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Angiographic architecture of intracranial vascular malformations and fistulas – pretherapeutic aspects

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Abstract

The authors describe the angio-architecture of intracranial vascular malformations. Several patterns can be identified thanks to the intracranial superselective angiograms that can now be performed. Schematically, the following features can be seen: 1. Direct arterial supply, 2. Indirect arterial supply, 3. Flow-related arterial ectasia (aneurysm), 4. Dysplastic aneurysm, 5. Direct arteriovenous fistula, 6. Intralesional arterial ectasia (aneurysm), 7. Intralesional venous ectasia (aneurysm), 8. Venous ectasia.

Each of these elementary arrangements are illustrated and their clinical significance outlined whenever possible. Finally, the dural AVM drainage into the cortical venous system serves as an almost experimental model for the appreciation of the role played by the venous congestive phenomenon in brain AVM symptoms.

Keywords: Aneurysm, brain AVM, cerebral venous drainage, vascular architecture and anatomy, venous varix.

1 Introduction

Vascular malformation, in contrast to tumors, is usually congenital, and its symptoms are (usually) not related to mass effect, but rather to hemodynamic phenomena. Arteriovenous malformation (AVM) do not, by definition have any proliferative activity; however, it is thought that AVM may be acquired, as expressed by mass effect and present angiogenetic activity [9]. They remain unpredictable despite the multiple attempts to analyze their natural history with epidemiological methods.

Personal style and dialectic, experience and physiopathological analysis have led to various treatment strategies. However, in the past 10 years, thanks to superselectivity in angiography in the external carotid branches and, more recently, in the internal carotid or vertebral ones, new questions have arisen. Simultaneously, some answers have been provided which have produced progress in treatment planning. Although generally long term follow-up is required to make definite conclusions from impressions or results; this is particularly true in AVMs where analysis of the natural history must be made over at least 10 to 15 years.

2 General description

For practical purposes and to separate specific clinical areas, AVM can be differentiated on a topographical basis:

- Brain AVM or AVM of the central system,
- Dural AVM (so-called),
- Maxillofacial AVMs or AVM of the visceral portion of the head.

Regardless of the topography, the vascular architecture of the AVM can be classified according to the following elementary features:

- Arterial or enlarged capillary nidus,
- Arterio-venous shunt with or without fistulas,
- Associated venous anomalies (agenesis or thrombosis),
- Ectasias.

Each of these can be encountered in the three territories mentioned above. Their combinations illustrate the vascular architecture of AVMs. The "venous malformation" will be handled separately.

Furthermore, angiographic analysis brings additional information which often relates to hemodynamics and, therefore, to physiopathology:

- High flow or slow flow lesion,
- Arterial steal (angiographic),

- Blush,
- Mass effect,
- Feeding vessel,
- Compartment.

The angiographic procedure should, in addition, provide information for all therapeutic teams involved [11, 13]. The quality necessary in the era of the surgery under microscope, stereoradiation-therapy and micro-embolization, cannot be obtained with average procedures such as intravenous, brachial or common carotid studies. Again the type of angiography performed conditions the type of questions that one can ask. Progress in understanding of these pathologies requires use of high quality pictures [10].

All arteriographic descriptions of AVMs are based on static pictures and depend on the quality of the interpretation; different interpretations may then lead to different analyses, explanations, and, therefore, decisions. Thus the angiographer who performs the examination and is accustomed to his technique and to the pictures obtained with his equipement is a priori the most able to interprete his results. However, several objectives should be listed prior to his procedure in the angiographic protocol. This requires close cooperation between the specialities involved and shows the importance of the pretherapeutic evaluation of AVM.

3 Brain AVMs

The angiographic analysis of the brain of a patient presenting an intracranial AVM must, in our mind, address several questions which concern the lesion itself and the surrounding brain. At present study of the internal carotid and vertebral arteries still remains too global, although selective [10]. However, superselective injection of the feeders to the lesions can allow the rebuilding of the AVM like pieces of a puzzle and thus allow segmental analysis of its vascular architecture (Figure 1). Dural supply and cerebral venous drainage represent, for us, additional information [4, 5] which is important for the understanding of symptomatology (Figure 2).

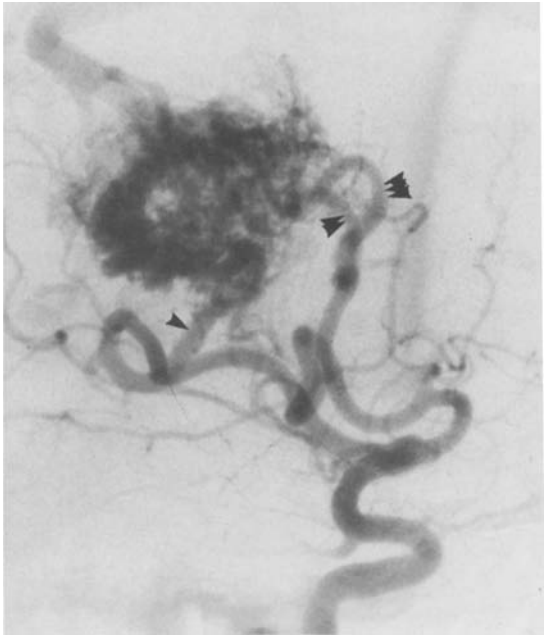


Figure 1A

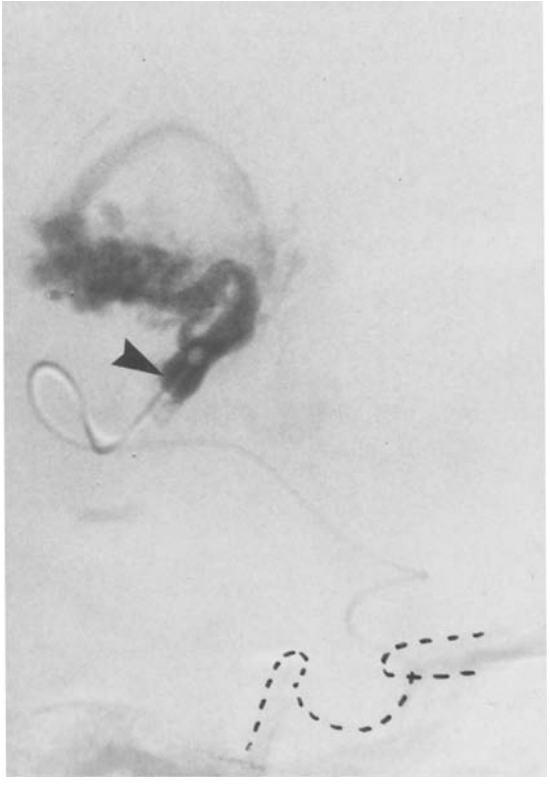


Figure 1B ▶

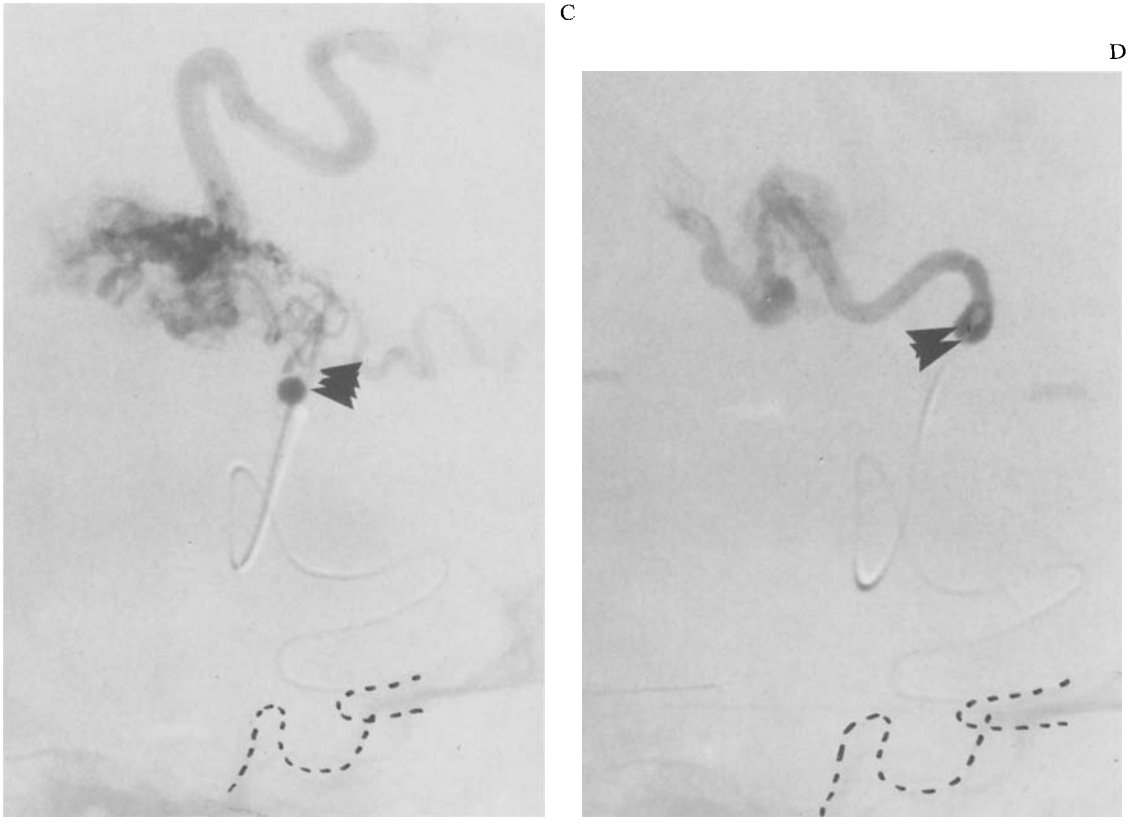


Figure 1. Selective injection of the internal carotid artery (A) in a case of Rolandic AVM. (B, C, D) Superselective injection before embolization of each of the three feeders arising from the middle cerebral artery. Note the nidus type of picture (B and D) and the direct arteriovenous fistula (C) inside the lesion.

In the MRI era, gyri and sulci mass effect and relationship of the feeders and the nidus to the functional areas can be better assessed by these new imaging modalities. Cerebral blood flow studies and neuropsychological tests can help evaluate the real arterial hemodynamic effects of the lesion on the adjacent brain [11, 14].

In our experience, we distinguish 5 locations of brain AVM: pure cortical, cortico-ventricular (which includes corpus callosum), deep hemispheric, brain stem and cerebellum. Although each presents different clinical behavior and therapeutic challenges, particularly for therapeutic angiography, they have no major differences in the angio-architecture.

In a given territory, various types of AVM can be encountered.

- Apparently unique direct arteriovenous fistula (Figure 3),
- Complex arterial nidus with shunts (Figure 1),
- Venous ectasias or thrombosis (Figure 4).

There is no evidence of transitional forms between types. Symptomatology does not appear to be related to either the type of feeder or to the characteristics of the nidus, but rather to the venous drainage patterns [2, 3, 6, 7, 12]. The importance given to the veins will be best justified after reviewing the dural AVM characteristics. The elementary pictures encountered in brain AVM are illustrated (Figure 5).

Purely spontaneous (congenital or acquired) arteriovenous fistulas are rare. They are usually seen in the pediatric population (dural AVM, vein of Galen malformation) [1] or as a complication of a vascular dysplasia (Recklinghausen, Ehlers Danlos syndrome).

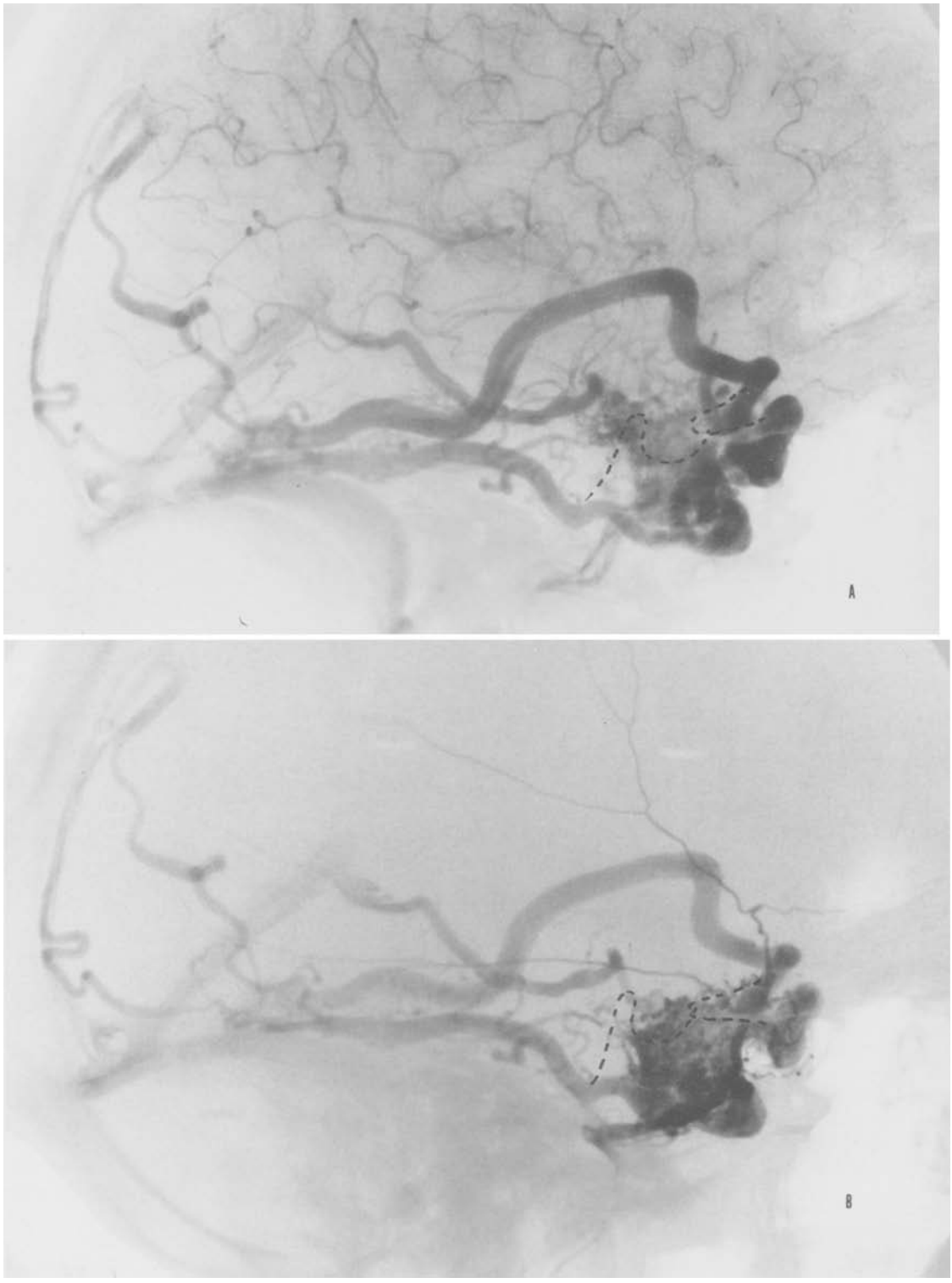
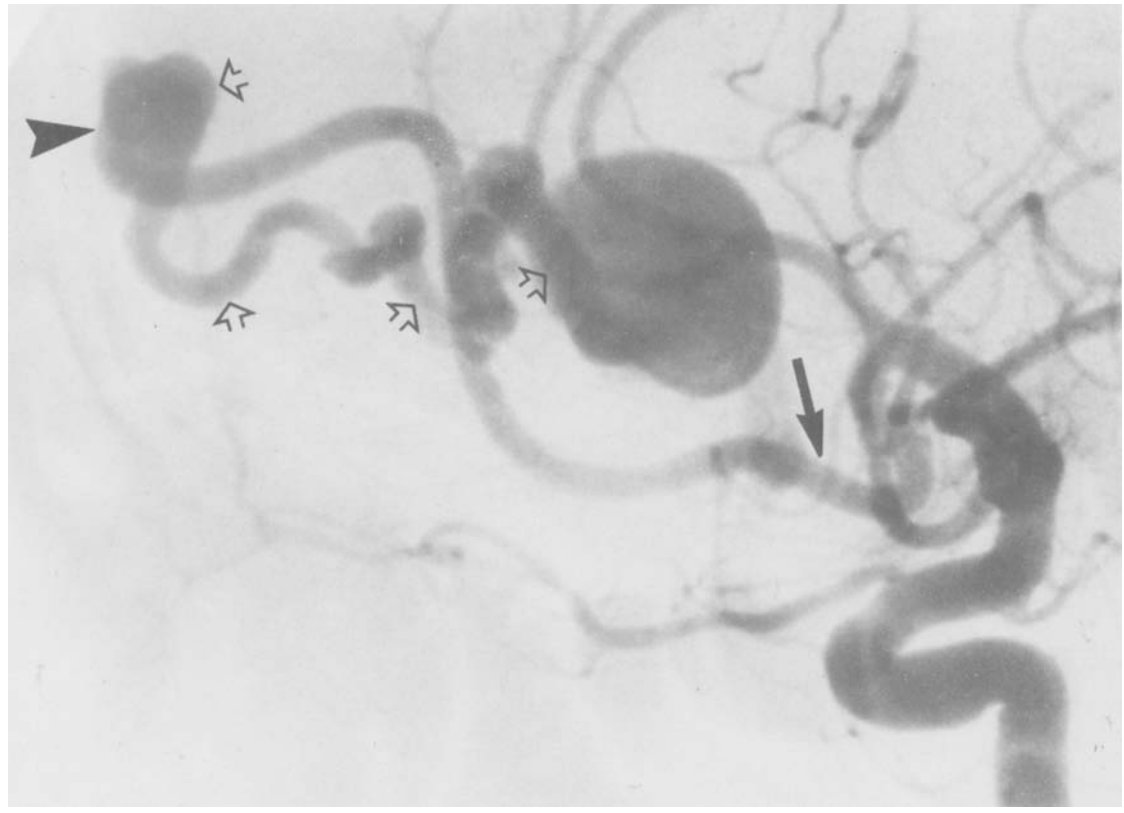
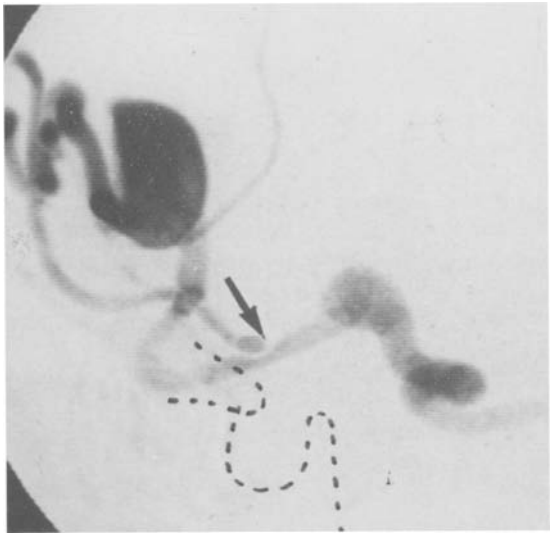


Figure 2. Selective injection of internal carotid (A) and middle meningeal artery (B) in a case of temporal pole AVM. No previous history of bleed; presenting symptom, intractable headaches.

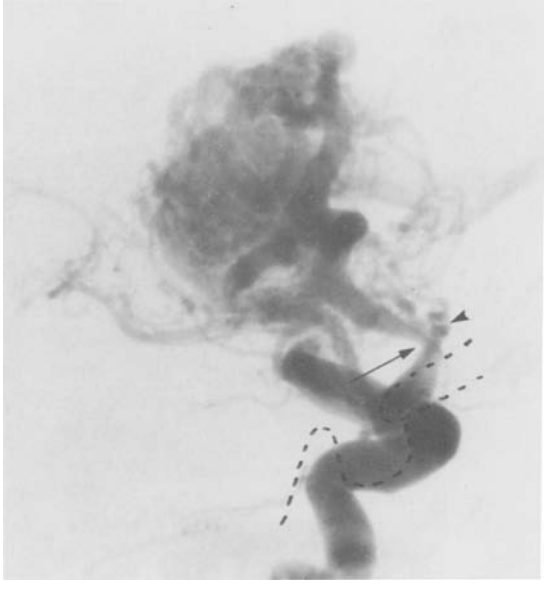


A



B

Figure 3. Selective injection of the internal carotid artery (A) in a case of intracerebral direct and isolated arteriovenous fistula (arrow head). (B) Superselective injection before embolization (arrow). Note the "normal" size of the draining vein until the pouch, in relation to a local stenotic kinking of the venous outlet in a sulcus.



A

Figure 4. Internal carotid injection, early (A) and late (B) phase, in a case of frontal AVM. A large venous pouch (arrow head) displaces posteriorly the topographic projection of the Rolandic fissure. The development of this ectasia also seems to be related to a venous obstacle.

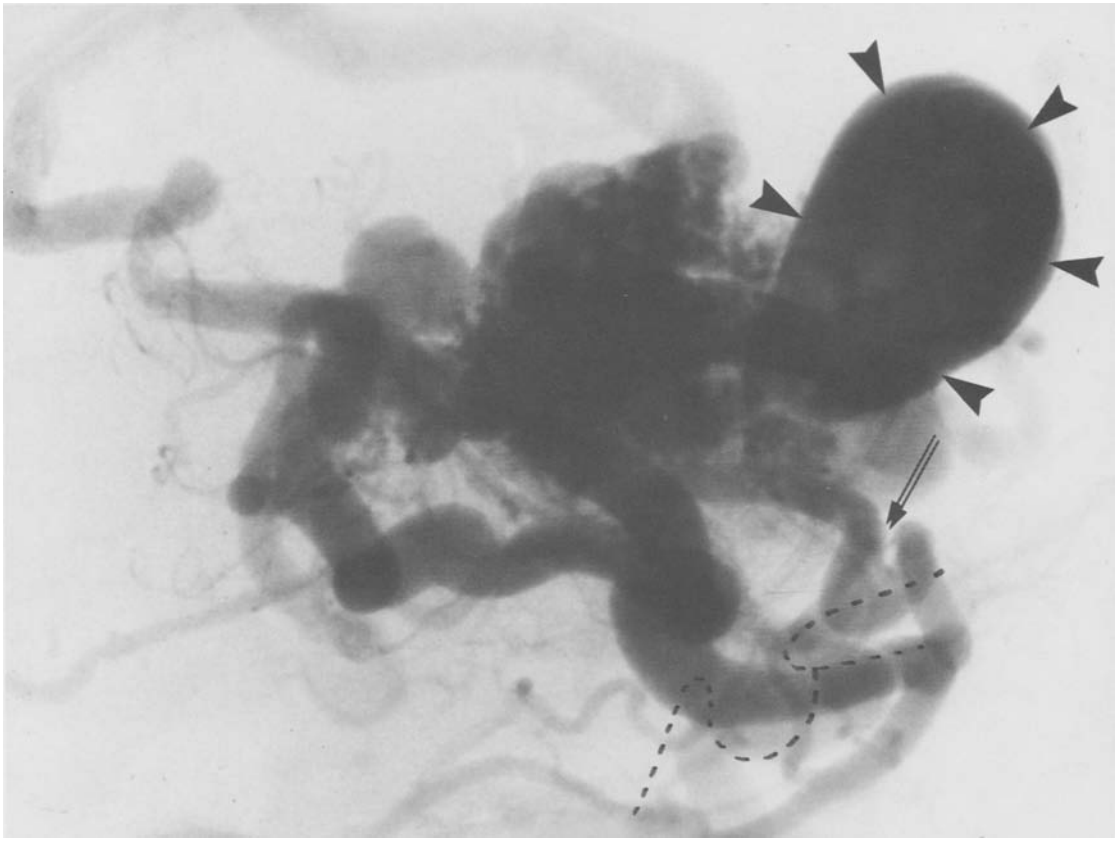


Figure 4B

4 Dural AVMs

Dural arteriovenous shunts correspond to two types of lesions, both acquired:

- Direct arteriovenous fistulas: carotid cavernous, carotid jugular, vertebro-vertebral, (traumatic or congenital) (Figure 6).
- Dural arteriovenous malformations (which may be related to a previous trauma, but which are considered to be spontaneous or acquired (Figure 7).

As with brain AVM, the arterial aspect of dural shunt is of less interest for the understanding of the clinical presentation than for planning of treatment.

Analysis of the relationship between the symptoms and the vascular characteristics of the dural AVM is particularly illustrative of the importance of the venous system (and, therefore, of its analysis) in the understanding of the intracranial AVMs, in general, and their neurological manifestations in particular.

4.1 Neurological findings and vascular architecture of dural AVM

4.1.1 Artery-related symptoms

Most dural AVM's (DAVM's) have a slow flow and no angiographic evidence of steal. Selective angiographic studies further demonstrate that the internal carotid or vertebral supply is insignificant. Therefore DAVM's cannot be considered to have any arterial hemodynamic effects on the central nervous system. On the other hand, meningeal arterial steal has always been neglected although dural arteries are known to supply the cranial nerves.

4.1.2 Nidus-related symptoms and venous related symptoms

Pulsatile tinnitus is partially related to the nidus. However, some locations are more commonly associated with this symptom than others, and it appears that tinnitus is perceived whenever the

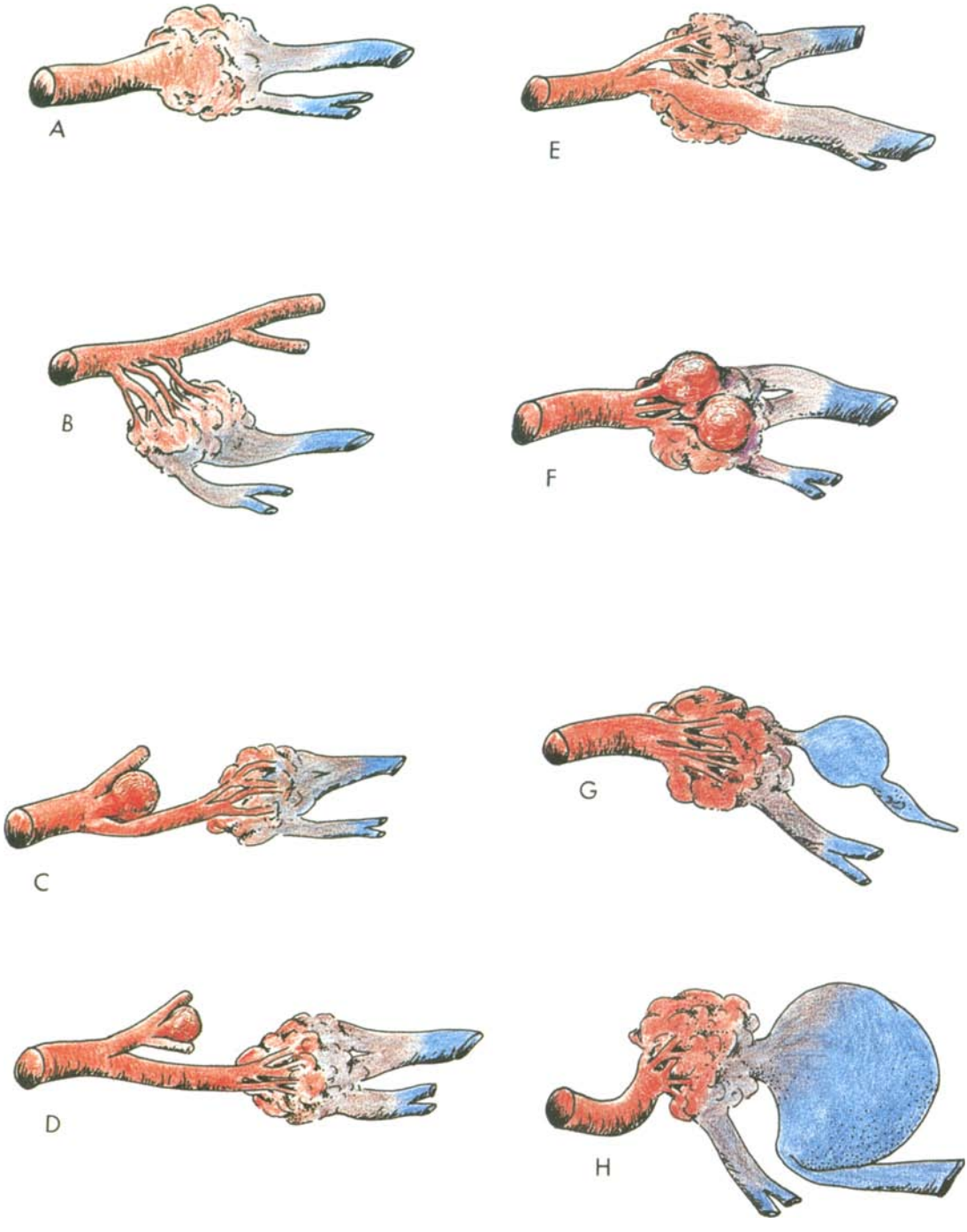


Figure 5. Schematic representation of the elementary aspects encountered in all vascular malformations of the head, brain and spinal cord areas. Each of them can be present by itself or associated to any other(s). A Direct arterial supply; B Indirect arterial supply; C High flow arterial ectasia; D Dysplastic aneurysm; E Arterio-venous fistula (with or without nidus); F Intra-lesional arterial ectasias; G Intra-lesional venous aneurysm; H Venous ectasia.

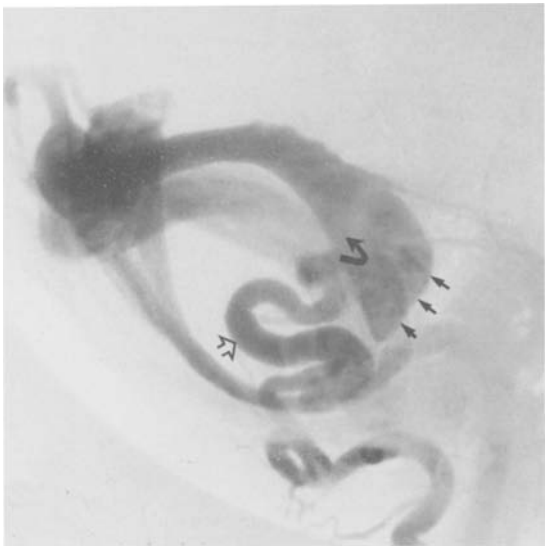


Figure 6. Vertebral injection in a case of spontaneous dural arteriovenous fistula (broken arrow). The child (3 year old) did not complain of any symptoms. The lesion was found incidentely during the search for an explanation to an objective bruit heard by his mother. Note the mastoïd artery (open arrow) and the jugular foramen obstacle (arrows). In this case no venous ectasia was encountered since the venous outflow was able to use the opposite jugular vein and reconstitute the ipsilateral one downstream of the obstacle (a diaphragm) via a medial occipital sinus.

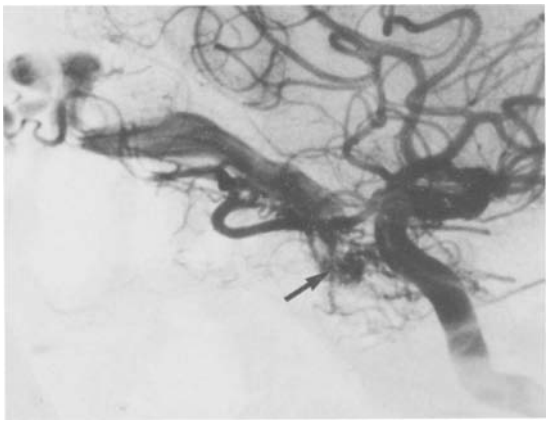


Figure 7. Selective internal carotid injection in a case of spontaneous but not immediately traumatic, dural arteriovenous malformation (shunt). Note the capillary aspect of the nidus (arrow).

drainage involves a dural sinus in direct contact with the petrous pyramid. The higher the flow in the nidus and the shunt, the more intense the tinnitus.

Venous high flow: 40% of cases with pulsatile tinnitus have objective bruits. They are usual in the pediatric group, presumably because high flow lesions occur most often in this population. It is of interest that, although the objective bruit is important, it may not be perceived by the child as an abnormality if it has been present since birth (Figure 6). In this particular group, high flow lesions may lead to cardiac complications and ultimately to death [1, 8]. Intracranially, these lesions often produce mass effect due to secondary dural venous lakes [1]. Longstanding high pressure in the superior sinus may create increased intracranial pressure. Two causes have been proposed:

- Chronic disturbance of the cerebrospinal fluid absorption due to venous sinus hypertension,
- Repeated episodes of minimal subarachnoïd hemorrhage (SAH) with subsequent meningeal reaction.

The former mechanism is the more frequent, as the latter can only occur in cases with the intradural type of drainage (see below).

Venous thrombosis: While the previous causes of increased intracranial pressure may be sufficient in themselves, secondary sigmoïd thrombosis may create an additional limitation on venous outflow leading to further increase of the intracranial pressure. Venous thrombosis may even modify the type of drainage of a given DAVM and, therefore, its clinical treatment (see below).

Venous ischemia: Focal C.N.S. symptoms are encountered exclusively in DAVM which drains into the cortical veins. Depending on the topography of the vein involved, various manifestations can be anticipated: aphasia, motor weakness, TIA, and seizures have all been reported. These symptoms are related to chronic passive congestion due to retrograde increased venous pressure towards the venous drainage of the normal brain, and can be reversed by treatment of the dural shunt (Figure 8).

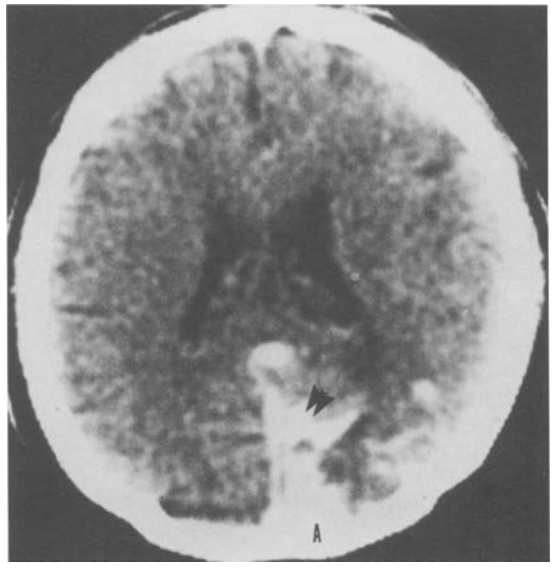
Venous mass effect: Although it is conceivable that a large venous lake may produce a mass effect, an engorged drainage vein from a DAVM is unlikely to cause mechanical compression. A mechanical phenomenon may also be responsible for the pseudo-extraocular motor nerve palsies in these cases; proptosis and muscle tension could be the cause of diplopia and poor movement of the eye-

balls. The presence of a venous lake *always* expresses a downstream obstacle (acquired or congenital).

Venous rupture: Another specific complication of DAVM drainage into the cortical vein is intradural hemorrhage. Depending on the type of hemorrhage, specific focal C.N.S. manifestations or their sequelae may result. Dural arteriovenous malformations in certain locations drain almost constantly into intradural veins, such as those of the anterior cranial fossa, and the tentorial ones. They are thus related to a high frequency of bleeding episodes. Many types of intradural bleeding may be encountered: subarachnoïd hemorrhage (SAH), subdural hematoma (SDH) and even intracerebral hematoma (ICH). Dural arteriovenous malformations draining into the transverse sinus or middle cranial fossa do not bleed unless they have cortical venous drainage (see: Table II).

Combined venous mechanism: Although episodes of headaches may result from minor bleeding, hemorrhagic complications may be overlooked and their frequency underestimated. Secondary thrombosis of the venous outlet of a pure sinusal DAVM reroutes the venous drainage to the cortical veins and can thus lead to potential focal C.N.S. symptoms and hemorrhagic complications.

The clinical importance of the symptoms described above is not equal; with the exception of cranial nerve involvement, the importance is related to the



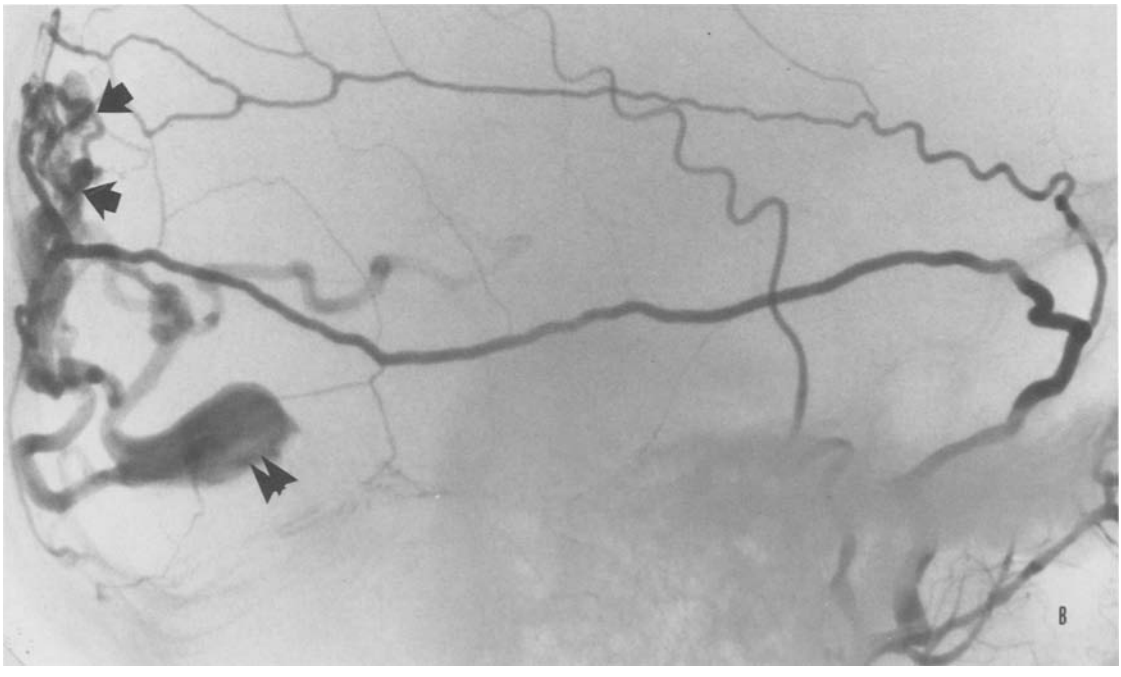


Figure 8. Enhanced CT examination (A) in a patient presenting generalized seizures with visual aura. Note the intraparenchymatous aspect of the abnormal pictures (double arrowhead). Selective internal maxillary artery (B) injection in the same patient. The shunt is located close to the superior sagittal sinus (arrows), the drainage reroutes the veins of the occipital cortex and produces a venous pouch at the calcarine fissure level (double arrow head).

Table I. Most common type of drainage of intracranial DAVM.

Location of intracranial DAVM	Sinusal venous drainage	Cortical venous drainage
Cavernous sinus	almost constant	rare
Sigmoid sinus	almost constant	rare
Superior Sagittal sinus	depending on type	depending on type
Transverse sinus	depending on type	depending on type
Torcular	depending on type	depending on type
Tentorium	rare	almost constant

Table II. Major symptoms related to the anatomic type of venous drainage of DAVM.

Sinusal venous drainage	Cortical venous drainage
Proptosis	intradural bleeding
Cranial nerve palsy	focal CNS deficits
Visual symptoms	seizures or other
ICP changes	transient symptoms
Bruits	

ICP: Intracranial pressure

venous character of the AVM (Tables I and II). However, the bleeding potential of a DAVM depends on its type of venous drainage; although if theoretically, all of them can drain into the cortical venous system (and bleed), some locations will bleed more often than others since the dural AVM has no venous outlet other than a cortical vein. It is of interest to note that no hemorrhage is reported in the epidural space regardless of the venous drainage patterns, i.e., at the level of the nidus.

5 Remarks

The similarity of the intradural symptoms of dural AVM to cortical venous drainage and brain AVM's is remarkable. However, neither the venous drainage of the brain AVM and its hemodynamic effect on the healthy brain nor its responsibility for neurological and hemorrhagic manifestations have been evaluated with modern tools [3, 6].

Spinal DAVM draining into medullary veins produces passive retrograde venous congestion. However, the normal venous drainage of the cord allows a slow progressive transmission of the retrograde venous pressure and thus leads to possible dilation of the entire medullary venous system. This could explain both the neurological deficits encountered and the absence of intradural bleeding complications in this type of AVM.

In practice proper analysis of its venous patterns is mandatory for the understanding of AVM symptoms. Although the arterial feeders are of relatively minor clinical significance, their role in the determination of a therapeutic strategy requires detailed angiographic studies. Their qualities will depend primarily on the skill of the angiographers available in a given institution. Although patients are referred by rhinolaryngologists, neurologists, neurosurgeons, ophthalmologists, and pediatricians, diagnostic neuroradiology constitutes an exceptional opportunity for a multidisciplinary approach to the investigation and treatment of these interesting, uncommon and ill-understood lesions.

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