Intracranial dural AV fistula- An unfolding enigma

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Abstract
An Intracranial dural arteriovenous fistulas (DAVF), is an abnormal direct connection (fistula) between a meningeal artery and a meningeal vein or a dural venous sinus &/ cortical vein.1 When there are multiple fistulas they are termed as dural arteriovenous malformations and comprise 10-15% of all intracranial arteriovenous malformations1. Here we present a case of 52yr old patient who presented to our casualty with history of sudden onset of headache, giddiness, and vomiting, followed by loss of consciousness. Neurological examination revealed impairment of consciousness, no verbal response, with left hemiparesis (E2V1M5). Bilaterally, pupils were 5mm and sluggishly reacting to light. Vitals were stable with B.P. recording of 140/80 mm of hg, and a pulse rate of 80 /min. Pre contrast computed tomography (CT) scan showed an intraparenchymal haemorrhage with perilesional oedema in the right fronto temporal region.

CT angiography brain showed a intracranial dural AVF with varix and adjacent intraparenchymal haemorrhage.

D.S.A. revealed a Borden type III & Cognard type IV – midline dural Arteriovenous fistula in the region of mid superior sagittal sinus and along the cerebral falx, with a large venous ectasia/varix along the anterior cerebral falx.

Management: Patient underwent a fronto parietal rectangular craniotomy Right posterior frontal parasagittal enorged bunch of focal cortical veins was cauterized. Underlying haematoma was evacuated and the underlying venous ectasia/ varix identified. It was found to be thrombosed. A large draining vein was found which was clipped and any consequent brain swelling was looked for, before finally exciting it in toto.

Post –op period was uneventful, with the patient gradually improving in sensorium with residual hemiplegia and aphasia. Although recent developments in catheter intervention has made it the preferred option in most of the cases, surgery is still preferred in some select cases.

Keywords: Intracranial dural AV fistula, Superior sagittal sinus, Cognard and borden classification, CT angiography, Digital subtraction angiography.

Introduction
An intracranial dural arteriovenous fistulas (DAVF), is an abnormal direct connection (fistula) between a meningeal artery and a meningeal vein or dural venous sinus &/ cortical vein.1 When there are multiple fistulas they are termed as dural arteriovenous malformations and comprise 10-15% of all intracranial arteriovenous malformations.2 Most of them are located in the posterior fossa or in the region of the cavernous sinus. Patients may be asymptomatic or symptoms may range from mild symptoms to fatal haemorrhage, depending on the location and venous drainage pattern, which ultimately dictates management strategy.3 Although recent developments in catheter intervention has made it the preferred option in most of the cases, surgery is still preferred in some select cases.2

Case report
A 52yr old patient presented to our institute with history of sudden onset of headache, giddiness, and vomiting, followed by loss of consciousness. Neurological examination revealed impairment of consciousness, no verbal response, with left hemiparesis (E2V1M5). Bilaterally, pupils were 5mm and sluggishly reacting to light. Vitals were stable with B.P. recording of 140/80 mm of hg, and a pulse rate of 80 /min. Pre contrast computed tomography (CT) scan showed an intraparenchymal haemorrhage with perilesional oedema in the right fronto temporal region, centrum semiovale and corona radiate measuring 5.2x4.1x4.5cms with extension into bilateral lateral ventricles and the 3rd and 4th ventricles. Diffuse subarachnoid hemorrhage noted in the bilateral fronto parietal regions, right sylvian cistern with mild generalized effacement of cortical sulci and basal cisterns suggestive of diffuse cerebral oedema. There is mass effect with midline shift to the left by 3.00mm.

Patient was intubated, anti-edema and all other supportive measures, instituted, and stabilized. CT angiography brain Fig 2 showed a dural avf with varix and adjacent intraparenchymal haemorrhage.

D.S.A Fig. 3 revealed a Borden type III & Cognard type IV –midline dural Arteriovenous fistula in the region of mid superior sagittal sinus and along the cerebral falx, with a large venous ectasia/varix along the anterior cerebral falx. The dural AVF is supplied by arterial feeders from the right middle meningeal artery, transosseous branches of right superficial temporal artery with direct cortical venous drainage. The rest of the dural and cortical venous sinuses are normal.

Due to the concerns of raised I.C.P., due to the large size of the hematoma, easy accessibility, informed choice of the patient relatives, and cost effectiveness, surgery was decided as the treatment of choice. Patient underwent a fronto parietal rectangular craniotomy
extending across the midline, exposing the superior sagittal sinus in the region of the dural avf, via a bicoronal skin incision behind the coronal suture.

Dura flap raised with a medial base towards the sinus, essentially cutting off the arterial meningeal feeders. Right posterior frontal, parasagittal engorged bunch of focal cortical veins was cauterized. (Fig. 4)

Underlying haematoma was evacuated and the underlying venous ectasia/varix identified. It was found to be thrombosed. (Fig. 5)

A large draining vein was found which was clipped Fig. 6 and any consequent brain swelling was looked for, before finally excising it in toto.

Duroplasty with fascia and wound closed in layers over a sub galeal drain. Post –op period was uneventful, with the patient gradually improving in sensorium with residual hemiplegia and aphasia.

**Fig. 1:** Showing right post frontal intraparenchymal hemorrhage with extension into lateral, 3rd and 4th ventricle

**Fig 2:** Angio brain showing dural AVF with adjacent varix

**Fig. 3:** DSA showing feeders from middle meningeal and superficial temporal artery with venous

**Fig. 4:** Intraop image of dural vessels and focal engorged cortical veins

**Fig. 5:** Venous varix and hematoma

**Fig. 6:** Clipped venous varix

**Fig. 7:** Postoperative CT brain showing resolution of hematoma

**Discussion**

Dural arteriovenous fistulas are anomalous arteriovenous communications developed within a venous space contained between the two layers of the duramater and typically supplied by multiple feeders from arteries that supply the relevant part of the meninges and regional scalp vessels which give transosseous branches. They account for 10%-15% of all intracranial vascular malformations. supratentorial and lateral: (external carotid artery)

Middle Meningeal Artery
Superficial Temporal Artery (transosseous branches)
Anterior cranial fossa: (internal carotid artery)
Ethmoidal branches: (internal and external carotid arteries)
Meningohypophyseal Trunk and inferolateral trunk
Accessory Meningeal Artery
Posterior cranial fossa: (vertebral and external carotid arteries)
Vertebral Arteries (both dural and muscular branches)
Occipital and ascending pharyngeal arteries

**Location**
- Transverse/sigmoid sinus
  - Most common
  - Least likely to have retrograde venous drainage
  - Cavernous Sinus (indirect carotico cavernous fistula)
  - Superior Sagittal Sinus
  - Straight Sinus
  - Other venous sinuses
  - Anterior Cranial Fossa
  - Typically only ICA supply due to meningeal supply of this region
  - Frequently associated with retrograde venous drainage
  - Tentorium: frequently associated with retrograde venous drainage.

![Fig. 1: Drawings (a = lateral view, b. = anteroposterior view) illustrate the most common location of dural AVFs: 1= cavernous sinus (CS) (20%-40% of cases), 2 = transverse-sigmoid sinus (TS, SS) (20%-60%), 3 = tentorium (12%-14%), 4 = superior sagittal sinus (SSS) (8%), and 5 = anterior fossa (2%-3%), IPS = inferior petrosal sinus, ISS = inferior sagittal sinus, JV = jugular vein, MS = marginal sinus, OS = occipital sinus, SPS = superior petrosal sinus.]

Dural arteriovenous fistulas are usually acquired lesions. Inciting events preceding the development of DAVF have included trauma, cranial surgery, otitis or sinus infection, hormonal changes (e.g., pregnancy, estrogen replacement), and an association with meningiomas.

DAVF are caused by angiogenic factors released from organizing thrombus leading to invasion of small dural arteries and formation of small arteriovenous shunts.

Venous hypertension may also increase angiogenic activity or lead to local tissue hypoxia that initiates neoangiogenesis and endothelial proliferation, leading to Davf formation.

Another theory suggests that DAVFs arise from naturally occurring dormant channels between dural arteries and sinuses, which opens when the sinus is occluded and venous pressure is increased.

In immune histochemical studies, expression of basic fibroblast growth factor (bFGF) and vascular endothelial growth factor (VEGF) has been identified in the wall of the dural sinuses in patients with davfs.

Patient symptoms may be characterized as either nonaggressive (benign) (e.g., tinnitus) or aggressive (e.g., intracranial haemorrhage) Table 1.

**Table 1: Symptoms of Intracranial Dural AVFs**

<table>
<thead>
<tr>
<th>AVFs Symptoms</th>
<th>Cavernous sinuses (%)</th>
<th>Transverse-sigmoid sinuses (%)</th>
<th>Tentorium (%)</th>
<th>Superior sagittal sinuses (%)</th>
<th>Anterior fossa (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ocular symptoms</td>
<td>80-97</td>
<td>-----</td>
<td>-----</td>
<td>----</td>
<td>----</td>
</tr>
<tr>
<td>Cranial nerve deficits</td>
<td>44-77</td>
<td>7-12</td>
<td>14-17</td>
<td>----</td>
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</tr>
<tr>
<td>Bruit, tinnitus</td>
<td>40-50</td>
<td>40-42</td>
<td>70-88</td>
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</tr>
<tr>
<td>Headache</td>
<td>-----</td>
<td>46-76</td>
<td>8-24</td>
<td>50</td>
<td>12-15</td>
</tr>
<tr>
<td>Visual symptoms</td>
<td>28-38</td>
<td>12-28</td>
<td>-----</td>
<td>----</td>
<td>----</td>
</tr>
<tr>
<td>Central nerve deficits</td>
<td>3</td>
<td>10-20</td>
<td>23-42</td>
<td>29</td>
<td>5-33</td>
</tr>
<tr>
<td>Intracranial hemorrhage</td>
<td>Rare</td>
<td>15-28</td>
<td>60-74</td>
<td>23</td>
<td>44-84</td>
</tr>
<tr>
<td>dementia</td>
<td>-----</td>
<td>Rare</td>
<td>-----</td>
<td>5</td>
<td>----</td>
</tr>
</tbody>
</table>

Although there have been many classification schemes suggested for DAVF, the classification proposed by borden and cognard are the most widely used.

The Borden classification system categorizes DAVF based on the site of venous drainage (dural sinus versus cortical vein) and the absence or the presence of cortical venous drainage.

The cognard system not only notes these features but also takes into account the direction of flow in an involved dural sinus (antegrade versus retrograde), as
well as the absence or presence of venous ectasia(s) in recruited cortical veins.\textsuperscript{10}

1. Borden classification
Type 1: venous drainage into a dural sinus with no CVD
Type 2: Venous drainage into a dural sinus with associated CVD
Type 3: Drainage into cortical veins (CVD)

2. Cognard classification
Type I: Venous drainage into dural sinus with antegrade flow
Type IIa: Venous drainage into dural sinus with retrograde flow
Type IIb: Venous drainage into dural sinus with retrograde flow & CVD
Type II (a+b): Venous drainage into dural sinus with antegrade flow and CVD
Type III: Venous drainage into cortical veins (CVD)
Type IV: CVD with associated venous ectasia(s)
Type V: Venous drainage into spinal Perimedullary veins.

Symptoms associated with dural avf can be due to either increased dural sinus drainage or the development of cortical venous hypertension.\textsuperscript{12} Anterior fossa lesions are typically supplied by ethmoidal arteries and drain into the cavernous sinus, these dafvs typically present with ocular symptoms including proptosis, chemosis, ophthalmoplegia, decreased visual acuity, or retro orbital pain because of their proximity to the orbit.

Middle fossa lesions commonly drain into the transvers or sigmoid sinus These fistulas typically cause symptoms of pulsatile tinnitus because of the close proximity to the auditory apparatus.\textsuperscript{12} Fistulas that drain into the superior sagittal sinus or deep venous system produce symptoms of global venous congestion and raised ICP, and may manifest with symptoms of hydrocephalus, papilloedema, seizures, or dementia.

Brainstem dafvs, though less common than other locations, can present with cranial neuropathies and /or quadriparesis.\textsuperscript{12} The presence of cortical venous hypertension in addition, typically causes more severe symptoms, including Intracranial haemorrhage and neurological deficits which include progressive dementia, seizures, parkinsonism, and other focal neurological deficits including aphasia, alexia, weakness, paraesthesias, and ataxia.\textsuperscript{12}

Non contrast computed tomography (CT) and conventional magnetic resonance (MR) imaging often seem unremarkable with benign DAVF. However, these studies may demonstrate the complications of aggressive lesions with CVD, including hemorrhage, venous congestion with edema, venous aneurysms, tortuous cortical veins in a pseudopheblitic pattern, and parenchymal or leptomeningeal enhancement.\textsuperscript{10}

CT and MR angiography (CTA and MRA, respectively) may be used to screen patients suspected with DAVF, can grade or classify these lesions when detected, as well as evaluate for response to treatment.\textsuperscript{10}

Both CTA and MRA may visualize the fistula itself as prominent vessels associated with the meninges or dural sinus wall, as well as detect enlarged feeding arteries, early dural sinus opacification, and prominent draining veins.\textsuperscript{10}

Despite these advances in MR and CT imaging, catheter angiography remains the definitive imaging study and the gold standard for evaluation of DAVF because of its superior spatial and temporal resolution. It can delineate both the arterial supply and the venous drainage of the fistula, as well as identify high-risk features including CVD, venous outflow obstruction, and arterial pedicle or venous aneurysms. Catheter angiography is also excellent for evaluation of any associated dural venous sinus thrombosis or occlusion. Finally it provides accurate information for planning endovascular or surgical treatment.\textsuperscript{10}

**General treatment approaches**

Management of DAVF should be based on patient characteristics, symptom severity, and risk of serious sequelae, the latter being primary determined by the presence or absence of cortical venous reflux.\textsuperscript{10} DAVF without high-risk features may be managed conservatively with an acceptably low rate of serious complications. In these instances, treatment should be tailored to palliation of intolerable symptoms.\textsuperscript{10}

Spontaneous regression of dural AVFs has been reported. However, patients with benign DAVF electing conservative management should undergo clinical and imaging follow-up given the small risk of conversion to an aggressive lesion.\textsuperscript{10}

Endovascular Interventions include Transarterial embolization (TAE) with particles, Transvenous Coil embolization, TAE with n-butyl -2-cyanoacrylate and Stent Placement.\textsuperscript{9}

Stereotactic radiosurgery has the advantage of decreased invasiveness and fewer short-term complications, whereas the disadvantage is the delayed response (approximately 6-12 months) after irradiation.\textsuperscript{9} Results from large centers have been mixed, with cure rates ranging from 50% to 93% for DAVF patients treated with SRS.\textsuperscript{13,17} The majority of studies used a median dose of approximately 20 Gy, with a range based on the location and whether adjunctive treatment with embolization or surgery was employed. In published studies, there is a trend toward a higher rate of success depending on location, with cavernous sinus lesions having the highest rate of successful obliteration.\textsuperscript{17}

Surgical treatment methods for DAVF include surgical excision of involved meningeal arteries and veins, packing of the diseased dural sinus, as well as skeletonization of the involved dural sinus with disconnection of draining leptomeningeal veins.\textsuperscript{10} Risks of surgical repair of DAVF include blood loss,
intracranial hemorrhage, arterial infarct, venous infarct, and cerebrospinal fluid leakage.\textsuperscript{10}

**Conclusion**

Davfs are venous-based rather than arterial–based lesions and treatment should focus on the venous side of the fistula.\textsuperscript{7} Microsurgical repair is a safe and effective treatment method for DAVF, either alone or in combination with endovascular embolization.\textsuperscript{10}

**References**