Case Report

Rare occurrence of dural arteriovenous fistula in a child: Multi-modality imaging and literature review ☆

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ABSTRACT

A dural arteriovenous fistula (AVF) is a rare condition in a child and is not evident clinically. It is a type of an acquired cerebral vascular malformation that usually occurs after a thrombotic event of the cerebral venous sinuses. Dural AVF is not suspected clinically and is revealed through imaging done for evaluation of cranial symptoms. Therefore, it is essential to revisit the pathophysiology and the clinical situations leading to intracranial dural AVF. Equally crucial is identifying the imaging findings on computed tomography, or magnetic resonance imaging brain scans done as a preliminary work-up in these patients. However, for optimal management decision and prognostication of dural AVF, a digital subtraction angiography is essential. As the entire burden of establishing the diagnosis rests on the radiologists, we would like to present this rare case report highlighting both the clinical and imaging aspects and the management options available for dural AVFs.

Introduction

Intracranial dural arteriovenous fistula (dAVF) is a type of cerebral vascular malformation with shunt formation between meningeal arteries and venules in the wall of dural venous sinus formed as a result of venous sinus thrombosis. According to the literature, dural AVFs usually present in adulthood.

Presentation in childhood is rare. We report a case of a 7-year-old boy who presented with a seizure, a history of occipital pain, and a history of an asymmetrical increase in head size. Magnetic resonance imaging (MRI) brain and selective digital subtraction angiography (DSA) of bilateral Internal Carotid Artery (ICA), External Carotid Artery (ECA), and vertebral arteries were suggestive of left transverse sinus/sigmoid sinus dural AVF. The current computed tomography (CT) angiography showed worsening of Cognard staging. We discuss the imag-
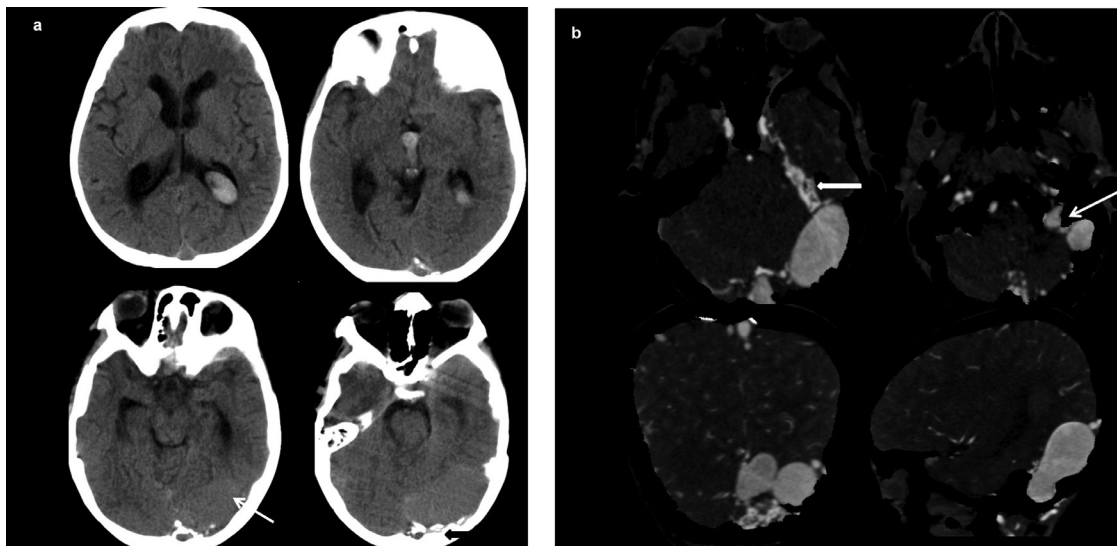
Fig. 1 – Contrast-enhanced CT scan head of the patient at 7 years of age. (a) Axial NCCT head images show a lobulated hyperdense (68 HU) extra-axial lesion along the left occipital convexity (white arrow) with few calcific densities within (black arrow) and scalloping of the inner table of overlying calvarium. Intraventricular hemorrhage was noted. (b) Axial, coronal, and sagittal CT angiography images show intense contrast enhancement of the lesion which measured 2.5 x 4.2 x 6.5 cm. A thin curvilinear area with calcifications on NCCT did not show enhancement s/o chronic partial venous sinus thrombosis. The left internal jugular vein was dilated (thin arrow). Feeders were seen along the left tentorium cerebelli (thick arrow). There was atrophy of the left occipital and left cerebellar hemisphere.

ing features of intracranial dural AVF in detail through this case report with a review of the literature to revisit its pathophysiology, clinical features, and management.

Case report

A 7-year-old boy presented to the emergency with complaints of seizure and loss of consciousness. His vitals were stable. On systemic examination, cardiomegaly was noted. Given the seizure, the NCCT head was done, which showed a hyperdense extra-axial lesion in the posterior fossa in the transverse sinus/sigmoid sinus region. Few calcific densities were also noted within (Fig. 1a).

CT angiography brain was performed subsequently. CT scan showed that the lesion was vascular and showing continuation with sagittal sinus at the confluence of sinuses (Fig. 1b). Multiple enlarged tortuous vessels arising from left ECA, ICA, and vertebral arteries were noted draining into the enlarged vascular structure. Left ECA was diffusely enlarged. Multiple enlarged cortical veins and subependymal veins were noted in both cerebral hemispheres. Venous dilatations were noted at various sites in these veins.

On inquiry, the patient's mother revealed a history consistent with perinatal hypoxia. It was a full-term normal vaginal delivery at home, and the baby did not cry for 5-7 minutes after delivery. However, the postnatal period and early development were normal according to the parents. The patient had a history of left-sided occipital and neck pain and asymmetrical enlargement of head circumference 4 years back, for which CE-MRI brain was done at the time (Fig. 2a, b). Findings

were suggestive of dural AVF of the left transverse/sigmoid sinus.

Selective angiography of bilateral ICA, ECA, and vertebral arteries was performed under general anesthesia post-MRI brain. Enlarged dural arteries were seen supplying the malformation (Fig. 3). The patient withstood the procedure well. Endovascular embolization therapy was advised but refused by the parents due to personal reasons.

In the present emergency admission, the patient was managed with IV fluids, phenytoin, and other supportive care. Intravenous phenytoin was administered slowly at a loading dose of 20 mg/kg body weight over 20 minutes, followed by a 5 mg/kg/day maintenance dose. The hospital stay remained uneventful. The patient was shifted to the ward the next day and discharged after 3 days in stable condition. The patient was advised Tab Acetazolamide (Diamox) 1 tab orally once daily for 7 days and to follow up in OPD after 1 week to review further management.

Discussion

Intracranial dAVF is a type of cerebral vascular malformation with shunt formation between meningeal arteries and venules in the dural venous sinus wall commonly formed as a result of venous sinus thrombosis. They account for 10%-15% of all intracranial vascular malformations [1]. Most dAVFs are found in the posterior fossa, with transverse/sigmoid sinus junction as the most common site. Caroticocavernous fistula is also a type of dural AVF in which fistulous communication is
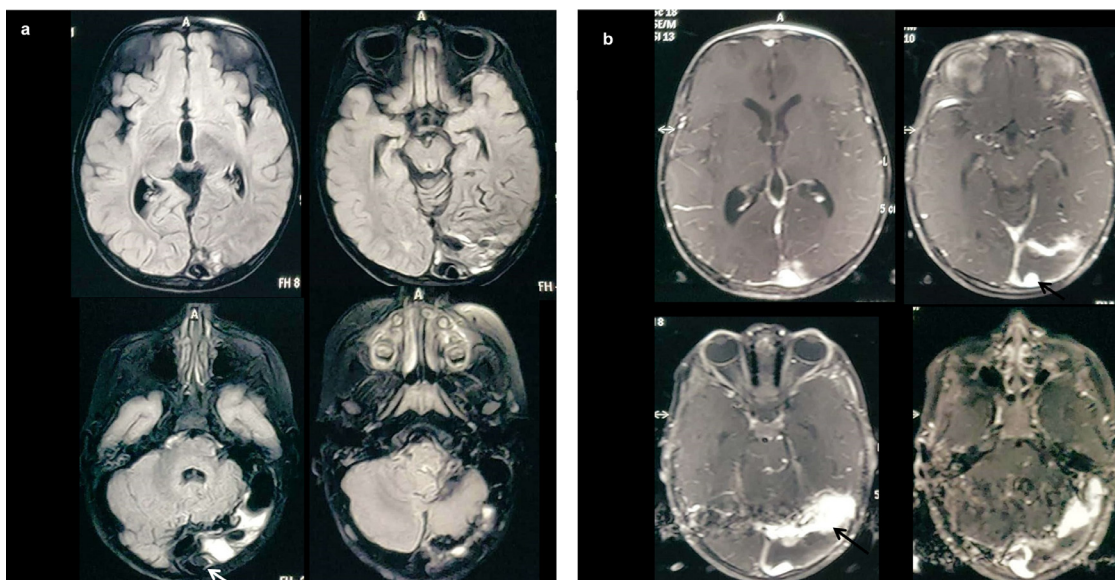
Fig. 2 – Contrast-enhanced MRI brain of the patient at 3 years of age. (a) Axial FLAIR images show areas of abnormal signal intensities (white arrow) within dilated left transverse sinus, sigmoid sinus, and superior sagittal sinus. (b) On postcontrast images, partial patency of left transverse and sigmoid sinus is seen (black arrow).

Fig. 3 – Selective DSA of bilateral ECA, ICA, and vertebral arteries of the patient performed at 3 years of age. Images show feeder vessels to the dural AVF from left ECA, vertebral artery, and ICA. The feeders were posterior meningeal, ascending pharyngeal, middle meningeal, left meningohypophyseal, and recurrent meningeal artery, and left Anterior Inferior Cerebellar Artery (AICA) branch of the basilar artery.

noted between ICA (commonly) or ECA or their branches and cavernous sinus.

Most dAVFs are found in adults with a peak age being 40-60 years. They are rarely seen in the pediatric age group, accounting for approximately 10% of all intracranial arteriovenous shunts in children, lower than the 10% to 20% estimated for adults [2]. Only a few case reports of intracranial dAVF in the pediatric population exist in the literature. The 2 largest reported series in pediatric patients are 29 cases by Lasjaunias et al [3] and 11 cases by Garcia-Monaco et al [2]. Unlike congenital intracranial arteriovenous malformations (AVMs), dAVFs are usually acquired [1].

Two significant hypotheses explain the pathogenesis of dAVFs. One is that the origin of dAVFs is from "dormant" channels between the external carotid circulation and the venous pathways within the dura mater, which open in response to venous hypertension. The second suggests that increased intra-sinus and tissue hypoxia resulting from sinus thrombo-

sis causes stimulation of angiogenic factors and the development of new vascular channels [4].

More than 40% of childhood cerebral venous sinus thrombosis occurs within the neonatal period, with an incidence of 2.6 per 100,000 children per year reported in one series [5]. The incidence of childhood cerebral venous thrombosis varies between 0.4 and 0.7 per 100,000 children per year [6,7]. This is probably an underestimation of the true incidence for several reasons. Often diagnosis may not be suspected since children with cerebral venous sinus thrombosis, notably neonates, commonly present with nonfocal neurologic signs, and symptoms. The lack of evidence supporting anticoagulation treatment may also be a contributing factor.

There are several conditions associated with neonatal cerebral venous thromboses such as meconium aspiration, Apgar ≤ 7 at 5 minutes, intubation at birth, neonatal infections, chorioamnionitis, polycythemia, severe dehydration, pneumonia, congenital heart disease, disseminated intravas-

cular coagulation, congenital diaphragmatic hernia, and maternal conditions like diabetes, hypertension [8]. In our case, sinus thrombosis was the cause of dAVF. Perinatal hypoxia (delayed onset of cry at 5-7 minutes) could have been the cause of neonatal cerebral venous sinus thrombosis, which was not detected.

Clinical presentation and prognosis vary with location and venous drainage. Uncomplicated dAVFs may present with bruit, tinnitus, or headache, whereas advanced disease with cortical venous reflux may present with seizures, focal neurological deficits, and intracranial hemorrhage due to venous hypertension. Among the pediatric age group, the most common presentation in infants is congestive heart failure, while that in children above 1 year of age is neurological deficits [9]. In our case, the patient complained of headaches and enlarged head size at the age of 3 years and seizure episodes later on. Cardiomegaly was also noted.

One of the most widely used classification systems for prognostication of dural AVFs is Cognard classification [10] based on the correlation of clinical findings with various angiographic features. Dural AVF is divided into 5 types (type I to type V) depending on the location of the fistula, presence of cortical venous drainage, the direction of blood flow, and presence of venous ectasia.

The natural history of dural AVF is variable and poorly understood. Progression of a dAVF from low to high grade does occur as in our case but is relatively uncommon [1]. The patient's recent CT angiography report is suggestive of Cognard Grade 4 (Fig. 1a, b). Intraventricular hemorrhage was also noted. On initial imaging 4 years back, it was Cognard grade 2A (Fig. 2a, b). No intracranial hemorrhage was seen then.

The natural course of untreated cerebral dural arteriovenous fistulas depends upon the grade of the fistula. The low-risk dural AVFs (Type I), that is, those draining antegradely into venous sinus with no drainage or reflux into the cortical veins, have a benign course, and some may spontaneously thrombose [11]. On the contrary, high-risk dural AVFs (type III to type V) with leptomeningeal/cortical venous drainage have a significant risk of hemorrhage and nonhemorrhagic neurological deficit [11], as was seen in our case.

Findings on both CT and MRI are variable from little or none to striking. The most common finding is thrombosis of the dural venous sinus with adjacent dilated tortuous vessels and no intervening nidus in the brain parenchyma.

On CT, these may present as iso- or hyperdense cortical tubular structures in the region of the involved dural venous sinus with striking enhancement on contrast-enhanced CT/CT angiography. Subcortical curvilinear calcifications may be seen in cases of chronic cortical venous reflux. Areas of adjacent hypodensity may be seen as a result of chronic venous congestion or infarction [12].

The advantage of CT angiography is rapid acquisition and diagnosis, which is of particular importance in uncooperative or restless patients. Bone removal techniques are available, which provide better image quality. However, these bone removal algorithms have some drawbacks: complexity of use, operator dependence, high dose of radiation, difficulties in the differentiation of arterial loops or infundibula from the aneurysm, and the obscuring of an aneurysm in the cavernous internal carotid artery [12]. Also, since dural AVFs are located adjacent to skull bones, it may be challenging to produce a good quality image sufficient to provide detailed information regarding dAVF lesions' anatomy.

MR imaging features depend on the disease stage and include dilated tortuous flow voids in the cortical region with prominent vascular enhancement. Dural sinus thrombosis may be seen as abnormal signal intensity within the sinuses and lack of normal flow void. White matter T2/FLAIR hyperintensities may be seen in late stages and are suggestive of chronic venous ischemia. MRI has the advantage of better soft tissue resolution with no interference from an adjacent skull bone. Phase-contrast MRA may detect the direction of flow. However, it cannot be performed in uncooperative patients or those with pacemakers [13].

DSA is the "gold standard" imaging modality for assessing sources of blood supply and pattern of venous drainage of dAVFs, particularly the presence of reflux into the cortical veins. However, DSA has a neurologic complication rate of 0.07%-1.3%. Super-selective catheterization of bilateral ECA, ICA, and vertebral arteries is done to complete the assessment. Early filling of venous sinuses is noted supplied by dural branches of these vessels.

The risk of intracranial hemorrhage rises significantly with cortical venous reflux and tortuous and dilated pial veins.

The goal of the treatment of dAVFs is to prevent the occurrence of intracranial hemorrhage or neurological deficits. The aim is to occlude the fistulous communication and arterial feeders. The diseased venous sinus may have to be sacrificed if it does not contribute to the brain's venous drainage. The treatment strategies currently available are – endovascular embolization, surgery, stereotactic radiosurgery, or a combination of these. Endovascular embolization therapy is the mainstay of treatment. In contrast, surgical resection of involved dural sinus wall or stereotactic radiosurgery is reserved for particular cases [9]. Patients with minimal or no symptoms and dAVFs with no cortical venous reflux (Cognard Grade 1 and 2A) on angiography can be followed up and managed conservatively, for example, compression therapy of ipsilateral carotid and occipital artery from contralateral hand 3 times a day [14].

A complete understanding of the fistula's anatomy and its arterial and venous components is essential before endovascular therapy because inappropriate embolization may result in abnormal flow dynamics and a further increase in cortical venous reflux.

The approach can either be transarterial or transvenous. Previously used embolization materials like detachable balloons, polyvinyl alcohol, silk sutures, and microspheres have been widely replaced by current embolic agents, including n-butyl-2-cyanoacrylate (glue), Onyx, Squid, precipitating hydrophobic injectable liquid, detachable microcoils, and flow diverters, such as the pipeline embolization device to treat AVFs and inhibit fistula recanalization [14].

A transarterial approach is a preferred treatment for high-grade dAVFs with direct cortical venous drainage (Cognard grade III-V) or cases where a transvenous approach is not possible. Advantages of transarterial embolization include decreased chance of flow redirection into an alternate venous pathway, ability to save functional venous system, avoidance of post-treatment de novo dAVF formation from venous

hypertension, and decreased complications associated with transvenous approaches, for example, abducens nerve palsy from catheterization of the superior petrosal sinus [14] The microcatheter must be placed as close as possible to the fistula. Onyx has shown higher cure rates and less operator dependency as compared to glue [14]. A significant complication of the transarterial approach is the retrograde filling of glue/Onyx with proximal vascular occlusion and infarction.

A transvenous approach is preferred when transarterial access is dangerous. This could be due to the following reasons: small tortuous arterial feeders, arterial supply of dAVF by branches directly from the ICA or vertebral artery, or when the feeder arteries are also nutrient arteries of cranial nerves. Therefore, the transvenous approach is the first line of treatment of an indirect CCF and Type I and II dAVFs of the hypoglossal canal [14]. The major complication of transvenous approach is venous rupture, hemorrhage, and cerebral infarction. Moreover, following transvenous embolization, cerebral venous drainage may be altered and result in intracranial hypertension [14].

Surgery is usually reserved for cases in which endovascular approaches have failed to cure the lesion completely. However, surgery is preferred in a few locations, such as the anterior cranial fossa and ethmoidal dAVF. Infection, hydrocephalus, hemorrhage, cerebrospinal fluid leak, stroke, and cranial nerve palsy are major surgery complications. Preoperative embolization can be helpful to reduce surgical blood loss [14].

Stereotactic radiosurgery is the last salvage option for the treatment of dural AVF when endovascular therapy and surgery have failed. Endothelial cell damage and thrombosis are suggested as the main mechanisms of dAVF occlusion by radiation. The mechanism of dural AVH occlusion by radiation is proposed to be endothelial cell damage and thrombosis. This technique's disadvantage is a long latent period of several months before an obliterative effect is noted. Complications include cranial nerve palsy, brain edema, hemorrhage during the latency period, and radiation effect [14].

Though a rare clinical entity, clinical suspicion, correct imaging diagnosis, and assessment help select the patient's appropriate treatment option.

Patient consent

Patient consent has been obtained.

Learning points

Childhood dural AVFs are uncommon; most of them are acquired during the neonatal period due to varied perinatal conditions.

Perinatal hypoxia may result in neonatal cerebral venous thrombosis.

Drainage of dural AVF by cortical venous reflux is a bad prognostic marker with an increased risk of intracranial hemorrhage.

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