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Surgical and Endovascular Treatment for Spinal Arteriovenous Malformations

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Abstract

Spinal arteriovenous malformation (AVM) is a broad term that constitutes diverse vascular pathologies. To date, various classification schemes for spinal AVM have been proposed in literature, which helped neurosurgeons understand the pathophysiology of the disease and determine an optimal treatment strategy. To discuss indications and results of surgical and endovascular interventions for spinal AVM, this article refers to the following classification proposed by Anson and Spetzler in 1992: type I, dural arteriovenous fistula (AVF); type II, glomus intramedullary AVM; type III, juvenile malformations; and type IV, perimedullary AVF. In general, complete obliteration of the fistula is a key for better outcome in type I dural and type IV perimedullary AVFs. On the other hand, in type II glomus and type III juvenile malformations, functional preservation, instead of pursuing angiographical cure, is the main goal of the treatment. In such cases, reduction of the shunt flow can alleviate clinical symptoms. Proper management of spinal AVM should start with neurological examination and understanding of angioarchitectures, which provide critical information that guides the indication and modality of intervention. Finally, close collaboration of the microsurgical and endovascular teams are mandatory for successful treatment.

Key words: arteriovenous malformation, endovascular treatment, spinal cord, surgery

Introduction

Spinal arteriovenous malformations (AVMs) are rare pathologies representing 3%–4% of all space-occupying lesions affecting the spinal cord.[1)] According to one of the simplest schemes based on the radiographical appearance of fistulas,[2)] spinal AVMs are classified as type I if the fistulas are located between the dural branch of the spinal ramus of the radicular artery and intradural medullary vein; as type II if intramedullary glomus malformations are present; as type III if extensive juvenile malformations, often expanding into the extradural space and involving multiple feeding arteries, are present; and as type IV if intradural perimedullary arteriovenous fistula (AVF) is observed.

This classification subdivides type I dural AVFs on the basis of the number of feeding arteries. However, it does not indicate extradural AVFs that are encountered in clinical practice.[3)] Having included extradural AVF as a new category, this article discusses indications and results of surgical and endovascular treatments for each type of spinal AVM.

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I. Type I, spinal dural arteriovenous fistula (DAVF)

Type I, spinal DAVF, is the most common type of spinal vascular malformation, constituting approximately 70% of all spinal AVMs.[4–6)] Aminoff and Logue published the first large clinical study of patients with detailed description of symptoms and established a famous clinical scale for spinal AVM (Table 1).[7)]

Fistulas are located in the dural root sleeve between the dural branch of the segmental artery and radicular vein.[8–10)] Increased intravascular pressure is transmitted from the radiculomeningeal arteries to radicular veins and venous system of the spinal cord,[11)] causing stagnation of the venous outflow from the spinal cord and resulting in intramedullary venous hypertension and ischemic insult to the spinal cord.[12)]

Spinal DAVF occurs most frequently in the thoracolumbar region, and progressive congestive myelopathy is the predominant clinical manifestation.[13,14)] When fistulas are located in the craniocervical junction or cervical spine, subarachnoid hemorrhage

Table 1 Aminoff-Logue scale

Grade	Definition
Gait	
0	normal
1	leg weakness, abnormal gait, or stance but no restriction of activity
2	restricted activity but not requiring support
3	requiring one stick for walking
4	requiring two sticks, crutches, or walker
5	confined to wheel chair
Micturition	
0	normal
1	hesitancy, urgency, frequency, altered sensation, but continent
2	occasional urinary incontinence or retention
3	total incontinence or persistent retention

(SAH) can occur as another clinical presentation of this disease.[15,16] If the venous drainage ascends cranially, there is an increased risk of hemorrhage.

Two effective treatment options, namely surgery and endovascular treatment, are available for type I spinal AVM. Surgery of DAVF is a simple procedure intending to disconnect a draining vein at the point where it intradurally emerges from the fistula (Fig. 1).[9,10] It is a safe and highly effective procedure with a reported success rate of approximately 98%.[17,18] In a series of 154 spinal DAVF patients treated by microsurgical obliteration, motor function improved in 82.2% patients and symptoms were stabilized in 14.4% patients. No patients suffered major neurological complications.[19] Recent technical advancement, including indocyanine green video angiography and 3D computer graphics, further contributed to making surgical intervention a safe and reliable option in treating spinal DAVF (Fig. 1).[20,21]

During endovascular treatment, which is another option for spinal DAVF, complete obliteration of the proximal portion of the draining vein is a key for success.[22] If complete obliteration is not achieved, recruitment of other arterial feeders and recanalization of the fistulas may occur. In addition, endovascular therapy may be contraindicated if the radicular artery supplies the anterior and/or posterior spinal arteries.[23] In one single institutional study, 69% patients treated by embolization achieved complete obliteration, and an additional procedure was required in 25% of cases during a 3-year follow-up period.[24] The ranges for success and recurrence rates of endovascular treatment were 75%–100% and 15%–20%, respectively.[25–27]

In spinal DAVF, clinical symptoms worsen without appropriate treatment, and it has been estimated that 50% of untreated patients become severely disabled within 3 years of the onset.[7] Considering that the degree of preoperative neurological deficit can be a strong prognostic factor of the outcome, prompt diagnosis and treatment play an important role in achieving good outcomes in symptomatic spinal DAVF cases.[18,28]

Spinal extradural AVF: We occasionally encounter patients whose magnetic resonance images disclose engorgement of epidural venous structures.[4,29] Patients suffer from congestive myelopathy and clinical presentations are similar to those of type I spinal DAVF.[3] However, angiograms demonstrate a direct fistulous connection between an "extradural" artery and veins. This feature does not fall into the aforementioned classification[2] because the location of the shunt is extradural and anatomically distinct from that of type I DAVF.[4] In a recent report, spinal extradural AVF was further divided into two subtypes: extradural AVF with retrograde parenchymal drainage (type A) and pure extradural AVF (type B).[30] Patients with type A extradural AVF have perimedullary draining veins which penetrate the dura and result in progressive myelopathy due to venous congestion.[3] Therefore, treatment for type A spinal extradural AVF should target both the epidural venous lake and intradural perimdullary draining veins. When an extradural drainer can be used as an access route, transvenous embolization (TVE) should be considered as the treatment of choice.[3] When a case does not have extradural drainers or if there is a risk for TVE, microsurgery can be an option. If a case has a single drainer, we recommend treatment with sole drainer occlusion as the first line treatment. Because patients suffer venous congestion due to perimedullary retrograde draining veins in type A extradural AVF, obliteration of the intradural drainer is advisable (Fig. 2). Meticulous coagulation and detachment of the epidural venous lake may be indicated for cases with multiple drainers penetrating the dura matter.[3,29] For type B DAVF, no intradural procedure is required. Rather, the purpose of the treatment is to reduce mass effect of the engorged epidural venous system on the spinal cord and spinal nerves.[30] In most cases, endovascular treatment successfully improves myelopathy in type B spinal extradural AVF.[29,31]

II. Type II, intramedullary AVM (glomus type)

Intramedullary AVM has a nidus, which is similar to intracranial AVM. A lesion has multiple feeders that originate from the anterior and/or posterior

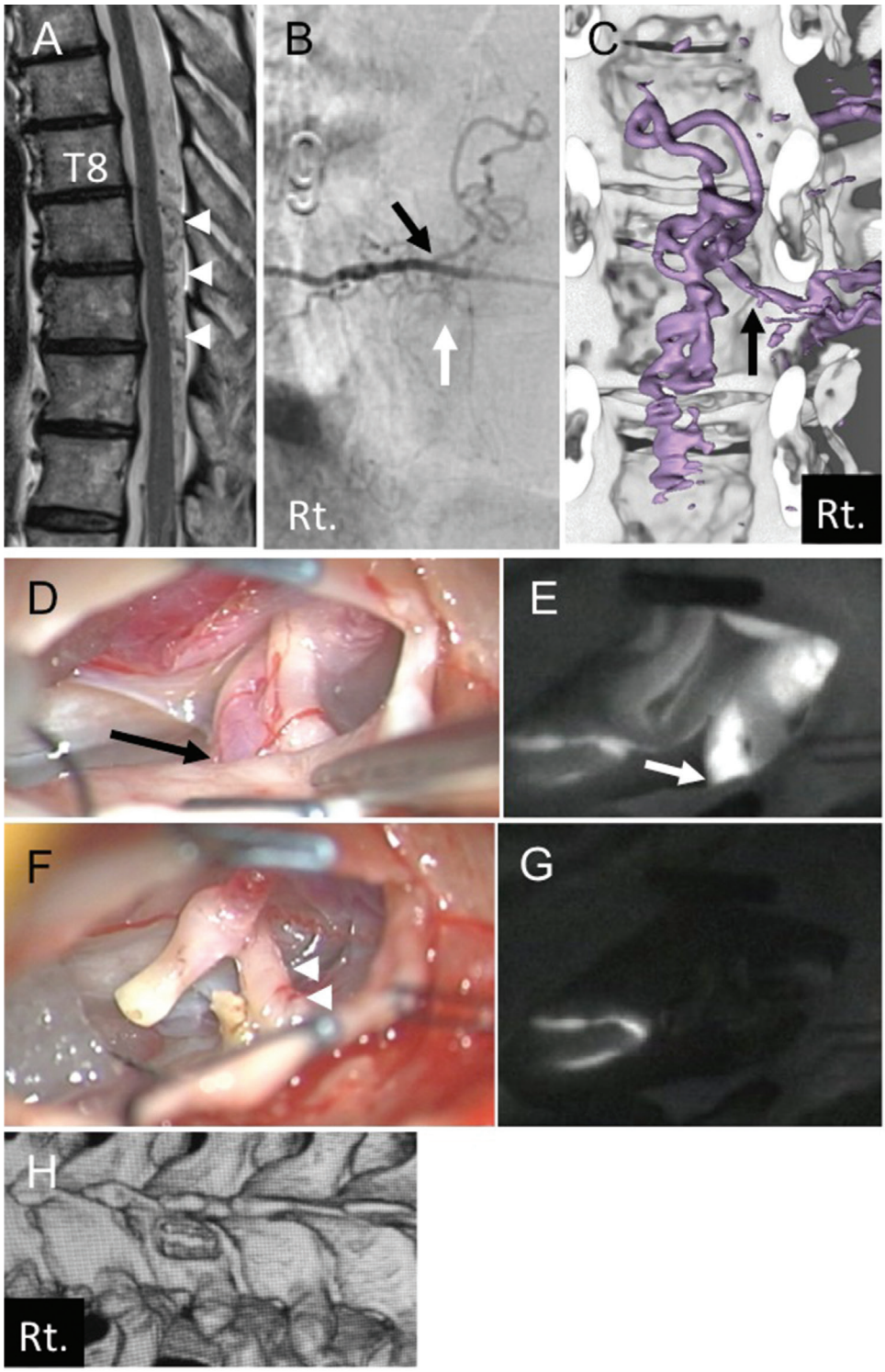

Fig. 1 A representative case of mid-thoracic type I spinal dural arteriovenous fistula. A 65-year-old man presenting with lower extremities weakness and sensory disturbance below T11 dermatome. A: T_2-weighted sagittal magnetic resonance image demonstrating abnormal flow voids dorsal to the spinal cord (*arrowheads*) below T8 vertebral level. B: A selective angiogram of the right T9 segmental artery demonstrating spinal DAVF (*white arrow*) fed by the radiculomeningeal artery. Perimedullary draining vein pierced the dura at the right T9 dural root sleeve (*black arrow*). C: Three-dimensional fusion image of rotational angiography and computed tomography of the thoracic spine. Dorsal view indicated the point where the draining vein entered intradurally (*black arrow*). D–G: Intraoperative images (D, F) coupled with images from indocyanine green videoangiography (E, G). Right T9 partial hemilaminectomy and dural incision identified the point where the draining vein entered intradurally (*arrows* in D and E). After the draining vein was cut (F), abnormal blood flow in the arterialized vein disappeared (G). Note, T9 dorsal nerve root was preserved (*arrowheads* in F). H: Three-dimensional reconstruction of computed tomography, a dorsal view of the thoracic spine, showing area of the bone window on the right T9 lamina. Postoperative course was uneventful. Patient's symptoms remarkably improved after the operation. Rt.: right.

spinal arteries.[32] Patients present with congestive myelopathy, SAH, or intramedullary hemorrhage.[33]

First-line treatment is embolization because surgical treatment aiming to remove the nidus from the spinal cord carries a significant intraprocedural risk.[33,34] In a surgical series of 20 type II intramedullary AVM cases, 20% patients suffered from worsening of postoperative neurological symptoms.[33] Although a few previous reports indicated that surgery is a feasible option in type II intramedullary AVM,[35,36] surgery should only be indicated for symptomatic cases when embolization is too hazardous or if

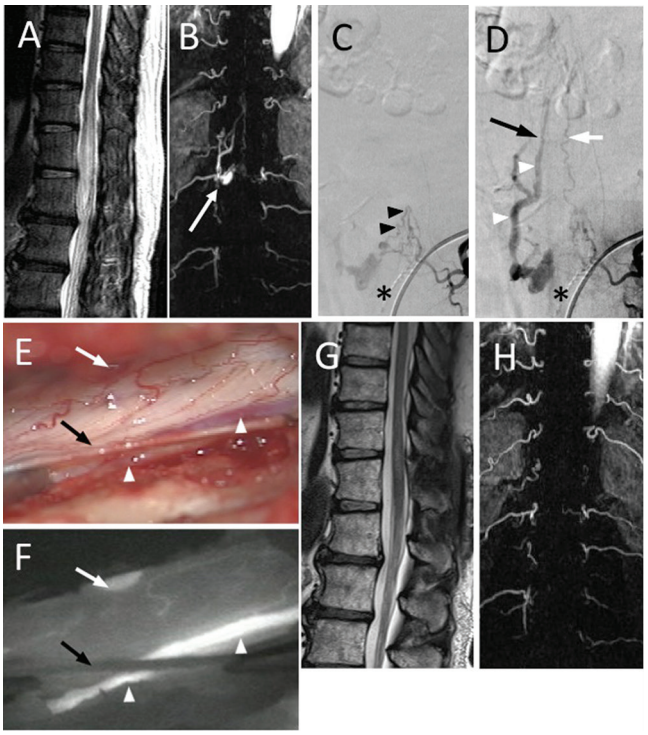

Fig. 2 A representative case of extradural spinal arteriovenous fistula (AVF) successfully treated by sole intradural drainer occlusion. A 59-year-old man presenting with progressive pain and weakness in the lower extremities as well as urinary retention. A: Preoperative sagittal T_2-weighted magnetic resonance (MR) image reveals intramedullary hyperintensity of the thoracolumbar spinal cord. B: Preoperative MR angiograpm reveals an epidural venous lake located outside the spinal canal (*arrow*). C, D: Time course images of the preoperative selective angiograms of the left L1 segmental artery. AVFs (*black arrowheads* in C), an epidural venous lake (*asterisks*), a draining vein (*white arrowheads* in D), and dilated spinal veins (*white arrow* in D) are visualized. *Black arrow* in D indicates the entry points of the draining vein crossing the dura. E, F: Intraoperative photographs coupled with images from indocyanine green (ICG) video angiography. The dura matter is indicated by *black arrows*. A draining vein penetrating the dura matter (*arrowheads*) and a dilated spinal vein (*white arrows*) were observed after hemilaminectomy of T12. ICG video angiography clearly identified the penetration site at the dorsolateral wall of the dural sac. The draining vein was coagulated and resected intradurally. G, H: Postoperative MR images 6 months after surgery. Sagittal T_2-weighted image (G) shows a decreased hyperintensity signal. MR angiogram reveals obliteration of the epidural venous lake, and the draining vein has disappeared. Figures were obtained and modified with permission from the following reference and rearranged: Niizuma et al., 2013.[3]

endovascular treatment results in an incomplete obliteration.

With recent advances in embolic materials and devices in endovascular treatment, transarterial embolization plays an increasingly important role in the treatment of AVM.[37,38] Complete angiographical obliteration of the nidus is not necessarily the goal of embolization, but rather, the treatment aims to reduce shunting volume and stabilize the symptoms. In a recent report utilizing Onyx for 17 intramedullary AVM patients, complete obliteration was attained in 37.5% patients, whereas 82% patients experienced clinical improvements.[38] A recent meta-analysis of type II intramedullary AVM has shown that the annual hemorrhage rate before the treatment was 4%, which increased to 10% for AVMs with a previous hemorrhage.[39] Both complete and partial endovascular treatments significantly decreased the risks of future hemorrhage.[39]

Radiosurgery is another treatment option for type II AVM. Hida et al. treated 10 patients with hypofractionated linear accelerator stereotactic radiotherapy,[40] all of whom presented with hemorrhage. However, no new hemorrhages were observed in the follow-up period. Although no angiographical cure of the AVM was observed, five patients displayed a reduction in nidus size and two patients had improved motor functions. With increased application of stereotactic radiosurgery to spinal cord pathologies, further accumulation of clinical evidence is anticipated to evaluate the efficacy of this treatment.[41]

Besides cervical and thoracic intramedullary AVMs, conus medullaris AVMs was first classified as a distinct entity in 2002.[4] This rare form of spinal AVM received arterial supplies from multiple feeding pedicles including the anterior and posterior spinal arteries. Their nidi located extra- and intramedullary at the level of the conus medullaris.[42] To achieve better clinical outcomes for this complex lesion, a combined endovascular and surgical treatments were required. In a clinical report including 16 patients of conus medullaris AVM, rates of angiographical cure and neurological improvement were 88% and 43%, respectively.[42] Care must be taken for possible recurrences related to multiplicity of feeding arteries and nidi.

III. Type III, juvenile malformations

This rare form of AVM is typically observed in adolescents and young adults.[32] In previous reports, this type of AVM has been variably referred to as Cobb syndrome, extradural-intradural AVM, and spinal arteriovenous metameric syndrome.[4,43–45] Hallmarks of juvenile malformation include multiple vascular

malformations that are derived from the same spinal metameric segment. AVM can be localized in the bone, epidural space, spinal cord, paraspinal soft tissues or muscles, subcutaneous tissues, and skin[4,43,46] Possible events creating vascular malformations in the same metamere include genetic mutations in the neural crest before migration.[47] Thus, this type of AVM is classified as "genetic non-hereditary" in the criteria proposed by Rodesch et al.[32,44]

Optimal treatment strategy for type III juvenile malformations remains to be established.[48,49] Complete obliteration of fistulas is not always possible because of complexity and diversity of AVMs.[43,50] Therefore, intervention is aimed at halting the progression of neurological symptoms and minimizing neurological sequelae (Fig. 3). In 21 reported cases of type III juvenile malformations, 14 cases were first treated by endovascular embolizations, and surgical excisions followed the embolization procedure in 7 cases.[51] Careful assessment of the symptoms and causative intra- and extradural vascular pathology based on the understanding of angioarchitectures is important for successful management of type III spinal AVM.

IV. Type IV, intradural or perimedullary AVF

Type IV intradural or perimedullary AVF was first described by Djindjian et al.[52] It is characterized by direct arteriovenous communications on the pial surface of the spinal cord without an intervening nidus.[53] Based on the number and diameter of the feeding and draining vessels, type IV lesions can be further divided into three subtypes: type IVa has a single shunting communication, type IVb has multiple dilated arterial feeders of intermediate size, and type IVc has multiple dilated arteries and ectatic draining veins.[2,54] This classification is important for clinical presentations and appropriate treatment strategies. For instance, patients with type IVc lesions are significantly younger and more commonly associated with hereditary hemorrhagic telangiectasia.[54] Type IVb cases have a higher risk of hemorrhage, although no statistical difference was observed in a recent meta-analysis.[54] Overall, annual hemorrhage risk was rated as 2.5%.

In general, embolization is considered difficult in type IVa AVF because of small caliber of the feeding vessels.[55] Hida et al. reported the efficacy of an anterior approach for treating type IVa cases located in the ventral cervical spinal cord.[56] On the other hand, type IVc cases having multipediculated feeding arteries as well as dilated draining vessels are more amendable to endovascular intervention.[53,57]

In type IVb cases, an optimal treatment strategy remains to be established. Although a relatively large

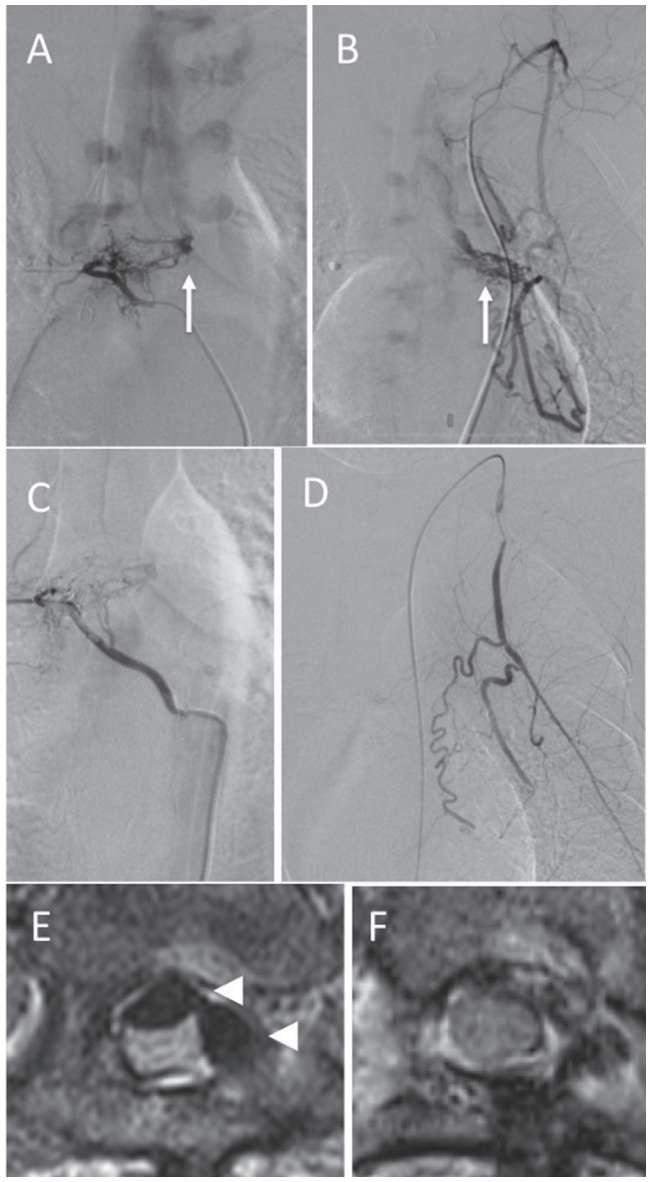

Fig. 3 A case of thoracic juvenile malformations treated by transarterial embolizations followed by open surgery. A, B: Preoperative selective angiograms through right T5 intercostal (A) and left descending scapular (B) arteries demonstrating arteriovenous fistulas along the left T5 root sleeve (*arrows*). An enlarged epidural venous plexus was apparent. C, D. Selective angiogram after endovascular interventions. Arteriovenous shunts and enlarged venous structures were not evident through selective angiography of the right T5 intercostal (C) and left descending scapular (D) arteries. E, F. Preoperative (E) and postoperative (F) axial T_2-weighted magnetic resonance images of the thoracic spinal cord. Note, engorged vein compressing the spinal cord (*arrowheads* in E) disappeared after the endovascular interventions (F). Open surgery followed the endovascular treatments and eliminated residual shunt flow. Figures were obtained and modified with permission from the following reference and rearranged: Elkordy et al. 2015.[51]

diameter of the feeding arteries makes endovascular treatment a feasible option,[1,57,58] embolization does not always provide complete obliteration because of multiple feeding arteries and fistulas.[57] As we recently summarized,[59] out of the five type IVb cases in the cervical spinal cord, three cases required an open surgery after endovascular interventions.[56,58,60,61] For 16 type IVb cases at our institute, surgical interventions were indicated and provided satisfactory clinical and angiographical results.[59] Although fistulas are often located on the ventral or ventrolateral cervical spinal cord surface, posterior approach with an aid of neuroendoscopy provides an adequate surgical view for safe obliteration of the perimedullary fistulas (Fig. 4).

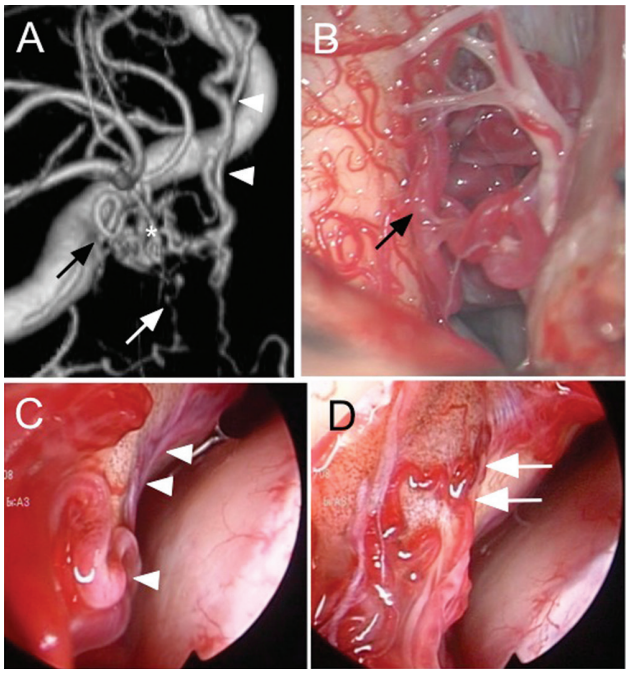

Fig. 4 A case of craniocervical junction type IVb perimedullary arterioveous fistula (AVF) presented with subarachnoid hemorrhage. A: Three-dimensional reconstruction image of the left vertebral artery rotational angiogram (posterior view) demonstrating perimedullary AVF (*asterisk*) fed by the anterior spinal (*white arrowheads*), C2 radiculomedullary (*white arrow*), and C1 radiculopial (*black arrow*) arteries. B: Open surgery was performed with suboccipital craniectomy and C1 hemilaminectomy (note, the rostral side of the patient is displayed at the bottom of the picture). A radiculopial artery was identified as one of the feeding arteries (*black arrow*). C, D: Ventrally located AVFs were successfully managed with endoscopic assistance. The anterior spinal (*white arrowheads* in C) and C2 radiculomedullary (*white arrows* in D) arteries were clearly visualized in endoscopic views. Figures were obtained and modified with permission from the following reference and rearranged: Endo et al. 2014.[59]

Overall, endovascular and surgical treatment offer comparable results with 74% and 88% obliteration rates, respectively.[54] In total, 70% patients improved after interventions.

Conclusion

Variable neurological symptoms and complex angioarchitectures make spinal AVM a challenging clinical entity. Classifying the spinal AVM cases based on angiographical characters is important for selecting proper surgical and endovascular treatments. Introduction of neuromonitoring, indocyanine green videoangiography, three-dimensional fusion images, and assisted use of endoscope made surgical intervention a safer and more reliable option. In endovascular treatment, embolic materials and techniques are continuously improved. Advancement of surgical and endovascular treatments can improve current treatment strategy. Neurosurgical and endovascular teams should collaborate to choose appropriate treatment options for individual cases.

Conflicts of Interest Disclosure

The authors declare no conflicts of interest.

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