INTRODUCTION

Cerebral vascular malformations comprise arteriovenous malformation (AVM), dural arteriovenous fistula (DAVF), cerebral cavernous malformation, developmental venous anomaly, and capillary telangiectasia. DAVF is an acquired vascular malformation that is different from parenchymal AVM. DAVF comprises direct shunting between dural feeding arteries and venous sinuses without a nidus [1-3]. It is found in various locations, including the cavernous sinus (CS), transverse–sphenoid sinus, tentorium/torcula, and cerebral convexity. Although endovascular embolization with transarterial or transvenous approach is the main treatment for the obliteration of DAVF and surgical resection is rarely used, stereotactic radiosurgery (SRS) alone or SRS in combination with embolization has recently been used more frequently as a treatment modality [4-7].

SRS has emerged as an alternative treatment to improve symptoms and occlude DAVFs. This article reviews patient selection, radiosurgical technique, and clinical outcomes of SRS as well as the pathogenesis, clinical presentation, and treatment options of DAVF.

PATHOGENESIS

DAVF is an acquired lesion resulting from events such as trauma, sinus thrombosis. One of the main theories relates to the development of dural sinus thrombosis and/or venous hypertension [8]. It is hypothesized that normal arteriovenous shunts in the dura enlarge following sinus thrombosis in response to venous hypertension. Increased blood flow via these shunts causes further venous hypertension. However, not all patients with sinus thrombosis develop DAVF, and some DAVF are seen without evidence of sinus thrombosis. Another theory is that an-
giogenic factors released from the brain or dura promote the development of blood vessels in response to local factors, such as venous hypertension, resulting in the formation of DAVFs [9]. Hypercoagulability may also predispose patients to venous sinus thrombosis and the development of DAVFs [10].

**TREATMENT MODALITY**

The standard diagnostic test for DAVFs is digital subtraction angiography. It allows the identification of the feeders and venous drainage as well as determination of the location and size of the fistulas. DAVFs are classified according to the Borden [11] and Cognard classification systems [12] (Table 1). The Borden classification system has three groups based on venous drainage site and cortical venous drainage (CVD). Cognard’s classification system includes five groups based on venous drainage site, flow pattern in sinus, and CVD. Borden type 1 or Cognard type I or Ia are considered benign or low-risk DAVFs. Borden type 2 and 3 as well as Cognard type IIb, IIa+b, III, and IV are aggressive and high-risk DAVFs.

DAVFs with an aggressive nature, such as cerebral hemorrhage or other severe symptoms, require active treatment. Some DAVFs without CVD follow a benign clinical course and respond to conservative therapy [13]. DAVF can cause cerebral hemorrhage, seizure, and progressive neurological symptoms. Symptoms of DAVFs depend on their location, ranging from pulsatile tinnitus associated with DAVFs in the transverse–sigmoid sinus, to ocular pain and chemosis associated with DAVFs in the CS. The choice of treatment depends on factors related to the lesion or the patient, such as fistula location, flow profile, angiographic features, and clinical symptoms or signs [14]. CVD, venous ectasia, hemorrhage, and progressive clinical course are critical factors that indicate the need for an active therapeutic intervention. To completely occlude fistulas, it is important to determine the treatment methods to be prioritized, the methods that can be used alone, and those that can be used in combination with others. Endovascular treatment is the main treatment method for DAVFs with venous hypertension or progressive neurological deficits [5,6]. Endovascular embolization of DAVFs is often incomplete because of difficult vascular approach, accidental migration of embolic materials, insufficient embolization, or the complex angioarchitecture of these fistulas. Incomplete embolization can cause surrounding vessels to pool, leading to an even more dangerous situation. SRS can be a suitable treatment modality for DAVFs without high-risk situations such as cerebral hemorrhage. SRS may be performed when a conventional approach is dangerous, or in case of incomplete results after previous treatments, and even when there are surgery or endovascular embolization-related risks. SRS alone has been recommended for asymptomatic patients or patients with minor symptoms and those without aggressive angiographic features such as CVD.

**STEREOTACTIC RADIOSURGERY**

Stereotactic radiosurgical technique is described in the Table 2. Stereotactic radiation targets both dural feeder arteries and fistulas, causing radiation-induced vessel hypertrophy, finally leading to occlusion of the fistulas (Fig. 1). Recently, SRS was recommended as

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**Table 1. Classification of dural arteriovenous fistulas**

| Borden classification | Cognard classification |
|-----------------------|------------------------|
| 1 Venous drainage directly into the dural venous sinus or meningeal vein | I Venous drainage into the dural venous sinus with antegrade flow |
| 2 Venous drainage into the dural venous sinus with CVD | IIa Venous drainage into the dural venous sinus with retrograde flow |
| 3 Venous drainage directly into subarachnoid veins (CVD only) | IIb Venous drainage into the dural venous sinus with antegrade flow and CVD |
| Cognard classification | Ia+IIb Venous drainage into the dural venous sinus with retrograde flow and CVD |
| I Venous drainage into the dural venous sinus with antegrade flow | III Venous drainage directly into subarachnoid veins (CVD only) |
| IIa Venous drainage into the dural venous sinus with retrograde flow | IV Type III with venous ectasias of the draining subarachnoid veins |
| IIb Venous drainage into the dural venous sinus with antegrade flow and CVD | V Direct drainage into spinal perimedullary veins |

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**Table 2. Stereotactic radiosurgical technique for DAVFs**

| Treatment course | Comment |
|------------------|---------|
| 1. Patient selection | All DAVFs were diagnosed using conventional cerebral angiography. |
| 2. Head frame fixation | Proper frame placement is important depending on the location of the DAVF. |
| 3. Stereotactic CT or MRI | Identifying adjacent critical structures. |
| 4. Stereotactic cerebral angiography | Targeting the DAVF. |
| 5. Dose planning | Proper dose selection and defining the DAVF in the wall of a venous sinus. |
| 6. Stereotactic radiosurgery | Gamma Knife, etc. |
| 7. Imaging follow-up | Cerebral angiography 2 or 3 years after radiosurgery. |

DAVF: dural arteriovenous fistula, CT: computed tomography, MRI: magnetic resonance imaging.
SRS for DAVF

a non-invasive, low-risk treatment option for DAVFs if a risk associated with conventional treatment exists or if the previous treatment has failed. In terms of SRS, it is often difficult to define complex fistulas in the venous sinus wall when planning treatment because DAVFs have a complicated angiography. Thus, selective angiography is required as part of treatment planning to account for multiple fistulas being fed by various arteries. Critical feeders may be missed if selective angiography is not performed. The radiosurgical target should be limited to the arteriovenous shunt; however, the surrounding venous sinus is likely to be irradiated. Nevertheless, unlike AVM, venous sinus irradiation does not appear to be a major concern, and similar treatment effect in noted for DAVF with transvenous embolization.

In high-risk patients with aggressive features, good treatment results are rarely reported in cases where SRS alone is performed according to patient’s preference. This may be owing to the low flow pattern of DAVFs. Except for the latency period, active treatment with non-invasive SRS is often helpful in improving neurological symptoms because the time to improvement of symptoms after SRS is relatively short. SRS often results in a higher complete occlusion rate with symptom improvement. Typically, DAVF occlusion occurs within 1 to 3 years after SRS. The latency period between SRS and complete occlusion is one of the main drawbacks of SRS. Compared with AVMs, DAVFs seem to more promptly react to SRS because the shunt size in DAVFs is smaller than that in AVMs [15,16]. Moreover, slow occlusion of DAVFs may reduce the likelihood of venous hypertension [17].

Complete occlusion was reported in 56% of DAVFs with CVD and in 75% of DAVFs without CVD [18]. DAVFs without CVD were associated with a significantly higher rate of complete occlu-

Fig. 1. (A) Magnetic resonance imaging (MRI) showed an intracerebral hemorrhage on the left basal ganglion. Before stereotactic radiosurgery, (B) an anterior angiographic view of the right internal carotid artery and (C) lateral view of the right external carotid artery (ECA) show dural arteriovenous fistulas (DAVFs) involving the cavernous sinus with cortical venous drainage (B). (D) Dose planning shows the integration of stereotactic angiography and MRIs. The DAVF was treated with a radiation dose of 16 Gy at the 50% isodose line. (E) A lateral angiographic view of the right ECA obtained 2 years after stereotactic radiosurgery revealed complete obliteration of the fistulas.

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sion after SRS than DAVFs with CVD. DAVFs with CVD have a higher risk of hemorrhage and a lower occlusion rate than DAVFs without CVD. CS DAVFs were associated with higher rates of obliteration than transverse–sigmoid DAVFs, which have a complex angioarchitecture [19,20]. A larger size, recanalization, and different shunting flow are associated with poor treatment outcomes in transverse–sigmoid DAVFs. CS DAVFs were associated with better clinical outcomes than DAVFs of the transverse–sigmoid sinus.

Asymptomatic, low-risk DAVFs may be conservatively treated. However, several DAVFs progress gradually, with symptoms worsening until a seizure or cerebral hemorrhage occurs. If they are located in the transverse–sigmoid sinus, active therapeutic intervention can be considered, even for DAVF with a benign course. DAVFs with CVD have a higher risk of hemorrhage and incomplete occlusion rate than those without CVD. Transverse–sigmoid sinus DAVF are more likely to be accompanied by CVD and have more aggressive angiographic features than CS DAVF.

Endovascular embolization has a potential risk of ischemic complications and migration of embolic material. Transvenous embolization affects patients at risk of cerebral hemorrhage owing to venous hypertension. CS embolization can cause cranial nerve damage. Cranial nerve signs after embolization may be due to progressive thrombosis of the CS, mass effect from the coils, or direct damage to the nerve by coils. Yoshida et al. [21] reported that complications such as cranial nerve palsy, brain stem infarction, and intracerebral hemorrhage were observed in the form of permanent morbidity in 7% of the patients and transient morbidity in 14% of the patients. There are few reports on radiosurgery-related complications after SRS for DAVFs. Cifarelli et al. [22] reported that T2-weighted radiation-induced changes were observed in 12% of the patients.

CONCLUSION

Depending on the patient’s condition and the angiographic features of DAVFs, SRS may be an alternative and safe treatment modality for DAVFs. SRS can be performed when it is difficult to treat DAVF endovascularly or surgically or when the patient prefer a non-invasive treatment.

CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

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