Combined surgical and endovascular treatment of a spontaneous diploic arteriovenous fistula

Case report

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The authors report on the case of a 28-year-old woman presenting with an intraosseous arteriovenous fistula (AVF) located in the left parietal bone. The fistula was formed by direct arteriovenous shunts connecting branches of the left middle meningeal and superficial temporal arteries with a parietal diploic vein. Drainage occurred through both the external and internal jugular venous systems. Therapy consisted of combined surgical and endovascular approaches. The results of a pathological examination of the resected AVF showed mild enlargement of the diploic space. The angiographic appearance, pathological anatomy, and treatment of this rare lesion are discussed, as is a possible relationship between diploic AVFs and the development of aneurysm bone cysts.

Key Words • arteriovenous fistula • surgery • endovascular management • diploic space

Diploic veins course within the diploë, that is, the cancellous bone located between the inner and outer tables of the skull vault. Their precise anatomy and function remain poorly understood. Diploic veins are found most abundantly within the parietal bone and normally communicate with dural sinuses and pericranial veins. Occasionally, they are demonstrated on cerebral angiography, their opacification being typically slightly delayed when compared with the rest of the cerebral venous system. Diploic veins may become prominent when the normal cerebral drainage pathways are compromised. For example, they commonly serve as collateral routes in the event of superior sagittal sinus obstruction by a meningioma. Arteriovenous fistulas involving diploic veins are extremely rare, with only a few posttraumatic cases having been reported thus far. We describe the angiographic appearance, pathological characteristics, and therapeutic approach in a case of spontaneous diploic AVF.

Case Report

History and Examination. This 28-year-old woman with a history of Type 1 diabetes, arterial hypertension, and Hashimoto thyroiditis presented with left parietal headaches followed several weeks later by a left pulsatile tinnitus. There was no history of trauma or other precipitating event. Note that the patient had given birth to her first child several months before the onset of symptoms. The pregnancy had been complicated by preeclampsia with persisting hypertension after birth. Results of general and neurological examinations were normal. A pulsatile bruit was confirmed by auscultation of the left retro-auricular region.

Neuroimaging Results and Treatment. The findings of carotid artery ultrasonography as well as those of cervico-cranial magnetic resonance imaging and angiography were nondiagnostic. Digital subtraction angiography revealed an AVF in the left parietal region (Fig. 1A and B). The AVF was fed by multiple branches of the left STA and MMA. Drainage initially occurred through a venous structure having the typical appearance and topography of a parietal diploic vein, but showing an abrupt change in morphological features and continuing as a subcutaneous vein. This latter vessel, identified as the left retro-auricular vein, drained into the left external jugular vein through a complex subcutaneous network but was also connected to the left sigmoid sinus via two small emissary veins.

The patient decided to undergo surgical treatment for the AVF. A preoperative angiogram was obtained in the operating room to determine the exact location of the fistula with the aid of skin markers. The prominent branches of the STA coursing through the temporalis muscle into the bone were cauterized using bipolar forceps and the exposed surface of bone was Bovied using monopolar cautery. Six burr holes were drilled around the site of the fistula, as determined on preoperative angiography. The burr holes were then waxed and connected using a high-speed drill. The bone flap was gently elevated, the edge of the craniectomy defect was promptly waxed, and the middle meningeal branches reaching the inner surface of the bone flap were cauterized. The piece of bone containing the AVF was excised with minimal complications.
blood loss, and several prominent feeding vessels coming from the MMA were cauterized. An intraoperative angiogram verified the absence of any residual arteriovenous shunt. Gross and microscopic examination of the specimen confirmed the diploic location of the fistula and the presence of an enlarged diploic vein with a thickened, arterialized wall (Fig. 1B). The AVF was not found to be directly involved with the dura mater.

**Postoperative Course and Additional Treatment.** The patient’s symptoms initially resolved completely, but the bruit and headache returned within 1 week of surgery. Results of a follow-up angiogram revealed a recurrent arteriovenous shunt that directly involved the retro-auricular vein. No evidence of direct involvement of this vein, which previously served as a drainage pathway for the fistula, had been found on either the initial diagnostic or the intraoperative angiography study. The diploic component of the lesion was no longer present but that there is a new or persisting AVF located in the retro-auricular vein (arrow) and fed by multiple small arterial branches from the left external carotid artery (occipital, posterior auricular, and superficial temporal arteries). Venous drainage occurred through the left sigmoid sinus (asterisk) via a mastoid emissary vein (white arrowhead) and toward the left external jugular vein (black arrowhead). D: A DS angiogram obtained after superselective injection of the venous side of the fistula. The microcatheter has been advanced through the sigmoid sinus (asterisk) and the mastoid emissary vein (white arrowhead) into the retro-auricular vein, with its tip at the site of the fistula (black arrowhead). E: A DS angiogram, left external carotid artery injection (lateral view), obtained following embolization of the retro-auricular vein with detachable microcoils, showing complete obliteration of the arteriovenous shunt. Note the placement of microcoils above and below the level of connection with the mastoid emissary vein.

**Discussion**

Arteriovenous fistulas involving the diploic venous system are rare and generally traumatic in nature; spontaneous diploic AVFs appear to be exceptional. Ikeda, et al., described one patient with a right frontoparietal aneurysm bone cyst, which was attributed to an AVF between the right MMA and a diploic vein with secondary enlargement of the diploic space.

The patient in the current report presented with an intrasosseous AVF localized to the left parietal region. The angiographic appearance of the main venous structure involved by the fistula strongly indicated a diploic vein, a finding later confirmed on surgical exploration and examination of the resected specimen. Unlike the traumatic diploic AVFs reported in the literature, which were vascularized by torn meningeal branches, the AVF in the present case had a dual arterial supply, with feeding arteries coming from both the left STA and MMA. The patient had no history of trauma and could not remember any instance of even minor head injury. We therefore assume that her lesion occurred spontaneously, unrelated to a traumatic event.

The possible causative relationship between diploic
Spontaneous diploic arteriovenous fistula

AVFs and aneurysm bone cysts posited by Ikeda and colleagues is interesting. It has been proposed that a primary bone lesion, such as fibrous dysplasia, may initiate an osseous AVF, which in turn creates the reactive lesion of the aneurysm bone cyst. Although the patient in the present case demonstrated no radiographic or anatomical evidence of an aneurysm bone cyst, analysis of the surgical specimen did reveal evidence of bone erosion and a widening of the diploic space at the site of the fistula. Note that the time elapsed between initial presentation and lesion resection was brief. Perhaps a longer evolution would induce pathological changes similar to an aneurysm bone cyst. In our experience, it is not unusual to find arteriovenous shunts in association with aneurysm bone cysts, and the possible role of AVFs in the development of aneurysm bone cysts has been previously reported. The lesion described in the current report could therefore represent the earliest expression of an aneurysm bone cyst.

Note that the pattern of venous drainage was unusual in the present case. Although the parietal diploic vein normally drains directly into the ipsilateral transverse sinus, the diploic vein harboring the fistula in our patient ended abruptly before reaching the sinus and instead drained into the extracranial, superficial vein. This scalp vein located in the left retro-auricular region was responsible for the pulsatile mass described by the patient. It drained into the left external jugular vein through a complex venous network but was also connected to the sigmoid sinus via two small emissary veins. The abrupt ending of the lower end of the parietal diploic vein coupled with the collateral appearance of the actual drainage through a superficial vein indicated thrombosis of the normal termination of the parietal diploic vein in the transverse sinus. Arteriovenous fistulas involving the dural sinuses have been linked, at least in some well-documented cases, to the recanalization of previously thrombosed dural sinuses. Based on the angiographic characteristics mentioned previously, we think that the lesion in the present case may have been caused by a similar mechanism, that is, initial thrombosis of the diploic vein with partial recanalization and AVF formation. Our patient’s history of a complicated pregnancy shortly before the onset of symptoms is compatible with this hypothesis. Increased estrogen levels during pregnancy increase the risk of thrombus formation, and dural sinus thrombosis is indeed a classic complication of the puerperal period. Alternatively, the fistula may have been created at some previous time and brought out of latency in response to the increased blood volume of pregnancy.

The recurrence of symptoms shortly after resection of the diploic AVF was unexpected given that the initial diagnostic angiogram revealed no direct involvement of the retro-auricular vein, and the angiogram obtained in the operating room immediately after resection documented the absence of any residual arteriovenous shunt. Although the formation of a new fistula within a draining vein that thrombosed as a result of the surgical treatment cannot be completely excluded, it seems that the period of time elapsed before the reappearance of pulsatile tinnitus (~1 week) is probably too brief to be compatible with such a mechanism. Instead, we think that dormant, angiographically undetectable fistulous connections most likely involved the retro-auricular vein from the beginning and that their sudden increase in size was stimulated by the surgical removal of the main arteriovenous shunt.

Finally, the unusual endovascular route used to access the fistula involving the retro-auricular vein merits mention. Because the quantity and complex anatomy of the feeding arteries rendered transarterial embolization impractical, a transvenous approach was elected. As mentioned earlier, however, the draining pathways of the fistulous vein coursed through an intricate network of small veins not easily negotiated. As an alternate route, retrograde catheterization of the left internal jugular vein and sigmoid sinus was chosen to take advantage of a mastoid emissary vein connecting the sigmoid sinus to the fistulous vein. This endovascular approach proved remarkably favorable and allowed fast and safe access to the fistula.

Conclusions

Spontaneous diploic AVFs appear to be exceptionally rare lesions. The case reported here includes angiographic and pathological characteristics that could be typical of this condition: 1) vascularization both by intracranial (meningeal) and extracranial (superficial temporal) arterial branches (as opposed to traumatic AVFs that derive their blood supply from a torn meningeal artery); 2) associated morphological anomalies consistent with venous thrombosis, indicating an etiopathogenic similarity between spontaneous diploic AVF and a dural AVF; and 3) secondary enlargement of the diploic space, possibly representing an early stage of aneurysm bone cyst formation.

References


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