




CASE REPORT

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Surgical management of cervical intramedullary cavernoma: case report and systematic review of the literature

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Abstract

Background Cavernous malformations can occur throughout the cerebral nervous system, most commonly located in the supratentorial compartment. The intramedullary location is rare and accounts for approximately 2.4% to 5% of all spinal vascular tumors. The cervical cord location and its clinical manifestations are underreported.

Methods The authors report a case of spinal cord compression at the C5–C6 level by a cervical intramedullary cavernoma with a systematic review of the literature according to the Preferred Reporting Items for Systematic Reviews and meta-analysis guidelines. Relevant studies (1980 to 2023) that reported patients with cervical intramedullary cavernoma were identified from PubMed databases.

Results A total of 29 studies reporting 423 patients were included in this study with a mean age of 40.5 ± 6.3 years old. The overall male-to-female ratio was 1:2.5 with a median duration of follow-up of 46 months [18.5–63.9]. The type of resection as well as the patient's outcome was also reported.

Conclusion Intramedullary cavernous angiomas are rare. Early surgical total resection of the symptomatic lesions in adequate conditions is crucial for good outcomes.

Keywords Cavernous angioma, Intramedullary lesion, Spinal cord, Case report

Introduction

Seldom do intramedullary cavernous malformations of the spine occur; they make up only 3–5% of cavernous malformations and 5–12% of all intraspinal vascular malformations [1]. The peak of the disease occurs in the fourth decade of life, which is comparable to the highest incidence of cerebral cavernomas [2]. Compared to men, women are impacted more frequently [3, 4].

The spaces inside spinal cavernomas are filled with blood and endothelium; they are surrounded by

thickened, hyalinized walls devoid of smooth muscle and elastic fibers [5, 6]. Cavernous malformations of the spinal cord are less well studied than those of the brain, especially when it comes to treatment and management. Because of their lower tolerance to mass lesions in the spine, they typically have a more aggressive clinical course than lesions in the brain. These lesions are prone to recurrent bleeding and progressive growth. A mechanism for the underlying acute episodes of neurological deterioration has been proposed: bleeding episodes. Gliosis and microbleeds can both lead to progressive myelopathy.

Ogilvy *et al.* [7] have reported that the most common symptoms include pain, weakness, and paresthesia. The literature has identified four distinct patterns of clinical presentation: (1) isolated episodes of neurological decline

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with varying degrees of recovery between episodes; (2) gradual neurological decline progression; (3) acute onset of symptoms with quick resolution; and (4) acute onset of mild symptoms followed by gradual resolution over weeks to months. In this study, we report the rare case of a 37-year-old man who underwent successful resection of cervical intramedullary cavernoma of the C5–C6 level.

Case presentation

Patient information

A 37-year-old man with a six-month history of progressively worsening neck pain and numbness in his right hand was admitted to our department because the pain became persistently unbearable and did not respond to standard pain medications. The patient is receiving medication for type 2 diabetes mellitus. He denied any alcohol or tobacco use, and there was no history of trauma.

Clinical findings

A neurological examination revealed no motor deficit in any extremities. He had decreased light touch and proprioceptive sensitivity in the right upper extremity. Deep tendon reflexes were exaggerated bilaterally in the upper and lower extremities. A bilateral Babinski and Lhermitte sign was positive.

Diagnosis assessment

Magnetic resonance imaging (MRI) scan of the spine revealed a 15-mm-sized intramedullary lesion at the C5–C6 level with mixed signal on T1WI and T2WI sequences. A popcorn-like lesion was characterized by a hyperintense core surrounded by a hypointense rim on the sagittal T1WI and axial T2WI suggesting the diagnosis of intramedullary cavernoma (Fig. 1).

Therapeutic intervention

The patient was operated on. The head was placed in the Mayfield head holder, while the patient was in the prone position and under general anesthesia. To protect peripheral nerves and reduce pressure on the chest and abdomen, all pressure points have been adequately padded. The posterior approach was used to perform laminectomies of C4, C5, and C6. Evoked somatosensory potentials are tracked. A baseline test was performed before dural opening to allow an informative interpretation of the intraoperative recordings. The dura mater was opened, a midline myelotomy was performed, and a well-circumscribed dark blue lesion was visualized spreading the posterior columns laterally to the right. The lesion was then completely removed “en bloc,” with the hemosiderin rim serving as a guide for the cleavage plane of the lesion. After the removal of the lesion, a thorough examination of the cavity revealed no evidence of residual deformity. Neither motor-evoked potentials nor somatosensory perception changed during the procedure. The histopathological findings were consistent with a cavernous angioma (Fig. 2).

Follow-up

Postoperatively, even though the severe neck pain has subsided, the patient experienced a weakness in the right upper extremity, which improved over eight months of physiotherapy. A month follow-up cervical spine magnetic resonance imaging demonstrated no residual lesion.

Methods

A systematic review of all the previously reported intramedullary cavernomas of the cervical spinal cord was performed according to the PRISMA guidelines

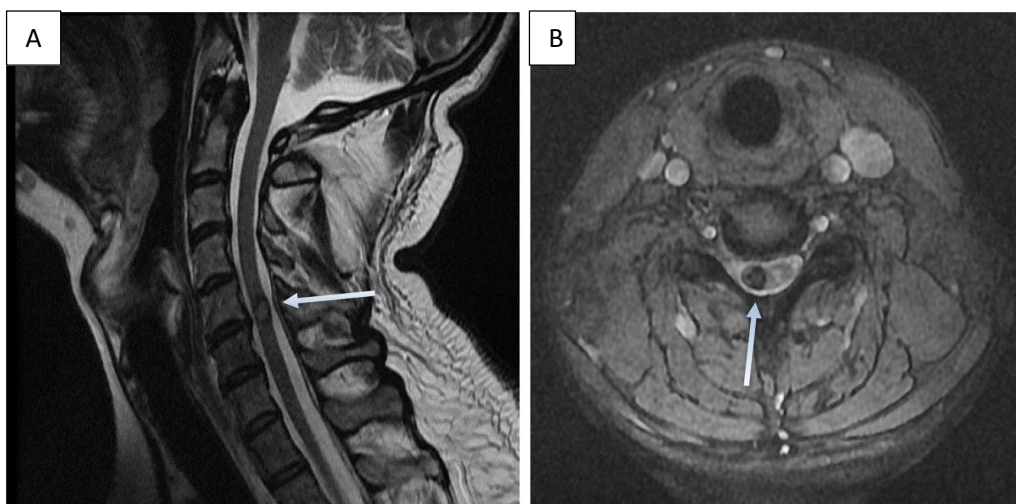


Fig. 1 Magnetic resonance image of the cervical spine, T2-weighted, **A** sagittal, **B** axial show cavernous malformation at the C5–C6 level (arrows)

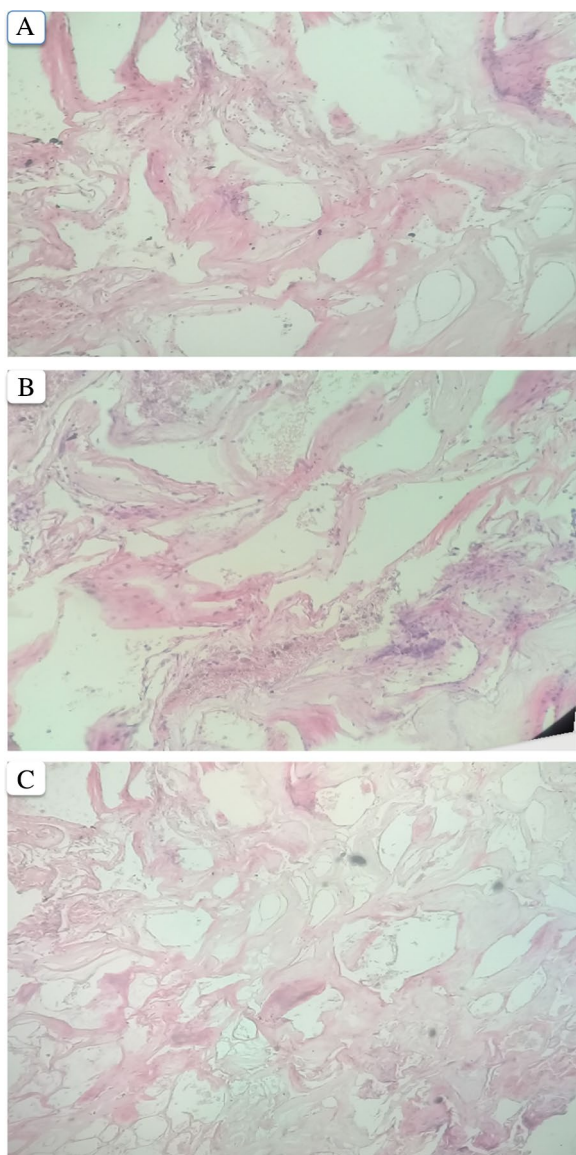


Fig. 2 A–C Benign tumor proliferation consisting of dilated vascular structures lined with regular endothelial cells, 40X

(Preferred Reporting Items for Systematic Reviews and Meta-Analysis). Two authors (Y.C.H.D. and S.E.M.) independently performed a search from 1980 to 2023 using the following mesh terms “cervical AND intramedullary AND hemangioma, cavernous” on PubMed (245 results).

According to PRISMA guidelines, the research order consisted of screening titles and abstracts first, and then the full-text screening (Fig. 3) (Table 1). Finally, the references of the articles included were screened. No automation software was used.

We included case reports and case series reporting patients affected by intramedullary cavernoma of the cervical spinal cord with definitive histopathological analysis. Only articles were published in the English language. All articles (i) not dealing with intramedullary cavernomas of the spinal cord in the cervical region, (ii) without full text or with no disaggregated data, (iii) written in a language other than English, about a retrospective study (iv) and duplicates (v) were included.

A database based on the previously selected case reports or case series was created. Descriptive statistical analyses were conducted by Jamovi 2.3.18 [3] (Fig. 4).

Results

A total of 29 studies reporting 423 patients with a mean age of 40.5 ± 6.3 years were included in this study. The overall male-to-female ratio was 1:2.5 with a median follow-up duration of 46 months [18.5–63.9]. Laminectomy and subsequent complete resection of the cavernomas were reported in 82.8% ($n=24$) of the included papers. Only one article (3.4%) reported complete resection of the lesion by an anterolateral partial vertebrectomy, although this information was not available in the remaining four 13.8% of papers. The outcome was favorable in 82.8% ($n=24$).

Discussion

It was previously believed that spinal cavernous malformations (SCM) were uncommon spinal cord lesions. However, over the past few decades, the prevalence of SCM has dramatically increased due to the widespread use of contemporary imaging techniques. The management of incidental and symptomatic findings is thus becoming increasingly important. However, experience with treatment and follow-up is minimal [8]. The same author reported only 7.1% of the cavernous malformations treated in their department over a decade were of spinal location. But surprisingly, 34.5% of these lesions were at the cervical spine. However, the authors do not provide any further details as to whether the location was extradural, intradural-extramedullary or intramedullary. Although the spine has been described as a rare site of cavernous malformations, the cervical spine may be the most common. However, the site of development showed an even distribution of cervical and thoracic locations, in contrast to previous reports documenting a thoracic predilection [7, 9]. Nevertheless, further studies on this topic are required to clarify precisely the intramedullary aspect, as is the case in our reported case.

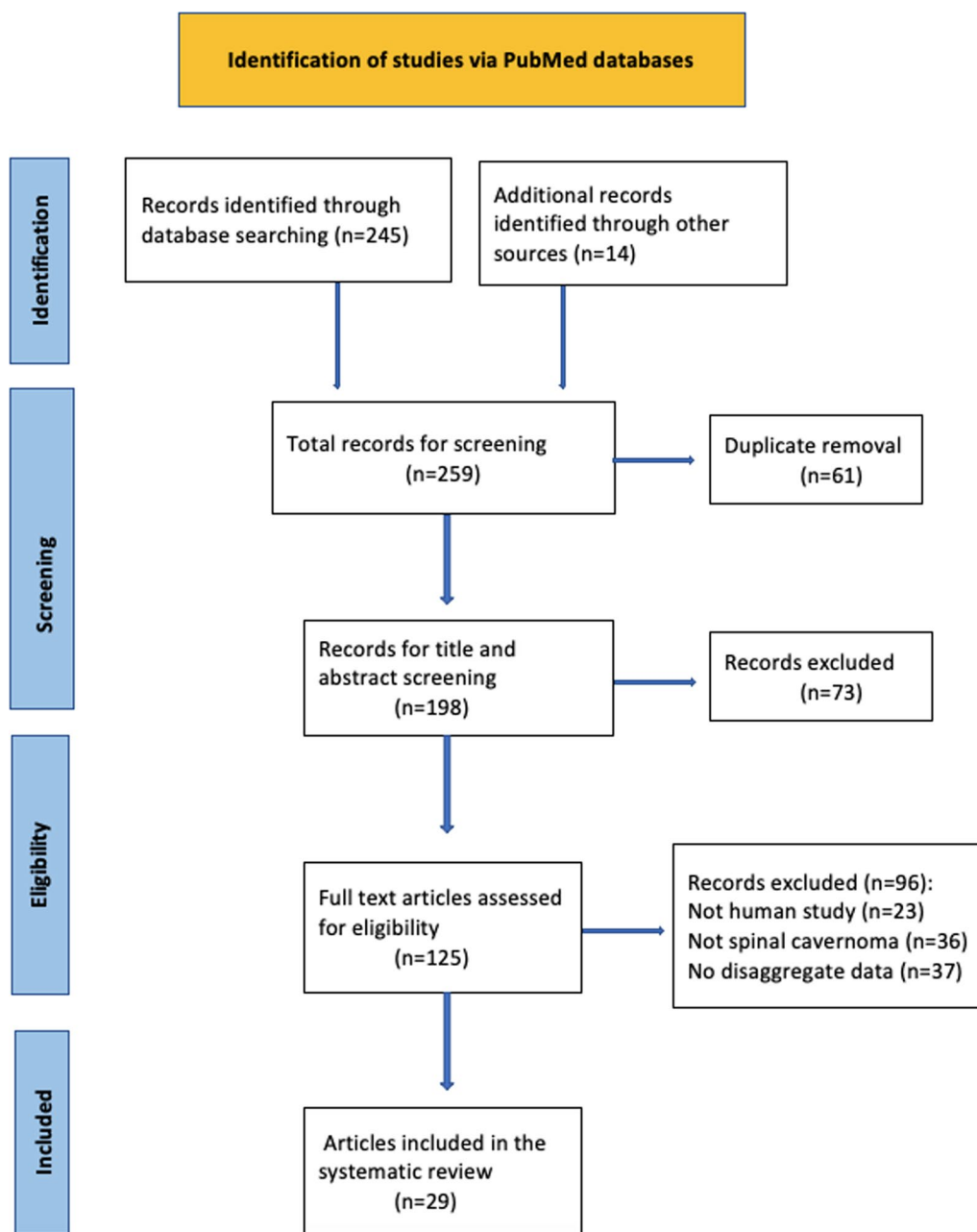


Fig. 3 PRISMA flowchart summarizing the selection of the studies

Because of the difficulty in diagnosing these lesions before the advent of advanced imaging techniques, the actual incidence of asymptomatic spinal cavernomas is unknown. In their series, Badhiwala et al. [4] reported the distribution of symptoms as 60% motor, 58% sensory, 34% pain, and 24% disturbance of bladder and/or bowel function. The symptom presentation of patients with CMs may be acute or slowly progressive. Acute neurological deterioration occurs in cases with significant

bleeding within the spinal cord, and slow progressive deterioration is related to multiple repeated microhemorrhages, subsequent thrombosis, hyalinization, and possible enlargement of the malformation [10]. The preferred imaging technique for identifying cavernous malformations is magnetic resonance imaging. Because cavernous malformations are angiographically occult and challenging to diagnose with traditional tomography and myelography, the majority of cases were documented

Table 1 General information about the included articles

References	Number of patients	Mean age(year)	M/F ratio	Cervical location	Treatment	Follow-up (months)	Outcome
Cosgrove [1]	5	41	2:3	1	TR	69	F
Mc Cormick [2]	6	32	4:2	1	TR	37	F
Vilani [3]	3	38	1:2	1	TR	NA	NA
Lunardi [4]	5	42	2:3	2	TR	NA	F
Contore [5]	6	53	5:1	2	TR	11,7	F
Turjman [6]	11	49	2:9	2	NA	NA	NA
Furuya [7]	4	45	2:2	2	-	49,5	-
Vishteh [8]	17	40	8:9	8	NA	NA	NA
Cristante [9]	12	33.5	4:8	8	TR	53,6	F
Chabert [10]	5	43	1:4	3	TR	12,8	F
Ghogawala [11]	9	44	5:4	6	TR	-	F
Tu [12]	7	30.1	5:2	4	TR	-	-
Zevgaridis [13]	9	38.2	4:5	3	TR	24	F
Santoro [14]	10	40,9	5:5	5	TR	68,3	F
Sandalcioglu [15]	10	34.5	3:7	5	TR	11,1	F
Weinzierl [16]	12	44.3	6:6	5	TR	15,2	F
Cohen-Gadol [17]	67	50	1:1	28	TR	116,4	F
Jallo [18]	26	38	1:1/2	8	TR	53,7	F
Nishikawa [19]	3	48	1:2	3	TR via anterolateral partial vertebrectomy	NA	F
Kharkar [20]	14	42	8:2	4	TR	42,5	F
Labauge [13]	53	40.2	1:1	12	TR	87,6	F
Bian [21]	16	38	1:1.2	9	TR	23,1	F
Matsuyama [22]	17	41.6	1:1.4	8	TR	66	F
Park [23]	14	34.3	1:1/2	4	TR	55	F
Maslehaty [24]	9	47	1:6	5	NA	17	F
Aoyama [25]	13	25	3:2	5	TR	94	F
Choi [12]	21	38.4	1:1.6	10	TR	38,4	F
Sun [26]	10	40		10	TR	12	F
Cai [27]	29	45.2	1:0.7	12	TR	57,6	F

NA, No available; F, Favorable; and TR, Total resection (after posterior midline incision and subsequent total laminectomy, cavernomas were excised totally through midline myelotomy)

following the introduction of MRI. A common observation is rounded areas of heterogeneous signal intensity on the T1- and T2-weighted images, which are caused by different age blood products and have a “popcorn appearance.” This is composed of a central lesion core that exhibits mixed heterogeneous signal intensity on both T1- and T2-weighted images, with minimal contrast enhancement, and a periphery with a hypointense ring of hemosiderin [11–13].

Most patients described in the literature were treated either surgically or conservatively. In their single-center serial analysis of intramedullary cavernous malformation of the spinal cord, Badhiwala et al. [4] reported

that 90% of patients underwent resection and only 10% underwent conservative treatment. Only the symptomatic patients benefited from complete resection of the lesion. The result was that in the surgical group, 30% of patients improved, 58% were neurologically stable, and 11% worsened their neurological status. Meanwhile, outcomes after five years of follow-up were reported to be less favorable for those who chose conservative treatment. These results have been confirmed by many other authors [3, 14–19]. Nevertheless, according to our systematic review, the surgical outcome was positive, and good results were reported in 82.8% ($n=24$) of all patients with totally resected cervical cavernoma.

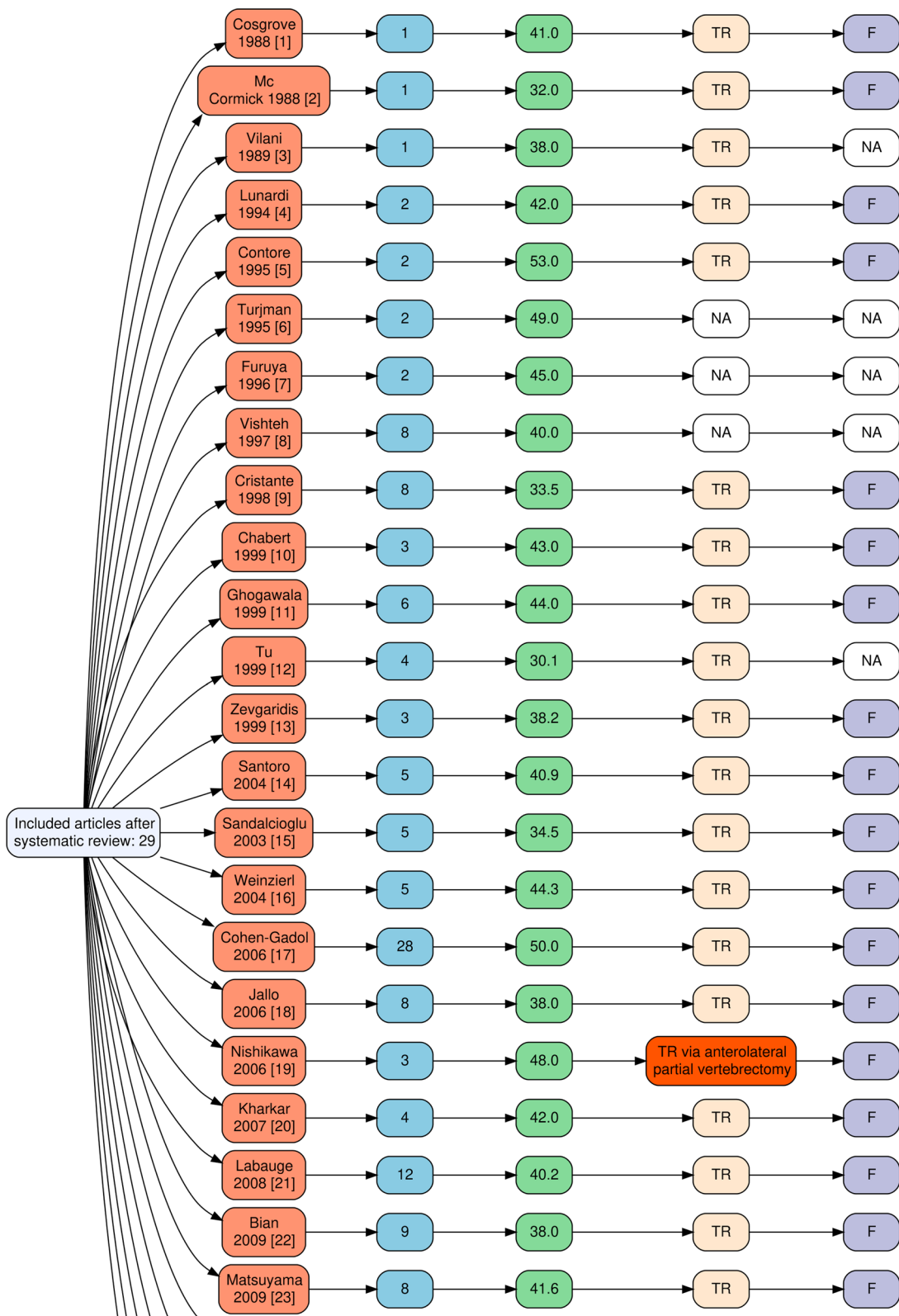


Fig. 4 A database based on the previously selected case reports or case series

Conclusion

Intramedullary cavernomas of the cervical spinal cord tend to be clinically progressive. Total macroscopic resection of the lesion is the gold standard treatment that should be performed whenever possible and is the better indication that guarantees a favorable outcome.

Abbreviations

T1WI T1-weighted images
T2WI T2-weighted images

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Not applicable.

Author contributions

YCHD contributed to conceptualization, writing draft, reviewing and editing, visualization, supervision, validation, and methodology. SM was involved in writing and review and editing, and FLOO contributed to writing and review and editing. MES was involved in writing and editing, NI contributed to writing and editing, and ACEA was involved in writing and editing, and MG contributed to supervision, validation, and review.

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Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Declarations

Ethics approval and consent to participate

Informed consent was obtained from the patient prior to the submission of this article. Also, this article respects both the Consensus-based Clinical Case Reporting Guideline and the Recommendations for the Conducting, Reporting, Editing, and Publication of Scholarly Work in Medical Journals.

Consent for publication

Informed consent was obtained from the patient to publish his case.

Competing interests

The authors declare that they have no competing interests.

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References

- Gross BA, Du R, Popp AJ, Day AL. Intramedullary spinal cord cavernous malformations. *FOC*. 2010;29:E14. <https://doi.org/10.3171/2010.6.FOCUS.10144>.
- Kharkar S, Shuck J, Conway J, Rigamonti D. The natural history of conservatively managed symptomatic intramedullary spinal cord cavernomas. *Neurosurgery*. 2007;60:865–72.
- Bakir A, Savas A, Yilmaz E, Savas B, Erden E, Caglar S, Sener Ö. Spinal intradural-intramedullary cavernous malformation. *Pediatr Neurosurg*. 2005;42:35.
- Badhiwala JH, Farrokhkar F, Alhazzani W, Yarascavitch B, Aref M, Algird A, Murty N, Kachur E, Cenic A, Reddy K. Surgical outcomes and natural history of intramedullary spinal cord cavernous malformations: a single-center series and meta-analysis of individual patient data. *J Neurosurg Spine*. 2014;21:662–76.
- Canavero S. Intramedullary cavernous angiomas of the spinal cord: clinical presentation, pathological features, and surgical management. *Neurosurgery*. 1993;32:693–4.
- Reitz M, Burkhardt T, Vettorazzi E, Raimund F, Fritzsche E, Schmidt NO, Regelsberger J, Westphal M, Eicker SO. Intramedullary spinal cavernoma: clinical presentation, microsurgical approach, and long-term outcome in a cohort of 48 patients. *Neurosurg Focus*. 2015;39:E19.
- Ogilvy CS, Louis DN, Ojemann RG. Intramedullary cavernous angiomas of the spinal cord: clinical presentation, pathological features, and surgical management. *Neurosurgery*. 1992;31:219.
- Velz J, Bozinov O, Sarnthein J, Regli L, Bellut D. The current management of spinal cord cavernoma. *J Neurosurg Sci*. 2018;62:383–96.
- 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;141:237–45. <https://doi.org/10.1007/s007010050293>.
- Kivelev J, Ramsey CN, Dashti R, Porras M, Tyninen O, Hernesniemi J. Cervical intradural extramedullary cavernoma presenting with isolated intramedullary hemorrhage: case report. *J Neurosurg Spine*. 2008;8:88–91.
- Mehdorn HM, Stolke D. Cervical intramedullary cavernous angioma with MRI-proven haemorrhages. *J Neurol*. 1991;238:420–6. <https://doi.org/10.1007/BF00314647>.
- Choi GH, Kim KN, Lee S, Ji GY, Oh JK, Kim TY, Yoon DH, Ha Y, Yi S, Shin H. The clinical features and surgical outcomes of patients with intramedullary spinal cord cavernous malformations. *Acta Neurochir*. 2011;153:1677–85. <https://doi.org/10.1007/s00701-011-1016-3>.
- Labauge P, Bouly S, Parker F, Gallas S, Emery E, Loiseau H, Lejeune JP, Lonjon M, Proust F, Boetto S, Coulbois S, Auque J, Boulliat J. Outcome in 53 patients with spinal cord cavernomas. *Surg Neurol*. 2008;70:176–81. <https://doi.org/10.1016/j.surneu.2007.06.039>.
- Utomo SA, Bajamal AH, Parenrengi MA. Multiple spinal intramedullary cavernous angiomas with bleeding episode mimicking an intramedullary tumor. *Radiol Case*. 2022;16:15–22. <https://doi.org/10.3941/jrcr.v16i3.4251>.
- Santoro A, Piccirilli M, Frati A, Salvati M, Innocenzi G, Ricci G, Cantore G. Intramedullary spinal cord cavernous malformations: report of ten new cases. *Neurosurg Rev*. 2004;27:93–8. <https://doi.org/10.1007/s10143-003-0302-9>.
- Pichierri A, Piccirilli M, Passacantilli E, Frati A, Santoro A. Klippel-Trenaunay-Weber syndrome and intramedullary cervical cavernoma: a very rare association. *Case report. Surg Neurol*. 2006;66:203–6.
- Giannattei L, Messerer M, Prada F, DiMeco F. Intramedullary cavernoma: a surgical resection technique. *Neurochirurgie*. 2017;63:426–9. <https://doi.org/10.1016/j.neuchi.2016.04.005>.
- Jallo GI, Freed D, Zareck M, Epstein F, Kothbauer KF. Clinical presentation and optimal management for intramedullary cavernous malformations. *FOC*. 2006;21:1–6. <https://doi.org/10.3171/foe.2006.21.1.1>.
- Nwachuku E, Duehr J, Kulich S, Marker D, Moossy J. Cervical intramedullary spinal cavernoma in setting of unresolved myelopathy: A case report. *Surg Neurol Int*. 2020;11:176.
- Kharkar S, Shuck J, Conway J, Rigamonti D. The natural history of conservatively managed symptomatic intramedullary spinal cord cavernomas. *Neurosurgery*. 2007;60:865–72.
- Bian LG, Bertalanffy H, Sun QF, Shen JK. Intramedullary cavernous malformations: clinical features and surgical technique via hemilaminectomy. *Clin Neurol Neurosurg*. 2009;111:511–7.
- Matsuyama Y, Sakai Y, Katayama Y, Imagama S, Ito Z, Wakao N, et al. Surgical results of intramedullary spinal cord tumor with spinal cord monitoring to guide extent of resection. *Clinical article. J Neurosurg Spine*. 2009;10:404–13.
- Park SB, Jahng TA, Chung CK. The clinical outcomes after complete surgical resection of intramedullary cavernous angiomas: changes in motor and sensory symptoms. *Spinal Cord*. 2009;47:128–33.
- Maslehay H, Barth H, Petridis AK, Doukas A, Mehdorn HM. Symptomatic spinal cavernous malformations: indication for microsurgical treatment and outcome. *Eur Spine J*. 2011;20:1765–70.
- Aoyama T, Hida K, Houkin K. Intramedullary cavernous angiomas of the spinal cord: clinical characteristics of 13 lesions. *Neurol Med Chir (Tokyo)*. 2011;51:561–6.
- Sun I, Pamir MN. Spinal Cavernomas: Outcome of Surgically Treated 10 Patients. *Front Neurol*. 2017;8:672.
- Cai Z, Hong X, Dai W, Zhang Z, Liang Q, Ding X, Sun W. Surgical outcomes of symptomatic intramedullary spinal cord cavernous malformations:

analysis of consecutive cases in a single center. *Neurospine* 2023;20(3):810.

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