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Surgical Management of Combined Intramedullary Arteriovenous Malformation and Perimedullary Arteriovenous Fistula within the Hybrid Operating Room after Five Years of Performing Focus Fractionated Radiotherapy: Case Report

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Abstract

Perimedullary arteriovenous fistula (AVF) shunts occur on the spinal cord surface and can be treated surgically or by endovascular embolization. In contrast, the nidus of an intramedullary arteriovenous malformation (AVM) is located in the spinal cord and is difficult to treat surgically or by endovascular techniques. The benefits of radiotherapy for treating intramedullary AVM have been published, but are anecdotal and consist largely of case reports. We present a case of combined cervical intramedullary AVM and perimedullary AVF which received surgical treatment within a hybrid operating room (OR) after 5 years of focus fractionated radiotherapy. A 37-year-old male presented with stepwise worsening myelopathy. Magnetic resonance imaging and spinal angiography revealed intramedullary AVM and perimedullary AVF at the C3 to C5 levels. In order to reduce nidus size and blood flow, we first performed focal fractionated radiotherapy. Five years later, the lesion volume was reduced. Following this, direct surgery was performed by an anterior approach using corpectomy in the hybrid OR. The spinal cord was monitored by motor-evoked potential throughout the surgery. Complete obliteration of the fistulous connection was confirmed by intraoperative indocyanine green video-angiography and intraoperative angiography, preserving the anterior spinal artery. We conclude that surgical treatment following focal fractionated radiotherapy may become one strategy for patients who are initially deemed ineligible for endovascular embolization and surgical treatment. Furthermore, the hybrid OR enables safe and precise treatment for spinal vascular disorders in the fields of endovascular treatment and neurosurgery.

Key words: arteriovenous fistula, arteriovenous malformation, intramedullary, perimedullary, radiation therapy

Introduction

Although spinal vascular malformations are rare and represent challenging lesions among spinal disorders, they are important clinical entities because they produce considerable morbidity and can be fatal if left untreated.[1)] Ideal treatment for spinal vascular malformations (AVMs) is complete interruption of arteriovenous (AV) shunts. Combinations of various treatment modalities, including conservative observation,[2)] endovascular embolization,[3,4)] surgical intervention,[5)] or radiation therapy,[6,7)] have been performed to address spinal vascular malformations based on angiographic features and pathophysiology. In particular, complete cure for intramedullary spinal arteriovenous malformation (AVM) lesions is challenging, because the nidus of the AVMs are located within the spinal cord parenchyma.[8,9)] Therefore, many intramedullary AVMs are not amenable to curative therapy, even with state-of-the-art endovascular and microsurgical techniques. This is primarily because the use of these sophisticated therapeutic approaches is precluded by either a complex angioarchitecture that can involve the anterior spinal artery (ASA)[1)] or a location within or immediately adjacent to critical ascending and descending sensory motor pathways. At our institution, we perform focal fractionated radiotherapy to treat intramedullary AVMs, because many patients are

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considered to be untreatable by other means, due to the size or location of their AVMs.[10] The underlying mechanism of action for radiotherapy is thought to be gradual endothelial hyperplasia of the abnormal vasculature, which, in turn, leads to progressive narrowing and eventual vessel occlusion.[11] Here, we present our experiences with a patient who presented with combined intramedullary AVMs and perimedullary arteriovenous fistulas (AVFs). After 5 years of focus fractionated radiation therapy, we treated his condition in the hybrid operating room (OR).

Case Report

The patient was a 37-year-old male who complained of hypesthesia in the lower trunk and lower right extremity in 1995. Magnetic resonance imaging (MRI) and spinal angiography led to a diagnosis of large cervical intramedullary AVMs, for which he received conservative treatment at another hospital. In May 2007, he experienced a sudden onset headache and motor weakness in both lower extremities. He was transferred to the hospital, where he was diagnosed with hematomyelia based on MRI findings. In August 2007, he was referred to our institution. On admission, he complained of sensory disturbance, hypalgesia at the left C5 and C6 level as well as at the T5 level and below, hypethesia at the T5 level and below, dysethesia in both palms as well as at the T5 level and below. MRI revealed multiple flow voids, both in front of and within the spinal cord. Low intensity signals, indicative of hemosiderin, associated with a previous hemorrhage within the spinal cord, were also noted (Fig. 1).

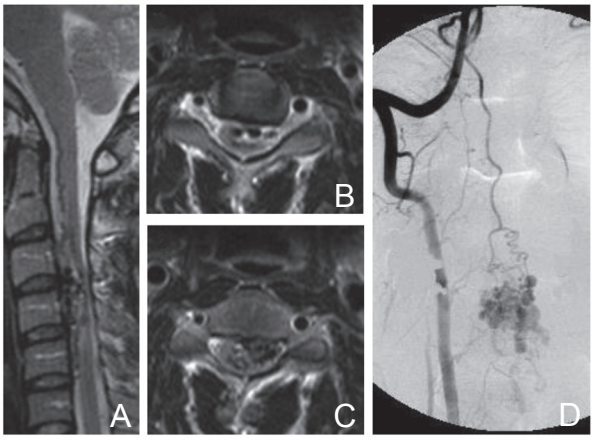

Fig. 1 Cervical spine MRI (A, sagittal T_2; B and C, axial T_2). Abnormal flow void signals in the ventral surface and inside of the cervical spinal cord at the C3 to C5 levels. D: Angiogram before treatment. Selective angiogram of the right subclavian artery (anteroposterior view) showing perimedullary AVF and intramedullary AVM fed by the anterior spinal artery from the right vertebral artery and the right deep cervical artery, respectively. AVF: arteriovenous fistula, AVM: arteriovenous malformation, MRI: magnetic resonance imaging.

Spinal angiography revealed an intramedullary AVM and perimedullary arteriovenous fistula (AVF), both of which were fed by the right vertebral artery (Fig. 2A) and the deep cervical artery via ASA (Fig. 2B). Type III AVMs were located from the surface of the spinal cord in the deepest part. Transarterial embolization and/or open surgery were not possible given the risk of ASA occlusion. Focal fractionated radiotherapy was performed using a total dose of 20 Gy in four fractions, and no complications related to radiation were noted. Following irradiation, the patient underwent a follow-up angiography every year. Five years after irradiation, angiography demonstrated that the lesion, which was fed by the right vertebral artery had disappeared (Fig. 2C), and that the other lesion, which was fed by the right deep cervical artery, had been significantly reduced to size of the nidus and the number of AV shunts (Fig. 2D). MRI revealed that flow void signals, varix, and nidus dramatically had been reduced at the C3 to C5 level of the spinal cord (Fig. 3). Dysethesia in both his palms became slightly worse during follow-up. Although transarterial embolization was considered, we gave it up because there was a feeder from deep cervical artery via the ASA. Hemorrhage due to cervical AVM has a risk of quadriplegia, therefore we planned direct surgical treatment.

I. Surgical procedure

Under general anesthesia, the patient was placed in the supine position within the hybrid OR with the neck slightly extended (Fig. 4). The spinal cord was monitored

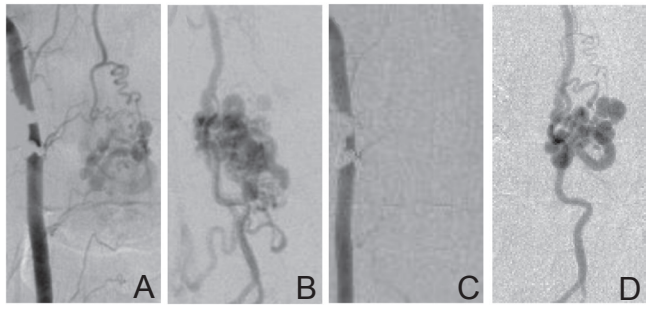

Fig. 2 A: Selective angiography of the right vertebral artery (anteroposterior view) before focal fractionated radiotherapy, revealing intramedullary AVM fed by the ASA with multiple feeders. B: Selective angiography of the right deep cervical artery (anteroposterior view) before focal fractionated radiotherapy revealing perimedullary AVF and intramedullary AVM fed by the ASA. C: Selective angiographyof the right vertebral artery (anteroposterior view) after 5 years from irradiation, revealing the disappearance of the lesion that was fed by the right vertebral artery. D: Selective angiographyof the right right deep cervical artery (anteroposterior view) after 5 years from irradiation, a reduction in size of the nidus and the number of AV shunts. ASA: anterior spinal artery, AV: arteriovenous, AVF: arteriovenous fistula, AVM: arteriovenous malformation.

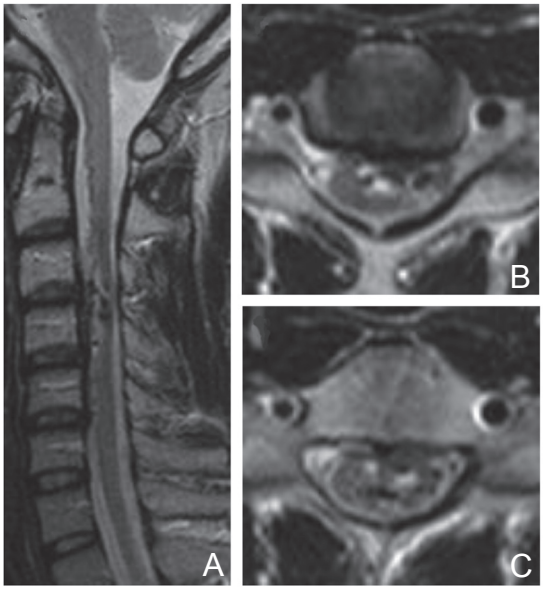

Fig. 3 Cervical spine magnetic resonance imaging (A, sagittal T_2; B and C, axial T_2). Flow void signals and varix in the ventral surface at the C3 to C5 levels, reduction of nidus within the spinal cord.

Fig. 4 Hybrid operating room at Hokkaido University Hospital, Sapporo, Japan. Single-plane neuroangiographic digital subtraction angiography units.

by motor-evoked potential (MEP) throughout the surgery. We elected to use an anterior approach because the shunt points were present in the anterior part of the spinal cord. We approached the cervical spine anteriorly on the right side through the transverse skin incision. After discectomies of C3/4 and C4/5 were performed under the operating microscope, a C4 corpectomy was performed using a high-speed drill. The inferior margin of C3 and the superior margin of C5 were also drilled out to obtain

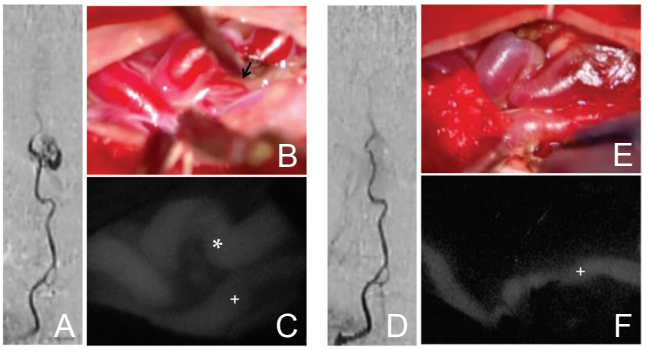

Fig. 5 A: Preprocedural anteroposterior angiogram of the right deep cervical artery confirming arterial supply to the spinal AVM via the ASA. B: Intraoperative photograph illustrating an extensive extrapial AVM nidus involving the anterior surface of the spinal cord and showing one of AV shunts (*arrow*). C: Indocyanine green videoangiography demonstrating early filling of the extra-pial draining vein (*) and ASA (+). D: Postprocedural anteroposterior angiogram of the right deep cervical artery, verifying complete AVM obliteration and patency of the ASA. E: Intraoperative photograph illustrating the color of the extra-pial draining vein which had changed from fresh red to dark red following coagulation of the AV shunts. F: Indocyanine green videoangiographycomfirms obliteration of the extra-pial draining vein and patency of the ASA (+). ASA: anterior spinal artery, AV: arteriovenous, AVF: arteriovenous fistula, AVM: arteriovenous malformation.

an adequate operative field. Gentle removal of the posterior longitudinal ligament exposed the dura matter. After opening the dura, the vascular lesion was exposed, and three AV shunts entering the venous pouches were identified by intraoperative indocyanine green-videoangiography (ICG-VA) and intraoperative angiography. ICG-VA revealed the vascular anatomy of the AVF. After coagulation of the shunt points, the color of draining veins changed from red to a normal color. Complete obliteration of the fistulous connection was confirmed by ICG and intraoperative angiography, and the ASA was preserved (Fig. 5). The dura was closed by suturing with CV-5 GORE-TEX® (WL Gore & Associates, Inc., Tokyo). Adjunctive sealants were used to avoid possible cerebrospinal fluid leakage. Simultaneous anterior fixation and reconstruction of C3–C5 were achieved using a titanium cage plus locking plate. MEPs were stable throughout the procedure.

II. Postoperative course

The patient showed a slight but noticeable weakness in his left limb after surgery, while the sensory disturbance did not change. Cervical spine X-rays showed a well-positioned titanium plate and screws. Angiography demonstrated that the fistula successfully occluded the spinal AVMs and that the ASA was preserved. The patient's neurological status improved gradually following surgery.

Discussion

Spinal vascular malformations are rare, and the prognosis for untreated lesions can be very bleak.[12] Patients with these lesions typically present with signs and symptoms of progressive myelopathy, while onset varies from slow progression to acute deterioration.[13] The goal of spinal AVM or AVF treatment is to interrupt arteriovenous communication while preserving the normal arterial supply and venous drainage of the spinal cord. Therapeutic options include surgery,[5] endovascular embolization,[3,4] radiosurgery,[6,7] or a combination of these,[9,14] and treatment strategy is often dictated by the subtype of spinal vascular malformation. For patients with spinal dural AVF, surgery can offer a permanent cure and satisfactory outcomes, with up to 98% obliteration rate and low complication rate.[15] In contrast, the optimal treatment strategy for intramedullary AVMs and perimedullary AVFs remains unelucidated. Conventionally, perimedullary AVFs were treated by surgery in which clips were placed,[16] but in recent years, endovascular treatment using particles,[17] cellulose acetate polymer solution,[18] glue,[19] or in combination with surgery[20–22] have become more prevalent. However, despite recent technological advances in endovascular devices and techniques, intramedullary AVMs are not always amenable to embolization as some arterial branches may be critical suppliers of the spine or the artery of Adamkiewicz. For this reason, microsurgical resection with or without adjuvant embolization remains the mainstay treatment for intramedullary AVMs.[5] Sinclair et al.[7] reported 15 patients with intramedullary spinal AVMs who were treated with CyberKnife radiosurgery. Follow-up angiography revealed complete obliteration in one patient and reduction in lesion size in four patients. Hida et al. reported 10 patients with intramedullary spinal AVMs treated with focal fractionated radiotherapy.[6] To date, follow-up angiography has been performed in seven patients. While none showed evidence of complete disappearance of intramedullary AVMs, the nidus size was reduced in five patients. Furthermore, the authors confirmed that neither hemorrhage nor adverse events had occurred during the follow-up period.

In our patient, the perimedullary AVF was localized to the anterior side of the cervical spinal cord, and the intramedullary AVM showed a large nidus at the C3 to C5 levels. Given the risk of ASA occlusion with endovascular embolization, we initially elected focal fractionated radiotherapy. Histopathological findings after stereotactic radiosurgery (SRS) in animal experiments as well as in clinical data suggest that SRS more successfully occludes small vessels of plexiform AVMs than larger high-flow vessels.[23,24] After MRI and angiography confirmed a reduction in nidus size and blood flow, we performed direct surgery. At 13-month follow-up, the patient showed no complications relevant to radiation therapy. Further careful follow-up is required, given the potential for radiation-related adverse effects.

We performed direct surgery and intraoperative spinal angiography for this patient in the hybrid OR. Routine intraoperative angiography during surgical procedures is more complicated when a portable digital subtraction angiography is used. Thus, combining tools from the catheterization laboratory and OR greatly enhances the options available to the neurosurgeon and endovascular surgeon as they operate on patients with spinal AVM and/or AVF.

Conclusion

Surgical treatment after focal fractionated radiotherapy could serve as one strategy for cases initially deemed ineligible for endovascular embolization and surgical treatment. The hybrid OR enables safe and precise treatment for spinal vascular disorders in the fields of endovascular treatment and neurosurgery.

Conflicts of Interest Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this article.

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