INTRACRANIAL DURAL ARTERIOVENOUS SHUNTS – DAVS (DAVF`S AND DAVM`S)

Introduction ;

Intracranial dural AVF`s are abnormal connections between the dural (occasionally pial) arteries and the venovascora with in the dura mater, comprising the walls of dural sinuses, the leptomeningeal veins, or the emissary veins within or adjacent to the dura mater.

Dural AVF`s were considered to be extremely rare disorders, but with advancement in vascular imaging techniques, there is a significant rise in diagnosis. At present they account for approximately 10-15% of intracranial vascular malformations. Exact incidence in Indian population is not known.

A majority of DAVF`s an thought to acquired lesions resulting from cerebral venous thrombosis, trauma, infections and at times they are iatrogenic secondary to neurosurgical intervention. In a significant number of patients, the etiology remains unknown.

Natural history of DAVF

Owing to the challenges involved in diagnosing the DAVF`s the natural history is not completely understood. The available data suggests that DAVF`s enlarge overtime in terms of recruitment or enlargement of arterial feeders or appearance of de novo fistulas. The pattern of venous drainage in the most important factor that determines the natural history and symptomatology. The presence of cortical venous drainage either via reflux from the fistulous sinus or fistula being located directly on the vein correlates with the clinical presentation. Based on the venous drainage, patients can present with intracranial hemorrhage, seizures or congestive venous encephalopathy. Other factors which are associated with increased risk of hemorrhage include male gender, old age at presentation , posterior fossa location and focal neurological deficits. Absence of cortical venous reflux is associated much less aggressive clinical course of DAVF`s.

Clinical Presentation :

The clinical presentation depends on the anatomic locations and venous drainage. For example, DAVF on cavernous sinus will cause conjunctival congestion, chemosis, proptosis, ophthalmoplegia and decreased visual acuity.t If idrains into the sigmoid or transverse sinuses, may present with pulsable tinnitus or hearing loss. Anterior cranial fossa DAVF`s have peculiar venous drainage pattern as there is no adjacent dural sinus and hence they can drain via transosseous emissary, there by presenting with epistasis, subdural hemorrhage or intracerebral hemorrhage. DAVF`s involving the skull base can cause cranial neuropathies. Venous drainage into the deep venous system can cause cognitive and personality changes leading to dementia

IMAGING

CT Scan

Non contrast enhanced CT (NCCT) is usually the first line imaging modality for most particular with suspected intracranial pathology, but had limited sensitivity and specificity for making diagnosis of DAVF. It may also show enlarged vascular structures which are likley to be draining veins, cerebral edema, SAH, parenchymal hemorrhages and bony abnormalities like enlarged vascular foramina at the skull base or calvarium indicating high flow vascular lesion.

CT angiography provides much more specific information with diagnostic sensitivity of almost 93% with new generation scanners. CTA give information about the approximate location of the fistula and advanced techniques like 4D CTA may also demonstrate cortical venous reflux. However, CTA is still not considered a definitive diagnostic test for DAVF since its capacity to demonstrate arterio venous shunting and characterize venous drainage is limited.

Magnetic Resonance Imaging (MRI)

MRI in patients with DAVF shows prominent vascular channels representing hypertrophic draining veins. Blooming of the draining veins due to high d deoxyhemoglobin content, cerebral edema due ti venous congestion and parenchymal/subdural/sub arachnoid hemorrhage. Contrast enhanced time resolved MRA has a sensitivity of 93% on diagnosing DAVF.

Digital Subtraction angiography (DSA)

DSA remains the gold standard for the diagnosis and characterization of DAVF. There are some DAVF, specially the ones without any venous reflux/retrograde flow, which can be picked up only on DSA. DSA also helps in planning the therapeutic strategy.

Classification of DAVF`s

Several classification systems have been proposed for example Borden et al proposed a system based on angiographic characteristics of the venous drainage where as Cognard et al presented a classification based on the clinical presentation and angiographic features . There are some other classification systems as well (by Ziptel et al, lasjaunias etc) but most of these systems lead to confusion without much help in planning the therapeutic strategy. Hence it is more practical to divide them in to aggressive (when there is cortical venous reflux and the once which are directly located on the leptomeningeal vein) and the non aggressive (when there is absence of cortical venous reflux).

Management

Considering the complexity and the potential risks involved in the treatment, the management of DAVF`s should be performed at the centers with adequate experience and expertise.

If the patient is asymptomatic and DSA doesn`t demonstrate any cortical venous reflux (non aggressive) conservative management can be considered. But these patients need to be under strict follow up as there is always a chance of transformation into an aggressive variety. However if the DAVF is aggressive, active intervention should be performed. Other indications for active intervention include secondary glaucoma, intractable headache, DAVF`s involving the cavernous sinus and clinically intolerable tinnitus. The goal of intervention is to obtain complete occlusion of the DAVF by completely obliterating the fistulous venous pouch/sinus.

Endovascular Treatment

Endovascular treatment remains the mainstay of treatment of DAVF. Endovascular treatment may be transarterial, transvenous or direct puncture. With the advent of liquid embolics, transarterial approach is usually the first line of treatment .It requires super selective microcatheterisation of the distal aspect of the feeding artery, allowing the embolic agent to pass on to the venous sinus/pouch. In most cases trans arterial approach alone is curative. Transvenous route and direct puncture is usually reserved from DAVF involving the cavernous sinus.

The liquid embolic agents, which are used at present, are ethylene vinyl alcohol copolymer-EVOH (i.e squid/onyx/phil depending on the parent company). EVOH is a non adhesive liquid embolic agent consisting of 2 subunits (ethylene and vinyl alcohol) suspended in an organic solvent, dimenthyl sulfoxide (DMSO). It popularity is mainly because of slow polymerisation allowing a longer duration of injection thereby resulting in higher rates of venous penetration. This increases the cure rates to more than 90% in experienced hands.

Coils (detachable or pushable) are the primary embolic agents for the transvenous endovascular approach for obliterating the DAVF.

Open surgery and Radio surgery :

Open surgical treatment is usually limited to patients presenting with large hematomas that require evacuation and anatomy favorable for surgical obliteration. Open surgery has higher procedure related morbidity with some studies reporting morbidity rates as high as 10%. Stereotactic radio surgery (SRS) can take upto 2 years to have an effort on the fistula closure and hence is not considered the first line treatment option. SRS is combination with endovascular treatment can be considered in selected cases.

Recommendations :

1. All patients with suspected DAVFs based on clinical presentation and/or non –invasive imaging findings should receive complete and high quality DSA in order to confirm and risk stratify their disease. (Class I, level of evidence C).
2. DAVFs with high risk features (eg. CVR) should be treated promptly to reduce the potential risk of intracerebral hemorrhage, venous hypertensive encephalopathy or other neurologic events. Endovascular treatment is considered as the preferred first line treatment is considered option with favorable anatomy. Open surgical treatment alone or combined endovascular and open surgical treatment should be considered for high risk fistulas not curable by endovascular means alone. SRS should be reserved as an adjunctive and/or complementary option for aggressive and symptomatic DAVS (Class I, level of evidence C)
3. Non aggressive but symptomatic DAVFs can be considered for definitive treatment. Endovascular treatment, open surgery and SRS can be considered for this type of DAVFs, but only if associated with very low treatment related risk in view of the benign natural history of these lesions (Class IIb, level of evidence C).
4. Non-aggressive asymptomatic (ie. Incidental) DAVFs lesions without CVR do not warrant active intervention and, if treatment is considered, treatment related risk versus the natural history of the disease should be thoroughly discussed between the practitioner and patient. Nonetheless, these patients should be followed both clinically and with non invasive imaging studies in regular fashion. An exception to this recommendation would be a patient who has become asymptomatic who was previously symptomatic, as a change in symptoms can portend a venous outflow thrombosis and hence, potential change in fistula angioarchitecture and venous drainage pattern that would warrant re-evaluation with DSA (Class I level of evidence C)
5. SRS is a reasonably effective and safe treatment option. Thus, it could be considered as a viable option for DAVFs that have a small compact shunt zone in patients who are not good candidates for endovascular or open surgical treatment or those who prefer a less invasive approach. (Class I level of evidence C)

Conclusions :

Intracranial DAVF`s are not very rare and the management has to be tailored to clinical symptomatology. Dynamic nature of DAVF`s warrants regular clinical and imaging follow up. Endovascular approach remains the mainstay of treatment with the aim being complete obliteration of fistulous sinus/pouch unless it is also being utilized by the brain