Arterio-venous fistulae of the neuraxis: an institutional experience

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Abstract

Introduction: Arteriovenous fistulae (AVF) are rare lesions. Patients typically present with slowly progressive myelopathy that is often mistaken for degenerative cervical or lumbar stenosis. Cranial presentations are even more innocuous ranging from seizures to tinnitus. The purpose of this study is to present a series of cases to aid in the assessment, diagnosis and treatment of this unusual pathology.

Case series: We present 11 cases of AVFs treated at our centre over an 4-year period. Seventy percent of patients were male. The mean age of presentation was 62.6 years. The most common lesion was a dural AVF emanating from the craniovertebral junction with single feeder. All patients underwent either microsurgical correction or endovascular embolization as the primary procedure. Eight patients showed improvement following treatment as graded by the Nurick system. Two patients failed to improve. None of the patients worsened. One patient had a cortical venous thrombosis after embolization that resolved well with anticoagulants.

Conclusion: The successful treatment of AVF requires a detailed understanding of clinical presentation and imaging findings to allow for precise treatment. Owing to the rarity of the condition, clinicians must continue to share their experiences to advance our knowledge.

Keywords: arteriovenous fistulae AVF; embolization; digital subtraction angiography DSA

Introduction

Arteriovenous fistulas (AVF) are the most common vascular shunts of the central nervous system. They occur predominantly in men (80%), commonly involve the thoracolumbar spine, and usually cause progressive myelopathy because of venous congestion of the spinal cord.[1,2] Patients with AVF often present with non-specific clinical features that are related to progressive myelopathy, seizures or isolated neurological deficits. Frequently they are misdiagnosed as having other more common pathologies, which can lead to a significant delay in diagnosis, treatment and poorer prognosis. The average patient will present, with 1–3 years before the diagnosis is made. Hence, understanding the radiological features found on MRI and angiography, and treating it earlier saves the patient significant morbidity. [3,4]

The aim of this study was to compare the angiographic and clinical characteristics of SDAVFs of the neural axis based on the patients treated at a quaternary care centre in south India over 4 years.

Methods:

All patients with dural and epidural AV fistulae within a period of 4 years were considered for the study. Details regarding demography as well as angiography were tabulated. Intervention and or surgery were studied and documented. The data was analysed, consolidated and presented.

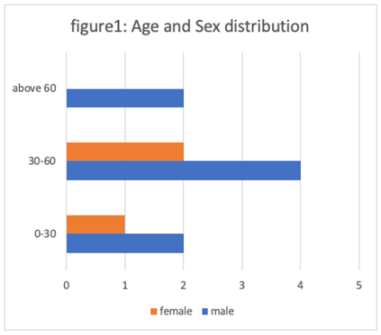
Results

Results are presented in 2 formats. General data regarding the trends in patient demographics and presentations along with modality used to treat the disease have been clubbed together. We also present individual data of each case, where radiological features, and follow up details are explained in brief.

Section 1: General data

11 patients presented with differing clinical features during the above mentioned period in 2 tertiary care centres in Bangalore, India for definitive care. The age and sex distribution are described in figure 1. As is evident from the figure 1 and its table, the maximum individuals were from 30-60 age group which coincides with the reports from literature, with a gross preponderance of males. These patients presented with varying symptomatology. These included features of compressive myelopathy to seizures. The symptomatology has been depicted in figure 2.

Figure1: age and sex distribution of the patient cohort

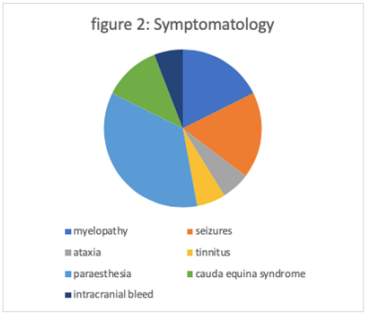


age in yrs.	male	female
0-30	2	1
30-60	4	2
above 60	2	0

Table1: showing demographic data, disease characteristics and treatment methodology

AGE/SEX	LEVEL	PRESENTATION	ANGIO	ENDO/SURG	MRI
52F	D6-8	MYELOPATHY WITH PARAESTHESIA	FEEDERS D8-D10 L>R WITH PROMINENT VEINS SEEN UPTO D6	SURGERY	MRI SHOWED SEVERE CORD OEDEMA FROM D6-D10
60M	CVJ EXTENDING UPTO TRANSVERSE SIGMOID Jn (L)	PARAESTHSIA IN (L) UL, FACE AND BACK RESEMBLING TRIGEMNAL PAIN	FEEDERS SEEN OVER THE TEMPORAL LOBE FROM THE MCA AND PCA	ENDO	
50M	TORCULAR AVF EXTENDING TO CEREBELLAR TONSILS	SEIZURES WITH (R.) LL PAIN AND B/L LL SPASTICITY	LEFT OCC ARTERY & SIGMOID SINUS WITH VARICOSITIES EXTENDING UPTO THE FM	ENDO	
26F	TEMPORO-OCCIPITAL EXTENDING INTO THE FM	ATAXIA WITH SEIZURES	(R.) PCA FEEDERS TO MAXILARY VEIN WITH VENOUS VARYX. CROSS CIRCULATION PRESENT	ENDO	MRI SHOWED MASS LESION WITH FLOW VOIDS EXTENDING UPTO THE RIGHT BASAL GANGLIA
36M	CONUS L1	CAUDA EQUINA SYNDROME	SPINAL AVF AT VENTRAL CONUS FED FROM (R.) L1 RADICULAR BRANCH	SURGERY	OPERATED ONCE IN DHAKA UNSUCCESSFULL
39M	D9	(R.) LL PAIN AND PARAESTHESIA	(L)ASA BEING FED FROM D8 (L.) D9 INTERCOSTAL ARTERY FEEDING AVF	SURGERY	MRI SHOWED CONUS ENHANCEMENT WITH FLOW VOIDS IN THE SUBARACHNOID SPACE
58M	D12	B/L LL SPASTICITY		SURGERY	MRI SHOWED RESIDUAL AVF FROM PROIR EMBOLIZATION
54F	L3	URGENCY WITH WEAKNESS AND PARAESTHESIA	LEFT L2 RADICULAR BRANCH FEEDING THE AVF	SURGERY	
26M	CVJ EXTENDING UPTO TRANSVERSE SIGMOID Jn (L)	(R.) LLPARAESTHESIA WITH TINNITUS	SAMLL DAVF SEEN EXTENDING FROM THE CVJ TO THE TRASNVERSE SINUS	SURGERY	MRI S/O DEMYELINATIVE LESIONS ACROSS THE DEEP NUCLEI
75M	FROM CVJ TO C5	WEAKNESS WITH PARAESTHESIA	LARGE EDAVF SEEN WITH FEEDERS FROM DEEP CERVICAL & VARTEBRAL ARTERIES	ENDO	
22M	RIGHT TEMPORAL BASE	SUDDEN SPONTANEOUS ICH WITH SEVERE CEREBRAL OEDEMA	SMALL FEEDERS FROM THE TEMPORAL DURA DRAINING THROUGH A LARGE BASAL VEIN	SURGERY	

Figure2: symptomatology represented as a pie chart

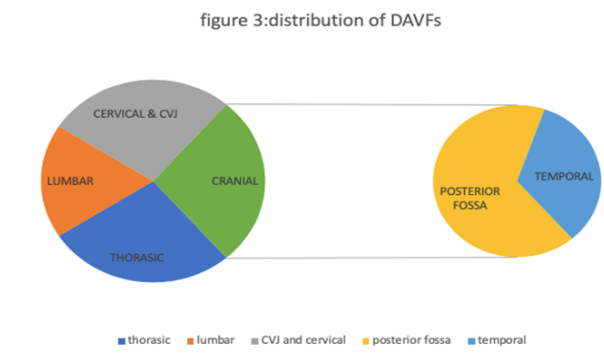


Symptomatology	number
myelopathy	3
seizures	3
ataxia	1
tinnitus	1
paraesthesia	6
cauda equina syndrome	2
intracranial bleed	1

Paraesthesia and compressive symptoms of the spine such as myelopathy, spasticity, limb weakness and bladder disturbances were the most common symptom.

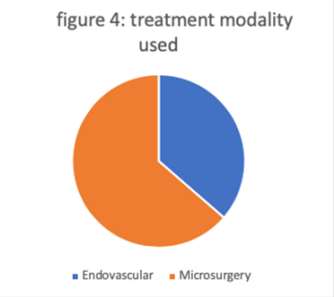
One patient had a sudden severe Intracranial bleed needing emergency decompressive craniectomy and subsequent surgery for his DAVF. All other patients had long standing complaints. The distribution of these lesions is also varied from, intracranial to spine to lesions straddling the craniovertebral junction extending into both spaces, as shown in figure 3. Here the majority of lesions are in the spine with only 3 out of the 11 lesions (27%) occurring intracranially. Of these lesions, 2 were from the posterior fossa around the transverse-sigmoid junction extending to the foramen magnum. The lone temporal DAVF presented with spontaneous intracranial bleed resulting in prostration requiring an emergency decompressive craniectomy.

Figure3: Distribution of lesions by location both intracranial and spinal are demonstrated in a layered pie chart



Distribution	number
Thoracic	3
Lumbar	2
CVJ and cervical	3
posterior fossa	2
temporal	1

Figure4: treatment modality used to treat the different AV fistulae

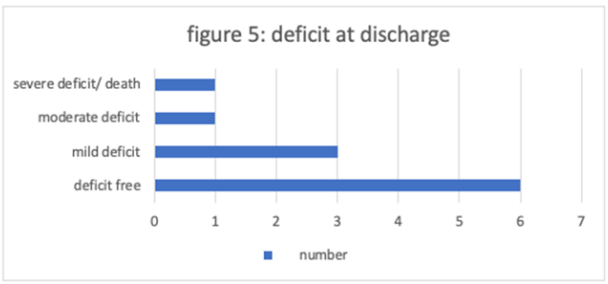


treatment	number
Endovascular	4
Microsurgery	7

Figure 4 shows the primary treatment modality employed in these lesions. There are occasions where Endovascular embolization of the lesion was employed before surgery, but this depiction is only for the main or primary modality involved. Intricacies of individual treatment strategies will be dealt with on a case to case basis later in the paper. Somewhat surprisingly, endovascular therapies are a minority in our data, with a preference for microsurgical resection, especially in those lesions presenting in the dorsal and lumbar spines. Cranial and cervical lesions were mostly treated endovascularly due to the sensitive nature of the location coupled with difficulties associated with approach and feeder detection.

The overall prognosis (figure5) was excellent with only 1 patient out of 11 (0.18%) had long tern deficits, which required nursing care, and this was due to the intracranial bleed which had occurred, for which only slow rehabilitation could be done. The patient however made a complete recovery 1 year later. The depiction below shows deficit at discharge. Another 2 patients were discharged with bladder incontinence (which was the presenting complaint) The condition sadly did not improve later on, even after 1 year. The precarious nature of bladder fibers in the conus however cannot be blamed on the intervention, but nevertheless, we present the data as it is. All others were discharged with mild weakness of limbs which improved on physiotherapy to normal functional status within 6 months of the procedure.

Figure5: deficits of patients at discharge

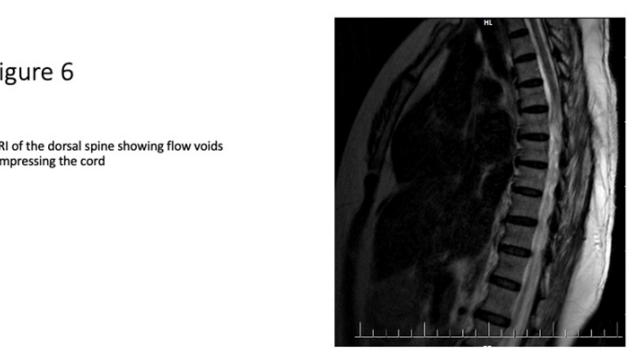


discharge status	number
deficit free	6
mild deficit	3
moderate deficit	1
severe deficit/ death	1

Section 2: Case Reports

Case1: A 52 year old lady presented with paresthesia and spasticity. MRI showed severe cord changes from D6 to D10. The angiogram showed feeders from the radicular arteries of D6 to D10 with prominent veins seen upto D6. Surgery was done and the vein clipped. The patient recovered well and was discharged deficit free. (figure 6)

Figure6: MRI of the dorsal spine showing flow voids compressing the cord



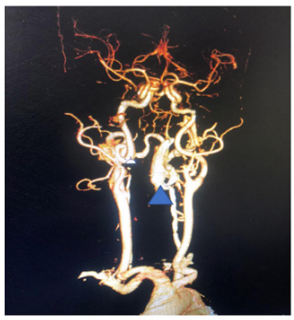
Case2: A 60 year old male patient presented with trigeminal neuralgia like pain across the left side of the face. An MRI showed flow voids extending from the left transverse sigmoid junction upto

the Cervical spine. The angiogram showed feeders from the MCA and PCA emptying into the left transverse sinus. Endovascular embolization was done successfully. (figure 7)

Figure7: CT angiogram with 3D reconstruction showing the AVF venous sac compressing the CVJ

Figure 7

This shows the CT angiogram with 3D reconstruction of the sequences. The arrow shows the large AVF which would have considerably compressed the cord.

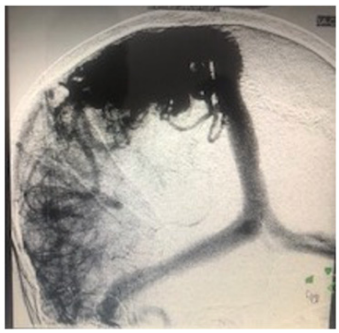


Case 3: A 50 year old male presented with new onset seizures. An MRI suggested enlargement of the torcula with extension into the cerebellar tonsils. An angiogram revealed a large DAVF fed from feeders from the left occipital artery and draining into the superior sagittal sinus with varicosities extending upto the foramen magnum. Endovascular embolization was done successfully using the venous route to approach the lesion. Post procedure there was a minor venous thrombosis in the sigmoid sinus, but since this side was non-dominant, there were no major symptoms. The patient was discharged on antiepileptics and anticoagulants which he continues to take. He remains seizure free till date. (figure 8)

Figure8: Large AVF draining into the sagittal sinus

Figure 8

Large vascular malformation draining into the sagittal sinus .

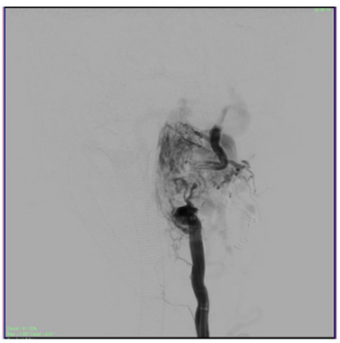


Case 4: A 26 year old female presented with seizures and episodic ataxia. An MRI of her brain revealed a mass lesion with significant flow voids in the right basal ganglia. An angiogram showed a large venous varyx fed from feeders from the Right PCA and draining into the right maxillary vein. Endovascular embolization was successfully done. The patient improved well after the procedure. (figure 9)

Figure9: Partial obliteration of the AVF showing stasis in the draining vein

Figure 9

Partial obliteration of the AVF after onyx injection of the feeders, showing stasis in the draining veins.



Case 5: A 36-year-old male presented with long standing incontinence and loss of sensation over the legs. He had been diagnosed to have a DAVF 2 years ago and had been operated for it unsuccessfully. An Angiogram revealed a ventral Conus DAVF fed from the L1 left radical branch. He was taken up for a redo surgery and improved well post op.

Case 6: A 39 year old male presented with spasticity and paresthesia in both lower limbs. His MRI showed conus enhancement with flow voids in the subarachnoid space. An angiogram showed radicular branches from the D9 level feeding a large left sided DAVF compressing the cord. A large venous channel was seen. Hence surgery was attempted, and the vein clipped. Post op recovery was good.

Case 7: A 58 year old male presented with significant ash worth grade 3 spasticity in both lower limbs coupled with radicular pain and paresthesia. He had been treated with embolization for the same problem 4 years ago. An MRI of the spine suggested a residual DAVF at the conus level. The angiogram of the spine confirmed this with feeders from the L1 and D12 radicular branches. Surgery was done and the venous channel blocked completely. He made a complete recovery. No recurrence has been noted since for the last 1 year.

Case 8: A 54 year old lady presented with progressive urgency and incontinence. She also had longstanding paresthesia of the lower limbs and mild weakness. An MRI of the lumbar spine showed a space occupying mass posterior to the conus and cauda equina compressing the structures in the spinal canal. An Angiogram revealed a DAVF fed from the L2 radical artery on the right side. The vessel was embolized and the patient made a good recovery as far as her pain and weakness were concerned. Her bladder symptoms marginally improved but remain after 1 year of follow up.

Case 9: A 26 year old male patient presented with pulsatile tinnitus and parenthesis of the

He right upper and lower limb. MRI done was suggestive of a space occupying lesion around the transverse sigmoid region extending onto the CVJ. An angiogram showed a small DAVF fed from branches of the PCA draining directly into the transverse sinus. As the DAVF was superficial, a small temporo-occipital craniotomy was done and the venous connection clipped, with excision of the nidus. The patient made a complete recovery and went home deficit free.

Case 10: A 75 year old male patient presented with weakness of the right upper and lower limb for 2 weeks. He had had these symptoms 1 year before, but the symptoms gradually improved, and no medical attention was sought then. This time, the weakness was progressive with spasticity, paresthesia and pain. An Angiogram showed a large epidural fistula with feeders from the vertebral arteries and the deep cervical arteries feeding a large venous lake in the epidural space compressing the cervical cord from C1 to C6. Endovascular glue injection was done immediately and successfully. The patient walked home after 2 days in hospital with no further discomfort. His spasticity and weakness gradually improved over the next 2 months to completely recover. (figure 10a & b)

Figure10a: showing MRI sagittal and axial views depicting the large venous sac compressing the CVJ leading to myelopathy

Figure 10a

MRI T2W images of the cervical spine showing the sagittal section of the compression of the cervical spinal cord secondary to large flow voids present inside and outside the dura.

The Axial image shows a large vessel compressing and displacing the cervical spinal cord with cord changes

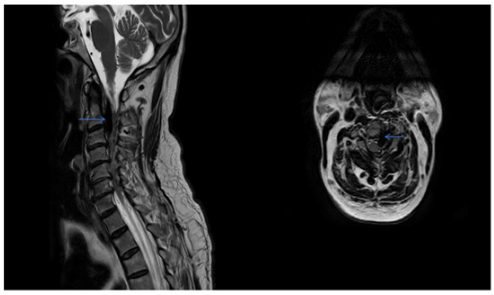
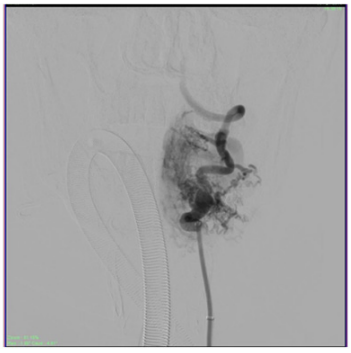


Figure10b: Embolization of the fistula through the vertebral artery.

Figure 10b

Microcatheter placed at the mouth of the feeder arising from the V2 segment of the ipsilateral vertebral artery



Case 11: A 22 year old male patient was brought in an air ambulance from a nearby city. The patient had had a sudden onset of loss of consciousness and prostration. He was taken to the local hospital, where a CT scan had shown a large right temporal lobe ICH with significant mass effect and impending uncal herniation. Due to the severity of the disease and the urgency of the situation, he was taken up for a decompressive craniectomy and hematoma evacuation. During the surgery, engorged venous channels were seen in the dura. The Neurosurgeon at this juncture, decided against hematoma evacuation and stopped after opening the dura decreasing the oedema and preventing further herniation. Post-surgery, the patient was airlifted to our centre.

A CT angiogram of the brain suggested a large Dural Arteriovenous fistula extending from the temporal base to the transverse sigmoid junction. The venous drainage was primarily through a large vein in the temporal base into the maxillary vein. A large number of small dural and pial feeders fed a large nidus occupying the centre of the hematoma. The patient was operated and the hematoma with the nidus and feeders were removed. The vein was clipped and cut thereby disconnecting the system. Post op, the patient spent 17 days in the ICU and gradually improved. He was sent home conscious and obeying commands, needing nursing care and physiotherapy. He rapidly improved and made a full recovery as seen during regular follow up. At the 1 year follow up, he was seen walking with no weakness and cognitive deficits.

Discussion

DAVFs are predominantly idiopathic, though a small percentage of patients have a history of previous craniotomy, trauma, or dural sinus thrombosis. [4-7] Two etiologic hypotheses based around sinus thrombosis have been put forward.

The first is that physiologic arteriovenous shunts between meningeal arteries and dural venous sinuses enlarge in response to elevated local venous pressure, resulting in a pathologic shunt. [4,5,8] The second is that venous hypertension due to outflow obstruction causes decreased cerebral perfusion and promotes neoangiogenesis.[4-9] Heritable risk factors for venous thrombosis, such as antithrombin, protein C, and protein S deficiencies, have furthermore been associated with DAVF occurrence. [10,11] These findings implicate the role of an underlying hypercoagulability in the development of DAVFs. The aetiology of paediatric DAVFs is thought to be congenital or a result of birth trauma, infection, in utero venous thrombosis, or maternal hormones.[3]

Classification

The Borden classification system stratifies lesions on the basis of the site of venous drainage and the presence or absence of venous drainage. Borden type I lesions have the direct communication of meningeal arteries with a meningeal vein or dural venous sinus and exhibit normal antegrade flow. Type II lesions have shunts between the meningeal arteries and dural sinus, with retrograde flow into the subarachnoid veins, causing venous hypertension. Type III lesions have direct drainage of meningeal arteries into subarachnoid veins or an “isolated” sinus segment. The latter phenomenon is the result of thrombosis on either side of the arterialized sinus segment, which directs retrograde flow into the subarachnoid venous system. The Borden classification scheme further subclassifies lesions as single-hole (a) or multiple-hole (b) fistulas. (table2) [12]

Table 2: Borden’s classification of Intracranial Dural Arteriovenous Fistulae

Type	Definition
1	Drains directly into major venous sinus or meningeal vein
2	Drains into sinus but high pressure within the sinus results in retrograde drainage via subarachnoid veins
3	Drains directly into subarachnoid veins

The Cognard classification [13] is based on the direction of dural sinus drainage, the presence or absence of CVD, and venous outflow architecture (nonectatic cortical veins, ectasia cortical veins, or spinal perimedullary veins). Type I lesions drain into the dural sinus, have an antegrade flow direction, and lack CVD. Type II lesions are subdivided in 3 subcategories: type IIa lesions drain retrogradely into a dural sinus with- out CVD, type IIb lesions drain antegradely into a dural sinus with CVD, and type IIa + b lesions drain retrogradely into a dural sinus with CVD. Types III, IV, and V lesions all have CVD, absent dural venous drainage, and varying cortical venous outflow architecture (Tables 3).[22]

Table 3: Cognard’s classification of cranial dural Arteriovenous Fistulae

Type	Venous Drainage	Flow Pattern in Sinus	Cortical venous Drainage
BENIGN			
1	Dural sinus	antegrade	No
2a	Dural sinus	retrograde	No
AGGRESSIVE			
2b	Dural sinus	antegrade	Yes
2a+b	Dural sinus	retrograde	Yes
3	Cortical vein		Yes
4	Cortical vein		Yes + venous ectasia
5	Cortical vein with spinal perimedullary drainage		Yes

Spinal DAVFs are classified by various methods and systems. The latest Takai Classification stands tall for merging both DAVFs and epidural fistulae as well. (table 4) [25]

Table4: Takai classification of Dural and Epidural AV fistulae of the spine (2016)

TYPE	NAME	SUBTYPE	DESCRIPTION
1	Dural AVF	As per the OLD Rosenblum Classification classically used	
2	Intramedullary Glomus AVM		
3	Intramedullary Juvenile AVM		
4	Perimedullary AVF	A	A single Feeder & Multiple small AVFs
		B	Multiple Feeders & Medium Size AVFs
		C	Multiple feeders & Giant AVFs
5	Extradural AVF	A	With Intradural Draining Vein
		B	Without Intradural Draining Vein

Clinical presentation

A majority of patients with DAVFs present in the fifth and sixth decades with symptoms related to lesion location and pattern of venous drainage. [11-18] Pulsatile tinnitus is a common symptom that results from increased blood flow through the dural venous sinuses, particularly in relation to transverse and sigmoid sinus lesions. [11-16] Cavernous sinus DAVFs can present with ophthalmoplegia, proptosis, chemosis, retroorbital pain, or decreased visual acuity. [12-22] Severe presentations include intracranial haemorrhage and nonhemorrhagic neurologic deficits such as seizures, parkinsonism, cerebellar symptoms, apathy, failure to thrive, and cranial nerve abnormalities, including rare cases of trigeminal neuralgia. [17-24] Some symptoms, including dementia and cognitive decline, may improve after treatment. [22] Haemorrhagic presentations are more frequent in high-grade (Borden types II and III, Cognard types IIb to IV) DAVFs. Unexplained subarachnoid or lobar haemorrhages should prompt consideration of a DAVF in the differential diagnosis. Spinal DAVFS present with compressive myelopathy like symptoms with paraesthesia and pain. Lower compressions may result in a cauda equina like picture with bowel and bladder involvement as well. [22-24]

Diagnosis:

Any suspicious flow void cluster around the dural venous sinus should prompt additional evaluation with dynamic CT Angiography (CTA), MR Angiography (MRA), or Digital Subtraction Angiography (DSA). [27, 26] CTA is particularly useful in treatment planning by precisely defining the arteriovenous shunt relative to surrounding brain and skull anatomy. Recent publications on 4D CTA by using 320-section multidetector row CT angiography have highlighted its potential to correctly diagnose, classify, and assist treatment planning for DAVFs. [25, 28, 29] Studies have reported, however, that CTA has reduced sensitivity versus MRA for the detection of DAVFs (15.4% versus 50%). [37, 36] Time-resolved MRA techniques are also promising and may be reliable for DAVF screening and surveillance in the future. [32, 31, 30] Due to current limitations of low resolution and saturation artefacts, the negative predictive value of MRA is inadequate to exclude DAVFs.[40]

Conventional angiography remains the most accurate method for detection and classification of DAVFs. [33- 38] The adjunct of FDCT to angiography is yielding previously unachievable high-resolution anatomic detail. Groups have demonstrated the utility of FPCT to precisely delineate the fistula site and provide superior visualization of arterial feeders and venous outflow. [37-38]

Treatment:

Endovascular approaches have become the mainstay of DAVF therapy, but the optimal approach for each case should involve discussions among a multidisciplinary team of interventional neuroradiologists, neurosurgeons, neurologists, and radiation oncologists. Careful assessment of a patient's clinical presentation, current status (age, medical condition, comorbidities), and type of lesion (location, classification, and angiographic features) should be conducted before embarking on any treatment. [37-39,]

Due to the efficacy of endovascular treatment, surgery is currently indicated in cases in which endovascular approaches have failed or are not feasible. [39, 44, 45] A variety of options is available, including direct intraoperative embolization of meningeal arteries or veins, resection of abnormal dura, packing of the diseased sinus, disconnection of the retrograde leptomeningeal venous drainage, and skeletonization of the dural sinus with disconnection of the dural arterial supply. [40] Certain anatomic locations of DAVFs are more amenable for surgery. These include the floor of the anterior cranial fossa and the superior sagittal sinus, where arterial access is difficult and/or sacrifice of the sinus is often undesirable. DAVFs that involve eloquent feeders are also better addressed by using a surgical or combined approach to ensure vessel preservation. [42, 44, 41]

Studies of SRS for DAVFs remain preliminary and have primarily involved low-risk lesions or those that are not amenable to endovascular or surgical approaches. [47, 45, 47] Lesions are irradiated with 20 –30 Gy, which causes vessel thrombosis and fistula closure during a latency period ranging from several months to a year. Until completion of vessel thrombosis, the haemorrhage risk remains elevated, so SRS is inappropriate as the primary treatment in DAVFs with CVD. [46, 48] Early results have been encouraging, with obliteration rates as high as 93% for combined endovascular embolization and SRS but have also demonstrated rates as low as 50% when only SRS is used. There have also been significant disparities in efficacy depending on the location of the fistula reflecting challenges for shunt targeting in complex lesions. [44,45,46,47,48]

Limitations of the study

As performed in different centres by different surgeons with differing degrees of competence in surgery and endovascular techniques, the procedures were not performed according to any protocol but to surgeon preference and convenience in order to ensure the best result for the patients. Another problem is that we operate in a third world country where financial constraints often dictate treatment protocols and hospital stay duration. We were also subject to the same forces. Since open surgery is cheaper significantly to endovascular procedures, we were obliged to opt for such a strategy despite evidence to the contrary.

Conclusion

Dural Av fistulae either cranial or spinal present a complex problem to today's patients. The advance of endovascular and surgical techniques along with advanced imaging modalities enable for early detection and prompt effective treatment which is so crucial in the prevention and minimising of neurological morbidity.

All patients have given written consents for their details to be used in this manuscript

There is no conflict of interest for any of the authors

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Authors contributions:

1. Dr. Sibhi Ganapathy: preparation of manuscript, data collection, analysis, review of features, approval of final draft, submission to journal
2. Dr. Swaroop Gopal: overview of case selection and images
3. Dr. Paritosh Pandey: Critical review of manuscript and final approval before submission

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