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Review Article

Intracranial Dural Arteriovenous Fistulas: A Brief Review on Classification and General Features

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ABSTRACT

Intracranial dural arteriovenous malformations are unique and rare pathological entities. They may present with a wide spectrum of clinical pictures and the natural disease course is also variable. The main determinant in prognosis and indication for and response to treatment is the venous architecture of the fistula. In this paper, two widely accepted classifications of dural arteriovenous fistulas, namely, a classification according to venous drainage and another one according to anatomic location will be discussed with relevant clinical and therapeutic implications.

KEY WORDS: Dural arteriovenous fistula, classification, endovascular treatment, surgery

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INTRODUCTION

Dural arteriovenous fistulas are abnormal arteriovenous connections within the dura and are usually located within the walls of a dural sinus or an adjacent cortical vein (3). In a sense, they may represent an arteriovenous malformation with the nidus inside the dura mater. The terminology concerning this malformation has been somewhat controversial. Many authors have preferred the term 'dural arteriovenous malformation' (DAVM) because it suggests a congenital etiology and also covers the frequently complex angioarchitecture involving many arterial feeders. Others have used 'dural arteriovenous fistula' (DAVF) implying an acquired nature for which there is cumulating evidence that will be discussed later in this text (5). In this paper, the term DAVF will be used.

DAVF comprise 10-15% of all intracranial arteriovenous malformations. There is a female preponderance with symptoms usually developing during middle to late adulthood (11). The initiating events which lead to development of a DAVF are not clear, but there are many papers reporting association with trauma, infection, recent surgery and dural sinus thrombosis (6,9,14,20). On the other hand, in many patients with this disorder, none of the stated causative factors can be probed from patient history (14). The causative factors listed above are quite common but DAVFs are rare lesions. Lasjaunias and Berenstein have suggested 'an underlying dural weakness' facilitating dural shunts in some individuals, when other people faced with the same insults do not develop DAVF (3).

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There are several theories attempting to explain the underlying mechanism leading to development of a DAVF. Propagation of existing micro arteriovenous fistulas in the dura and formation of new ones under the impact of an angiogenetic/fibroblastic factor are among the suggested pathways. Also, venous hypertension that develops upstream a venous occlusion may account for the opening or enhancement of arteriovenous shunts (3).

Natural disease course in DAVFs is also variable with some cases regressing spontaneously, as in some angiographically documented cases of carotid cavernous fistulas. Other DAVFs may progress into aggressive lesions leading to severe clinical pictures with recruitment of arterial supply from additional vessels and hypertrophy of existing ones (19).

As the angioarchitecture of DVAFs were documented more precisely and many cases were followed up over time, it was clearly stated by many authors that DAVFs were 'venous' lesions, meaning that natural history, symptomatology as well as prognosis of the disease were dependant on venous features (5,8).

There have been many attempts to classify DAVFs, and presently two main classification schemes have gained acceptance: A classification according to venous drainage pattern which has many important implications for prognosis and another one classifying DVAFs according to location that is more practical and used more frequently in daily practice. Both classification systems will be acknowledged in this paper.

Symptoms and signs of DAVFs are reportedly headache, pulsatile tinnitus and/or bruit while some cases with an aggressive clinical course may present with signs of raised intracranial pressure due to intracranial hemorrhage or infarction (2,17). After a thorough neurological evaluation, these patients usually undergo a neuroradiological examination, which is a head CT or MRI with or without angiographical sequences. Venous thrombosis, parenchymal changes or the presence of enlarged vascular structures near cortical brain surfaces may suggest the diagnosis of DAVF; however the mainstay of diagnosis is catheter angiography. DAVFs can be supplied by any and all meningeal branches of external and internal carotid and vertebral arteries and rarely by cortical vessels. Various angiographical findings will be discussed in the following sections of the text.

DAVFs can vary from very simple to extremely complex vascular lesions and the natural history of different subtypes of this disease can therefore be very divergent. Therapeutic approaches are also manifold; conservative follow up, carotid-jugular compression, arterial or venous embolization, surgery and combinations. A complete and correct understanding of arterial supply, nidus architecture/location and venous drainage pattern of a DAVF is very important in therapeutic decision-making (11)

Classification according to venous drainage pattern

As stated earlier in this text, venous drainage pattern of DAVFs is very important. As the high arteriovenous shunting in DVAFs continues, there is progressive pathology on the venous side. A high flow vascular malformation draining into intracranial veins and dural sinuses, especially in the presence of additional disturbing factors such as venous occlusion and reflux into cortical veins, results in intracranial venous hypertension which is a crucial factor predisposing to an aggressive disease course. The following classification scheme originally forwarded by Djinjian and Merland (later modified by Cognard) is established on venous drainage (Table I) (10,13).

Table I: Classification of DAVF according to venous drainage

TYPE	VENOUS DRAINAGE
Type I	Drainage into a dural sinus, with normal antegrade flow
Type II	Drainage into a dural sinus, with reflux into *II a:* (other) sinuses *II b:* cortical veins *II a+b:* sinuses + cortical veins
Type III	Drainage into cortical veins
Type IV	Drainage into cortical veins with cortical ectasia
Type V	Drainage into spinal perimedullary veins

Type I DAVF

Type I DAVFs are clinically benign lesions. They drain antegradely into a dural sinus with normal flow and architecture (Figure 1 A-F). Intracranial

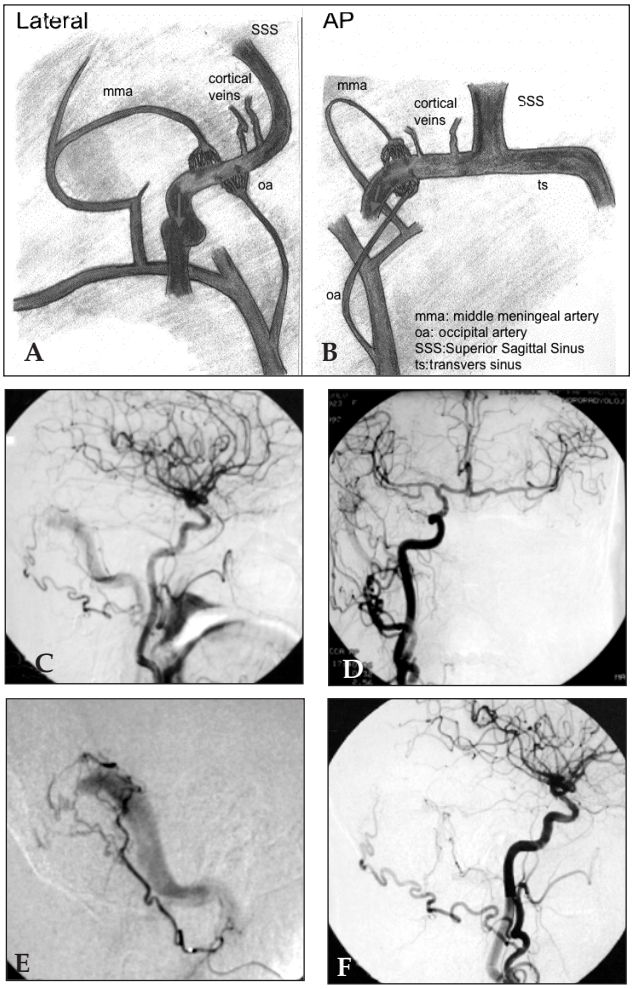

Figure 1 A-F: In type I DAVF, the fistula drains into the dural sinus with normal antegrade flow as depicted in the graphics (**A-B**). In a middle aged female patient with right-sided pulsatile tinnitus, there is a DAVF supplied by right occipital artery, draining into the right sigmoid sinus (**C-D**). The antegrade flow in the sinus is undisturbed as shown in the selective injection during transarterial embolization (**E**). Postembolization angiogram shows no residual fistula flow and the symptom has disappeared(**F**).

venous flow is also undisturbed. The symptoms may be tinnitus, retroauricular pain and ocular signs and are usually due to increased flow in the sinus. Since type I DAVFs are clinically and angiographically benign lesions, decision to treat must be made individually and therapy must be risk-free. The intention of treatment may be control of the functional symptoms when they are disturbing for the patient. Conservative follow-up, carotid jugular compression to decrease flow and induce thrombosis of the fistula or transarterial embolization of the arterial feeders with the same purpose are the options for treatment (11).

Type II DAVF

Type II DAVFs also drain into sinuses like type I, but reflux into other sinuses and/or cortical veins complicate the disease (Figure 2 A-F). The reason for reflux may be a stenosis or occlusion in the venous system downstream the venous drainage of the fistula or extremely high flow in the shunt exceeding the drainage ability of the local veins. The intracranial circulation is affected in varying degrees depending on the severity of the reflux and the symptoms are therefore due to raised intracranial pressure. Headache, decreased visual acuity and diplopia may ensue.

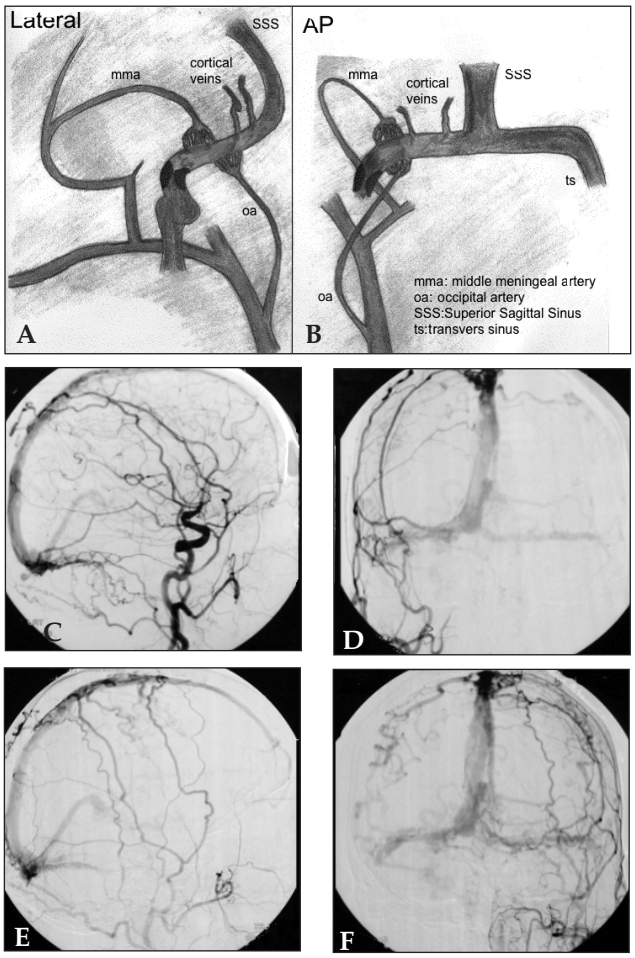

Figure 2 A-F: In type II DAVF, there is reflux into the other dural sinuses or cortical veins due to occlusion downstream of the fistula site as shown in the picture (**A-B**). In a 47-year-old female admitted with headache, DSA has shown a multicentric DAVF (located in both the superior sagittal and both transverse-sigmoid sinus junctions). Angiographically there was retrograde flow in both dural sinuses and cortical veins, compatible with type II a+b fistula (**C-F)**. (the red arrow in d depicts retrograde flow in the cortical vein)

In type II a DAVF, treatment must be performed especially if there are signs of raised intracranial pressure. The goal of therapy is flow reduction and downgrading of the fistula. Transarterial embolization is the treatment of choice. It is most frequently performed by selective catheterization of arterial feeders of the fistula and embolization with particle or liquid agents. In type II b and a+b DAVFs, reflux into cortical veins is more likely to cause an increase in intracranial pressure. In this type of fistula there is a 10% risk of raised ICP and 29% risk of neurological sequel. Consequently, treatment must be performed and result in a definite cure of the lesion. The lesion may be cured by endovascular occlusion of the defective segment of the dural sinus responsible for the arteriovenous shunts, or surgery may be performed after transarterial devascularization of the fistula (5,11)

Type III and IV DAVF

These types of DAVF are located more frequently in the anterior cranial fossa and the tentorium. The presence of varicosities on the draining cortical veins increases the risk of bleeding and neurological deficit from 40 to 65% and from 76 to 96%, respectively. Location of these DAVFs and high percentage of bleeding and neurological deficit calls for prompt and definitive treatment. (Figure 3 A-B and Figure 4 A-B). The patient in Figure 8 with a tentorial DAVF is a typical example of Type III and IV DAVF. There is direct drainage into the pial veins (with varicosities). The usually complex angiographical features of these lesions usually complicate both endovascular and surgical treatment. Combination of transarterial embolization and surgery or transarterial and transvenous embolization may be performed. Complete cure of the fistula should be aimed since inadequate or incomplete treatment may cause (re)bleeding (3, 5, 11).

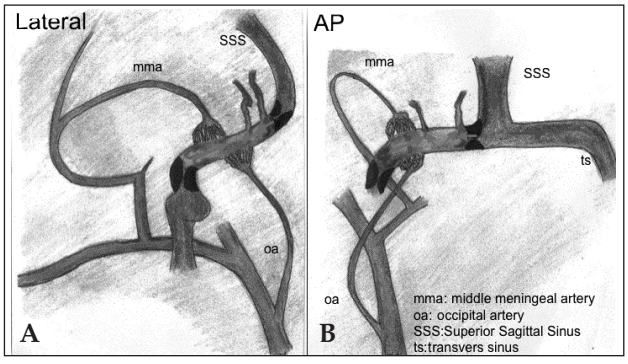

Figure 3 A-B: Type III DAVF drains directly into the cortical veins as depicted in the graphics.

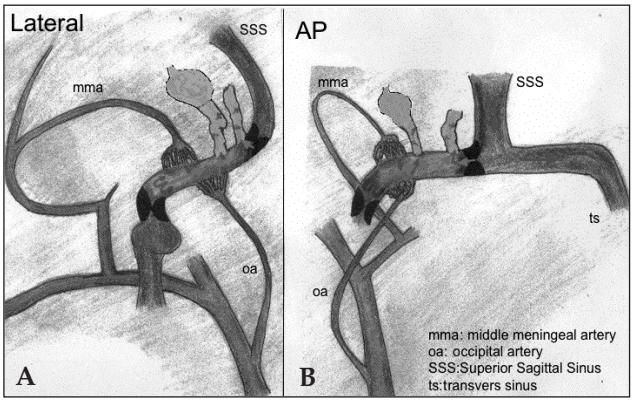

Figure 4 A-B: In type IV DAVF there are varicosities in the cortical veins draining the fistula.

Type V DAVF

These DAVFs drain into spinal veins. The underlying physiopathological mechanisms of the disease may generally be attributed to spinal venous hypertension. Clinical symptoms are due to the resultant myelopathy (Figure 5 A-G). This type of fistula has the same risk of neurological deficit and haemorrhage as in types III and IV DAVFs and must be treated in the same fashion (7,21). In a patient with progressive myelopathy, type V DAVF must be included in the differential diagnosis, enlargement and edema of the involved segment of the spinal cord and congested perimedullary vessels in cross-sectional imaging should be considered suggestive while catheter angiography must be performed for definitive diagnosis.

Classification according to location

When classified according to location, transverse-sigmoid sinus DAVFs make up the majority of DAVFs. Cavernous sinus DAVFs (10-16%), tentorial DAVFs (8-12%), superior sagittal sinus DAVFs (8%) and anterior cranial fossa DAVFs (5%) and rare locations (torcula, foramen magnum, deep venous fistula) are other types of DAVFs (11).

Transverse - sigmoid sinus DAVF

DAVF located in the transverse-sigmoid system make up the majority of DAVFs. There are usually multiple arterial feeders originating from external carotid artery branches or dural branches of the internal carotid artery (ICA), vertebral artery (VA) and posterior cerebral artery (PCA) (Figure 6 A-F). Venous drainage may be in as in any type discussed in the above classification of DAVFs. DAVFs located in the foramen magnum are considered a specific subtype of this group; however, they are much rarer than their counterparts located in the transverse-

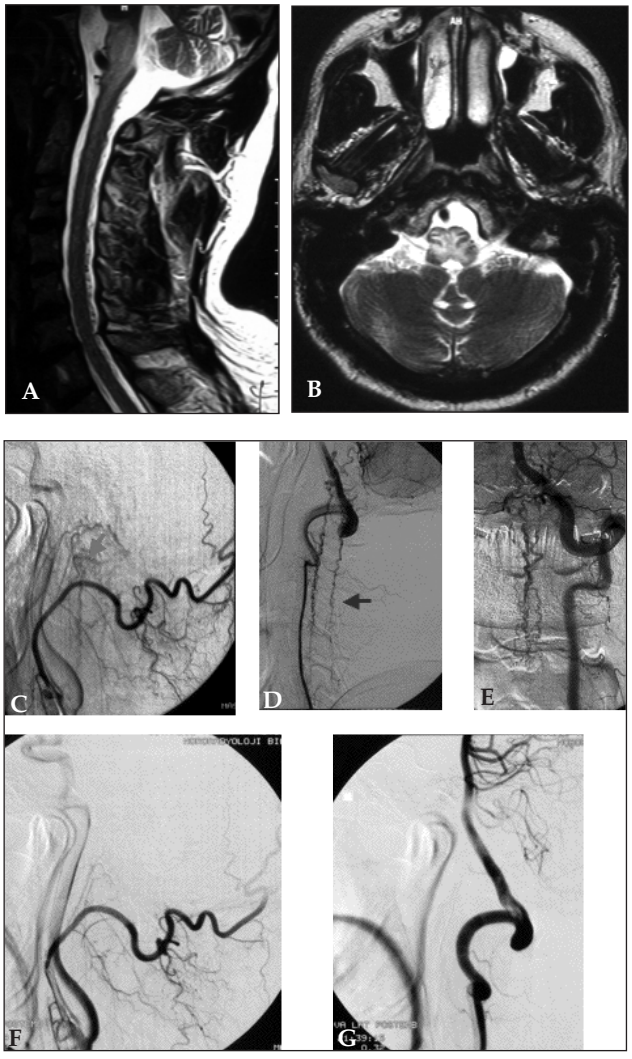

Figure 5 A-B: 55-year-old male presented with ataxia and disability in walking. Cervical MR revealed edema in the brain stem as well as enlarged perimedullary vessels (**A-B**). DSA with the presumptive diagnosis of spinal vascular malformation revealed a type V DAVF, supplied by the occipital artery and meningeal branches of the vertebral artery and draining into spinal perimedullary veins (**C-E**). After transarterial embolization there was no residual fistula and the symptoms resolved (**F-G**). (the red arrow shows fistula location and the blue arrow points to perimedullary venous drainage).(**D**)

sigmoid area. Brain MR studies must be carefully examined for parenchymal edema suggesting venous hypertension, hydrocephalus, or intracerebral or subarachnoid hemorrhage. Choice of treatment depends on the type of venous drainage (16).

Cavernous sinus DAVF

Cavernous sinus DAVFs are usually encountered in females and association with pregnancy, sphenoiditis and trauma have been reported. The

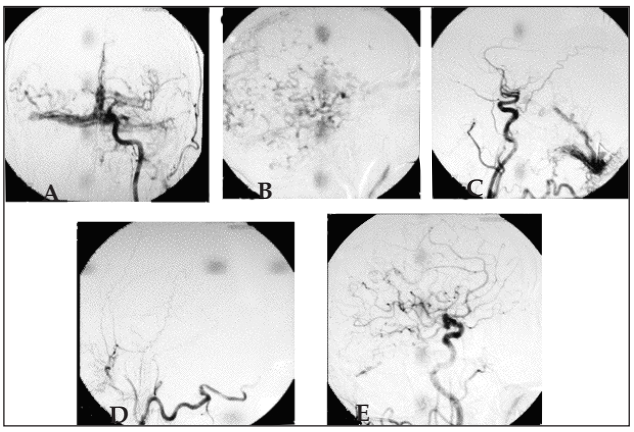

Figure 6 A-E: 50-year-old man was admitted with signs of raised intracranial pressure: Figures (**A-C**) show occlusion of both sigmoid and disturbed flow in the right transverse sinuses. There is prominent cortical reflux together with a DAVF located in both transverse-sigmoid areas. Transarterial embolization failed to cure the fistula and transvenous approach was unsuccessful. The fistulous sinuses were embolized during surgery with excellent angiographical (**D-E**) and clinical cure. At one-year follow-up, the patient was in good clinical condition; control DSA could not be performed since the patient refused any further invasive test.

symptoms depend on the type of venous drainage and are usually ocular (proptosis, chemosis, diplopia). Loss of visual acuity is also common and variable in degree. According to the classification by Barrow and colleagues, cavernous sinus fistula type A are direct and high flow arteriovenous fistula usually secondary to trauma, whereas type B, C, D are dural AVFs supplied by meningeal branches of ECA and ICA (Figure 7 A-D) (4,18). Venous drainage may be 'anterior', involving the ophthalmic veins where the symptoms are usually ocular, or it may be 'posterior' via the petrosal veins, where the risk of hemorrhage and neurological deficit is higher. The more frequent ocular sign and symptoms are generally reversible if not longstanding. Suggested methods of treatment are compression, transarterial embolization, transvenous embolization and surgery.

Tentorium cerebelli DAVF

DAVF located at the tentorium cerebelli are usually type III and IV fistulas and therefore almost always present with haemorrhage, neurological deficit, seizure or myelopathy. Treatment must definitely be performed and result in total cure. The main feeders are meningohypophyseal and inferolateral trunks of ICA. There is perimesencephalic venous drainage with venous ectasia. The arterial feeders from ICA may not be

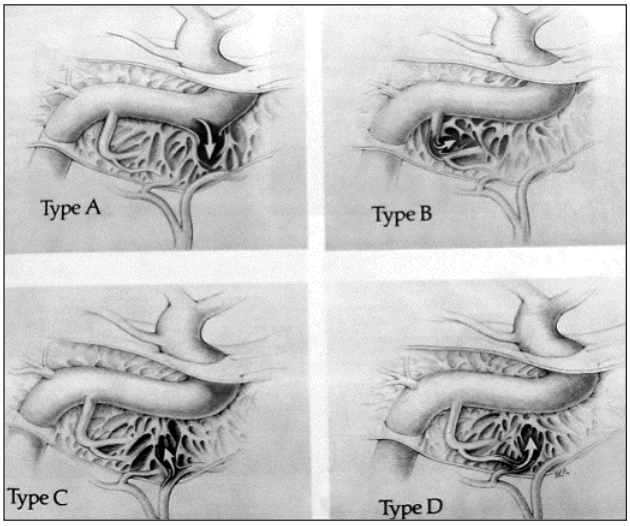

Figure 7 A: Barrow classification of carotid cavernous fistula. In type A, there is laceration in the wall of ICA due to trauma or aneurysmal rupture. Type B, C and D fistulas are dural type.

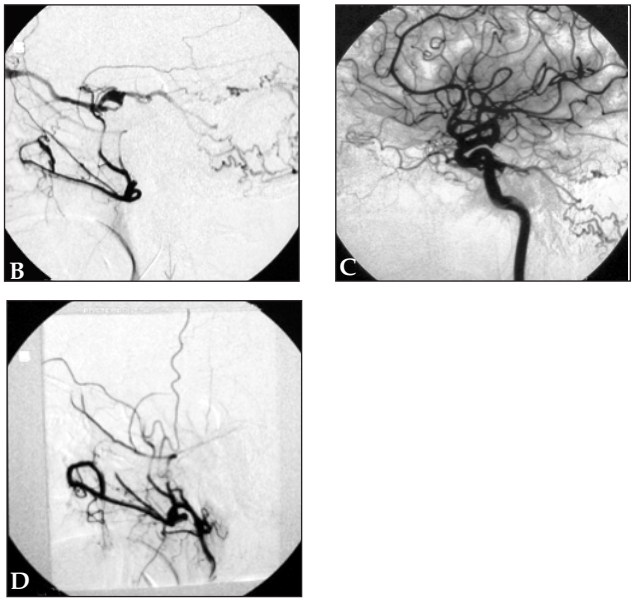

Figure 7 B-D: In a middle-aged female with exophthalmus, DSA has shown a type D fistula with supply from both ECA (**B**) and ICA (**C**). Venous drainage is both anterior into ophthalmic veins and posterior into the occipital cortical veins. ECA injection after embolization of the ECA feeders (**D**). (the red arrow: ophthalmic vein, blue arrow: cortical veins)

embolized safely. Therefore, endovascular treatment has limited use in this type of malformation. In the authors' opinion, the best treatment is surgical clipping of the draining vein and resection of the involved dura segment (12). In very complicated cases, radiosurgery combined with serial embolizations have been suggested as useful (Figure 8 A-G).

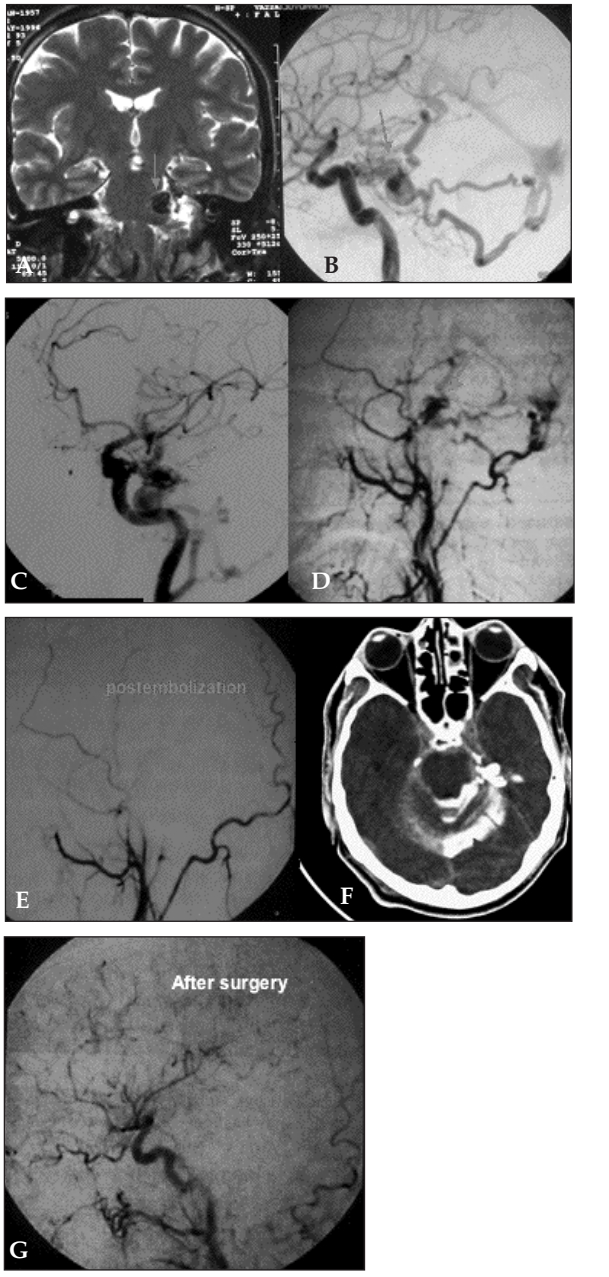

Figure 8 A-G: In this patient with tentorial DAVF, the fistula site is marked by the perimesencephalic venous pouch (red arrow). After transarterial embolization of the ECA feeders, a venous approach has been attempted but the draining vein has ruptured with SAH (blue arrow). After surgical clipping of the draining vein, there is complete obliteration of the DAVF(**G**).

Superior sagittal sinus DAVF

This type of DAVF usually presents with intracranial bleeding or dementia (12). Since the lesion is located in the midline, there are bilateral feeders from dural branches of ICA and ECA. The fistula site is frequently in the middle or posterior third of the superior sagittal sinus Endovascular

treatment via the arterial route is usually successful in this fistula

Anterior cranial fossa DAVF

This type of fistula is usually a type III or IV DAVF. They are supplied by ethmoidal arteries and drain into the frontal cortical or olfactory vein. The frequency of venous dilatation is predisposes to intracranial bleeding. The aim of treatment is total cure and, surgical clipping of the draining vein can safely succeed (1,15).

CONCLUSION

The scope of this review does not allow an extensive discussion of the DAVFs. DAVFs are intriguing lesions that may present with a varying clinical course. Natural disease course of DAVFs located in the anterior cranial fossa, superior sagittal sinus and tentorium cerebelli may be aggressive with a high risk of intracranial haemorrhage. No matter what the natural disease course may be, the following issues are crucial for optimal clinical management of DAVFs: The key feature in both diagnostic and therapeutic decision-making is the venous drainage pattern of the lesion. Indication for treatment as well as choice of therapeutic alternative must be individualized for each patient as the existing classification schemes are not perfect and subject to change over time. The risks and benefits of therapy must be treatment carefully in each patient and considered together with the possible natural course or present complications of the disease as dictated by the venous features previously emphasized in this text.

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