outcome. J Neurosurg (Spine) 93:65–70
7. Frankel H (1969) The postural reduction in the initial management of closed injuries of the spine with paraplegia and tetraplegia. Paraplegia 7:179–192
8. Harrison MJ, Eisenberg MB, Ullman JS, Oppenheim JS, Camins MB, Post KD (1995) Symptomatic cavernous malformations affecting the spine and spinal cord. Neurosurgery 37:195–204
9. Lopate G, Black JT (1990) Cavernous hemangioma of the spinal cord. report of 2 unusual cases. Neurology 40:1791–1793
10. Lunardi P, Acqui M, Ferrante L, Fortuna A (1994) The role of intraoperative ultrasound imaging in the surgical removal of intramedullary cavernous angiomas. Neurosurgery 34:520–523
11. McCormick PC, Michelsen WJ, Post KD, Carmel PW, Stein BM (1988) Cavernous malformations of the spinal cord. Neurosurgery 23:456–463
12. Mehdorn HM, Stolke D (1991) Cervical intramedullary cavernous angioma with MRI-proven haemorrhage. J Neurol 238:420–426
13. Ogilvy CS, Louis DN, Ojemann RG (1992) Intramedullary cavernous angiomas of the spinal cord: clinical presentation, pathological features, and surgical management. Neurosurgery 31:219–230
14. Porter PJ, Willinsky RA, Harper W, Wallace MC (1997) Cerebral cavernous malformations: natural history and prognosis after clinical deterioration with and without hemorrhage. J Neurosurg 87:190–197
15. Porter RW, Detwiler PW, Spetzler RF, Lawton MT, Baskin JJ, Derksen PT, Zabramski JM (1999) Cavernous malformations of the brainstem. Experience with 100 patients. J Neurosurg 90:50–58
16. Sandalcioglu IE, Wiedemayer H, Secer S, Asgari S, Stolke D (2002) Surgical removal of brain stem cavernous malformations: surgical indication, technical considerations, and results. J Neurol Neurosurg Psychiatry 72:351–355
17. Spetzger U, Gilsbach JM, Bertalanffy H (1995) Cavernous angiomas of the spinal cord. Clinical presentation, surgical strategy, and postoperative results. Acta Neurochir (Wien) 134:200–206
18. Tu YK, Liu HM, Chen SJ, Lin SM (1999) Intramedullary cavernous hemangiomas: clinical features, imaging diagnosis, surgical resection and outcome. J Clin Neurosci 6:212–216
19. Vishteh AG, Sankhla S, Anson JA, Zabramski JM, Spetzler RF (1997) Surgical resection of intramedullary spinal cord cavernous malformations: delayed complications, long-term outcomes, and association with cryptic venous malformations: Neurosurgery 41:1094–1100
20. Vishteh AG, Zabramski JM, Spetzler RF (1999) Patients with spinal cord cavernous malformations are at an increased risk for multiple neuroaxis cavernous malformations. Neurosurgery 45:30–32
21. Zentner J, Hassler W, Gawehn J, Schroth G (1989) Intramedullary cavernous angiomas. Surg Neurol 31:64–68