ORIGINAL ARTICLE

The clinical outcomes after complete surgical resection of intramedullary cavernous angiomas: changes in motor and sensory symptoms

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Study design: Evaluation of the surgical effects of patients with intramedullary spinal cord cavernous angiomas (CAs).

Objectives: To assess the sensory and motor deficits in the clinical symptoms of patients with intramedullary CAs after complete surgical resection.

Setting: Seoul National University Hospital, Korea.

Methods: The authors reviewed 14 patients with intramedullary spinal cord CAs who underwent surgical resection between 1995 and 2006. The average follow-up duration was 55.0 ± 45.1 months. To analyze the changes in the patients' symptoms after surgery, the ASIA (American Spinal Injury Association) and JOA (Japanese Orthopedic Association) scoring systems were used at different time points: preoperative, postoperative and during the final follow-up period.

Results: The patients' mean ASIA score at follow-up was increased in comparison to the preoperative score (97.2 ± 5.3 vs 95.2 ± 7.6 , $P = 0.201$), but this difference was not significant.

The mean JOA motor score at follow-up was slightly increased in comparison to the preoperative score (7.2 ± 1.3 vs 6.8 ± 1.3 , $P = 0.107$). There was a significant difference between the patients' preoperative and follow-up JOA sensory score (4.3 ± 1.0 vs 4.9 ± 0.9 , $P = 0.007$). The rates of complete resolution of motor and sensory deficits were 45.5% (5/11 patients) and 7.1% (1/14 patients), respectively.

Conclusion: Although complete surgical resection could improve the clinical symptoms of intramedullary CAs, the sensory deficits usually remain in the long-term period after complete resection.

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Introduction

Intramedullary cavernous angiomas (CAs) account for 5–12% of all spinal vascular abnormalities.¹ Spinal intramedullary CAs usually occur at the thoracic levels during the third and fourth decade of life, with a slight female predominance of 1.5:1.² The hemorrhage rate of symptomatic intramedullary CAs is 1.4–4.5% per lesion and per year.^{1,3}

There might be several treatment strategies in spinal intramedullary CAs. Although radiosurgery has been used in the treatment of intracranial CAs, no data exist for spinal intramedullary CAs.⁴ Because the natural history of

spinal intramedullary CAs is uncertain yet, surgical removal of asymptomatic spinal intramedullary CAs is still under debate.¹ However, only surgical removal of spinal intramedullary CAs in symptomatic patients provides a way to stop progression and to reduce the neurological symptoms.^{1,5}

In terms of surgical outcome, 66% of patients showed improvement following the removal of spinal intramedullary CAs and 28% of patients showed no change.¹ However, long-term control or improvement in pain severity was achieved in only 50% of patients.⁶ Most reports concerning the surgical resection of intramedullary CAs have not addressed the changes in motor and sensory symptoms separately.^{1,5,6} To fully determine the realistic picture of surgical outcome, we evaluated the changes in motor and sensory symptoms separately after complete surgical resection using quantitative measures.

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Table 1 Summary of 14 patients with intramedullary cavernous angiomas

Case	Age (yrs), Sex	Symptom ^a	Symptom duration (months)	Location	Clinical presentation	Myelotomy/hemorrhage	Operation	ASIA (JOA)			Sensory disturbance		FU (months)
								Preop.	Postop.	FU	Postop.	FU	
1	40, M	M, S	6	C1/C	B	Midline/Y	GTR	95 (15)	93 (15)	98 (15)	Aggravated	Stationary	19
2	34, F	S	48	C2-3/DC	A	Midline/Y	GTR	100 (14)	100 (15)	100 (16)	Improved	Improved	130
3	21, M	M, S	2	C4-5/DC	C	Midline/Y	GTR	96 (14)	96 (15)	100 (16)	Improved	Improved	57
4	44, M	M, S	120	C5-6/DC	B	Midline/N	GTR	95 (13)	100 (15)	100 (16)	Improved	Improved	9
5	42, F	S	48	T1-2/DC	B	Midline/Y	GTR	100 (15)	100 (16)	100 (16)	Improved	Stationary	130
6	35, M	M, S	48	T3/C	B	DL/N	GTR	97 (14)	74 (9)	87 (12)	Aggravated	Stationary	86
7	34, F	S	16	T5-6/L	B	DL/Y	GTR	100 (16)	99 (15)	99 (15)	Improved	Stationary	69
8	47, M	M, S	32	T5-6/DC	C	Midline/Y	GTR	78 (11)	90 (12)	99 (16)	Improved	Improved	48
9	46, M	M, S	4	T9/L	C	Midline/Y	GTR	99 (14)	100 (16)	100 (16)	Improved	Stationary	109
10	25, F	M, S	48	T9-10/C	B	Midline/N	GTR	78 (7)	84 (8)	84 (8)	Improved	Stationary	6
11	23, M	S	48	CM/DC	A	Midline/N	GTR	100 (16)	98 (15)	100 (16)	Improved	Improved	40
12	33, M	S	10	CM/DC	A	Midline/N	GTR	100 (15)	100 (16)	100 (16)	Improved	Stationary	4
13	42, F	M, S	14	CM/L	A	DL/N	GTR	99 (15)	99 (15)	100 (17)	Improved	Intact	6
14	14, M	M, S	10	T5-6/VC	B	Midline/Y	GTR	96 (14)	78 (7)	94 (13)	Aggravated	Improved	57

Abbreviations: ASIA, American Spinal Injury Association; C, most cross-section area of spinal cord; CM, conus medullaris; D, dorsal side of spinal cord; DC, dorso-central side of spinal cord; DL, Dorsolateral; FU, follow-up; GTR, gross total removal; JOA, Japanese Orthopedic Association; L, lateral side of spinal cord; midline, dorsal midline; Preop, preoperative status; Postop, postoperative status; VC, ventro-central side of spinal cord; Y, yes; N, no.

^aM, motor weakness; S, sensory disturbance (paresthesia, hypesthesia).

Figure 1 Schematic illustrations of myelotomy (arrow) according to cavernous angioma (CA) location. (a) If intramedullary CA was located dorsocentral or ventrocentral areas, dorsal midline myelotomy was performed. (b) If intramedullary CA was located laterally, dorsolateral myelotomy was performed. (c) If intramedullary CA occupied most of the cross-sectional area of cord, either dorsal midline or dorsolateral myelotomies was chosen depending on the abnormality of its cord surface.

Materials and methods

The clinical records of all patients with intramedullary spinal cord CAs who underwent surgical resection between 1995 and 2006 were retrospectively reviewed. In total, 14 patients were pathologically proven to have CAs after surgery. This study included 9 men and 5 women, with a mean age of 34.3 ± 10.2 years (range 14–72 years).

A dorsal approach was performed by laminoplastic laminotomy or laminectomy. Dorsal midline or dorsolateral myelotomies were performed mainly over discolored dorsal midline or dorsolateral surfaces of the spinal cord, and the lesion was totally removed under microscopic view.

Clinical outcomes were evaluated during the preoperative, postoperative and final follow-up periods using the motor

scale of the ASIA (American Spinal Injury Association) impairment classification system and JOA (Japanese Orthopedic Association) scoring system.^{7,8} The motor function in ASIA scale is evaluated through the testing of a key muscle in 10 paired myotomes and the strength of each muscle is graded on a six-point scale (0–5 score).⁷ Therefore the highest score (100) corresponds to normal active movement and the lowest score (0) corresponds to total paralysis.⁷ The severity of the spondylotic myelopathy is assessed according to the JOA scoring system that consists of three parts; the motor functions of upper and lower extremities, the sensory functions of upper and lower extremities and trunk, and the bladder function.⁸ The highest score (17) corresponds to no myelopathy and the lowest score (0) corresponds to complete cord injury.⁸

Clinical information was gathered by outpatient review and telephone interviews. The mean follow-up duration was 55.0 ± 45.1 months (4–130 months).

Statistical analyses were performed using the Wilcoxon signed-rank test. The significance level was defined as $P=0.05$.

Results

Clinical presentations and location of lesions

The overall median duration of pretreatment symptoms was 32.4 ± 31.5 months (range 2–120 months). All patients suffered from progressive clinical symptoms and signs. We identified the three patterns of presentation published by Zevgaridis *et al.*⁹ Four patients (A, 29%) showed multiple episodes of discrete neurological deterioration with varying degrees of recovery between acute attacks. Seven patients (B, 50%) showed slow progression of neurological deterioration and three patients (C, 21%) experienced sudden-onset symptoms with rapid decline or gradual worsening. Nine patients had motor and sensory deficits preoperatively. Five patients had only sensory deficits during the preoperative period. None of the patients showed motor deficits alone during the preoperative period.

Magnetic resonance imaging (MRI) scans on admission revealed four CAs located in the cervical area, seven in the thoracic area and three in the conus medullaris area. The locations of lesions within the spinal cord were ventrocentral in one patient (case 14), dorsocentral in seven patients and lateral in three patients (cases 7, 9 and 13). Three patients (cases 1, 6 and 10) had intramedullary CAs (CAs) occupying in majority of the cross-section area of the spinal cord (Table 1; Figure 1). Eight patients (57.1%, 8/14 patients) had hemorrhages on MRI or in the surgical field.

Surgical management

Dorsal midline myelotomy was performed in 11 cases, and dorsolateral myelotomy was performed in 3 cases. The myelotomy site was dependent on the location of the intramedullary CA with discoloration of the spinal cord surface on dural opening (Figure 1).

Dorsal midline myelotomy

In total, 11 patients with intramedullary CAs (8 cases were located in dorso- or ventrocentral area, 1 in the lateral area and 2 primarily in the cross-section area) underwent dorsal midline myelotomy.

Case 9 underwent dorsal midline myelotomy because the lateral surface of the spinal cord appeared to be normal. Cases 1 and 10, in which dorsal midline myelotomy was performed, had the intramedullary CAs located primarily in the whole cross-sectional area with discoloration of the spinal cord surfaces in the dorsal midline area.

Dorsolateral myelotomy

Three patients (cases 6, 7 and 13) underwent dorsolateral myelotomy. In case 6, in which the lesion was occupying the entire spinal cord, the right side of the spinal cord surface

Figure 2 Graph represents the change of clinical deficit measured by JOA (Japanese Orthopedic Association) (a) and ASIA (American Spinal Injury Association) (b) scoring system after surgery compared with last follow-up. Asterisk depicts statistically significant differences between preoperative status and follow-up status ($P<0.05$). Preop, preoperative status; Postop, postoperative status; FU, follow-up status.

had a discolored and bulging appearance in comparison to the left side. Dorsolateral myelotomy was performed, and the CA was completely removed.

Case 7 underwent microsurgical resection at another hospital 10 years earlier and had a recurrent mass laterally in the spinal cord. There was a severe adhesion between the cord and dura. The spinal cord was rotated so that the dorsolateral roots would be visible in the midline. Discoloration of the spinal cord surface was seen laterally. In case 13, having the intramedullary CAs located laterally, discoloration of the spinal cord surfaces was identified laterally, and the CA was completely removed after dorsolateral myelotomy.

Figure 3 Graph represents the change of motor (a) and sensory (b) deficits measured by JOA (Japanese Orthopedic Association) scoring system after surgery compared with last follow-up. Asterisks depict statistically significant differences ($P < 0.05$). Dagger depicts statistically insignificant difference ($P > 0.05$). Preop, preoperative status; Postop, postoperative status; FU, follow-up status.

Following a small myelotomy, the lesions were removed in an inside out fashion until the gliotic white matter was identified. The lesions were carefully examined after resection to ensure that no residual CAs remained. All patients underwent complete microsurgical removal.

Clinical outcomes

The patients' JOA and ASIA scores decreased postoperatively and increased during the follow-up period (Figure 2). There was a significant difference between the preoperative and long-term follow-up JOA scores (13.8 ± 2.3 vs 14.9 ± 2.4 , $P = 0.046$). The long-term follow-up ASIA score increased more than the preoperative score (97.2 ± 5.3 vs 95.2 ± 7.6 , $P = 0.201$), but this difference was not significant.

The follow-up JOA motor score was slightly higher than the preoperative score (7.2 ± 1.3 vs 6.8 ± 1.3 , $P = 0.107$). There was a significant difference between the preoperative and follow-up JOA sensory scores (4.3 ± 1.0 vs 4.9 ± 0.9 , $P = 0.007$) (Figure 3).

With regard to long-term outcomes, the ASIA scores of 11 (78.6%, 11/14) patients increased or remained the same (50%, 7/14 patients and 28.6%, 4/14 patients, respectively), and the JOA scores of 11 (78.6%, 11/14) patients increased or remained the same (64.3%, 9/14 patients and 14.3%, 2/14 patients, respectively).

Two patients (cases 7 and 11) who showed only sensory deficits during the preoperative period had postoperative motor weakness. The postoperative motor weakness in case 7 persisted until the last follow-up but the postoperative motor weakness in case 11 had completely resolved at the final follow-up.

Four patients (cases 1, 6, 7 and 14) deteriorated over the course of the follow-up period. The deteriorations in motor function observed postoperatively in cases 6, 7 and 14 did not improve to the preoperative level. Case 6 could not move without a wheelchair for paraparesis and required occasional assistance. Case 7 complained that he felt the shuffle of left foot during walking. Case 14 walked with

axillary crutches for paraparesis and had paresthesia below T6 dermatome. Similarly, the deterioration in sensory function observed postoperatively in cases 1 and 14 did not recover to the preoperative level. Although case 1 had paresthesia on left upper and lower extremities, he could carry on normal daily activity.

The rate of complete resolution of motor and sensory deficits was 45.5% (5/11 patients) and 7.1% (1/14 patients), respectively.

Discussion

Intramedullary CA is relatively rare and constitutes 5% of vascular malformations.^{10,11} Patients can present with acute neurological deficits that occur because of a hemorrhage within the spinal cord parenchyma.¹² The bleeding rate is $< 5\%$ per lesion per year and it is assumed that these lesions were present at birth in most patients.^{1,3} In our study, eight bleedings (57.1%, 8/14 cases) in 14 cases resulted in an average bleeding rate of 1.7% per lesion per year. The mechanisms of slow progressive myelopathy are microhemorrhages and resultant reactive gliosis.⁵

Case 4, having the longest symptom duration in this series, presented with slow progressive symptoms during 120 months. Under operative view after myelotomy, there was no gross hemorrhage inside CA, though mass looked yellow and brown in color. Pathologic examination showed fibrous soft tissue with hemosiderin pigment and blood. These findings suggest that the causes of slow progressive myelopathy might be microhemorrhages and resultant reactive gliosis.

Clinical outcomes

Because the patients' response to surgical treatment for intramedullary CAs are allegedly to be favorable, many reports recommend surgical treatment in symptomatic intramedullary CAs.^{1,5,11,13–15} In previous reports, the long-term outcome rates after surgical resection were good

in 61–66% of patients, fair in 28–31% of patients and poor in 6–8% of patients.^{1,5} However, another study reported that the levels of pain improved in 11 (52%) patients, remained at the preoperative baseline level in 9 (43%) patients and worsened in 1 (5%) patient during the follow-up period.⁸ In our study, the surgical outcomes were good or fair in 78.6% of the patients and poor in 21.4% of the patients, and the rate of complete resolution of motor and sensory deficits were 45.5% (5/11 patients) and 7.1% (1/14 patients), respectively. Most of the patients' sensory symptoms persisted throughout the long-term follow-up period.

Factors related to poor outcome

Certain causes are associated with poor outcomes. The prognostic factors of poor outcomes reported in the literature are preoperative status and symptom duration.^{1,4} However, three of the patients (cases 6, 7 and 14) with poor outcomes in our study did not have a long symptom duration or poor preoperative status in comparison to the other patients. Although approach through a short distance to the lesion is a basic concept of surgical removal, dorsolateral myelotomy could induce postoperative neurological deficits by insulting dorsal root afferents and the lateral corticospinal tract. In our study, two (cases 6 and 7) of the three patients with dorsolateral myelotomies had poor outcomes. CAs located on the ventral side of the spinal cord are associated with a risk of additional neurological deficits after surgery.¹⁶ In case 14, with a ventrally located CA, dorsal midline myelotomy was performed and complete removal was achieved. Additional neurological deficits were detected during the follow-up period in some patients. Previous studies have good outcomes using the ventral approach at the cervical and thoracic levels.^{16–18} If CAs are located ventrally, especially when present superficially, ventral approach could provide direct approach into CA without myelotomy and avoid additional neurological deficits following dorsal or dorsolateral myelotomy. However, when CAs are deep to the surface, ventral approach also has risks damaging corticospinal tract, resulting in additional neurological deficit. Also ventral approach leads to reconstruction following corpectomy. There are also some risks such as possible bleeding from the venous plexus and damage to the vertebral artery.¹⁶

CAs with a well-defined gliotic plane between the lesion and normal cord along with discoloration of the spinal cord surface could be completely removed without surgical insult. The remaining gliotic rim could be the cause of the low recovery rate because it may induce adjacent neural firing after surgery.⁹ However, we believe that the removal of the gliotic rim could cause postoperative neurological deficits by the injuring the normal cord.

The following two explanations might explain the remaining sensory symptoms after complete resection of intramedullary CAs. Following dorsal or dorsolateral myelotomy, the dorsal columns are more vulnerable than the ventrally or laterally located motor fibers. Also the hemosiderin-laden macrophages in gliotic rim left after surgery could induce

further damage to the sensory tracts resulted in haphazard neuronal firing that can result in paresthesia.

Because this was a retrospective study with a small number of patients, it had some limitations that make it difficult to draw a definitive conclusion from the study results. However, our results suggested that the surgical outcomes in patients with intramedullary CAs may not be as good as the outcomes reported in previous studies especially in terms of sensory deficits.

Conclusion

Surgical removal of an intramedullary CA can be used to reduce the severity of neurological deficits. Although the clinical outcome showed significant improvement, sensory symptoms remained in the majority of patients after complete resection.

References

- 1 Zevgaridis D, Medele RJ, Hamburger C, Steiger HJ, Reulen HJ. Cavernous haemangiomas of the spinal cord. A review of 117 cases. *Acta Neurochir (Wien)* 1999; **141**: 237–245.
- 2 Polymeropoulos MH, Hurko O, Hsu F, Rubenstein J, Basnet S, Lane K *et al*. Linkage of the locus for cerebral cavernous hemangiomas to human chromosome 7q in four families of Mexican-American descent. *Neurology* 1997; **48**: 752–757.
- 3 Sandalcioğlu IE, Wiedemayer H, Gasser T, Asgari S, Engelhorn T, Stolke D. Intramedullary spinal cord cavernous malformations: clinical features and risk of hemorrhage. *Neurosurg Rev* 2003; **26**: 253–256.
- 4 Canavero S, Pagni CA, Duca S, Bradac GB. Spinal intramedullary cavernous angiomas: a literature meta-analysis. *Surg Neurol* 1994; **41**: 381–388.
- 5 Jallo GI, Freed D, Zareck M, Epstein F, Kothbauer KE. Clinical presentation and optimal management for intramedullary cavernous malformations. *Neurosurg Focus* 2006; **21**: e10.
- 6 Kim LJ, Klopstein JD, Zabramski JM, Sonntag VK, Spetzler RF. Analysis of pain resolution after surgical resection of intramedullary spinal cord cavernous malformations. *Neurosurgery* 2006; **58**: 106–111; discussion 106–111.
- 7 Maynard Jr FM, Bracken MB, Creasey G, Ditunno Jr JF, Donovan WH, Ducker TB *et al*. International standards for neurological and functional classification of spinal cord injury. American Spinal Injury Association. *Spinal Cord* 1997; **35**: 266–274.
- 8 Okada K, Shirasaki N, Hayashi H, Oka S, Hosoya T. Treatment of cervical spondylotic myelopathy by enlargement of the spinal canal anteriorly, followed by arthrodesis. *J Bone Joint Surg Am* 1991; **73**: 352–364.
- 9 Dey DD, Landrum O, Oaklander AL. Central neuropathic itch from spinal-cord cavernous hemangioma: a human case, a possible animal model, and hypotheses about pathogenesis. *Pain* 2005; **113**: 233–237.
- 10 Bakir A, Savas A, Yilmaz E, Savas B, Erden E, Caglar S *et al*. Spinal intradural-intramedullary cavernous malformation. Case report and literature review. *Pediatr Neurosurg* 2006; **42**: 35–37.
- 11 Spetzger U, Gilsbach JM, Bertalanffy H. Cavernous angiomas of the spinal cord clinical presentation, surgical strategy, and postoperative results. *Acta Neurochir (Wien)* 1995; **134**: 200–206.
- 12 Ogilvy CS, Louis DN, Ojemann RG. Intramedullary cavernous angiomas of the spinal cord: clinical presentation, pathological features, and surgical management. *Neurosurgery* 1992; **31**: 219–229; discussion 229–230.
- 13 Kondziella D, Brodersen P, Laursen H, Hansen K. Cavernous hemangioma of the spinal cord—conservative or operative management? *Acta Neurol Scand* 2006; **114**: 287–290.

- 14 Obermann M, Gizewski ER, Felsberg J, Maschke M. Cavernous malformation with hemorrhage of the conus medullaris and progressive sensory loss. *Clin Neuropathol* 2006; **25**: 95–97.
- 15 Vishteh AG, Sankhla S, Anson JA, Zabramski JM, Spetzler RF. Surgical resection of intramedullary spinal cord cavernous malformations: delayed complications, long-term outcomes, and association with cryptic venous malformations. *Neurosurgery* 1997; **41**: 1094–1100; discussion 1100–1091.
- 16 Nishikawa M, Ohata K, Ishibashi K, Takami T, Goto T, Hara M. The anterolateral partial vertebrectomy approach for ventrally located cervical intramedullary cavernous angiomas. *Neurosurgery* 2006; **59**: ONS58–ONS63; discussion ONS58–63.
- 17 Cantore G, Delfini R, Cervoni L, Innocenzi G, Orlando ER. Intramedullary cavernous angiomas of the spinal cord: report of six cases. *Surg Neurol* 1995; **43**: 448–451; discussion 442–451.
- 18 Fontaine D, Lot G, George B. Intramedullary cavernous angioma. Resection by oblique corpectomy. *Surg Neurol* 1999; **51**: 435–441; discussion 432–441.