Cerebral Arteriovenous Fistulas Induced by Dural Arteriovenous Shunts

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Summary: Dural arteriovenous shunts and pial arteriovenous fistulas are uncommonly associated. Their etiology, pathogenesis, and natural history are still unclear and are likely different. We present three cases of high-flow dural arteriovenous shunts associated with pial arteriovenous fistulas and discuss their pathogenesis, anatomic association, and angioarchitecture. We propose that venous steal effect in the dural sinus secondary to the high-flow dural arteriovenous shunt induced the pial arteriovenous fistulas. Treatment of the high-flow dural arteriovenous shunts and the induced pial arteriovenous fistulas are discussed.

Dural arteriovenous shunts (DAVSs) associated with pial arteriovenous fistulas (AVFs) have been rarely reported (1–3). Cerebral pial AVFs induced by high-flow DAVSs are an exceptional subgroup of this uncommon association.

Case 1

A male patient presented at age of 17 months with intracerebral hemorrhage at the age of 3 years in September 1998. Angiography showed multifocal DAVSs supplied by dural arteries at both sides. The main supply was from branches of the right middle meningeal artery to the anterior superior sagittal sinus, from the mastoid branch of the right ascending pharyngeal artery to the right transverse sinus, and the ethmoidal branch of the left ophthalmic artery to the anterior superior sagittal sinus. There was cortical venous congestion and reflux. The right transverse sinus showed stenosis and subsequently occluded, resulting in a residual venous pouch. There were bilateral pial AVFs supplied by branches of the anterior cerebral arteries draining via cerebral cortical veins into the superior sagittal sinus. The patient was treated, from the end of 1998, by multiple sessions of transarterial glue embolization to several of his DAVSs, followed by embolization of the residual pouch of the right transverse sinus, and the ethmoidal branch of the left ophthalmic artery to the anterior superior sagittal sinus. There was cortical venous congestion and reflux. The right transverse sinus showed stenosis and subsequently occluded, resulting in a residual venous pouch. There were bilateral pial AVFs supplied by branches of the anterior cerebral arteries draining via cerebral cortical veins into the superior sagittal sinus. The patient was treated, from the end of 1998, by multiple sessions of transarterial glue embolization to several of his DAVSs, followed by embolization of the residual pouch of the right transverse sinus. Because of recurrent neurologic deterioration, the high-flow pial AVFs at both anterior cerebral artery territories were embolized with glue, resulting in transient improvement. Finally, the patient died from posterior fossa venous congestion and hemorrhagic infarction in November 2003 at the age of 8 years.

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cases. The similarity of the angioarchitecture and the close anatomic relationship of the multiple DAVSs to the pial AVFs in our cases lead us to propose that these lesions are pathogenetically linked.

Steal Phenomenon

An arterial steal phenomenon is often discussed when pial arteriovenous shunts (AVSs) appear to divert flow toward a low-resistance vascular bed, away from adjacent regions. Intracranial pressure measurements and transcranial velocity studies have shown regional hemodynamic alterations and evidence from single-photon emission CT scans suggest a relationship between regional hypoperfusion and neurologic deficits (4). Similar steal effects have been proposed to account for the pial AVFs associated with infantile DAVSs (5).

Pial AVFs

Pial AVFs are considered to be congenital in nature (6). By contrast, there is little evidence that the pial AVF diagnosed in adults are present in the same form at birth. In addition, cases of de novo pial AVF have been reported (7–10). With knowledge of endothelial cell origin (such as the study of genetic vascular diseases including hereditary hemorrhagic telangiectasia and Rendu-Osler-Weber disease) and the biology of the remodeling process of the vascular system, Lasjaunias (6) proposed that the congenital event is primarily involving the vascular modeling and remodeling process at the cellular and structural level. It is likely to affect the endothelial cells at the venous side of the capillaries, resulting in a quiescent dysfunction. The structural manifestation of this dysfunction depends on revealing triggers such as mechanical, hormonal, pharmacologic, hemodynamic, thermal, radiation, viral, infective, and metabolic factors. The diverse nature and timing of the various triggers causes the different time and form of presentation of the abnormal AVS. This postulation can also be extended to explain an acquired lesion to occur as a postnatal acquired event. Under these circumstances, the trigger was applied for a certain length of time and had the same consequence as the postulated congenital event on the target cells related to venous vascular remodeling. The pathogenesis of pial AVF also includes abnormal expression of various angiogenic factors (11–13) such as vascular endothelial growth factor, basic fibroblast growth factor, and alpha transforming growth factor. Suppression of vascular cell growth modulators, including endoglin1, has also been found (14). Cases of acquired pial AVFs in Moya-moya disease and following cerebral vein thrombosis (8, 9) suggest ischemia or hypoxia is an important etiologic factor. Hypoxia is a powerful trigger for the up-regulation of angiogenic factors expression (15).

DAVSs

DAVSs are a heterogeneous group of lesions that were initially considered to be of congenital origin. They can now be classified as dural sinus malformations, infantile DAVSs, and adult types of DAVSs (16). Only the dural sinus malformation is truly congenital in origin. Many reports have documented the
acquired nature of DAVS after dural sinus thrombosis, intracranial infection, trauma, surgery, and occlusion of the dural sinuses by surgery or tumor (17–23). DAVSs are also known to be associated with pregnancy and hypercoagulation states (24). Experiments in a rat model have suggested that chronic venous hypertension is the most important etiologic factor (25). It has been implied that abnormal angiogenic activity is induced directly or indirectly by venous hypertension through decreasing cerebral perfusion and increasing ischemia (26). These activities are likely mediated through abnormal expression of endothelial-cell vascular growth factors (e.g., vascular endothelial growth factor, basic fibroblast growth factor), which are demonstrated in DAVS (27).

**The Association of DAVSs and Pial AVFs**

**Dural AVSs Inducing Pial AVFs.** Anatomically, both dural and cortical veins open directly into the dural sinus. Abnormal AVSs between dural arteries and veins without intervening capillaries are formed in the wall of the dural sinus triggered by increased venous pressure or thrombosis (28). Increased flow of blood into the low-resistance system of dural sinus can exert a venous steal effect on the upstream cortical veins opening directly into the dural sinus (16). The exact mechanism of the opening up of the pial shunt into these cortical veins remains unknown. The result of steal effect may cause hypoxia in the regions drained by these cortical veins. The hypoxia can result in up-regulation of the angiogenic factors that act on the capillarovenous endothelial cells and other cellular process involved in the remodeling of the vascular system for a significant period of time triggering the formation of the pial AVFs. Because these induced pial AVFs seem to locate preferentially at certain parts of the dural sinus (e.g., at the superior sagittal sinus in our cases) there may be additional regional factors to account for it. The influence on cortical veins by the high flow in the dural sinus is affected by factors such as the orientation of the cortical vein with respect to the dural sinus, the size and shape of the vein, function of the venodural junction, the flow characteristic in the dural sinus near the opening of the cortical veins, and so forth. The multiple anatomic factors and the various hemodynamic situations in the vicinity of the opening of these cortical draining veins dictate whether the steal effect is efficient as a trigger for the pial AVFs and where the location of these fistulas form upstream from the high-flow DAVSs.

**Pial AVF Inducing DAVS.** By contrast, the possibility of pial AVFs inducing DAVSs has been proposed particularly in some high-flow pial AVFs associated with DAVSs upstream from their drainage into the dural sinus (29). This could be caused by a similar sump effect created by the high-flow venous drainage of the pial AVF downstream. The venous changes produced by high-flow pial AVS on the venous sinuses, such as increased venous pressure or venous outflow obstruction, can also be triggering factors. But this is not the situation in our cases, because they were high-flow DAVSs anatomically closely related to the pial AVFs that were upstream from the DAVSs. The dominant shunts in our cases were high-flow DAVSs rather than pial AVFs.

**Thrombosis of Local Cortical Vein Inducing Pial AVF.** Local thrombosis of cortical veins has been reported in one case to precede the de novo formation of pial AVF (8). It is not possible for us to postulate the same mechanism to account for our cases, because we do not have evidence whether there was thrombosis of the cortical veins before the formation of the pial AVFs. DAVS may rarely have local pial supply, which is more often in the occipito-temporal region (21). This may be related to previous venous thrombosis, hemorrhage, or cortical venous ischemia. Our cases do not seem to fit into this type of association.

The DAVSs of all our cases were draining into the superior sagittal sinus. The induced pial AVFs are from branches of anterior cerebral arteries. This association may indicate that the superior sagittal sinus is a common site for high-flow DAVSs to induce pial AVFs through the venous steal mechanism.

**Clinical Course and Management**

The clinical course of high-flow multiple infantile-type DAVSs is often progressive, and their prognosis is poor (16). They require multiple endovascular interventions and/or operations, as in our cases. Case 1 required multiple repeated arterial embolizations for the DAVSs, transvenous coil obliteration of the draining venous pouch, venous stent placement, and dilation of sinus strictures. The natural history of the induced pial AVFs is not known, but they can subside after occlusion of the triggering DAVSs (16). It is interesting to note that in case 1 the pial AVFs continued to persist after coiling of that part of the superior sagittal sinus. This may be secondary to the remaining steal effect and the local hemodynamic of the dural sinus near the cortical veins draining these pial AVFs. The triggered dysfunction creating the pial AVFs may persist despite the cessation of the triggering factors. These pial AVFs appear to contribute to the clinical deterioration of the patient in case 1 by contributing to the venous hypertension. Embolization of the pial AVFs in case 1 has led to clinical improvement of the patient. The primary consideration of treatment, however, is of the high-flow DAVSs because they represent the primary lesion mainly responsible for the clinical conditions of the patients. Some of the induced pial AVFs disappear after treatment of the high-flow DAVSs (16). The risk of these induced pial AVFs is unknown. Their treatment decision is based on the knowledge extrapolating from the behavior of pial AVFs in general and on their specific contribution to the clinical symptoms. The induced pial AVFs can cause secondary arterial changes as in case 2, who had an aneurysm (Fig 4) related to the high flow in the shunt, formed in the arterial feeder supplying the fistulas. Case 2 is an example that high-flow DAVSs can also occur in
AVFs can be useful to improve the clinical symptoms. In that case, endovascular embolization of these pial AVFs both draining to the superior sagittal sinus (large arrow). Reflux into cortical vein is noted (small arrow).

adults and induce pial AVFs similar to those in children or infants.

Conclusion

Our cases were all high-flow DAVSs with anatomically closely related pial AVFs both draining into the superior sagittal sinus. We postulated the high-flow DAVSs cause a venous steal (sump) effect that, in a suitable combination of anatomic and hemodynamic factors, induces pial AVFs. Treatment should focus on treating the DAVS, which is the primary lesion and likely to be the main cause of symptoms. These pial AVFs can subside after successful treatment of the high-flow DAVSs, but they can persist and contribute to the clinical symptoms of the patients. In that case, endovascular embolization of these pial AVFs can be useful to improve the clinical symptoms.

References