ORIGINAL RESEARCH

Dural Arterio-Venous Fistulae: Aetiology, Clinical and Radiological Features with Recent Management Trends

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ABSTRACT

Introduction: Dural arteriovenous fistula (DAVF) is an abnormal connection of vessels in the tissues around the brain or spinal cord. Exact etiology of DAVF is not known. They can cause multiple symptoms as per location and presents with Haemorrhages. Diagnosis - Includes CT and MR, dynamic CTA, MRA or DSA.Treatment ranges from conservative treatment to endovascular therapy, surgery, and Radiation therapy.

Aim and Objectives: To study aetiology, clinical and radiological features and management of Dural Arterio-venous fistulae.

Methods: Total 33 patients of DAVF studied which divided in three groups as ncsDAVF (18), csDAVF (8) and spinal DAVF (7) and were investigated and treated as per radiological finding.

Results: Mean age of presentation of ncsDAVF is 44 years, csDAVF is 37 years and spinal DAVF is 56 years. Male preponderance present in both. In intracranial DAVF, in spinal DAVF, thoraco-lumbar region is the most common site. On angiographic follow up of 11 Onyx treated patients, 10 patients were achieved complete cure (90.9%) in a single session (9.09%).

Conclusion: In csDAVF, endovascular treatment has good results, In spinal DAVF also endovascular embolisation using Glue+ lipoidal solution. complete occlusion is seen.

Keywords: Spinal Dural Arteriovenous Fistula, Cranial Dural Arteriovenous Fistula, Subdural Hemorrhage, Intracranial Hemorrhage, Subarachnoid Hemorrhage, ONYX, Coiling.

INTRODUCTION & BACKGROUND

A dural arteriovenous fistula (DAVF) is an abnormal connection of vessels in the tissues around the brain or spinal cord in which one or more arteries are directly connected to one or more veins or venous spaces called sinuses. Intracranial DAVFs are rare vascular abnormalities, account for 10-15% of all intracranial vascular abnormalities.[1] Though spinal dural arteriovenous fistulas (SDAVFs) are most commonly encountered spinal vascular

malformation, still remain underdiagnosed and rare entities.[2,3] The exact etiology of DAVF is not known. There are two etiologic hypotheses based around sinus thrombosis. 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. The second is that venous hypertension due to outflow obstruction causes decreased cerebral perfusion and promotes neoangiogenesis. The most common (70%-90%) cause of direct CCF is trauma from a basal skull fracture resulting in tear in the internal carotid artery (ICA) within the cavernous sinus.[4,5,6]. However, spontaneous fistulas usually have multiple dural feeders and numerous microfistulas within the cavernous sinus wall. Location of DAVFs : Intracranial : Transverse/sigmoid sinus , Cavernous sinus, Tentorium, Superior sagittal sinus and anterior cranial fossa. In Spine: Thoracolumbar region though rarely cervical and sacral regions affected. Patients with DAVFs may be completely asymptomatic. Symptoms, when present, may range from mild symptoms to fatal hemorrhage. The symptoms depend on the location and venous drainage pattern of the DAVF.[1]Headache, Tinnitus, Ocular symptoms, Cranial nerve palsie, Stroke-like Symptoms- Symptoms of venous hypertension, raised intracranial pressure, Paresthesias, diffuse or patchy sensory loss, but also radicular pain that may affect both lower limbs or initially only one limb. Bowel and bladder incontinence, erectile dysfunction, and urinary retention are more often seen late in the course of the disease.They can cause Hemorrhage, Subdural hemorrhage (SDH), Intracranial hemorrhage (ICH), Subarachnoid hemorrhage (SAH), Intraventricular hemorrhage (IVH), Intracranial hypertension, Venous congestion and oedema, Spinal myelomalacia, They are classified as per Cognard classification[8,9], Borden classification[10] and Barrow classification.[11]
DIAGNOSIS: Initial radiologic evaluation includes CT and MR imaging. Any suspicious flow void cluster around the dural venous sinus should prompt additional evaluation with dynamic CTA, MRA or DSA.[12]
TREATMENT: Conservative treatment , endovascular therapy, surgery, and Radiation therapy.

AIM AND OBJECTIVES

To study aetiology, clinical and radiological features and management of Dural Arterio-venous fistulae.

METHODS

Total 33 patients of DAVF studied which divided in three groups as ncsDAVF (18), csDAVF (8) and spinal DAVF (7) and were investigated and treated in a territory care center with help of the radiology and the interventional department of radiology by the neurosurgeon.

RESULTS AND OBSERVATIONS

Table-1 shows age among the groups. There was no significant ($p>0.05$) difference in the age among the groups.
Table-2 shows gender among the groups. There was no significant ($p>0.05$) association of gender among the groups.
Table-3 shows comparison of etiology among the groups. Traumatic was the most common etiology in csFAVF (75%). However, idiopathic was the most common etiology in ncsDAVF and sDAVF. There was no significant ($p>0.05$) association of etiology among the group.
Table-4 shows comparison of location among the groups.
Table-5 shows chief complaints in csDAVF. Proptosis and Eye Congestion were the main clinical complaints each constituting 87.5%. Decreased/loss of vision was the most common neurological symptom.

Table-6 shows neurological deficit in csDAVF. Neurological deficit was present in 75% patients of csDAVF.
Table-7 shows clinical examination in csDAVF. Proptosis and Chemosis of eye were in majority of patients of csDAVF each constituted 87.5%.
Table-8 shows CT brain with Angio done in 4 patients of csDAVF. Prominent Cavernous sinus /draining vein was seen all the patients of csDAVF. Venous reflux was seen in 50% patients of csDAVF.
Table-9 shows MR brain contrast/MR-angio in csDAVF. Prominent draining sinus/vessels was seen all the patients of csDAVF.
Table-10 shows digital subtraction cerebral angiography findings in csDAVF. Left cavernous sinus location was seen in majority of patients (87.5%) of csDAVF. One no. of feeder (62.5%) was found in more than half of patients of csDAVF. Unilateral (87.5%) was observed in majority of patients of csDAVF. ICA branch (75%) was found in majority of patients of csDAVF.
Table-11 shows venous drainage in DSA in csDAVF. Intracranial was the most common (87.5%) of csDAVF and Extracranial was the second most common venous drainage in DSA. Venous Thrombosis was least common in patients of csDAVF.
Table-12 shows direct/indirect in csDAVF. Direct and indirect was in 50% each of csDAVF.
Table-13 shows treatment in csDAVF. Coilling was done in majority of patients of csDAVF (87.5%). Average follow-up: 2.62 months
Table-14 shows clinical follow-up results in csDAVF. Better clinical finding was found in all the patients of csDAVF.
Table-15 shows radiological follow-up results in csDAVF. Cured was in 12.5% patients of csDAVF on both CT-Brain with Angio and MRI-Brain with Angio. Total Obliteration was in all the patients of csDAVF on DSA.
Table-16 shows chief complaints in ncsDAVF. Headache was the main clinical complaint (72.2%) in ncsDAVF. Limb weakness (38.9%) was the most common neurological symptoms in ncsDAVF. Tinnitus was the second most common neurological symptom (16.7%) in ncsDAVF. Neurological deficit was present 55.6% (10) patients of ncs DAVF.
Table-17shows clinical examination in ncsDAVF. Limb weakness was the most common (38.9%) patients of ncsDAVF and Altered sensorium was the second most common (16.7%).
Table-18 shows CT brain in ncsDAVF. Localized Subarachnoid Hemorrhage was seen in 30% and Diffused was in 10% patients of ncsDAVF. SDH was in 10% patients. Cerebellar intra axial was found in 30% patients of ncsDAVF.
Table-19 shows CT Angio in ncsDAVF. Visible Vascular Malformation and Prominent draining veins were the most common finding on CT angio in patients of ncsDAVF each constituted 71.4%.
Table-20 shows MRI brain contrast/MR-angio in ncs DAVF. Abnormal Flow voids was found most common on MRI brain (84.6%) and Prominent draining vessels/sinus (46.2%) was the second most common finding on MRI of ncsDAVF patients
Table-21 shows location of ncsDAVF. Transverse-Sigmoid sinus location of ncsDAVF was most common (27.8%) followed by Tentorium & Superior Saggital sinus (22.2%).
Fig.1 shows DSA finding inncsDAVF. One feeder was most common (33.3%) on DSA in ncsDAVF. Unilateral was in majity of patients (72.2%). ECA was in all the patients in ncsDAVF. Superficial Venous System was in all the patients in ncsDAVF.
Fig.2 shows treatment of ncsDAVF.Endovascular was done in more than half of patients (66.7&). Conservative treatment was done in 11.1% patients of ncsDAVF.
Fig.3 shows endovascular treatment in ncsDAVF. ONYX was done in majority of patients (91.7%) in ncsDAVF.

AVERAGE FOLLOW-UP

3.69 month

Radiological follow-up results in ncsDAVF.Cured was seen in all the patients on MRI brain. Total Obliteration was seen in 76.9% patients of ncsDAVF on DSA.

Fig 5 shows chief complaints in sDAVF. Limb weakness and Bowel/ Bladder alteration were most common complaints each constituted 85.7% in sDAVF. Limb Sensory changes was the second most common complaint (71.4%) in sDAVF.

Clinical examination in sDAVF. Neurological Deficit and Limb weakness were most common findings on clinical examination each constituted 85.7% in sDAVF. Sensory loss was the second most common clinical examination finding (57.1%) in sDAVF. MRI-spine contrast in sDAVF. Abnormal Flow voids was most common findings on MRI-spine contrast in sDAVF (71.4%). Cord edema was the second most common finding (57.1%) on MRI spine contrast in sDAVF. Location of sDAVF. Thoraco-Lumbar location was the most common in sDAVF (71.4%).

Fig. 6 shows location of sDAVF. Single arterial feeder was most common (71.4%). Unilateral was in all the patients of sDAVF. These feeders most commonly arising from lumbar artery in 3(43%) cases, intercostal artery in 2(29%) cases, segmental arteries from right iliac artery in 1 (14%) case. Single draining vein was in more than half of patients (57.1%). All the patients in sDAVF were treated with Glue with Lipoidal solution No complication was found in sDAVF. Average follow-up=4.14 months. Better clinical outcome was found in majority of patients (85.7%). Obliteration on DSA was found in majority of patients (71.4%).

STATISTICAL ANALYSIS

The results are presented in frequencies, percentages and mean±SD. Chi-square test was used to compare categorical variables. One way analysis of variance was used to compare continuous variables. The p-value<0.05 was considered significant. All the analysis was carried out on SPSS 16.0 version (Chicago, Inc., USA).

Table 1: Age among the groups

Groups	Age in years (Mean±SD)
csDAVF	37.62±12.23
ncsDAVF	44.94±11.20
sDAVF	56.57±10.61
p-value[1]	0.08

Table-2: Gender among the groups

Gender	csDAVF (n=8)		ncsDAVF (n=18)		sDAVF (n=7)		p-value[1]
	No.	%	No.	%	No.	%	
Male	7	87.5	17	94.4	7	100.0	0.11
Female	1	12.5	1	5.6	0	0.0	

Table-3: Comparison of Etiology among the groups

Aetiology	csDAVF (n=8)		ncsDAVF (n=18)		sDAVF (n=7)		p-value[1]
	No.	%	No.	%	No.	%	
Idiopathic	1	12.5	17	94.4	7	100.0	0.23
Traumatic	6	75.0	1	5.6	0	0.0	

Iatrogenic	1	12.5	0	0.0	0	0.0	

Table-4: Locations of DAVF in all 3 groups in the groups

Location of DAVF	No of patients (n=33)	%
1]**Intra-cranial [ncsDAVF+csDAVF]**	**(n=26)**	78.8
Cavernous sinus	8	30.8
Transverse-Sigmoid sinus	5	19.2
Tentorium	4	15.4
Superior Saggital sinus	4	15.4
Sigmoid sinus	1	3.8
Superior Petrosal sinus	1	3.8
Falx cerebri	1	3.8
Basi Frontal region	1	3.8
Foramen Magnum	1	3.8
2] Spinal DAVF [sDAVF]	**(n=7)**	21.2
Thoraco-Lumbar	5	71.4
Cervical	1	14.3
Sacral	1	14.3

Table-5: Chief complaints in csDAVF

Chief complaints*	csDAVF (n=8)	
	No.	%
Clinical complaints		
Proptosis	7	87.5
Eye Congestion	7	87.5
Headache	2	25.0
Vomiting	1	12.5
Neurological symptoms		
Loss of Consciousness	1	12.5
Tinnitus	4	25.0
Limb weakness	1	12.5
Diplopia	2	25.0
Decreased/loss of vision	4	50.0

Table-6: Neurological deficit in csDAVF

Neurological deficit	csDAVF (n=8)	
	No.	%
Present	6	75.0
Absent	2	25.0

Table-7: Clinical Examination in csDAVF

Clinical examination*	csDAVF (n=8)	
	No.	%
Altered sensorium	1	12.5
Proptosis	7	87.5
Chemosis of eye	7	87.5
Decreased visual Acuity	4	50.0
Orbital Bruit	4	50.0

Cranial nervesinvolvement	3	37.5
Limb Weakness (Motor Power)	1	12.5

Table-8: CT Brain with Angio (Carotids and cerebral arteries) in csDAVF

CT Brain with Angio	**csDAVF (n=4)**	
	No.	**%**
Intracranial Hemorrhage [ICH]	1	25.0
Prominent Cavernous sinus /draining vein	4	100.0
Venous reflux	2	50.0
Dural sinus opacification	0	0.0
Feeding arteries	0	0.0

*Multiple response

Table-9: MRI-Brain contrast / MR-AngioincsDAVF

MRI-Brain contrast / MR-Angio *	**csDAVF (n=5)**	
	No.	**%**
Abnormal Flow voids	2	40.0
Feeding arteries	0	0.0
Prominent draining sinus/vessels	5	100.0
Venous congestion	2	40.0

Table-10: Digital Subtraction Cerebral Angiography Findings in csDAVF

Digital Subtraction Cerebral Angiography	**csDAVF (n=8)**	
	No.	**%**
Location		
Left cavernous sinus	7	87.5
Right cavernous sinus	1	12.5
Arterial Feeders*		
No of Feeders		
1	5	62.5
2	1	12.5
3	2	25.0
U/L or B/L		
U/L	7	87.5
B/L	1	12.5
Branches Arising from		
ICA	6	75.0
ECA	1	12.5
Both ICA + ECA	1	12.5

Table-11: Venous Drainage in DSAin csDAVF

Venous drainage*	**csDAVF (n=8)**	
	No.	**%**
Intracranial	7	87.5
Extracranial	6	75.0
Cortical Venous Reflux	3	37.5
Venous Thrombosis	1	12.5

Table-12: Direct/Indirect in csDAVF

csDAVF	No of pts (n=8)	%
Direct	4	50
Indirect	4	50

Table-13: Treatment in csDAVF

Treatment	csDAVF (n=8)	
	No.	%
ONYX	0	0.0
Coiling	7	87.5
Combined ONYX + coiling	1	12.5

Table-14: Clinical Follow-up results in csDAVF

Clinical	csDAVF (n=8)	
	No.	%
No Change	0	0.0
Better	8	100.0
Worst	0	0.0

Table-15: Radiological Follow-up results in csDAVF

Radiological	csDAVF (n=8)	
	No.	%
CT-Brain with Angio		
Cured (Post-coiling status with complete obliteration of CCF with resolved ICH)	1	12.5
MRI Brain with Angio		
Cured (Significant reduction in caliber of left SOV and left extra-occular muscles)	1	12.5
DSA		
Total Obliteration	8	100.0

Table-16: Chief complaints in ncsDAVF

Chief complaints*	ncsDAVF (n=18)	
	No.	%
Clinical		
Headache	13	72.2
Vomiting	5	27.8
Vertigo	2	11.1
Neurological symptoms		
Seizures	2	11.1
Loss of consciousness	2	11.1
Speech difficulty	1	5.6
Decreased vision	2	11.1
Tinnitus	3	16.7
Limb weakness	7	38.9
Sensory loss	1	5.6
Bladder/Bowel alteration	1	5.6
Gait disturbance	2	11.1

*Multiple response

Table-17: Clinical Examination in ncsDAVF

Clinical examination*	ncsDAVF (n=18)	
	No.	%
Altered sensorium	3	16.7
Altered Speech	1	5.6
Decreased Visual Acuity	2	11.1
Cranial nerves involvement	0	0.0
Increased Tone	1	5.6
(Motor Power) Limb weakness	7	38.9
(Sensory Loss)Numbness	1	5.6
Brisk Deep Tendon reflexes	1	5.6
Cerebellar signs present	2	11.1

*Multiple response

Table-18: CT Brain (Plain) in ncsDAVF

CT brain	ncsDAVF (n=10)	
	No.	%
Subarachnoid Hemorrhage		
Diffused	1	10.0
Localised	3	30.0
Intra-Cerebral Hemorrhage		
Extra-axial		
SDH	1	10.0
Intra-axial		
Temporal	2	20.0
Cerebellar	3	30.0
Parietal	1	10.0
Intraventricular Hemorrhage		
Only IVH	0	0.0
IVH + ICH	2	20.0
Lateral/Fourth ventricle location	2	20.0

Table-19: CT -Angio (Carotids and cerebral arteries) in ncsDAVF

CT angio*	ncsDAVF (n=7)	
	No.	%
Visible Vascular Malformation	5	71.4
Feeding art	0	0.0
Prominent draining veins	5	71.4
Venous reflux	1	14.3
Dural sinus opacification	1	14.3

Table-20: MRI-Brain contrast/MR-Angio in ncsDAVF

MRI-Brain contrast / MR-Angio *	ncsDAVF (n=13)	
	No.	%
Abnormal Flow voids	11	84.6
Feeding art	1	7.7
Prominent draining vessels/sinus	6	46.2

Venous congestion	3	23.1

Table-21: Location of ncsDAVF

Location of ncsDAVF	No of patients (n=18)	%
Transverse-Sigmoid sinus	5	27.8
Tentorium	4	22.2
Superior Saggital sinus	4	22.2
Sigmoid sinus	1	5.6
Superior Petrosal sinus	1	5.6
Falxcerebri	1	5.6
BasiFrontal region	1	5.6
Foramen Magnum	1	5.6

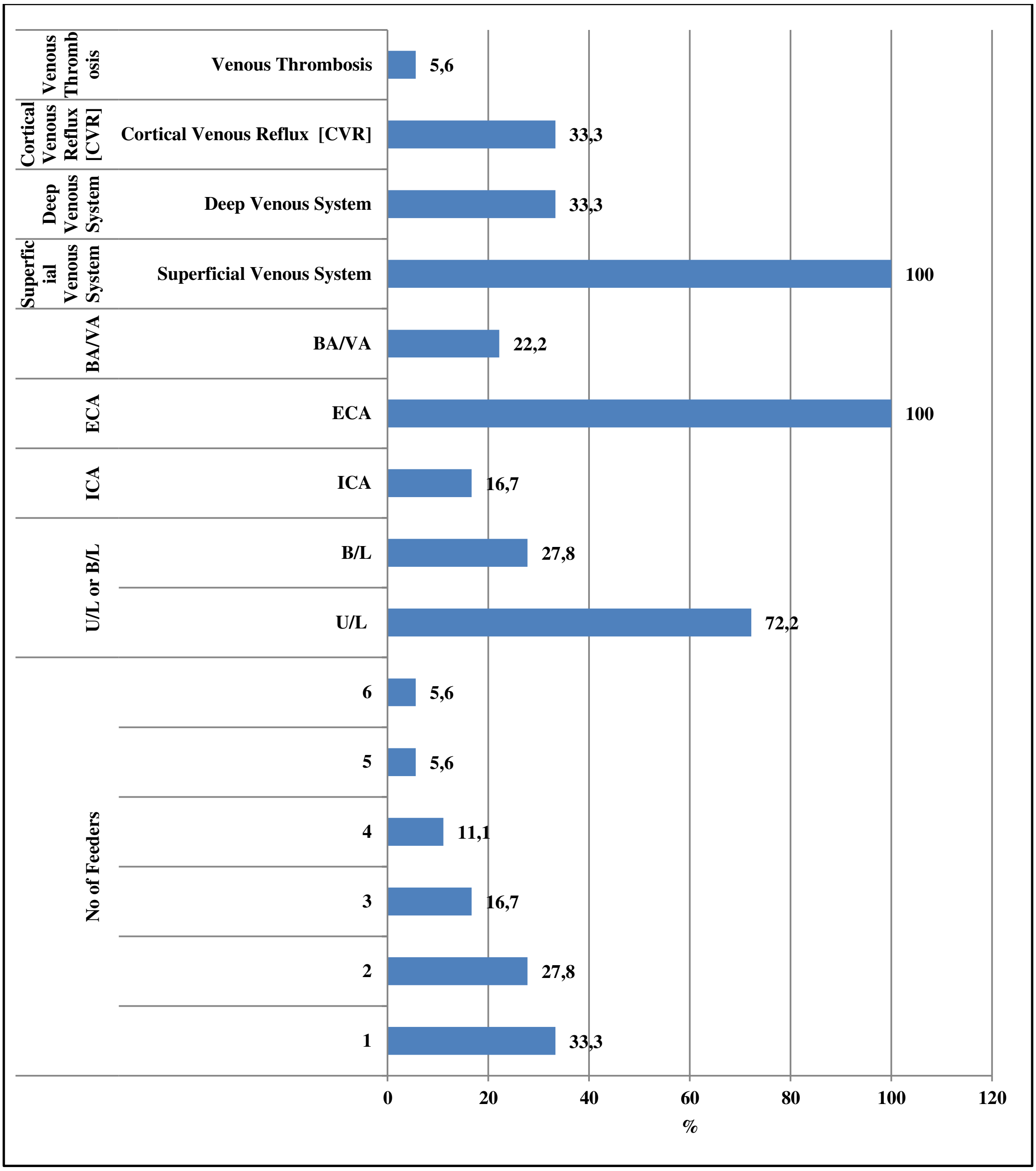

Fig.1 shows DSA finding in ncsDAVF

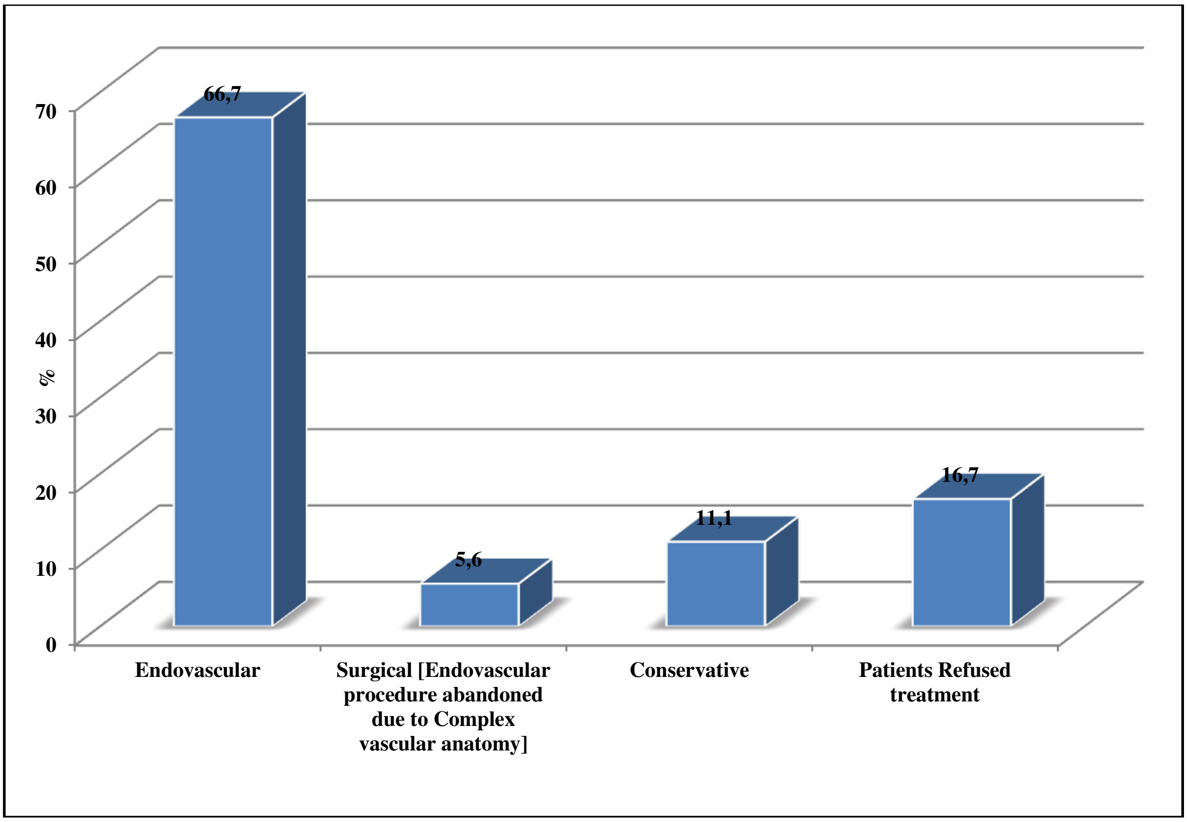

Fig. 2: of Treatment of ncsDAVF

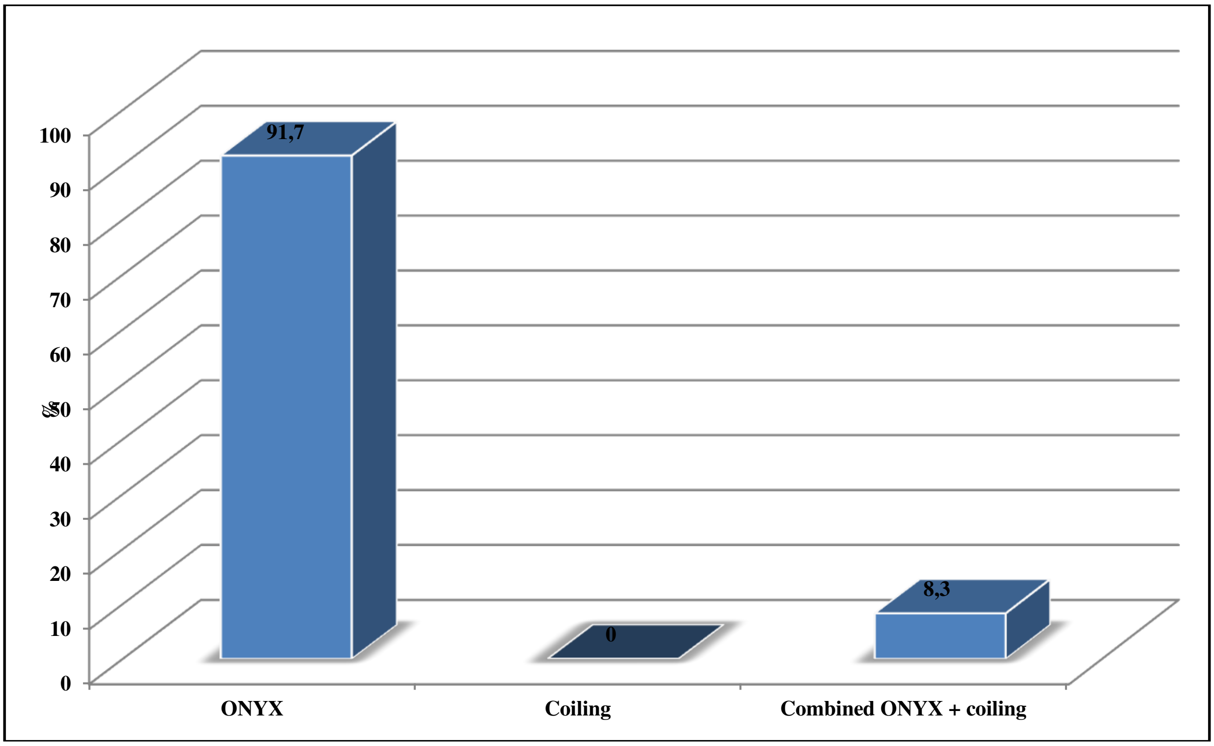

Fig. 3: Endovascular Treatment in ncsDAVF

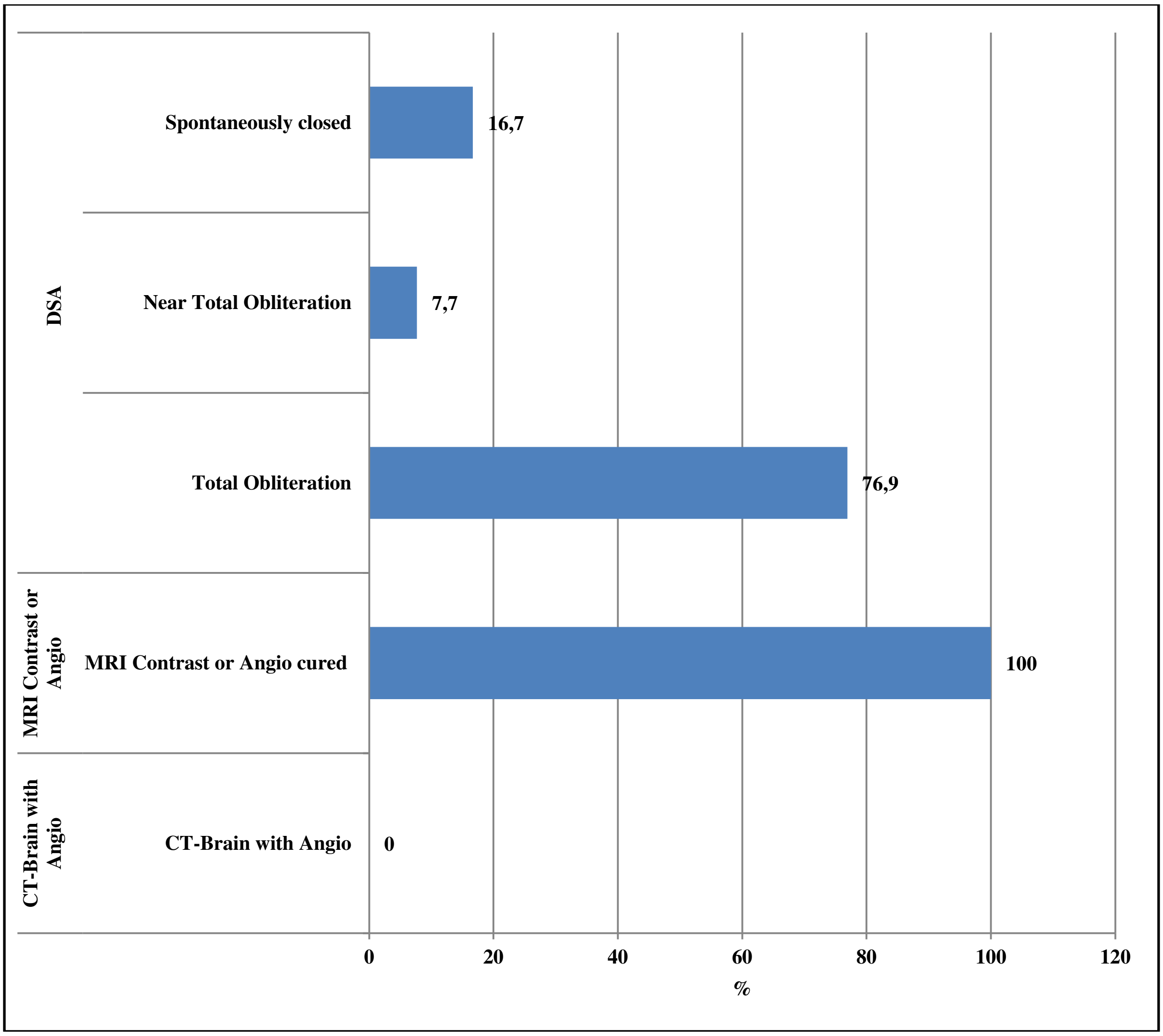

Fig. 4: Radiological Follow-up results in ncsDAVF

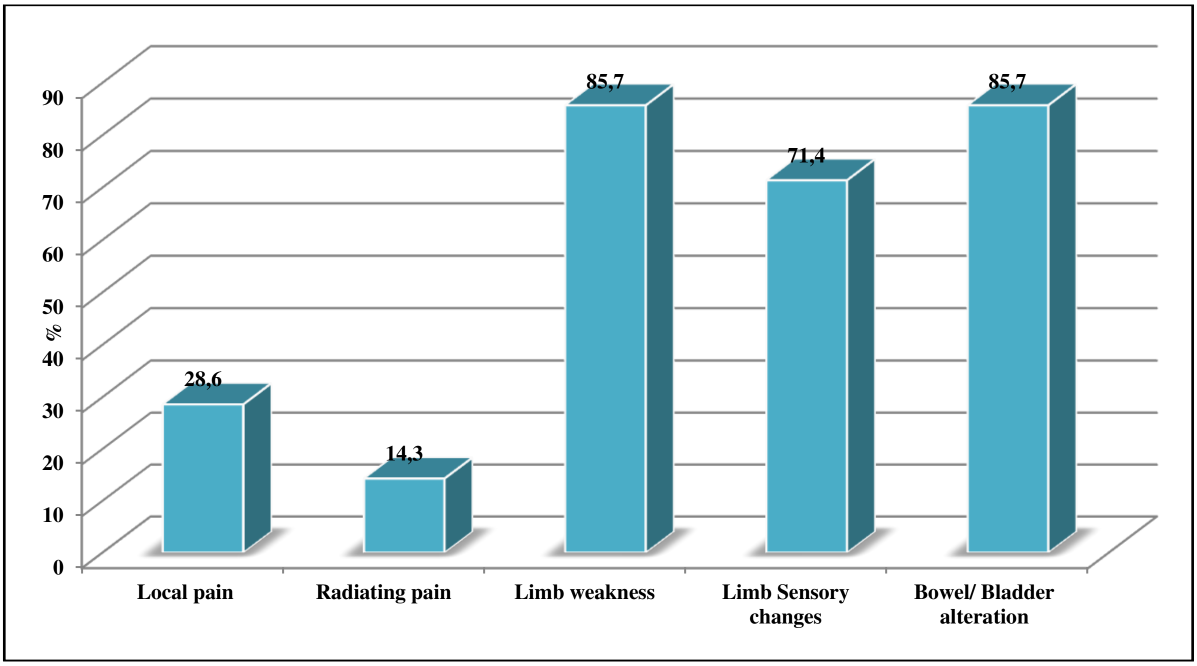

Figure 5: Chief complaints in sDAVF

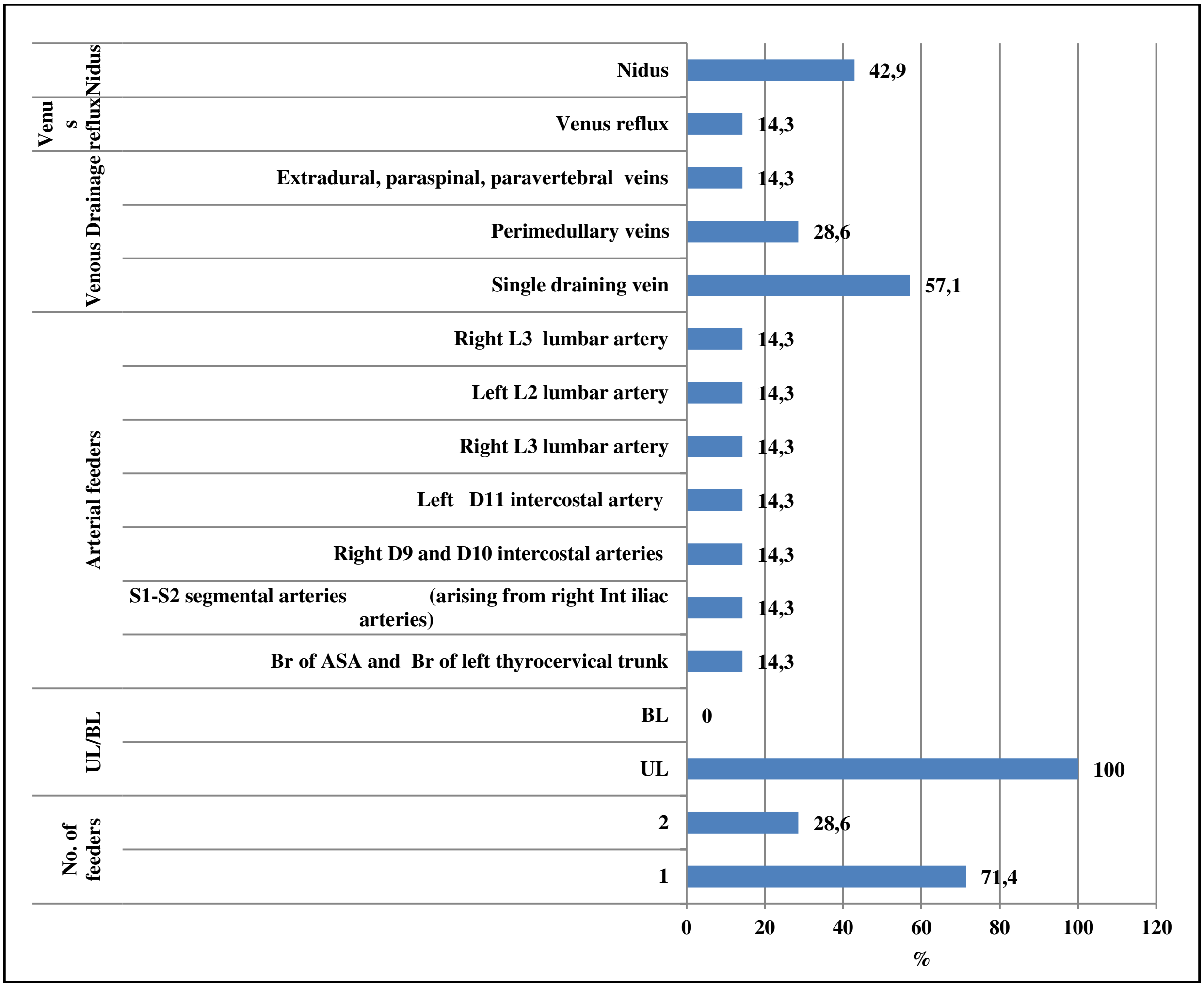

Fig. 6: Spinal Digital Subtraction Angiography in sDAVF

DISCUSSION

The study sample consisted of 33 patients (n = 33). For the purpose of study, they were broadly divided into two groups: intracranial and spinal dural AVFs. Of all the intracranial fistulae, the cavernous sinus fistulae were grouped and studied together because of their unique presentation. The data were analyzed in three groups:

a. Non-cavernous sinus dural AV fistula (ncsDAVF, n=18)
b. Cavernous sinus dural AV fistula (csDAVF, n=8)
c. Spinal AV fistula (DAVF, n=7)

AGE AND SEX

The mean age of patients of ncsDAVF is 44.94 (±11.20) years. Cooper CJ et al[13] reported that DAVF are very rare and more common over the age of 40 years. In our study, mean age of presentation of csDAVF patients is 37.62 (±12.23) years with maximum patient between age group 30-40 years. A review article by Ellis JA et al[4] mentioned that traumatic CCFs are the most common type, accounting for up to 75% of all CCFs. Spinal DAVF patients had a mean age of 56.57 (±10.61) years. Several studies in literature reported male preponderance in DAVF.[2,4]

ETIOLOGY

In our study, 17 patients (94.4 %) of ncsDAVF, one patient (12.5%) of csDAVF and all 7 patients (100%) of spinal DAVF were idiopathic in nature without any precipitating factor.

One patient from ncsDAVF group had traumatic etiology. On the other hand, We reported trauma as most common predisposing factor for csDAVF which was present in 6 patients (75%). One patient (12.5%)of csDAVF had history of previous endoscopic pituitary surgery.

CLINICAL FEATURES

NCS DAVF -

In our study, 18 patients of ncsDAVF clinically presented as headache in 13 (72%), intracranial hemorrhage in 7 (38%), limb weakness in 7(38%), altered sensorium in 3(16.7%), tinnitus in 3 (16.7), visual symptoms, vertigo, seizures, gait disturbance in 2 patients (11%), sensory loss and bowel/bladder disturbance in 1(5.5%). In our study, out of 7 patients of intracranial hemorrhage, 4 patients had subarachnoid hemorrhage [1 had diffuse SAH and 3 had localised SAH], 1 had subdural hematoma and 5 patients had intraparenchyamal hematoma; two hematomas had intraventricular extension. DAVFs with cortical venous drainage (CVD) may present with progressive dementia, seizures, parkinsonism, ataxia, hemorrhage and neurological deficits due to local venous congestion, brain edema and or ischemia. Particularly superior sagittal sinus DAVFs and fistulas that drain into the deep venous system cause these symptoms. Hu YC et al[15] studied 50 cases of DAVF and found clinical presentation of DAVF as headache 11(17%), intracranial hemorrhage in 14 (22%), limb weakness in 2(4%), altered sensorium in 3(6%), visual symptoms and vertigo in 1 (2%), seizures 3 (5%).

CSDAVF

In our study, indirect CCF (4 patients) presented as chemosis and proptosis in 75%, tinnitus 50%, orbital bruit 50%, diplopia 50%, decreased or loss of vision 50%, other cranial nerves involvement presents in 25%, headache in 25 %. One patient (25%) presented with ICH who presented with history of loss of consciousness and limb weakness. In our study, most common clinical presentation of CCF was proptosis and chemosis present in 7 of 8 patients (88%). Gupta AK et al[1] reported that direct CCFs often present acutely. They typically progress rapidly, necessitating urgent treatment. The most common presenting signs and symptoms include proptosis in 72%–98%, chemosis in 55%–100%, orbital bruits in 71%–80% and headache in 25%–84% of patients. Visual disturbances, including diplopia reported in 88% of patients, blurry vision, and orbital pain, while other cranial nerve deficits have been reported in 17%–44% of patients. In our study, direct CCF (4patients) presented as proptosis and chemosis in 100%, tinnitus in 50%, orbital bruit 50%, decreased or loss of vision 50%, other cranial nerves involvement presents in 50%, seizures in 25% and headache in 25 %. Meyers PM et al[16] in the series of 135 patients with indirect CCFs, common signs and symptoms included chemosis in 87%, proptosis in 81%, diplopia with ophthalmoparesis in 68%, orbital bruit in 49%, headache in 34% and decreased visual acuity in 31%.

SPINAL

In our study, most common clinical presentation at the time of diagnosis were Paraparesis (85.7%), bowel/bladder alteration (85.7%) followed by sensory changes in limb (71.4%). All 3 symptoms were present in 57.2 % of patients. Similar clinical presentation was reported by Jellema K et al1[7] in which most common symptoms were micturition problems (80%), leg weakness (78%), and numbness in the legs or buttocks (69%). Combination of all 3 symptoms were present in 58% of patients. Pain was present in 3 patients, 2 patients (28.6%) had local pain and 1 patient (14.3%) had radiating pain in our study

LOCATIONS

DAVFs are most frequently located in the region of transverse, sigmoid and cavernous sinuses.[12] In our study, cavernous sinuses and transverse-sigmoid sinus were the commonest sites of intracranial DAVFs which were similar to the study results given by Saraf R et al.[3]In the study of Natarajan et al[18] out of 32 patients of DAVF , 10 DAVFs (31%) were located at the transverse/sigmoid sinus region, 7 (22%) were located at the tentorial region, 3 (9%) were located at the falcine/superior sagittal sinus region, 7 (22%) were csDAVF 3 (9%) were in the middle fossa, and 1 each (3%) located at the torcular/transverse sinus and anterior fossa region.Most of the spinal DAVFs were located in Thoracolumbar regions in our study (5 patients,71.4%), Cervical region (1 patient, 14.3%) and sacral region (1 patient,14.3%) involvement was less common. Review by Krings T et al[2]mentioned that spinal DAVF are solitary lesions and found in thoracolumbar region with >80 % cases located between T6 and L2.

RADIOLOGY

CT AND MRI IMAGING

Initial radiologic evaluation includes CT and MR imaging. Noncontrast CT is limited to identifying intracranial hemorrhage and edema due to venous congestion. MR imaging is more helpful because it can demonstrate dilated vessels, venous pouches, vascular enhancement, and signs of venous hypertension in high-grade lesions (eg. white matter hyperintensity, intracranial hemorrhage or venous infarction). Van Rooij et al[19] studied 91 patients of intracranial DAVF in which 29 (32%) had cortical venous reflux and presented with intracranial hemorrhage in 18 (20%). In our study total 26 cases [18 ncsDAVF + 8csDAVF], cortical venous reflux seen in 9 cases (35%) and presented with aggressive symptoms like ICH, seizures, neurological deficit, positive cerebellar signs in 8 cases (31%) [6 ncsDAVF + 2 csDAVF]. This suggests that intracranial DAVF patients with retrograde cortical venous reflux are aggressive lesions which presents with intracranial hemorrhages. In the present study, clinical presentation of ncsDAVF was intracranial hemorrhage in 7 of 18 patients (39%), and 5 of these 7 patients had an intraparenchymal hematoma and 1 patient with a subdural hematoma. On CT scan intraparenchymal hematoma present in the following locations: cerebellum in 3, temporal lobe in 1, parieto-temporal lobe in 1. Four patients presented with subarachnoid hemorrhage. In all patients who presented with hemorrhage, the DAVF was discovered on subsequent angiography. In previous study of van Rooij et al, [19] the clinical presentation was hemorrhage in 18 of 29 patients (60%), and 16 of these 18 patients had an intraparenchymal hematoma on CT scanning with the following locations: cerebellum in 4, occipital lobe in 4 (in 1 patient with a concomitant subdural hematoma), temporal lobe in 3, frontal lobe in 2, parietal lobe in 2, and basal ganglia in 1. Two patients presented with subarachnoid hemorrhage.

In csDAVF, ICH found to be less 5%.[13]. In our study, one patient out of 8 patients showed intracranial hemorrhage. In our study, CT-Brain with Angiography was done in 7 patients of ncsDAVF, vascular malformation and prominent draining veins/sinuses seen in 5 patients, venous reflux and dural sinus opacification seen in 1 patient. While MRI-Brain with Angiography done in 13 patients of ncsDAVF, abnormal flow voids seen in 11 patients, feeding arteries in 1 patient, prominent draining veins/sinuses in 6 patients and venous congestion in 3 patients.It is concluded by many authors that 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. CT and MR angiography may be used to screen patients suspected of harboring DAVF, grade or classify these lesions when detected, as well as evaluate for response to treatment. [19, 20,21] Study on MR-imaging of DAVF done by Chang SY et al[24] showed 11 of 12 patients (92%), showed the presence of signal voids and dilatation of the involved dural sinus. In our study, abnormal flow voids seen in 11 of 13 patients (84.61%) and dilatation of dural sinuses/vessel in 6 patients [46.15%]. In same study, dilatation of the superior ophthalmic vein and cortical venous system and high signal intensity on T2-weighted images were observed in three cavernous lesions (50%), In our study, MR-imaging done in 5 cases of csDAVF and prominent superior ophthalmic vein in 3 cases (60%) and prominent cavernous sinus in 2 cases (40%), venous congestion seen in 2 cases (40%).

For the diagnosis of DAVFs, cerebral angiography is the gold-standard imaging modality, patients typically undergo noninvasive cerebral imaging with CT scanning, MRI, or CT/MR angiography first. Evidence of cavernous sinus enlargement, proptosis, extraocular muscle enlargement, superior ophthalmic vein dilation or dilation of cortical or leptomeningeal vessels, as well as associated skull fractures, may be seen on CT or MRI and are suggestive of CCF.[1,] However, the absence of abnormalities on noninvasive imaging studies does not exclude the diagnosis of CCF. Any suspicious flow void cluster around the dural venous sinus should prompt additional evaluation with dynamic CTA, MRA.[19] Spinal DAVF is a lesion that can be easily misdiagnosed.

DSA

Digital Subtraction Angiography (DSA) remains the most accurate method for detection and classification of DAVFs. Natarajan et al.[18] The fistula was supplied exclusively by branches of the ECA in 11 patients (34%), including the occipital artery, posterior auricular artery, and ascending pharyngeal artery in lesions of the transverse sigmoid sinus and from the middle meningeal artery (MMA), accessory meningeal artery, artery of foramen rotundum, and ascending pharyngeal artery in lesions of the cavernous sinus. Nineteen fistulas (59%) were also supplied from cavernous or intradural ICA branches, and two (6%) had feeders from the dural branches or cerebellar arteries arising from the vertebrobasilar system. The main ICA branches supplying the DAVF were the meningohypophyseal trunk and the tentorial branch. In cavernous sinus lesions, draining veins were mainly the superior ophthalmic vein and the IPS, and leptomeningeal venous drainage was through the superficial sylvian vein and the basal vein of Rosenthal. In transverse sigmoid sinus lesions, the drainage system was the ipsilateral jugular vein and the contralateral transverse sinus–internal jugular vein. The main leptomeningeal venous drainage was through the veins of Labbé or the occipital veins. Leptomeningeal venous drainage was present in 24 patients (75%), with venous stenoses and ectasia identifiable in most of these patients.

In our study for csDAVF, unilateral arterial feeders seen in 7 (87%) and bilateral feeder seen in 1 (12.5%), The number of feeders found single in 5(62%) cases, double in 1(12%) case, three in 2(25%). These feeders arise from ICA in 6(75%) cases, ECA in 1(12.5%) case and both in 1 (12.5%) case. Venous drainage occurred intracranially in 2(25%) cases and extracranially in 1 (12.5%) case and both in 5(62.5%) cases. Cortical venous drainage seen in 3(37.5%) cases. Venous thrombosis seen in 1(12.5%) case.

In ncsDAVF, nidus was seen in 2(11.1%) cases, unilateral feeders seen in 13 cases (72.2%) and bilateral arterial feeders seen in 5(27%); 6(33%) cases had single arterial feeder, 5(27%) cases had double feeders and rest 7(30%) cases had multiple (>2) feeders. Arterial feeders coming from ECA in all 18 (100%) cases, ICA in 3 (16%) cases and VA/BA in 4 (22%) cases. Venous drainage into superficial venous system:18(100%) and deep venous system in 6(33%) cases. Cortical venous reflux seen in 6(33%) cases and venous thrombosis in 1(5.6%).

In spinal DAVF, nidus seen in 3(43%) cases, all 7 (100%) had unilateral arterial feeders. 5(71%) had single feeder while 2(29%) had 2 feeders. These feeders most commonly arising from lumbar artery in 3(43%) cases, intercostals artery in 2 (29%) cases, segmental arteries from right iliac artery in 1 (14%) case, branch from anterior spinal artery and left thyrocervical trunk in 1 (14%) case. Venous drainage as follows- in 4 (57%) cases, into single draining vein, 2(29%) perimedullary veins and 1(14%) had extradual, paravertebral veins. Venous reflux seen in 1 (14.3%) case.

TREATMENT

In our study, total 33 cases diagnosed DAVF [18 ncsDAVF, 8csDAVF and 7sDAVF] of which 30 patients [15ncsDAVF, 8 csDAVF and 7 sDAVF] were received treatment and 3 patients were refused treatment.

In 15 ncsDAVF patients, 12 treated by endovascular approach,1 by surgical approach and 2 were conservatively managed. In csDAVF(8) and sDAVF(7), all were managed by endovascular approach.

During the past 2 decades, embolisation by using transarterial, transvenous, or, occasionally, combined approaches has become a first-line treatment for DAVFs. [22] The use of Onyx has been increasingly reported for the treatment of DAVFs. A major advantage of Onyx is the ability to cure complex multifeeder fistulas via a single pedicle. Excellent cure rates have been reported with this agent, with a high proportion of treatments completed in a single session. In a series of 30 patients with DAVFs with CVR, Cognard et al[23] achieved a complete cure in 24 patients. Of these 24 patients, 20 (83.33%) cures were achieved after a single procedure.In the study of Saraf et al[3], 33 (92%) out of 36 were cured by Onyx embolisation while in our study, 10 (90.9%) of 11 patients cured by Onyx embolisation. Natarajan et al.[9] reported surgery achieved complete occlusion of four fistulas after failed or incomplete endovascular treatment. Due to the efficacy of endovascular treatment, surgery is currently indicated in cases in which endovascular approaches have failed or are not feasible. [2] In our study, one (7.6%) patient of ncsDAVF treated by surgical approach due to inaccessible DAVF at the time of endovascular treatment. Hu YC et al.[15] stated that when Onyx was used as the sole embolic agent, in 29 (87%) of 33 patients achieved angiographic cure while 5 (8.3%) DAVF require surgical disconnection. In our study**,** angiographic cure found in 10 (90.9%) of 11 patients while one (7.6%) patient require surgical ligation. In the study of Saraf R et al[24], out of 36 Onyx treated patients, 2 (5.5%) patients had complications and no death occurred. In our study, 11 Onyx treated patients, complications like hemorrhage (1) and cerebral edema (1) occurred in 2 (18%) patients and death occurred in one patient of ncsDAVF who treated with Onyx + coiling of venous pouch had complication of increase in size of previous bilateral cerebellar hematoma. This patient underwent surgical evacuation of bilateral cerebellar hematoma, but patient could not be survived in post-operative period. In our study, 12 cases of ncsDAVF treated by endovascular approach, 11 by Onyx and 1 by Onyx +coiling. On angiographic follow up of 11 Onyx treated patients, 10(90.9%) patients were achieved complete cure and 1 (9.09%) patient had near total obliteration in the first session. The patient improved significantly. Since this was a slow-flow fistula and the patient had become asymptomatic after the first session of treatment, it was decided to manage him conservatively and keep him under observation. Close follow-up is necessary to assess the development of new symptoms or progression of existing ones.[2] In our study, clinical follow up, all 11 patients who received Onyx embolisation treatment were symptomatically better (100%) The patient who treated surgically was better on clinical follow up and showed complete cure on MR imaging

Kannath et al[25]Thirty-two patients were included in study. Five patients (15.7%) showed spontaneous resolution in the follow-up period while awaiting definitive treatment. Van Rooij et al[19] showed 2 patients (9.6%) of 29 patients have spontaneous obliteration of DAVF. In our study, out of 23 patients [15 ncsDAVF and 8csDAVF], 2 patients (8.6%) showed spontaneous closure of ncsDAVF, on follow up. These 2 patients of ncsDAVF had slow flow, Type-I DAVF at the time of diagnosis and managed conservatively. Gupta AK et al[1] reported that endovascular embolisation by using transarterial or transvenous approach is the first line treatment modality for the treatment of most CCFs. Metallic coils and/or liquid embolic agents are now most commonly used for this purpose. Natarajan et al.[18] All seven patients with carotid-cavernous sinus fistulas were treated by tranvenous Guglielmi detachable coil [GDC]embolization, with one patient receiving additional transarterial Onyx therapy. All 7 carotid-cavernous fistulas were completely obliterated. In our study, complete fistula occlusion seen in all 8 patients (100%) treated by using coiling in endovascular treatment. While on follow up cerebral angiography, one patient of completely obliterated indirect CCF found small new indirect CCF. This was not visible earlier. This patient was asymptomatic, and it was a slow flow fistula, so it was conservatively managed.

On clinical follow up of all treated 8 patients of csDAVF in our study, proptosis and chemosis improved in all 8 patients, diplopia and cranial nerves paresis improved in 2 patients, Vision improved in 2 patients and remained same (loss of vision) in 2 patients. After successful intervention with complete closure of a csDAVF, symptoms such as chemosis and proptosis generally resolve within hours to days. Cranial nerve palsies typically resolve over the course of several weeks. The degree of vision recovery, if vision loss was experienced prior to intervention, is largely dependent on the pathogenesis, severity, and duration of the preintervention deficit. In our study, all 7 patients of spinal DAVF were treated by using Glue+ lipoidal solution in endovascular approach. In our study, all 7 patients of spinal DAVF were treated by endovascular embolisation using Glue+ lipoidal solution. No complications reported during and after each procedure. Patients were followed up for average period of 4.14 months (range 1 to 6 months) both clinically and radiologically. Six patients (85.7%) showed clinical improvement while one patient (14.3%) showed no change clinically. Radiologically, on DSA, there was complete occlusion in 5 patients 71.4%), near total occlusion in one patient (14.3 %) and partial occlusion in one patient (14.3%). There was a small slow flow fistula at same location in patient of near total occlusion. As clinical improvement was there in neck pain and limb parasthesia in same patient, he was managed conservatively while another patient with partial occlusion, re-embolised in same setting. This patient cured clinically as well as angiographically on further follow up.

CONCLUSION

Total 33 patients of DAVF studied which divided in three groups as ncsDAVF (18), csDAVF (8) and spinal DAVF (7). Mean age of presentation of ncsDAVF is 44 years, csDAVF is 37 years and spinal DAVF is 56 years. Male preponderance present in both. ncsDAVF(94.4%) and sDAVF(100%) predominantly found to be idiopathic. Traumatic was the most common etiology in csFAVF (75%)

In intracranial DAVF, cavernous sinus (30%) was the most common location followed by transverse-sigmoid region (19%) which was second most common location. In spinal DAVF, thoraco-lumbar region is the most common site.

ncsDAVF most commonly presented with headache (72%), intracranial hemorrhage (38%), limb weakness (38%), tinnitus (16%), altered sensorium (16%), decreased vision (11%)

Proptosis (87%), chemosis (87%), decreased /loss of vision (50%) and diplopia (25%) was the clinical presentation that occur in carotid-cavernous fistula

In spinal DAVF, limb weakness (85%), bladder-bowel alteration (85%) and sensory loss (57%) were commonest presentation.

71% and 100% Prominent draining sinus or veins seen on CTA in ncsDAVF and csDAVF respectively. On MR-Brain with angio showed abnormal flow voids 84% and 40% in ncsDAVF and csDAVF respectively. In spinal DAVF, abnormal flow voids (71%) and cord edema (57%) found on MR-spine with angiography.

DSA findings in csDAVF, unilateral7 (87%), single 5(62%) arterial feeders were most common. These feeders mostly arise from ICA in 6(75%) cases. Venous drainage occurred in both intracranially and extracranially in 5(62.5%) cases.
DSA findings in ncsDAVF, unilateral arterial feeders seen in 13(72.2%) cases; Number of arterial feeders found to be single in 6(33%) cases, double feeders in 5(27%) cases and rest 7(40%) cases had multiple (>2) feeders. Arterial feeders most commonly arising from ECA in all 18 (100%) cases. Venous drainage most commonly seen in superficial venous system 18(100%).
DSA findings in spinal DAVF, all 7(100%) had unilateral arterial feeders. 5(71%) had single feeder. These feeders most commonly arising from lumbar artery in 3(43%) cases, intercostals artery in 2 (29%) cases. Venous drainage found to be single draining vein in 4 (57%) cases.
In ncsDAVF, out of 15 cases, 12 cases treated with endovascular embolisation, 1 case treated with surgical intervention and 2 cases managed conservatively. In csDAVF (8) and sDAVF(7), all cases treated by endovascular coiling and glue embolisation respectively.
In ncsDAVF, 12 cases endovascularly, 11 by Onyx and 1 by Onyx +coiling.
On clinical follow up, all 11 patients who received Onyx embolisation treatment were symptomatically better (100%). On angiographic follow up of 11 Onyx treated patients, 10 patients were achieved complete cure (90.9%) in a single session (9.09%).
Out of 11 Onyx treated patients, complications like intracranial hemorrhage (1) and cerebral edema (1) occurred in 2 (18%) patients and death occurred in one patient of ncsDAVF who treated with Onyx + coiling and had post-procedure increase in previous intracranial hemorrhage. In ncsDAVF, out of 15 treated patients, 2 patients (13.3%) of Type-I DAVF who managed conservatively showed spontaneous closure of intracranial DAVF on follow up.
In csDAVF, on follow up cerebral angiography, complete fistula occlusion seen in all 8 patients (100%) treated by using coiling in endovascular treatment.
In spinal DAVF, all 7 patients of spinal DAVF were treated by endovascular embolisation using Glue+ lipoidal solution. No complications reported during and after each procedure. On follow up angiography, complete occlusion seen in 5 patients (71.4%).

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