Pediatric Traumatic Dural Arteriovenous Fistula

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ABSTRACT: Background: Dural arteriovenous fistulas are uncommon lesions in children, with traumatic lesions suspected to arise from incomplete arterial injury in proximity to a vein or draining sinus. Management of symptomatic acquired lesions requires evaluation of patient presentation, neurological status, and pathoanatomic configuration, with special consideration required for surgery secondary to failed endovascular technique. Case Report: A 12-year-old male sustained a bicycle fall causing a right temporo-parietal skull fracture associated with non-surgical right epidural hematoma and left contre-coup parietal contusion. Six-weeks later, he complained of a right temporal bruit with subsequent cerebral angiography demonstrating a dural-based fistula between the right middle meningeal artery and a dural vein draining into the sigmoid sinus. Intervention: Endovascular treatment of this lesion with glue embolization and coiling was unsuccessful, with angiographic illustration of previously unobserved collateral vessels and coils occupying the sigmoid sinus. A right temporo-parietal craniectomy was required to excise the dural-based fistula, followed by dural defect repair with bovine pericardium and subsequent cranioplasty. Six years later the patient remains neurologically intact with no headaches or bruit. Conclusions: Dural arteriovenous fistula can uncommonly occur following traumatic injury in children. Partial injury to the middle meningeal artery may have established arterial communication with the draining vein that became ectatic and tortuous under high pressure. Failure of primary endovascular treatment may complicate secondary surgical intervention.

RÉSUMÉ: Fistule artério-veineuse durale d'origine traumatique chez l'enfant. Contexte : Les fistules artério-veineuses durales sont des lésions rares chez les enfants et on soupçonne que les fistules traumatiques résultent d’un traumatisme artériel à proximité d’une veine ou d’un sinus de drainage. La prise en charge du patient qui présente une lésion acquise symptomaticque doit tenir compte de la symptomatologie, inclure une bonne évaluation clinique afin d’établir l’état neurologique et préciser la configuration anatomopathologique en portant attention à certains éléments importants si une chirurgie était requise suite à l’échec d’une intervention endovasculaire. Cas clinique : Un garçon de 12 ans a subi une fracture du crâne temporo-pariétale droite, avec hématome epidural droit ne requérant pas de chirurgie, et une contusion pariétale gauche par contrecoup lors d’une chute à bicyclette. Il s’est plaint six semaines plus tard d’un bruit temporal droit et une angiographie cérébrale a montré la présence d’une fistule durale entre l’artère méningée moyenne droite et une veine durale se drainant dans le sinus sigmoïde. Intervention : Le traitement endovasculaire de cette lésion par embolisation de colle et de spirales métalliques (coils) n’a pas été noté antérieurement et des coils dans le sinus sigmoïde. Nous avons dû procéder à une craniectomie temporo-pariétale droite pour exciser la fistule dure-mérienne, réparer la brèche dure-mérienne avec du péricarde bovin et procéder à une cranioplastie. Le patient ne présente pas de séquelles neurologiques, de céphalée ou de bruit 6 ans plus tard. Conclusions : Chez les enfants, les fistules artério-veineuses durales sont rares suite à un traumatisme. Une lésion partielle à l’artère méningée moyenne peut avoir provoqué une communication entre l’artère et la veine de drainage qui est devenue ectasique et tortueuse lorsque soumise à une pression élevée. L’échec du traitement endovasculaire peut compliquer une chirurgie subséquente.


Intracranial dural arteriovenous fistulas (AVFs) account for 10-15% of all intracranial arteriovenous lesions,1 and in the pediatric population these lesions are often multiple and more complex in configuration than in the adult population.2-4 The lesion consists of an abnormal direct connection between a meningeal artery and either a meningeal vein or dural sinus. High mortality is observed among pediatric patients, with presentation in congestive heart failure among neonates and neurological deterioration in older children reported as high as 67% and 38% for these two groups respectively.5 Kincaid and coworkers describe a series of seven likely prenatally acquired lesions with a review of the surrounding literature describing evidence against true congenital anomaly to include no genetic abnormality or association with other vascular anomalies alongside a mature vascular configuration.6,7 They advocate for expeditious endovascular embolotherapy as the mainstay of therapy for life-saving result in the setting of cardiovascular failure and symptomatic improvement in neurological dysfunction. Indeed, these lesions also exhibit a more aggressive...
clinical course than in adults,\textsuperscript{3,4} although the natural history of traumatic acquired pediatric lesions is less well defined and may have a degree of indolence more characteristic of older individuals.

We present a case of a 12-year-old male who developed a benign acquired dural arteriovenous fistula following traumatic head injury. The patient was symptomatic from pulsatile tinnitus, failed endovascular therapy, and required surgical excision of the lesion to provide for adequate symptomatic relief.

**CASE REPORT**

A 12-year-old male sustained a closed head injury by falling unhelmed from a bicycle. Following a short staring spell that lasted two minutes, he began to speak coherently and had intact motor and sensory function. Upon presentation to the hospital, he complained of headaches, had thrice vomited, and was amnestic to the period surrounding the accident. Subgaleal swelling was observed in the right temporo-parietal region. Level of consciousness was maintained at Glasgow Coma Scale

![Figure 1: CT Head at the index head injury. (A) Non-depressed fracture of the right squamous temporo-parietal skull. (B) Underlying epidural hematoma associated with mild mass effect and no midline shift. Left contre-coup parietal lobe contusion and subarachnoid hemorrhage.]
of 15 in the emergency room and the neurological examination was normal.

Computed tomography of the head (Figure 1) revealed a non-depressed skull fracture in the squamous portion of the right temporal bone. This fracture was overlaid on an 8 mm epidural hematoma associated with mild mass effect and no midline shift. There was also a contre-coup left parietal contusion and traumatic subarachnoid hemorrhage. The patient was treated conservatively and discharged home three days following injury.

At six weeks, the patient complained of mild occipital headaches and pulsatile tinnitus that lasted for two weeks. This was most prominent on the right and in the bending position. Physical examination revealed a bruit immediately above the right ear. The remainder of the neurological examination was normal.

Magnetic resonance imaging revealed traces of hemosiderin in the right temporal and left parietal lobes. Magnetic resonance angiography showed a tangle of vessels in the right posterior temporal and inferior parietal region to originate from the external carotid artery. Magnetic resonance venography showed a small right transverse sinus, sigmoid sinus and internal jugular vein. Cerebral angiography demonstrated a large high-flow fistula from branches of the right middle meningeal artery emptying into a large vein that drained into a patent sigmoid sinus.

Endovascular treatment was attempted with selective transarterial placement of Gugliemi detachable coils (GDCs) (Boston Scientific) and Vortx fibered coils (Boston Scientific) into a feeding branch of the right middle meningeal artery (Figure 2). A second branch was infused with N-bucrylate and Lipiodol. These maneuvers resulted in reduced flow through the AVF. Supplemental transvenous catheterization was attempted with placement of Vortx fibered coils into the venous side of the fistula, although this was unsuccessful at reducing patency. Diagnostic cerebral angiography one month later showed persistence of the dural AVF with interval hypertrophy of the middle meningeal artery branches.

The dural AVF was successfully excised with the surrounding dura, through an elective right temporo-parietal craniectomy (Figure 3). The dura was circumferentially coagulated and transected peripheral to the bone flap, found to be adherent to the dura in the region of the AVF. Hemostasis was achieved from multiple arteries within the dura, but the venous outflow at the posterior and inferior margins was more difficult to coagulate because of the endovascular coils. Once the bone flap was lifted with the centrally adherent dura, there was no cortical component identified to the AVF. The brain was stained with hemosiderin, consistent with the patient’s history of contusion. The dural defect was repaired with bovine pericardium (DuraGuard, Synovis) and a cranioplasty was performed using hydroxyapatite bone cement (Mimix, Walter Lorenz Surgical). The postoperative course was unremarkable, with resolution of the right pulsatile bruit. Symptoms have not recurred in six years follow-up.

Figure 2: (A) Right external carotid angiogram revealed a dural arteriovenous fistula, supplied by a hypertrophied middle meningeal artery and two posterior branches and draining early through a dural vein into the sigmoid sinus. Through the larger of the two posterior branches, GDC and Vortx fibered coils were positioned through the fistula into the vein. Transvenously, Vortx fibered coils were deposited into the fistula, which displaced into the sigmoid sinus not allowing future microcatheter passage. (B) Scout view demonstrating position of coils.
Pathological examination of the surgical specimen was consistent with an AVF undergoing partial obliteration after embolization with a granulomatous foreign body response. Firm fibrous tissue was observed on the inner surface of the excised rectangular bone flap (Figure 3). Histological evaluation revealed a gap in the bone occupied by non-inflammatory dense fibrous tissue and a blood vessel with mural chronic inflammatory changes (Figure 4). The adjacent dura mater had a cluster of blood vessels with elastin stain showing a very well defined internal elastic lamina illustrative of arteries. Other vascular structures revealed a mixture of appearance with larger arteries characterized by absent internal elastic lamina, and regions of focally thickened intima in which embolization material was occasionally absent or present. Elastin stain showed elastic lamina remnants in the embolized vascular structures, with iron stain showing dura mater to contain iron pigment. Recent hemorrhage was also observed alongside siderophages.

Discussion

Dural AVFs in pediatric patients have been classified by Lasjaunias and coworkers to include dural sinus malformations in neonates, infantile dural arteriovenous shunts in children, and adult type of dural arteriovenous fistulas which are less common but still occur in pediatric patients. The most common arterial sources are the middle meningeal artery and the occipital artery, with the most common draining sinus system being the transverse-sigmoid complex. Dural AVFs have been defined as benign or aggressive based on the direction of cortical venous flow, with reversal of flow being suggestive of a more aggressive phenotype with higher likelihood of intracranial hemorrhage and nonhemorrhagic neurological deficit. Patients with benign dural AVFs present based on location either asymptomatic or with pulsatile bruit, orbital congestion, cranial nerve palsy, or chronic headache. Patients with aggressive dural AVFs may also be asymptomatic or present with intracranial hemorrhage, nonhemorrhagic neurologic ischemic deficit, dementia, papilledema, or death. In more recent reports, the aggressive dural AVF group has been subdivided by symptomatology at presentation with symptomatic cortical

Figure 3: (A) Right temporal bone fracture (asterisk) after circumferential craniectomy and partial dural cauterization. Dural middle meningeal branches are exposed anterior, superior, and inferior to the craniectomy and contain the underlying adherent dural arteriovenous fistula. A dural vein leads to the sigmoid sinus, posteriorly and inferiorly situated relative to the craniectomized bone. (B) Specimen of the craniectomized bone and dura after circumferential bipolar cauterization and excision of the vascular dura. Endovascular coils protrude through inner dura.

Figure 4: Low-power photomicrograph with hematoxylin-eosin stain demonstrating a fibrous gap in the bone at the previous fracture site. One adjacent vessel demonstrates chronic mural inflammation (H+E, 10X magnification).
venous drainage (CVD) portending a ten-fold higher event rate than asymptomatic counterparts. Other angiographic features suggestive of aggressive course with intracranial hemorrhage include aneurysmal dilatation of the draining vein or a venous varix.

Morita and coworkers report a series of 81 pediatric patients with dural AVFs, three of whom had involvement of posterior dural sinuses. Those lesions involving the posterior dural sinus were proposed to be more frequently acquired; and compared with adult lesions that have female preponderance, the neonatal and infantile onset lesions are more frequently reported to occur in males. The common modes of presentation included heart failure, hydrocephalus, macrocrania, dilated scalp veins, and cranial bruits; venous hypertension is rare with the involved sinus often forming a dilated sac, by way of contrast with adult patients in whom the sinus becomes stenosed or occluded in the adult variety. The presence of venous hypertension predisposes to hemorrhage, non hemorrhagic neurological deficit, and death, and is associated with reversal of flow in veins draining normal brain causing venous ischemia. The patient in this report presented with a benign dural AVF, symptomatic from an intolerable pulsatile tinnitus. This tinnitus may have resulted from the fistula proximity to the petrous temporal bone with its contained auditory apparatus.

Acquired dural AVFs in both adult and pediatric patients have been theorized to result from partial arterial injury following traumatic insult. Kitahara and coworkers described the need for limited arterial damage and small volume of extravasated blood, although such conclusions are often skewed because patients with more extensive hemorrhage may be more likely to undergo surgical evacuation with secure control of the damaged artery. This patient presented with a head injury including an epidural hematoma with tempo-parietal fracture, with a mechanism likely to involve tearing of the middle meningeal artery parent or branch. Such vessels injured in close proximity to a meningeal vein or dural venous sinus can complete formation of a dural AVF. Conversely, the middle meningeal artery is anatomically accompanied by laterally situated paired veins, and sometimes these veins may form an intradural venous sinus with the middle meningeal artery situated within this sinus. Establishment of CVD transforms the dural AVF into a more aggressive lesion and may be initiated by development of venous sinus thrombosis. This promotes the growth of dural arteries and subsequent communication with the patent portion of the sinus. The dura mater has a rich network of arterial anastomoses accounting for why AVFs supplied solely by external meningeal branches can quickly recruit blood supply following embolization. Indeed, in this case, sequential angiography after attempted embolization revealed persistence of the dural AVF with newly-onset meningeal vessel hypertrophy and collateral vessel recruitment.

Common treatment modalities for dural AVFs include observation, endovascular intervention, surgery, and radiosurgery. Kincaid and coworkers report on seven cases of pediatric dural AVFs, all likely developing prenatally, of whom six were managed by endovascular therapy alone and one required secondary surgical intervention. The variety of agents available to embolize dural AVFs via transarterial route includes GDCs, fibered platinum microcoils, polyvinyl alcohol particles, and liquid adhesive agents. Onyx liquid embolic system is a non-adhesive agent with the advantage of longer setting time over which injection can be performed, providing the potential for arterio-arterial retrograde feeder filling and thus accomplishing multiple feeder embolization without the need for multiple catheterizations. Complications of transarterial embolization include ischemic cranial nerve palsies, transcollateral embolization of normal cerebral arteries, and systemic venous embolization. A transarterial approach may be unsuccessful when the lesion is fed by multiple arteries, though a transvenous approach may be more effective in these cases with the best indication for such treatment being when the involved sinus no longer contributes to drainage of normal brain parenchyma. Such technique provides for high rates of occlusion although it requires sacrifice of sinus flow with potential complications including alteration of AVF venous drainage to an aggressive lesion and potential of venous infarction or intracranial hemorrhage of normal cerebral tissue by outflow obstruction.

Close observation may be selected because benign dural AVFs have low potential approximating 2% of developing CVD and becoming symptomatic and aggressive lesions. Conversely, early treatment is justified to prevent the development of this sequela, with other indications in patients with benign lesions including ophthalmological sequelae and intolerable bruits. Surgery may be offered based on failed endovascular therapy as a viable alternative to radiosurgery in order to achieve a rapid therapeutic effect, although careful assessment of the surgical risk must be made based on lesion location, patient medical and neurological status, and vascular configuration. Surgery provides effective treatment of dural AVFs, with craniotomy or craniectomy used to expose the lesion and enable direct obliteration. The optimal lesion for surgery should be an aggressive dural AVF that drains through a single cortical vein. Fistulas at the superior petrosal sinus and petrous ridge or in the anterior fossa related to the anterior inferior aspect of the falx are examples of surgical lesions where the AVF almost always drains into cortical veins through a single venous channel. Often, only obliteration of the draining vein is necessary to treat most of these lesions, although resection of the lesion when possible may be of value to protect against recurrence. Among adults having surgical management of dural AVFs, Kawaguchi and coworkers describe that the presence of sinus occlusion with retrograde cortical vein flow supports that total lesion removal is possible, and the utilization of preoperative coil embolization did not affect the volume of procedural blood loss. Ushikoshi and coworkers advise direct sinus packing for lesions draining directly into the dural sinus and for surgical interruption of draining veins when the lesions drain first into cortical veins. Surgery can also be considered with staged or intraoperative endovascular procedures.

Gamma knife radiosurgery has been used to treat dural AVFs when immediate devascularization is not required. A delay on the temporal order of years is required to achieve vascular occlusion, thereby minimizing the role of this modality as primary therapy for lesions with CVD. Radiosurgery has been used in the treatment of benign dural AVFs, particularly for patients with subjectively intolerable bruit, or for aggressive dural AVFs that are not amenable to endovascular treatment and surgery. More recently, stereotactic radiosurgery has been combined with transarterial embolization to treat low-risk dural
AVFs, with embolization providing immediate symptom relief and radiosurgery engendering a higher likelihood of permanent fistula obliteration.31-33

In conclusion, dural AVFs are a rare lesion in the pediatric population and are most often acquired prenatally after maturation of the cardiovascular system. While the vascular configuration in the pediatric age group can be more complex than in acquired adult lesions, endovascular treatment remains the mainstay of therapy with success in both cardiovascular and neurological relief. The presented case was of a benign lesion with no cortical venous reflux and symptomatic from pulsatile tinnitus, but endovascular therapy failed to provide for lesion obliteration and surgical excision was ultimately required. The natural history and management of traumatic pediatric AVFs remains unclear and patients are best managed aggressively as in adults to prevent longstanding venous hypertension and irreversible brain injury.

REFERENCES