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Review Article

Spontaneous closure of a superior sagittal sinus dural arteriovenous fistula with an extensive angioarchitectural network: A case report and systematic review of the literature

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ABSTRACT

Background: Intracranial dural arteriovenous fistulas (DAVFs) have been documented to occasionally spontaneously regress. However, the mechanism responsible for this occurrence remains speculative.

Methods: We present a case of a Borden II – Cognard IIa+b DAVF involving the superior sagittal sinus (SSS) with bilateral external carotid artery supply that regressed spontaneously. A systematic literature review was conducted to explore the current theories explaining the spontaneous regression of DAVFs according to Preferred Reporting Items for Systematic reviews and Meta-Analyses guidelines.

Results: A total of 26 studies and 54 cases were included in our results. Of the included cases, 57.14% of cases were Borden I, 16.33% were Borden II, and 26.53% were Borden III. Ruptured status or intracranial hemorrhage was documented in 24.1% of all cases, the majority of which (69.2%) were in cases with aggressive lesions (Borden II or greater). The most commonly involved location was the transverse sinus (38.89% of cases, $n = 21$), and the SSS was only involved in 12.96% of all cases. 50% of included cases proposed a mechanism responsible for spontaneous regression. The most frequently proposed mechanisms were thrombosis of the involved sinus/chronic inflammatory changes or direct endothelial injury, endoluminal stasis, and thrombogenic effects of contrast medium during angiography. We present the case of a 54-year-old woman with an aggressive ruptured DAVF that likely developed following a pediatric traumatic brain injury that was left untreated before she presented to our institution after significant delay. Her DAVF regressed on repeat angiography before neurovascular intervention without a clear identifying mechanism as proposed by the current literature.

Conclusion: Our results suggest that spontaneous regression is not necessarily associated with lower risk DAVFs. The present case offers a unique long-term insight into the natural history of an aggressive ruptured DAVF of the SSS that regressed without intervention. Further research into the natural history of DAVFs will be helpful in deducing key factors leading to spontaneous regression.

Keywords: Angiography, Dural arteriovenous fistula, Intracerebral hemorrhage, Spontaneous closure, Superior sagittal sinus

INTRODUCTION

Intracranial dural arteriovenous fistulas (DAVFs) are vascular malformations that form within the dura involving abnormal connections between dural or meningeal arteries and dural, arachnoid, or cortical veins.[1,4,13] The pathogenesis of DAVFs is thought to involve previous craniotomy, trauma, or thrombotic diseases with subsequent angiogenesis and the creation of arteriovenous shunts within the dura itself.[4,9] DAVFs account for 5–15% of all intracranial vascular malformations and their associated symptoms usually relate to the location of the fistulas and venous drainage patterns.[13,15] However, some intracranial DAVFs may remain asymptomatic or spontaneously regress.[4]

Spontaneous regression of DAVFs was classically thought to be associated with less aggressive lesions (Borden I, Cognard IIa or lower), yet recent studies have highlighted that more aggressive DAVFs may regress as well.[15] While a variety of factors have been theorized to contribute to the spontaneous disappearance of DAVFs, those mechanisms remain a matter of speculation. To better understand the current theories explaining spontaneous regression and the factors that may precipitate this phenomenon, we performed a systematic literature review and present an institutional case of a ruptured high-risk DAVF that spontaneously regressed at 1-year repeat angiogram before neurovascular intervention.

SYSTEMATIC REVIEW METHODOLOGY

We conducted a systematic review of the literature regarding the spontaneous resolution of DAVFs in accordance with the guidelines set forth by the Preferred Reporting Items for Systematic reviews and Meta-Analyses statement.[23] The systematic review was performed using PubMed, from date of database inception to February 2023, combining the terms (Dural OR intracranial) AND arteriovenous AND ("fistula" OR "fistulas" OR "malformation" OR "malformations" OR "shunt") AND (spontaneous OR natural OR conservative OR isolated OR angiographic) AND (closure OR closed OR obliteration OR regression OR disappearance OR conversion OR thrombosis). Two independent authors (HC and JK) screened abstracts and, subsequently, evaluated full-text articles; a third author resolved discrepancies (MS). We included articles if they included the spontaneous regression of intracranial DAVFs. We excluded all non-English articles and articles that did not involve intracranial DAVFs or their spontaneous regression. Baseline demographic statistical analysis was performed in R (R Core Team, 2020); a $P < 0.05$ was set as the threshold for statistical significance.

RESULTS

Case report

A 54-year-old woman with a history of motor vehicle accident at the age of 4 years, complicated by traumatic brain injury requiring craniectomy and evacuation of hematoma, presented to an outside institute with an acute episode of vomiting associated with elevation in blood pressure, right-sided weakness, intermittent diplopia, and seizure. The patient was admitted to the intensive care unit and then discharged after a week but suffered from a repeat episode of seizure 2 months later with new onset receptive and expressive aphasia.

A brain magnetic resonance imaging (MRI) revealed hyperintensity in the left temporal and parietal lobes, which was thought to be caused by congestion secondary to venous hypertension. Blood products were also noted on the left parietal lobe with gyral enhancement, presumably from prolonged venous congestion with cortical vein/dural thrombosis or hemorrhage in the past. Notably, the superior sagittal sinus (SSS) filled irregularly and was suggestive of a thrombotic process.

A cerebral angiogram was then performed which revealed a DAVF (Borden II – Cognard IIa+b) involving the middle to posterior thirds of the SSS supplied by multiple arterial sources [Figure 1]. Notable fistulous connections included a connection from the right superficial temporal artery (STA) with distal dural connections into the middle third of the SSS with significant retrograde flow into the left vein of Trolard. In addition, the middle third of the SSS was also supplied by the right and left middle meningeal artery (MMA) and the left anterior falcine artery. Two more fistulous connections were noted: one was at the posterior third of the SSS (supplied by the posterior meningeal artery from the left posterior inferior cerebellar artery) and another at the junction between the middle and posterior third of the SSS (supplied by the left occipital artery and the occipital branch of the left MMA). Significant cortical venous drainage and occlusion of the SSS were noted as well.

Brain computed tomography (CT) 1 month later demonstrated chronic infarct of the left parietal lobe with cortical laminar necrosis in both the left parietal and temporal lobes. These results remained stable at 2-month and 6-month repeat brain CTs. The patient was started on dabigatran for her venous sinus thrombosis based on decision from the patient's outside neurologist and interventionalist.

When the patient presented to our institute 1 year later never having received treatment for the DAVF, she had residual weakness on the right side, mild gait imbalance, and short-term memory deficit. Given the complexity of the DAVF and the patient's age, alongside history of stroke/hemorrhage, the decision was made to proceed with a repeat cerebral angiogram to evaluate for changes in the angioarchitecture of the DAVF in an effort to plan for neurovascular intervention. However, on repeat angiogram, there was no evidence of fistula noted; the DAVF had self-obliterated without treatment. The SSS remained occluded, and drainage was

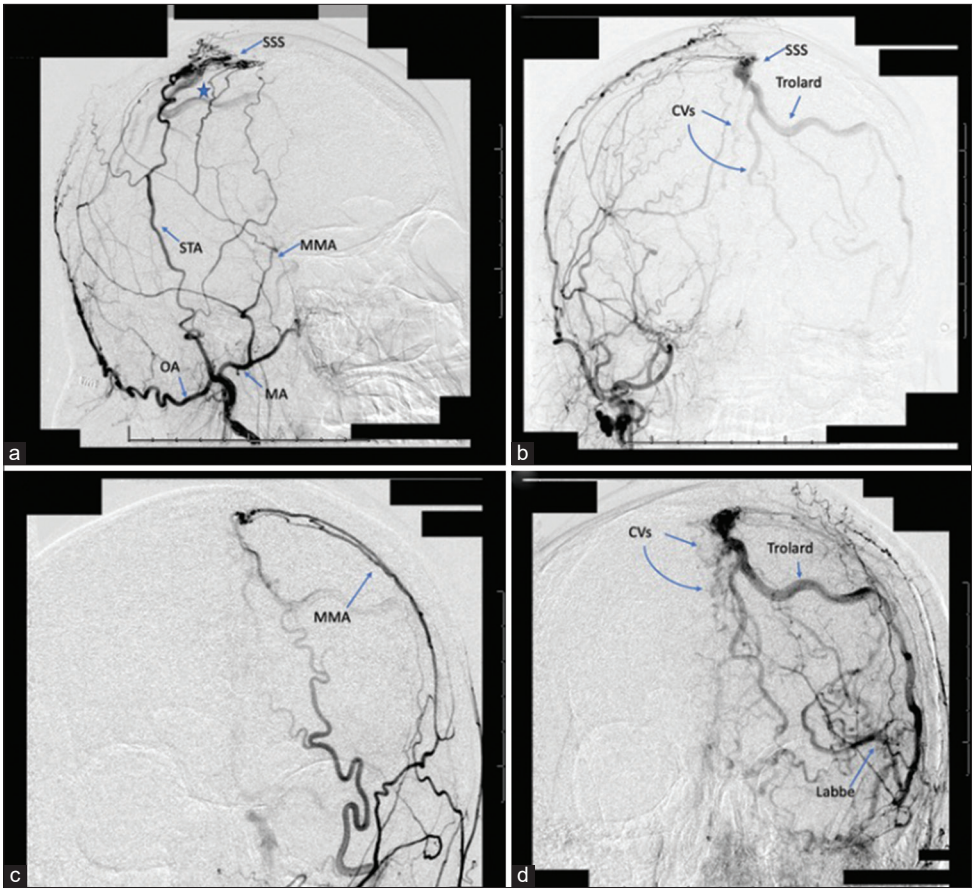

Figure 1: A 54-year-old woman with a dural arteriovenous fistula (DAVF) involving the superior sagittal sinus (SSS). (a) Right external carotid artery (ECA) angiogram, arterial phase, demonstrating a long segment DAVF (Borden II – Cognard IIa+b) involving the middle to posterior thirds of the SSS with supply from the right superficial temporal artery and right middle meningeal artery (MMA) into the middle third of the SSS (star). (b) Right ECA angiogram, late arterial phase, demonstrating reflux into cortical veins (CVs) and occlusion of the SSS. (c) Left common carotid injection, arterial phase, demonstrating a fistulous connection between the left MMA and the middle third of the SSS. (d) Left common carotid injection, venous phase, revealing reflux into CVs and the left vein of Trolard. Labbe: Inferior anastomotic vein of Labbe, MMA: Middle meningeal artery, OA: Occipital artery, STA: Superficial temporal artery.

noted through extensive dilatated cortical veins as collaterals [Figure 2]. The patient was then referred to ophthalmology and neurology for the assessment of papilledema and potential need for shunt placement, along with plans for repeat MRI/magnetic resonance angiography (MRA) at 3-month follow-up.

REVIEW OF THE LITERATURE

A systematic review was conducted to examine other cases of intracranial DAVFs with spontaneous resolution in the scientific literature. Our search querying PubMed yielded 1338 journal articles. Of our results, we included 26 articles detailing 54 cases of intracranial DAVFs spontaneously regressing [Figure 3 and Table 1]; details of each case and their respective clinical courses may be found in Supplementary Table 1.[2,3,5-8,10-12,14-22,24-31] The mean age was 50.29 years (range: 19–57 years) with equal male/female prevalence (27:27 cases each). The average lapse in time from initial diagnosis of DAVF to regression was 33.59 months (range: 0–240 months). There were no significant associations between sex and age with the time lapse between initial diagnosis of DAVF to regression among included cases ($P = 0.645$, $P = 0.414$, respectively, linear regression).

Based on Borden classification, 57.14% of cases were Borden I, 16.33% were Borden II, and 26.53% were Borden III; 34.48% of DAVFs were Cognard I, 10.34% were Cognard IIa, 10.34% were Cognard IIb, 17.24% were Cognard IIa+b, 20.69% were Cognard III, and 6.90% were Cognard IV. Neither patient age nor sex were found to be associated

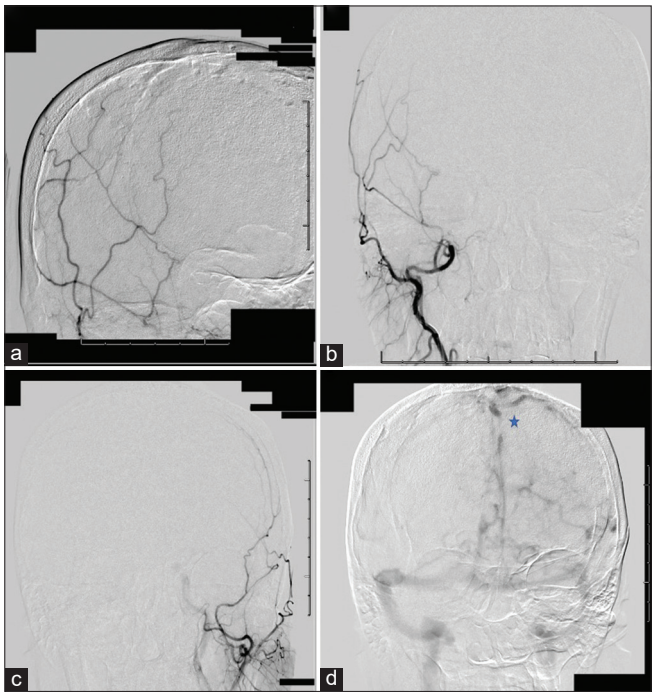

Figure 2: Angiogram performed 1 year later. Right external carotid artery (ECA) angiogram, lateral (a) and anterior-posterior (b) views, demonstrating patent filling of the right ECA and its branches. No evidence of early venous drainage to suggest dural arteriovenous fistula (DAVF). (c) Common carotid injection, arterial phase, revealing normal filling and no evidence of fistula. (d) Left internal carotid injection, venous phase, demonstrating no evidence of DAVF. Superior sagittal sinus thrombosis and venous engorgement of cortical veins (star) are observed.

with Borden ($P = 0.754$, age; $P = 0.818$, sex; ordinal logistic regression) or Cognard distributions ($P = 0.554$, age; $P = 0.7$, sex; ordinal logistic regression). The most frequently involved sinus was the transverse sinus (38.89% of cases, $n = 21$); the SSS was involved in 7 cases (12.96%). Intracranial hemorrhage was observed at the time of DAVF diagnosis in 13 cases (24.1%). Neither involvement of the SSS nor rupture status had significant associations with lapse between DAVF diagnosis and spontaneous regression ($P = 0.626$, $P = 0.127$, respectively, linear regression). The relationship between hemorrhagic or ruptured presentation and higher-risk lesions (Borden II or greater, Cognard IIb or greater) had a trend toward significance ($P = 0.063$, logistic regression).

Of the included cases that proposed a reason for spontaneous regression, the most commonly cited factors were thrombosis of involved sinus/chronic inflammatory changes and fibrosis (24.07% of reported cases, $n = 13$) and arteriography/attempted neurovascular intervention-associated reasons (18.52% of reported cases, $n = 10$). Of all included cases, 50% ($n = 27$) identified no definitive factors responsible for spontaneous regression of DAVF. Of cases that reported sinus status (patent vs. occluded) at the time of DAVF diagnosis and the time of regression ($n = 24$), 41.67% of cases ($n = 10$) reported sinus patency throughout the period when DAVF regression occurred, 29.17% ($n = 7$) reported persistent sinus occlusion, 25% of cases ($n = 6$) demonstrated recanalization of involved sinus, and 4.17% ($n = 1$) documented new onset sinus occlusion at the time of DAVF regression.

DISCUSSION

From our literature review, the cases documenting the spontaneous regression of intracranial DAVFs were nearly balanced between low-risk lesions (57.14% Borden I, 44.82% Cognard IIa or lower) and aggressive high-risk lesions (42.58% Borden II or greater and 55.18% Cognard IIb or greater). This supports the notion that lower risk DAVFs do not necessarily portend to higher rates of spontaneous regression.[15] Ruptured status or intracranial hemorrhage was documented in 24.1% of all included cases from our systematic search; however, the majority of which (69.2%) were in cases of high-risk lesions. The subset of ruptured cases had a varying length of time between DAVF diagnosis to eventual spontaneous regression, ranging from 0 days to 5 years. DAVFs involving the SSS only represented 12.96% of all cases and demonstrated a similar range of time between diagnosis and regression, ranging from 0 days to 7 years. The subgroup of DAVFs of the SSS was all high-risk lesions and 71.4% of them had a ruptured or hemorrhagic presentation. The association between high-risk DAVFs and ruptured or hemorrhagic presentation exhibited a trend but failed to reach statistical significance. However, neither SSS involvement nor rupture/hemorrhagic presentation had statistically significant associations with the length of time to spontaneous regression, suggesting that other factors might be involved in the mechanism behind regression.

Several mechanisms underlying the spontaneous regression of DAVFs have been proposed in the literature. Magidson and Weinberg initially proposed the involvement of sinus thrombosis in the mechanism for the spontaneous regression of a Borden I DAVF involving the transverse sinus in 1976.[21] From the results of our systematic review, 12 other cases also posit a similar mechanism as the sole or primary factor behind the disappearance of their respective DAVFs.[3,8,14,16,17,24,25,28,30] In a report by Kannath *et al.*,[14] the authors speculated that DAVFs with sparse angioarchitectural networks were associated with spontaneous thrombosis and regression of DAVFs; similarly, Voormolen *et al.*[30] posited that small fistulas with single draining veins were more susceptible to venous outflow obstruction and subsequent lesion thrombosis.

A report by Kataoka and Taneda[16] detailed the role of fistula tortuosity, suggesting that the prothrombotic environment due to intravascular turbulence promoted spontaneous regression

Table 1: Literature review on the spontaneous regression of intracranial DAVFs.

Author	Year of study	Case	Age (years)/Sex	Feeders/ Draining veins	Borden/ Cognard classification	Time lapse from DAVF diagnosis to regression	Sinus patency at diagnosis/ follow-up	Posited mechanism of regression
Ahn *et al.*	2003	1	45/F	OA, MMA, APA/TS	Borden I - Cognard I	1 year	Patent/Patent	Thrombosis during intervention; loss of tumor angiogenic stimulators
Al-Afif *et al.*	2014	1	73/F	OA, MMA, APA, MHA/ TS, cerebral veins	Borden II - Cognard IIa+b	8 weeks	Occluded (SS)/ Occluded	Gradual closure noted on each subsequent angiography until complete closure on week 8; elevated intracranial pressure and compression by hematoma + hemodynamic changes from intervention + thrombogenic potency of the contrast agents during angiographies
Basheer *et al.*	2008	1	25/M	MMA/ subarachnoid veins, TS	Borden II	1 year	Patent/Patent	Intrinsic compression of the arteriovenous shunts within the sinus wall
Beniwal *et al.*	2019	1	19/M	OA, MMA, APA, MHA/ TS, CV (right and left DAVF)	Borden II - Cognard IIa+b	3 months	Occluded (TS, SSS)/occluded	Potentially post-traumatic
Blomquist *et al.*	1998	1	43/F	APA, PAA/ epidural venous plexus	Borden I	3 days	NR	Thrombosis after angiography
Chaudhary *et al.*	1982	1	56/F	OA, MMA/ SS (initial); OA, APA/ paravertebral venous plexus (at 1 year FU)	Borden I - Cognard I	30 months	Irregular/ Irregular	Gradual fibrosis of the intraluminal thrombus and inflammation of sinus walls
		2	50/M	MMA/LS	Borden I - Cognard I	1 month	Irregular/ Irregular	Gradual fibrosis of the intraluminal thrombus and inflammation of sinus walls
		3	38/M	OA, MMA, APA/CV (SSS)	Borden III - Cognard III	2 years	Patent/Patent	Thrombosis and occlusion at junction with sinus

(Contd...)

Table 1: *(Continued).*

Author	Year of study	Case	Age (years)/Sex	Feeders/ Draining veins	Borden/ Cognard classification	Time lapse from DAVF diagnosis to regression	Sinus patency at diagnosis/ follow-up	Posited mechanism of regression
Clarençon *et al.*	2013	1	45/M	MMA/LS	Borden I - Cognard IIa	3 months	Irregular/ Patent	Unique arterial feeder compression by hematoma
		2	60/M	OA, MMA, APA/LS, CV	Borden II - Cognard IIb	3 months	Irregular/ occluded	Potentially associated with DSA and contrast media
		3	61/M	PMA/CV	Borden III - Cognard III	6 months	NR	Unique arterial feeder compression by hematoma; potentially associated with DSA and contrast media
Endo *et al.*	1979	1	26/F	OA, MMA/ TS, SS, CV	Borden II - Cognard IIa+b	2 years	NR	NR
Hansen and Sogaard	1976	1	23/F	OA, VA/Torc	Borden I - Cognard IIa	15 months	NR	NR
Kannath *et al.*	2017	Overall series summary (*n*=5)	45.8 (mean)/ 3:2 (M:F)	Sparse network noted in 80% of cases	Borden II (20% of cases), III (80%) - Cognard IIb (40% of cases), IIa+b (20%), III (40%)	49.2 days (mean)	NR	Sparse network promoting stasis and thrombosis of DAVF
		1	36/F	MMA/TSJ, CV	Borden III - Cognard IIb	10 days	NR/Occluded	
		2	54/M	OA, MMA/ TS, CV	Borden II - Cognard IIa+b	6 weeks	NR/Occluded	
		3	NR	NR	Borden III	4.75 months	NR	
		4	NR	TSJ	Borden III	1.75 months	NR	
		5	NR	TSJ	Borden III	17 days	NR	
Kashiwagi	2020	Overall series summary (*n*=9)	66 (mean)/ 4:5 (M:F)	TSJ (6), ACC (3)	Borden I (5), III (1) NR (3) - Cognard I (3), Iia (2), III (1), NR (3)	<3 months (2), 3 months–1 year (2), >1 year (5)		Noninvasive nature of 3D TOF MRA enabled the authors to do more frequent imaging of all patients and could explain the higher rate of spontaneous closures; lower sensitivity of 3D TOF MRA, compared to arteriography, potentially led to missed residual small/low-flow DAVFs

(Contd...)

Table 1: *(Continued).*

Author	Year of study	Case	Age (years)/Sex	Feeders/ Draining veins	Borden/ Cognard classification	Time lapse from DAVF diagnosis to regression	Sinus patency at diagnosis/ follow-up	Posited mechanism of regression
		1	87/F	OA, APA/SS, CV	Borden III - Cognard III	4 years	Occluded (TSJ)/occluded (TSJ)	
		2	67/F	APA/ACC	Borden I - Cognard I	5 weeks	NR	
Kataoka and Taneda	1984	1	56/M	OA/TS (DAVF 1); STA/SSS, superficial cerebral vein (DAVF 2)	Borden I (DAVF 1); Borden II (DAVF 2)	59 weeks	Occluded/ occluded (TS); patent/patent (SSS)	Venous sinus thrombosis and prothrombotic intravascular turbulence from elongation and tortuosity of fistula
Kim *et al.*	2010	1	47/M	TS	Borden I	76 months	Patent/NR	Sinus thrombosis/ occlusion
		2	62/M	TS	Borden I	140 months	Patent/NR	
		3	66/M	CS	Borden I	23 months	Occluded/NR	
		4	63/F	CS	Borden I	112 months	Patent/NR	
		5	62/M	CS	Borden I	74 months	Occluded/NR	
		6	41/F	TS	Borden I	55 months	Irregular/NR	
		7	51/M	NR	Borden I	12 months	Patent/NR	
		8	37/F	TS	Borden I	36 months	Patent/NR	
		9	52/F	JB	Borden I	17 months	Patent/NR	
		10	38/F	NR	Borden I	8 months	Patent/NR	
		11	42/M	CS	Borden I	22 months	Occluded/NR	
		12	43/M	NR	Borden III	114 months	Patent/NR	
		13	45/M	SSS	Borden III	1 month	Patent/NR	
		14	60/F	TS	Borden I	119 months	Patent/NR	
Kutluk *et al.*	1991	1	51/F	OA, tentorial arteries/TS, sylvian veins	Borden II - Cognard IIa+b	12 months	Occluded/ Patent	Potentially related to sinus recanalization
Landman and Braun	1985	1	60/F	APA/JV	Borden I - Cognard I	6 weeks	Patent/Patent	Venous stasis and subsequent thrombosis secondary to patient positioning during imaging studies
Luciani *et al.*	2001	1	42/M	Bilateral OA, MMA, PMA/ TS, Torc	Borden I - Cognard I	14 years	Patent/Patent	Compression of arteriovenous shunts in sinus walls
		2	57/F	MMA, PMA, VA/TS	Borden I - Cognard I	20 years	Patent/Patent	Compression of arteriovenous shunts in sinus walls
		3	28/M	MHA/SPS	Borden I - Cognard I	1 year	Occluded (TS)/Patent	Post-traumatic (e.g., fibrosis, inflammation)
Magidson and Weinberg	1976	1	39/F	OA, MMA, MHA/TS	Borden I	29 months	Patent/ Occluded (TS)	Spontaneous sinus thrombosis

(Contd...)

Table 1: *(Continued).*

Author	Year of study	Case	Age (years)/Sex	Feeders/ Draining veins	Borden/ Cognard classification	Time lapse from DAVF diagnosis to regression	Sinus patency at diagnosis/ follow-up	Posited mechanism of regression
Manabe *et al.*	2008	1	62/M	OA, APA, VA/ ACC, JV	NR	3 months	NR	NR
		2	59/F	OA, APA, VA/ ACC, JV	NR	3 months	NR	NR
		3	67/F	OA, APA, VA/ ACC, JV, IPS	NR	1 month	NR	NR
		4	59/M	APA/ACC, JV	NR	2 months	NR	NR
Olutola *et al.*	1983	1	50/M	MMA/ pterygoid plexus	Borden I	4 months	Patent/Patent	Thrombosis of DAVF
Pritz and Pribram	1991	1	44/F	OA/CV, SSS	Borden III	7 years	Occluded (TS, SS)/Patent (TS)	Spontaneous thrombosis of DAVF
Reul *et al.*	1993	1	24/M	OA/olfactory and CV, basal vein Rosenthal, SSS	Borden III	0 days	Patent/Patent	Occurred during angiography
Saito *et al.*	2008	1	60/M	OA, MMA, APA, tentorial arteries/TS	Borden I - Cognard I	5 years	Patent/Patent	Potentially due to flow changes from reduced supply via external carotid (occipital and ascending pharyngeal) coupled with new supply from internal carotid (tentorial branches)
Tsuji *et al.*	2014	1	61/M	APA/ACC	Borden I - Cognard I	5 days	NR	Potentially due to vascular endothelial injury and prothrombotic effect of ionic/ hyperosmolality of undiluted gadoterate meglumine
van Beijnum *et al.*	2010	1	62/M	OA, MMA/ CV, SSS	Borden II - Cognard IIb	NR	Occluded/ Partial recanalization	The treatment of polycythemia and Factor V Leiden led to the resolution of prothrombotic state and subsequently the recanalization of the SSS, resulting in the regression of DAVF

(Contd...)

Table 1: *(Continued).*

Author	Year of study	Case	Age (years)/Sex	Feeders/ Draining veins	Borden/ Cognard classification	Time lapse from DAVF diagnosis to regression	Sinus patency at diagnosis/ follow-up	Posited mechanism of regression
Voormolen *et al*.	2009	1	53/F	OA, MMA/ osteodural vein, CV	Borden III - Cognard IV	0 days	Occluded (TS, SS)/NR	Possibly due to small fistulas with single draining veins leading to venous outflow obstruction and fistula thrombosis, also might be associated with thrombogenic effects of contrast medium during DSA
		2	63/F	Ethmoid branches of bilateral OtA, ECA/ osteodural vein, CV, SSS	Borden III - Cognard IV	Days (unspecified)	NR	Possibly due to small fistulas with single draining veins leading to venous outflow obstruction and fistula thrombosis, also might be associated with thrombogenic effects of contrast medium during DSA
Warren *et al*.	2010	1	51/F	OA, MMA, bilateral PMA/TS (contralateral)	Borden I - Cognard IIa	10 years	Occluded (SS)/ Patent	Recanalization of sigmoid sinus reduced the venous hypertension that was maintaining the patency of the DAVF, promoting DAVF regression

ACC: Anterior condylar confluence, APA: Ascending pharyngeal artery, AVF: Arteriovenous fistula, CS: Cavernous sinus, CV: Cortical veins, DAVF: Dural arteriovenous fistula, DSA: Digital subtraction angiography, ECA: External carotid artery, FU: Follow-up, ICA: Internal carotid artery, IPS: Inferior petrosal sinus, JB: Jugular bulb, JV: Jugular vein, LS: Lateral sinus, MHA: Meningohypophyseal artery, MMA: Middle meningeal artery, NR: Not reported, OA: Occipital artery, OtA: Ophthalmic artery, PAA: Posterior auricular arteries, PMA: Posterior meningeal artery, SPS: Superior petrosal sinus, SS: Sigmoid sinus, SSS: Superior sagittal sinus, STA: Superficial temporal artery, Torc: Torcular, TS: Transverse sinus, TSJ: Transverse-sigmoid junction, VA: Vertebral artery, 3D TOF MRA: 3-dimensional time-of-flight magnetic resonance angiography, n: Sample size, M: Male, F: Female, number in bracket/parentheses: number of cases

of DAVFs. Chaudhary *et al.*[8] built upon the concepts discussed by Magidson and Weinberg and posits that, in addition to the initial thrombus, the gradual fibrosis of the intraluminal thrombus with chronic inflammation and stenosis of the sinus led to the eventual occlusion and regression of the DAVFs in their two cases. It is worth noting that the involved sinus in those two cases remained irregular/occluded throughout the period of DAVF disappearance (time lapse ranging from 1 month to 30 months). Finally, Luciani *et al.*[20] propose that post-traumatic DAVFs – particularly those with small or sparse angioarchitecture – are more likely to spontaneously regress.

Another commonly cited mechanism for spontaneous regression involves attempted neurovascular intervention or the impact of angiography and associated contrast medium. From our systematic review, the two main suggested mechanisms involve either direct endothelial injury and endoluminal stasis induced by neurosurgical intervention or thrombogenic effects of contrast medium during imaging.[2,3,7,10,16,26,28,30] A prototypical case reported by Al-Afif *et al.*[3] documented both mechanisms in conjunction leading to the regression of the DAVF in their case: hematoma evacuation led to hemodynamic changes and partial closure

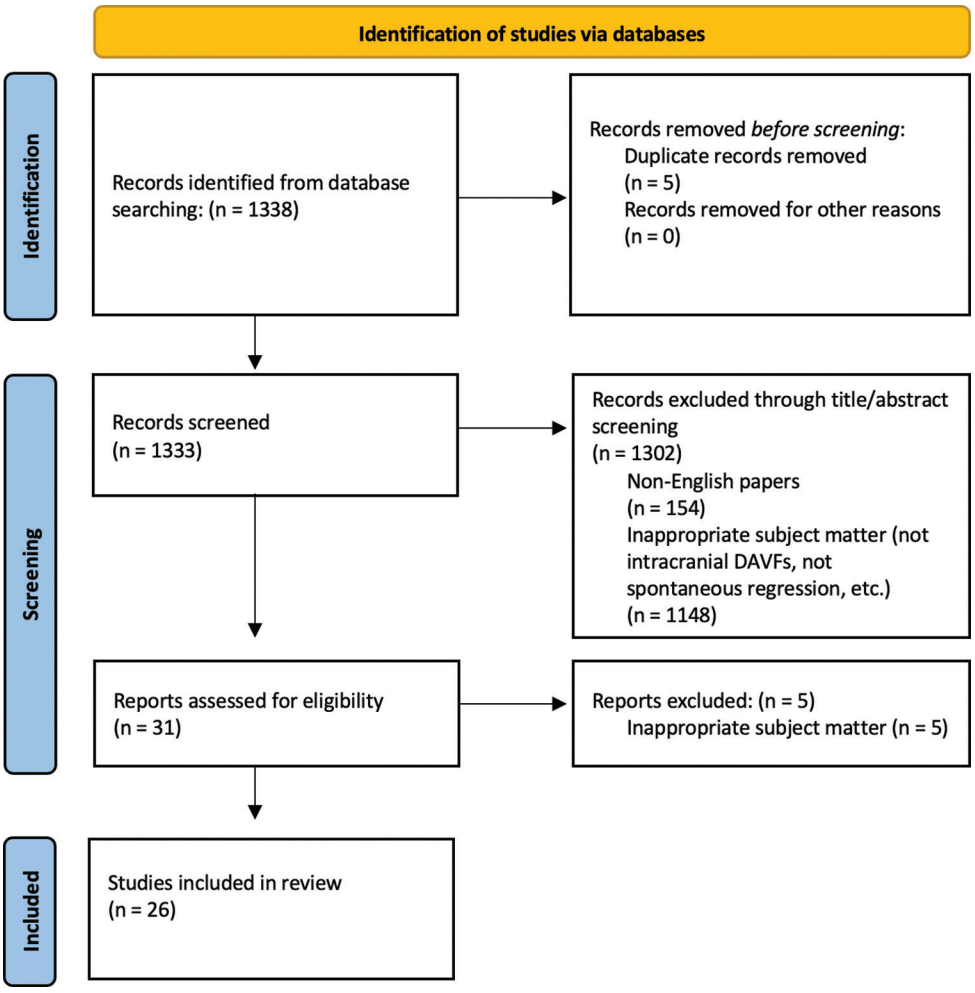

Figure 3: Flow diagram depicting literature review process according to Preferred Reporting Items for Systematic reviews and Meta-Analyses guidelines. Total studies included: 26. n: number of studies, DAVF: dural arteriovenous fistula

of DAVF feeders, and subsequent follow-up angiographs contributed to chronic thrombosis and eventual regression. Selective catheterization of feeding arteries with resultant blood flow stagnation and endothelial damage during angiography[2,3,30] or attempted endovascular intervention[10] have also been cited as potential precipitators of DAVF closure. Landman and Braun also proposed that in cases with drainage into the jugular vein, hyperextension of a patient's neck may lead to functional obstruction of the jugular vein, resulting in stasis of venous blood due to compression and subsequent thrombosis of DAVF.[19]

The thrombogenic effects of contrast media have also been documented and emphasized in the cases of DAVFs with unique feeding arteries or those with a small nidus and single draining vein,[10,30] which is thought to combine synergistically with the prothrombotic hemodynamic changes that occur during the angiography leading to the eventual closure of DAVFs.[3] Spontaneous closures of DAVFs have been documented to occur as rapidly as the same day of/during angiography itself[26,30] or even following extracranial angiography (i.e., cardiac catheter angiography).[27] These principles were built upon in the case reported by Tsuji *et al.*,[28] who posited that the undiluted gadoterate meglumine they utilized in their case – due to patient allergy with iodine contrast – further promoted thrombosis due to its ionic and hyperosmotic nature. Ionic contrast may lead to cytotoxicity and hyperosmolality may lead to crenation, damage, and apoptosis of endothelial vascular cells, resulting in platelet deposition and further promoting a thrombogenic environment.

Compression of DAVF feeders or shunts by a hematoma, hematoma-associated mass effect, or edema has also been proposed as a possible mechanism for spontaneous DAVF occlusion.[10] Compression of arteriovenous shunts in the involved sinus walls is another mechanism that has been suggested to be associated with spontaneous closure of DAVFs, namely, that focal expansion in size of the sinus

could compress the DAVF shunt connections.[5,15,20] This is distinct from the mechanism proposed by Warren *et al.*[31] and also observed in Kutluk *et al.*:[18] The recanalization of the involved sinus in their cases was posited to have resolved the venous hypertension that was necessary in keeping their respective DAVFs patent, without which their cases each, respectively, regressed. This phenomenon is also based on the idea that venous hypertension decreases cerebral perfusion, inducing angiogenesis and further promoting the formation of DAVFs.[29] It is conceivable that venous sinus pressures and conditions have a complex interaction with DAVF pathogenesis and regression.

An interesting case was reported by van Beijnum *et al.*[29] The authors documented a satisfactory spontaneous regression of a DAVF following the treatment of polycythemia and Factor V Leiden in a patient. The authors speculate that the resolution of the prothrombotic state, and subsequently the recanalization of the involved sinus, resulted in the regression of the patient's DAVF.[29] This mechanism closely aligns with the principles outlined by Kutluk *et al.* as discussed above.[18,29] Saito *et al.*[27] also posit that DAVFs are dynamic, with continually changing arterial flow and arteriosinus channels as a part of their natural history, which may be responsible for their development and spontaneous closure. This is an interesting conjecture as it would explain the heterogenicity of mechanisms proposed for DAVF spontaneous regressions.

In the cases that involved the SSS ($n = 7$), the most often cited mechanism remained venous sinus or arteriosinus shunt thrombosis ($n = 4$), followed by angiography or contrast media-related thrombogenic effects ($n = 2$). Of note, two of those cases speculated that angioarchitecture played a contributing role in the spontaneous occlusion of their DAVFs: Intravascular turbulence from fistula tortuosity[16] and small fistulas with single draining veins,[30] respectively. Finally, one report (van Beijnum *et al.*[29]) discussed the impact of polycythemia and Factor V Leiden prothrombotic states as discussed above.

In the present case, our patient suffered a traumatic brain injury from a motor vehicle accident as a child, requiring a craniectomy and hematoma evacuation that likely led to the occlusion of the SSS. The resultant venous congestion/hypertension and infarct of the parietal lobe likely promoted DAVF angiogenesis and development. Subsequently, the chronic venous hypertension likely contributed to maintaining the patency of the DAVF. It is also possible that the venous sinus hypertension ultimately played a role in compressing shunt connections, leading to spontaneous regression; however, this mechanism is less probable given our patient's extensive and bilateral angioarchitecture network. Therefore, if venous hypertension was responsible for the maintenance of the DAVF in our patient, we considered the possibility that the initiation of dabigatran and subsequent resolution of the SSS occlusion could explain the regression of the DAVF. However, the SSS remained occluded at repeat angiography, suggesting that recanalization was not the mechanism behind our patient's DAVF regression. It is possible that the thrombus occluding the SSS gradually fibrosed and led to chronic inflammation and stenosis, leading to the eventual occlusion of the DAVF; the patient's hemorrhagic presentation – with resultant inflammatory and hemodynamic changes – alongside prior angiogram 1 year before presenting to our institute could have also had prothrombotic effects that further acutely contributed to the spontaneous occlusion of the DAVF.

Our case documents a DAVF with an extensive network, involving supply from the internal carotid (anterior falx/falcine artery), vertebral artery (posterior inferior cerebellar artery), and bilateral external carotids (MMA, STA, occipital artery), that spontaneously regressed without a clear identifying mechanism as proposed by the current literature. One other SSS DAVF with bilateral supply was identified in our systematic review; however, the patient had a single draining vein that was speculated to have had a contributing role in the eventual regression of their DAVF.[30] The multitude of factors discussed as potential contributors to the disappearance of our patient's DAVF highlight the principles set forth by Saito *et al.*[27] and the overall heterogenicity of the literature: DAVFs are dynamic and respond variably to each of the mechanisms described thus far in the literature. Moreover, while neurovascular intervention is recommended for aggressive high-risk DAVFs, our patient resided in another country where the fistula was left untreated and she experienced significant delay before arriving at our institute. Our case provides a unique long-term insight into the natural history of an aggressive ruptured DAVF that would normally be treated. During our literature search, we found cases where the duration between the diagnosis of DAVF and spontaneous regression ranged from 10 to 20 years.[17,20,31] However, all of those cases involved Borden I DAVFs and none had ruptured or involved the SSS. Furthermore, our patient's DAVF had an extensive angioarchitectural network which suggests that sparse networks, small fistulas, or single draining veins are not necessarily a requirement for spontaneous regression. Despite the probability of spontaneous regression, conservative measures or observation are not recommended in the management of aggressive DAVFs. It is also worth noting that, in cases involving DAVFs that regress in the setting of venous thrombosis, the most cited mechanism from our systematic review, those patients may develop venous hypertension due to the resultant venous outflow obstruction and therefore should be evaluated for papilledema with consideration of treatment with medical therapy or surgery. The complex interplay of

factors responsible for natural resolution demand further exploration. Identification of reliable predictors of regression could lead to improved treatment strategies and outcomes in this patient cohort.

Several limitations of our study should be acknowledged. First, our review included predominantly case reports and case series due to the low documented incidence of DAVF with spontaneous regression. Furthermore, high-risk DAVFs are generally recommended for intervention and therefore the long-term insight into their natural history is limited in the literature. Second, the limited number of reported DAVF with spontaneous regression and the lack of consistent and detailed reporting among the included studies (e.g., sinus patency before and after spontaneous resolution, and proposed mechanism explaining spontaneous resolution) limited the power of our statistical analysis. Third, the inherent limitations of a systematic review based on published articles in the literature include publication bias.

CONCLUSION

In our systematic review of 26 articles and 54 cases of spontaneous regressions of DAVFs, 57% were Borden I and 43% were Borden II or greater. DAVFs that involved the SSS were all Borden II or greater and 71% of them presented with hemorrhage. The most often proposed mechanisms for spontaneous regression – among the total cohort and the subset of cases involving the SSS – involved sinus thrombosis and chronic inflammatory changes or fibrosis, followed by angiography/attempted neurovascular intervention associated factors. However, there is no consensus in the literature regarding the mechanisms responsible for the natural regression of DAVFs, with 50% of our included cases without an identifiable mechanism proposed. Our report on a 54-year-old woman provides a distinctive and extended perspective on the natural progression of a high-risk ruptured DAVF that remained untreated for a considerable duration. The regression of her DAVF occurred without a clear identifying mechanism as proposed by the current literature. DAVFs are dynamic diseases, and further research into their natural history will be needed to deduce key factors in their spontaneous regressions.

Declaration of patient consent

Patients' consent not required as patients' identities were not disclosed or compromised.

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Conflicts of interest

There are no conflict of interest.

REFERENCES

1. Agnoletto GJ, Hoover JM, Monteiro A, Hanel RA. Catch me if you can: Disappearing and reappearing posterior fossa dural arteriovenous malformation. BMJ Case Rep 2019;12:e229437.
2. Ahn JY, Lee BH, Cho YJ, Joo JY, Lee KS. Dural arteriovenous fistula associated with meningioma: Spontaneous disappearance after tumor removal. Neurol Med Chir (Tokyo) 2003;43:308-11.
3. Al-Afif S, Nakamura M, Gotz F, Krauss JK. Spontaneous closure of a dural arteriovenous fistula. BMJ Case Rep 2014;2014:bcr2014011255.
4. Alkhaibary A, Alnefaie N, Alharbi A, Alammar H, Arishy AM, Alshaya W, *et al.* Intracranial dural arteriovenous fistula: A comprehensive review of the history, management, and future prospective. Acta Neurol Belg 2022;123:359-66.
5. Basheer N, Kasliwal MK, Gaikwad S, Sharma BS. Spontaneous closure of dural arterio-venous fistula. Neurol India 2008;56:207.
6. Beniwal M, Saini J, Somanna S, Deepesh A, Rao KV, Vazhayil V, *et al.* Spontaneous closure of dural arteriovenous fistula; a visual specter. Neurol India 2019;67:1376-9.
7. Blomquist MH, Barr JD, Hurst RW. Isolated unilateral hypoglossal neuropathy caused by dural arteriovenous fistula. AJNR Am J Neuroradiol 1998;19:951-3.
8. Chaudhary MY, Sachdev VP, Cho SH, Weitzner I Jr., Puljic S, Huang YP. Dural arteriovenous malformation of the major venous sinuses: An acquired lesion. AJNR Am J Neuroradiol 1982;3:13-9.
9. Chen PM, Chen MM, McDonald M, McGehrin K, Steinberg J, Handwerker J, *et al.* Cranial dural arteriovenous fistula. Stroke 2018;49:e332-4.
10. Clarençon F, Biondi A, Sourour NA, Di Maria F, Iosif C, Nouet A, *et al.* Spontaneous closure of intracranial dural arteriovenous fistulas: A report of 3 cases. Clin Neurol Neurosurg 2013;115:971-5.
11. Endo S, Koshu K, Suzuki J. Spontaneous regression of posterior fossa dural arteriovenous malformation. J Neurosurg 1979;51:715-7.
12. Hansen JH, Sogaard I. Spontaneous regression of an extra- and intracranial arteriovenous malformation. Case report. J Neurosurg 1976;45:338-41.
13. Iampreechakul P, Wangtanaphat K, Lertbutsayanukul P, Wattanasen Y, Siriwimonmas S. Spontaneous closure of a cavernous sinus dural arteriovenous fistula with spinal perimedullary drainage (cognard V) during attempted transvenous embolization. Asian J Neurosurg 2019;14:1268-74.
14. Kannath SK, Rajan JE, Mukherjee A, Sarma PS. Factors predicting spontaneous thrombosis of aggressive cranial dural arteriovenous fistulas. World Neurosurg 2017;103:821-8.e2.
15. Kashiwagi N, Miyazaki K, Takahashi H, Tsuji K, Fujiwara M, Arisawa A, *et al.* Spontaneous closure of non-cavernous sinus dural arteriovenous fistulas: A case series and systematic review of the literature. J Neuroradiol 2022;49:94-100.

16. Kataoka K, Taneda M. Angiographic disappearance of multiple dural arteriovenous malformations. Case report. J Neurosurg 1984;60:1275-8.
17. Kim DJ, TerBrugge K, Krings T, Willinsky R, Wallace C. Spontaneous angiographic conversion of intracranial dural arteriovenous shunt: Long-term follow-up in nontreated patients. Stroke 2010;41:1489-94.
18. Kutluk K, Schumacher M, Mironov A. The role of sinus thrombosis in occipital dural arteriovenous malformations--development and spontaneous closure. Neurochirurgia (Stuttg) 1991;34:144-7.
19. Landman JA, Braun IF. Spontaneous closure of a dural arteriovenous fistula associated with acute hearing loss. AJNR Am J Neuroradiol 1985;6:448-9.
20. Luciani A, Houdart E, Mounayer C, Maurice JP, Merland JJ. Spontaneous closure of dural arteriovenous fistulas: Report of three cases and review of the literature. AJNR Am J Neuroradiol 2001;22:992-6.
21. Magidson MA, Weinberg PE. Spontaneous closure of a dural arteriovenous malformation. Surg Neurol 1976;6:107-10.
22. Manabe S, Satoh K, Matsubara S, Satomi J, Hanaoka M, Nagahiro S. Characteristics, diagnosis and treatment of hypoglossal canal dural arteriovenous fistula: Report of nine cases. Neuroradiology 2008;50:715-21.
23. Moher D, Liberati A, Tetzlaff J, Altman DG, PRISMA Group. Preferred reporting items for systematic reviews and meta-analyses: The PRISMA statement. PLoS Med 2009;6:e1000097.
24. Olutola PS, Eliam M, Molot M, Talalla A. Spontaneous regression of a dural arteriovenous malformation. Neurosurgery 1983;12:687-90.
25. Pritz MB, Pribram HF. Spontaneous closure of a high-risk dural arteriovenous malformation of the transverse sinus. Surg Neurol 1991;36:226-8.
26. Reul J, Thron A, Laborde G, Bruckmann H. Dural arteriovenous malformations at the base of the anterior cranial fossa: Report of nine cases. Neuroradiology 1993;35:388-93.
27. Saito A, Furuno Y, Nishimura S, Kamiyama H, Nishijima M. Spontaneous closure of transverse sinus dural arteriovenous fistula: Case report. Neurol Med Chir (Tokyo) 2008;48:564-8.
28. Tsuji K, Nakagawa N, Fukawa N, Kato A. Spontaneous closure of a dural arteriovenous fistula immediately after cerebral angiography using a gadolinium contrast agent. J Stroke Cerebrovasc Dis 2014;23:e449-52.
29. Van Beijnum J, Klijn CJ, Lo TH, van der Zwan A, Kappelle LJ. Spontaneous obliteration of a dural arteriovenous fistula after treatment of polycythemia in a patient with factor V Leiden mutation: Case report. J Neurol 2010;257:1573-5.
30. Voormolen V, Geens K, Van Den Hauwe L, Parizel PM. Spontaneous closure of cerebral dural arteriovenous fistulas with direct cortical venous drainage. A case report. Interv Neuroradiol 2009;15:359-62.
31. Warren DJ, Craven I, Romanowski CA, Coley SC. Spontaneous closure of a Type 2a dural arteriovenous fistula following late recanalization of the occluded sinus. Interv Neuroradiol 2010;16:282-5.

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Supplementary Table

Supplementary Table 1: Clinical courses for included cases of spontaneously regressed DAVFs.

Author	Year of Study	Case	Age (years)/Sex	Presentation	Diagnosis	Clinical Course
Ahn *et al.*	2003	1	45/F	Dull headache	Meningioma with coexistent DAVF	Preoperative embolization of anterior branch of left middle meningeal artery + tumor resection; 1 year FU angiography with disappearance of DAVF
Al-Afif *et al.*	2014	1	73/F	Generalized seizure	DAVF	Conservative management, anti-epileptic medication; re-admission 1 week later with large ICH involving temporal, occipital, and parietal lobes requiring evacuation and decompressive hemicraniectomy; 2 weeks later at scheduled embolization for DAVF, decreased flow was observed and intervention was postponed; at 4 week FU, further decrease in DAVF flow noted on angiogram; at 8 week FU angiogram, DAVF had disappeared
Basheer and Kasliwal	2008	1	25/M	Headache, pulsatile tinnitus	DAVF	Planned embolization but patient lost to follow-up; re-presented 1 year later with resolution of tinnitus and residual mild headache
Beniwal *et al.*	2019	1	19/M	Headache, vision disturbance, loss of consciousness	History of TBI 18 month prior to initial presentation, DAVF (bilateral)	Patient was lost to FU after initial TBI; patient re-presented after an unspecified amount of time and DSA revealed bilateral DAVF and patient was scheduled for embolization but was again lost to FU; patient returns 3 months later with worsening headache and DSA at that time demonstrated disappearance of DAVF
Blomquist *et al.*	1998	1	43/F	Right 12th nerve palsy, headache, pulsatile tinnitus	DAVF	Planned endovascular therapy; 24 h after initial diagnosis patient had improvement in headache; repeat angiogram at 72 h demonstrated thrombosis of DAVF and resolution of cranial nerve palsy
Chaudhary *et al.*	1982	1	56/F	Trauma, tinnitus, bruit over mastoid region	Trauma, multiple DAVFs	6 months after initial TBI multiple DAVFs discovered on angiogram during work-up of persistent symptoms, surgical ligation of meningeal branches of the occipital artery and the mastoid emissary vein was performed; at 18 month FU from TBI, arteriography revealed persistent DAVFs; 3 year FU from TBI, arteriography revealed regression of DAVFs

(Contd...)

Supplementary Table 1: *(Continued)*.

Author	Year of Study	Case	Age (years)/Sex	Presentation	Diagnosis	Clinical Course
		2	50/M	Trauma, disorientation	Trauma, scalp AVF, multiple DAVFs	3 weeks after initial TBI, arteriography revealed left frontoparietal SDH and a right scalp AVF during work-up for persistent disorientation, requiring hematoma evacuation; patient was readmitted at 6 months FU following TBI, repeat angiogram demonstrated multiple DAVFs without evidence of previous scalp AVF and the patient declined treatment; patient returned 1 month later (7 month following TBI) for embolization, repeat angiogram revealed regression of DAVFs
		3	38/M	Trauma, seizure	Trauma, temporal fracture, ICH, DAVF	3 weeks after initial TBI, patient developed seizure and workup revealed DAVF, the patient was managed conservatively; angiography at 2 year FU revealed DAVF had closed spontaneously
Clarençon *et al*.	2013	1	45/M	Headache	Temporal hematoma (non-traumatic), DAVF	Patient declined treatment, 3 month FU DSA demonstrated spontaneous closure of DAVF, 6 month FU redemonstrated no residual deficit
		2	60/M	Headache	DAVF	Aborted embolization attempt at 1 month following diagnosis, final control DSA revealed slower venous drainage of DAVF with no spasm of feeders; at 3 month FU complete closure of DAVF was observed
		3	61/M	Headache, dyskinesia, gait disturbance	ICH (non-traumatic), DAVF	Repeat angiogram 15 days after DSA diagnosis of DAVF at scheduled endovascular intervention demonstrated marked slowdown of draining veins, intervention subsequently canceled; FU control DSA at 6 month demonstrated complete closure of DAVF
Endo *et al*.	1979	1	26/F	Tinnitus, vertigo, retroauricular bruit, homonymous hemianopsia, seizure	DAVF	Conservative management, anti-epileptic medication; 2 year FU angiogram revealed complete regression of DAVF, patient's symptoms resolved
Hansen and Sogaard	1976	1	23/F	Headache, bruit	DAVF	Conservative management; 1 year later the patient experienced gradual resolution of bruit over 2 weeks; at 15 month after initial diagnosis, patient was readmitted and repeat arteriography demonstrated complete regression of DAVF with no residual symptoms
Kannath *et al*.	2017	Overall series summary (n=5)	45.8 (mean)/3:2 (M:F)	Hemorrhage in 60% of cases	DAVFs	Thrombosis of draining vein in 40% of cases

(Contd...)

Supplementary Table 1: *(Continued).*

Author	Year of Study	Case	Age (years)/Sex	Presentation	Diagnosis	Clinical Course
		1	36/F	Headache, seizure, tinnitus	Temporo-occipital hematoma, DAVF	20 days after initial presentation and imaging demonstrating ICH, DSA was performed and DAVF was diagnosed; embolization was planned 10 days after DSA, however, repeat angiogram showed spontaneous disappearance of DAVF
		2	54/M	Headache, loss of consciousness	Occipital hematoma, DAVF	Patient consented to intervention at 6 weeks after initial diagnosis of DAVF, at which time control angiogram revealed spontaneous disappearance of DAVF
		3	NR	Seizure, headache	DAVF	NR
		4	NR	Headache, visual blurring	DAVF	NR
		5	NR	NR	Temporal hematoma, DAVF	NR
Kashiwagi	2020	Overall series summary (n=9)	66 (mean)/4:5 (M:F)	Pulsatile tinnitus (7), headache (2), impaired consciousness (1)	DAVF	Symptoms resolved (4), improved (4), persisted (1)
		1	87/F	Headache, loss of consciousness	Occipital hematoma, DAVF	Patient declined treatment, followed with regular 3D TOF MRA; 4 year FU revealed complete closure of DAVF
		2	67/F	Pulsatile tinnitus	DAVF	Patient reported immediate improvement in tinnitus after initial 3D TOF MRA; repeat 3D TOF MRA at 5 weeks demonstrated closer of DAVF
Kataoka and Taneda	1984	1	56/M	Loss of consciousness, upper motor neuron signs	Third and lateral ventricle IVH, tentorium cerebelli ICH, DAVF ×2	25 h after initial presentation, the patient regained consciousness and was neurologically intact at which time angiogram revealed the two DAVF; 18 days later, the right occipital artery was ligated and subsequently the transverse sinus DAVF decreased in size whereas the superior sagittal sinus DAVF increased in size; patient was readmitted 2 years post-operatively, and angiogram revealed disappearance of both DAVFs
Kim *et al.*	2010	1	47/M	Bruit	NR	Resolution of symptoms at final FU
		2	62/M	Bruit	NR	Resolution of symptoms at final FU
		3	66/M	Diplopia, bruit	NR	Improvement of symptoms at final FU
		4	63/F	Bruit	NR	Persistent fluctuating symptoms at final FU
		5	62/M	Headache, vision loss, bruit	NR	Improvement of symptoms at final FU
		6	41/F	Pontine hemorrhage	NR	Improvement of symptoms at final FU
		7	51/M	Bruit, 12th nerve palsy	NR	Persistent fluctuating symptoms at final FU
		8	37/F	Bruit, occipital pain	NR	Resolution of symptoms at final FU
		9	52/F	Bruit	NR	Resolution of symptoms at final FU
		10	38/F	Bruit	NR	Resolution of symptoms at final FU
		11	42/M	Diplopia, bruit	NR	Resolution of symptoms at final FU

(Contd...)

Supplementary Table 1: *(Continued).*

Author	Year of Study	Case	Age (years)/Sex	Presentation	Diagnosis	Clinical Course
		12	43/M	Bruit	NR	Resolution of symptoms at final FU
		13	45/M	Parietal hemorrhage	NR	Improvement of symptoms at final FU
		14	60/F	Bruit	NR	Resolution of symptoms at final FU
Kutluk *et al.*	1991	1	51/F	Headache, pulsatile tinnitus	DAVF	Patient declined treatment initially, elected for embolization at 12 month FU, at which point control angiography revealed spontaneous closure of DAVF
Landman and Braun	1985	1	60/F	Pulsatile tinnitus, bruit	DAVF	6 week FU revealed spontaneous disappearance of DAVF
Luciani *et al.*	2001	1	42/M	Pulsatile tinnitus, bruit	DAVF	12 year FU redemonstrated DAVF during work-up of bilateral tinnitus; at 14 year FU from initial diagnosis of DAVF the patient had resolution of tinnitus and bruit, angiogram revealed disappearance of DAVF
		2	57/F	Pulsatile tinnitus, bruit	DAVF	Patient underwent ×2 embolization, however, post-operative angiogram revealed persistent DAVF supplied by vertibral branches; at 20 month FU repeat angiogram redemonstrated DAVF supplied by vertibral and external carotid arteries; at 17 year FU the DAVF remained unchanged on angiogram, however, patient experience resolution of tinnitus 8 months later; at 20 year FU the patient experienced a headache and angiogram revealed spontaneous closure of DAVF
		3	28/M	Trauma	Trauma, frontonasal hematoma, pneumocephalus, post-traumatic carotid-cavernous fistula, DAVF	Patient improved rapidly from TBI and was discharged; patient presented 5 months later with exophthalmos and work-up revealed carotid-cavernous fistula and a DAVF, subsequently underwent embolization of carotid-cavernous fistula and no intervention for DAVF; repeat control angiogram 1 year later demonstrated closure of DAVF
Magidson and Weinberg	1976	1	39/F	Tinnitus, bruit	DAVF	Patient discharged without intervention, re-presents 29 months later with headache and vertigo, the tinnitus and bruit had abated at this time, and angiogram demonstrated disappearance of DAVF
Manabe *et al.*	2008	1	62/M	Tinnitus	Hypoglossal canal DAVF	Previous trauma >10 years prior
		2	59/F	Tinnitus	Hypoglossal canal DAVF	Previous trauma 1 month prior
		3	67/F	Tinnitus	Hypoglossal canal DAVF	Previous trauma 1 day prior
		4	59/M	Tinnitus	Hypoglossal canal DAVF	Previous trauma 5–6 years prior
Olutola *et al.*	1983	1	50/M	Headache, vomiting, pulsatile tinnitus, nuchal rigidity, temporal bruit	Epidural hematoma, DAVF	Initially managed conservatively; 4 month FU angiogram revealed spontaneous closure of DAVF

(Contd...)

Supplementary Table 1: *(Continued)*.

Author	Year of Study	Case	Age (years)/Sex	Presentation	Diagnosis	Clinical Course
Pritz and Pribram	1991	1	44/F	Tinnitus	DAVF	No initial intervention; repeat angiogram at 7 year FU demonstrated spontaneous closure, patient remained with persistent tinnitus, episodic loss of consciousness responsive to anticonvulsant therapy, memory difficulties
Reul *et al.*	1993	1	24/M	Headache, seizure, memory disturbance	Frontobasal ICH, DAVF	DAVF closed spontaneously during angiography
Saito *et al.*	2008	1	60/M	Headache, nausea, diplopia	Right lateral ventricle IVH, DAVF	Initially presented with IVH managed conservatively and eventually discharged; 2 years after, patient presented with right 4th nerve palsy and angiogram revealed DAVF, managed conservatively; 2 more years later (4 years from initial admission), patient presented with pulsatile tinnitus and repeat angiogram re-demonstrated DAVF but now with additional feeders from the internal carotid; 1 year later patient underwent cardiac cath, and subsequently 1 month later his tinnitus resolved, and angiography revealed disappearance of DAVF
Tsuji *et al.*	2014	1	61/M	Tinnitus, bruit	DAVF	Angiography was performed with gadolinium due to patient's history of anaphylactic shock with iodine contrast, immediately the patient reported resolution of tinnitus; 5 days later, MRS showed disappearance of DAVF
van Beijnum *et al.*	2010	1	62/M	Episodes of visual disturbance, headache, dysesthesia, generalized shaking (EEG negative)	Polycythemia, Factor V Leiden, SAH, DAVF	CT revealed SAH and DSA demonstrated DAVF; patient underwent phlebotomy until hemoglobin and hematocrit returned to normal limits; embolization was planned for the DAVF, however, on localizing DSA the DAVF was found to be occluded
Voormolen *et al.*	2009	1	53/F	Headache, visual loss, aphasia, hemiparesis	Transverse sinus thrombosis, DAVF	Several hours post-DSA, patient experienced sudden headache, aphasia, hemiparesis; patient declined repeat DSA, but MR imaging was suggestive of DAVF occlusion; 3 months after, control DSA confirmed occlusion of DAVF
		2	63/F	Auditory disturbances, dizziness	DAVF	Several days after DSA, patient experienced sudden onset headache without neurological deficit; subsequently at the time of endovascular intervention, pre-embolization DSA revealed occlusion of DAVF
Warren *et al.*	2010	1	51/F	Bilateral tinnitus, headache, bruit	Sigmoid sinus thrombosis, DAVF	Conservative management; patient re-presents at 10 year FU with occipital headaches, which abate along with the tinnitus after 9 days, repeat angiogram demonstrates occlusion of DAVF

3D TOF MRA: 3-dimensional time-of-flight magnetic resonance angiography, AVF: Arteriovenous fistula, DAVF: Dural arteriovenous fistula, DSA: Digital subtraction angiography, FU: Follow-up, ICH: Intracerebral hemorrhage, IVH: Intraventricular hemorrhage, NR: Not reported, SAH: Subarachnoid hemorrhage, SDH: Subdural hematoma, TBI: Traumatic brain injury, CT: Computed tomography, MRS: Magnetic resonance spectroscopy, M: Male, F: Female.