Radiological and pathological aspects of dural arteriovenous fistulas

Case report

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A case of dural arteriovenous (AV) fistula is presented with detailed radiological and pathological findings. The complex hemodynamic alterations that may result from dural AV fistulas are described. Pathological examination in this case demonstrated widespread occlusion of the superior sagittal sinus with multiple abnormal fistulous communications between abnormal arteries and arterialized veins. A portion of the lesion resembled a recanalized blood clot, in support of the theory proposed by others that dural AV fistulas are acquired lesions.

KEY WORDS • arteriovenous fistula • dural lesion • dural sinus thrombosis • superior sagittal sinus • thrombosis

Dural arteriovenous (AV) fistulas are uncommon lesions in which meningeal and extracranial arteries shunt blood directly into the dural sinuses or, less commonly, into the meningeal or pial veins. These lesions most often afflict women over the age of 40 years, and predominantly involve the transverse sinuses, although any of the intracranial dural sinuses including the cavernous sinus may be affected. The etiology is controversial. Initially, most authors considered these lesions to be congenital, but convincing evidence has recently been presented that at least some of them are acquired.

Pathological descriptions of dural AV fistulas are rare. We present here a case report with a detailed account of the radiological and pathological findings.

Case Report

Clinical Summary

This 41-year-old man was well until September, 1981, when he developed focal seizures arising in the left hemisphere. There was no history of trauma, infection, or surgery. No abnormalities were found on neurological examination. A computerized tomography (CT) scan at this time was interpreted as normal. The patient was treated with Dilantin (phenytoin) and remained seizure-free until November, 1982, when he again had a convolution. The postictal neurological examination revealed bilateral limb hyperreflexia. A repeat CT scan was abnormal and cerebral angiography was performed (see below). A few weeks later, while riding as a passenger in a motorcar, the patient suffered another seizure, aspirated massively, and died despite emergency resuscitation.

Radiological Findings

The patient's radiological data were reviewed. In retrospect, the initial contrast-enhanced CT scan obtained in September, 1981, showed an 8-mm area of increased density surrounded by a thin rim of low density in the posterior left frontal lobe. The second CT scan, obtained in November, 1982, also with intravenous administration of contrast material, showed areas of nodular and linear enhancement in the wall of the right lateral ventricle; these were not present on the previous examination and were shown on angiography to represent enlarged thalamostriate and internal cerebral veins (see below). Small areas of enhancement were also seen in the white matter of both hemispheres, consistent with enlarged medullary veins. The density in the posterior left frontal lobe noted on the first scan was no longer apparent. We now believe that this represented a small intraparenchymal hemorrhage of venous origin.

The abnormal CT findings prompted the perform-
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Fig. 1. Left: Right common carotid angiogram, lateral view of the arterial phase, showing enlarged branches of the superficial temporal and middle meningeal arteries communicating directly with a short segment of the superior sagittal sinus at the vertex (arrow). Branches of the enlarged occipital artery fill a large midline occipital sinus (two arrows). Right: Right internal carotid angiogram at 12 seconds after injection of contrast medium, lateral view. The superior sagittal sinus and its tributary cortical veins are not seen. A tortuous medullary venous network is apparent in the parietal and frontal regions. The thalamostriate (short arrow) and internal cerebral (long arrow) veins are enlarged.

Fig. 2. Left: Right internal carotid angiogram at 9.5 seconds after injection of contrast material, anteroposterior view. The superior sagittal sinus and its tributary cortical veins are not seen. The transmedullary venous network is well demonstrated. Right: Right common carotid angiogram at 4.5 seconds after injection of contrast material, anteroposterior view. The superior sagittal sinus is opacified, and retrograde filling of the cortical veins is seen bilaterally with eventual filling of the left transverse and sigmoid sinuses (arrow).

ance of cerebral angiography. Right internal carotid, right and left common carotid, and left vertebral angiograms were performed (Figs. 1 to 3). A left internal carotid artery injection could not be performed due to vascular tortuosity.

The middle meningeal and superficial temporal arteries were enlarged bilaterally and communicated directly with a short segment of the superior sagittal sinus at the vertex. This portion of the sinus was also supplied from an enlarged anterior falcial artery. From this segment blood flowed retrograde to the superior cortical veins bilaterally, as well as to the anterior portion of the superior sagittal sinus. The posterior portion of the superior sagittal sinus from the vertex to the venous confluence was never visualized, despite filming to 20 seconds after injection of contrast medium.

Both occipital arteries, the tonsillohemispheric branch of the left posterior inferior cerebellar artery, and the posterior meningeal branches of the vertebral arteries were also enlarged and demonstrated direct fistulous
FIG. 3. Left vertebral angiogram showing posterior meningeal branches and the tonsillohemispheric branch of the posterior inferior cerebellar artery directly filling the occipital sinus (arrow). An enlarged meningeal branch of the ambient segment of the left posterior cerebral artery ascends on the falx to feed the fistula at the vertex.

communications to a large occipital sinus in the falx cerebelli which drained to the right transverse sinus. The internal carotid arterial circulation was markedly delayed in both hemispheres, with the arterial phase persisting to 8 seconds after injection. This delay was most marked in the right parietal region.

In the venous phase of the right internal carotid angiograms, no opacification of the superior sagittal sinus or superficial cortical veins was noted. Instead, a network of fine transmedullary veins was seen carrying blood to the middle cerebral vein and thence to the vein of Labbé and right transverse sinus. A similar pattern of venous drainage was present in the left hemisphere. There was also delayed opacification of enlarged right thalamostriate and internal cerebral veins which corresponded to the enhancement noted in the lateral wall of the right ventricle on the second CT scan.

Pathological Findings

The autopsy was performed elsewhere and only the brain with the attached dura was available to us for examination. The dura covering the hemispheres was grossly normal. The superior sagittal sinus was patent anteriorly, but small and surrounded by a thick lining. At the level of the parietal lobe, it was abruptly occluded by solid gray tissue which further back became partly red (Fig. 4). From the occlusion backward, large vascular channels could be seen around the sinus and extending into the falx cerebri (Figs. 4 and 5). The sinus was totally obliterated right down into the venous confluence. The transverse sinuses were patent.

Numerous microscopic sections were prepared from transverse cuts of the sinus. Sections from the normal sinuses of other cadavers and also from a sinus recently thrombosed due to carcinomatous invasion were used for comparison. Microscopically, the entire superior sagittal sinus was abnormal. The original outline of the sinus was established by a thin circumferential elastic

FIG. 4. Transverse sections of the superior sagittal sinus. At the vertex (upper specimen), a thick-walled lumen is present. At the level of the posterior parietal lobe, the sinus is occluded (center specimen). At this level and more posteriorly (lower specimen), abnormal vascular channels are present.

FIG. 5. Transverse section showing the sinus occluded by an organizing blood clot and surrounded by large atypical vessels which penetrate into the falx. Trichrome stain, X 4.
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lamina. The cavity of the anterior half of the sinus was smaller than normal. A thin layer of loose fibrous tissue lined the inside of the limiting elastic lamina, and in some segments this formed denser collagenous septae which divided the lumen into one or more channels. Loose fibrous tissue extended into and obliterated the normal lateral cavernous extensions of the sinus and also the arachnoid granulations anteriorly. Two large arteries flanked the anterior portion of the sinus. Their walls were normal in some places, but long segments had defects of the medial muscle coat which were overlaid by thick elasticized fibrous plaques.

At the level of the vertex the lumen of the sinus became enlarged, but was still lined by loose fibrous tissue. At this level, several large abnormal vascular channels appeared in the dura adjacent to the sinus. At some points, the structure of their walls was arterial, with smooth muscle and internal elastic lamina (Fig. 6).

In the sinus posterior to the vertex, the lumen was virtually obliterated by a sheet of loose fibrous tissue. This condensed into walls around vascular spaces which communicated with thick-walled venous channels outside the sinus lumen. In the occipital area, the loose fibrous tissue contained numerous endothelialized vascular spaces, free red cells, and small new vessels; this area resembled a revascularized blood clot (Fig. 7). The confluence of sinuses was converted into a leash of abnormal thick-walled vessels. The transverse sinuses just beyond the confluence had a completely normal structure.

The brain was congested and the arachnoid over the vertex was slightly thickened. The right frontal lobe contained a cluster of small abnormal dilated vessels and an adjacent 5-mm round red spot at the upper end of the head of the caudate nucleus (Fig. 8). The right thalamostriate vein was twice the size of the left. Microscopically, the abnormal vessels were venules which extended from the subarachnoid space to the thalamostriate vein. The blood spot in the center was surrounded partly by a fibrous wall and partly by vascular granulation tissue and hemosiderin-filled macrophages. The dilated venules were assumed to have enlarged to act as alternative routes of venous drainage following the occlusion of the superior sagittal sinus. The blood spot was interpreted as a vein that had leaked due to increased pressure within it.

Discussion

This case concerns a man who developed seizures at 41 years of age. In retrospect, the CT scan obtained on presentation showed what was probably a small hemorrhage in the left frontal lobe. We surmise that this was a venous hemorrhage caused by occlusion of the superior sagittal sinus. Angiography 14 months later demonstrated thrombosis of the posterior half of the superior sagittal sinus and multiple fistulous communications from meningeal and extracranial arteries to the dural sinuses at two sites, indicating a dural AV fistula.

These abnormalities produced rather complex alterations in cerebral hemodynamics. The marked delay in arterial circulation through both cerebral hemispheres reflects the generalized impedance of venous drainage caused in part by occlusion of the posterior half of the superior sagittal sinus. The enlarged right thalamostriate and internal cerebral veins and the abnormal transmedullary venous network undoubtedly developed in response to increased pressure and flow in these vessels, which were providing an alternative route of venous drainage. The enlargement of the right thalamostriate and internal cerebral veins was shown by the CT scans to be an acquired phenomenon.

A second factor impeding venous drainage of the hemispheres is the rapid shunting of blood from the meningeal and extracranial arterial branches into the

![Fig. 6. Photomicrograph of the wall of an abnormal vessel in the dura. The wall consists partly of arterial structure, with internal elastic lamina and muscle, and partly of only fibrous tissue. Verhoeff stain, x 140.](image)

![Fig. 7. Photomicrograph of the posterior part of the sinus showing an organizing blood clot occluding the residual lumen. H & E, x 12.5.](image)

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superior sagittal sinus. The resulting increased pressure and retrograde flow in the superficial cortical veins rendered these veins incapable of contributing to venous drainage of the right hemisphere, as was shown by the right internal carotid angiograms. A similar phenomenon probably occurred in the left hemisphere but is not proven because a selective left internal carotid angiogram could not be performed.

Postmortem pathological examination confirmed occlusion of the posterior half of the superior sagittal sinus by loose fibrous tissue containing large vascular channels which appeared to communicate with arterialized veins in the posterior part of the dura. In the occipital region, a spongy network of sinusoidal spaces within the sinus lumen had the appearance of a recanalizing clot. The confluence of sinuses was converted into a leash of abnormal thick-walled vessels. Even the anterior portion of the superior sagittal sinus, although still patent, was partially obstructed by fibrous tissue.

The occasional occurrence of dural AV fistulas in children or in association with cerebral AV malformations and berry aneurysms has suggested that these lesions are congenital. Other evidence, however, is beginning to accumulate to indicate that many of these lesions are acquired. Patients with this disease have an increased incidence of antecedent trauma, infection in the mastoid or paranasal sinuses or elsewhere, and previous surgery. Our patient was very active in sports, and he could easily have suffered minor trauma. Second, reports are to be found in the recent radiological literature that clearly document the development of a dural AV fistula as an acquired phenomenon and suggest the importance of dural sinus thrombosis as an etiological factor. Houser, et al., described two cases in which initial angiography demonstrated occlusion of a dural sinus 13 months and 33 months, respectively, prior to subsequent angiographic demonstration of a typical dural AV fistula in the same or in an immediately adjacent sinus. These were the only two patients in their series who had undergone angiography both early and late in the course of their disease. Of their remaining 12 patients, eight had either stenosis or occlusion of a dural sinus in addition to a dural AV fistula. Chaudhary, et al., described four patients with a history of significant head injury. In one, an angiogram performed for other reasons 2 months prior to injury was normal; angiography 1 week after trauma demonstrated occlusion of the right internal jugular vein, and an angiogram 6 months later for evaluation of complaints of tinnitus showed a typical dural AV fistula. In another patient, bilateral common carotid angiography soon after an injury which produced diastasis of the right lambdoidal suture showed no evidence of a dural AV fistula; however, a repeat angiogram 6 months later revealed a typical dural AV fistula involving the right transverse sinus. The dural sinuses in both these patients were clearly abnormal.

Abnormalities of the dural sinuses are very frequent in cases of dural AV fistula. In our own unpublished series, irregularity of blood flow, stenosis, or occlusion with retrograde flow were found in nine of 11 cases. Thus, thrombosis of the dural sinuses is an integral feature of dural AV fistulas, and in at least some cases clearly precedes the development of these AV communications.

In 1984, Piton, et al., proposed a different pathogenesis. Citing the anatomical studies of Baló, who described cavernous vascular structures within the walls of the dural sinuses reminiscent of the corpora cavernosa of the penis, they suggested that AV shunting in the wall of the sinus leads to engorgement of the cavernous system which “progressively reduces the venous lumen and may end in complete obstruction.” In our case, while a meshwork of cavernous spaces was seen next to the main lumen of the superior sagittal sinus, there was no muscle or extensive elastic tissue comparable to the erectile tissue of the corpora cavernosa. Obstruction was definitely due to intraluminal thrombosis and fibrosis in both the sinus and the cavernous spaces. Moreover, the sequence of events postulated by Piton, et al., would be difficult to reconcile with the cases in the literature referred to above in which venous sinus occlusion preceded the development of the dural AV fistula.

The normal dura is, however, an extremely vascular structure, receiving a rich network of meningeal branches not only from the external carotid artery but

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Fig. 8. Section showing a small encapsulated hematoma at the upper end of the head of the right caudate nucleus. Dilated small veins are collateral vessels draining to the deep cerebral veins.
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also from the internal carotid and vertebral arteries. Postmortem injection studies by Rowbotham and Little,18 later confirmed by Kerber and Newton,13 demonstrated AV shunts 50 to 90 μ in diameter in normal dura. These are particularly numerous near the superior sagittal sinus. It has been postulated that thrombosis or thrombophlebitis of the dural sinuses is the initial event in the development of dural AV fistulas. The multiple fistulous channels that are seen angiographically are thought to represent a pathological enlargement of these normal AV shunts in response to thrombosis.

Thrombosis of the superior sagittal sinus was a prominent feature of the pathology in our case. Some of the thrombosis was obviously long-standing, but in other places it appeared more recent. At least portions of the malformation had the appearance of recanalized blood clot, implying that the lesion was acquired and developed secondary to thrombosis. A similar observation was made by Houser, et al.9 Even if one accepts the acquired nature of these lesions, many intriguing questions persist. Most patients with thrombosis of a dural sinus do not appear to develop a dural AV fistula,11,12 and the factors that stimulate such development in a few patients remain unknown. The function of normal physiological dural AV shunts is also not known. Presumably, these shunts are regulated in some way as to prevent them from enlarging under normal circumstances, and one can only speculate regarding the factors associated with thrombosis that overcome this regulation and lead to fistula formation. Alternatively, one might propose that the fistulous vessels demonstrated angiographically are not enlarged physiological shunts but represent a pathological neovascularity. One can question why similar lesions are not seen more frequently in response to thrombophlebitis in other parts of the body. Is there an absence of AV shunts in the walls of normal systemic veins? (Interestingly, one of us has noted an acquired lesion affecting an internal iliac vein which resembles a dural AV fistula on angiography.)

Many of these lesions resolve spontaneously, probably on thrombosis of the fistulous connections.16 In lesions that drain entirely into the transverse or sigmoid sinus, the only symptoms may be headache and bruit, and one may hope for spontaneous resolution unless the symptoms are so distracting to the patient that definitive therapy is requested. Lesions in the region of the cavernous sinus may require treatment for such symptoms as deterioration of vision, elevation of intracranial pressure, severe pain, or chemosis.

Cases such as the present one in which cortical venous drainage and/or altered cerebral hemodynamics are present constitute an important subgroup, as neurological deficit, seizures, elevated intracranial pressure, or hemorrhage may result.7 Treatment is definitely indicated in these cases. Embolization may be effective in occluding external carotid artery branches. Patients with significant supply from the internal carotid or vertebral artery circulations may require surgical isolation or even excision of the involved sinus.10

Acknowledgment

We gratefully acknowledge the careful clinical management and support provided by Dr. Barbara Allan, who referred this patient to us.

References


Manuscript received September 4, 1985. Accepted in final form November 22, 1985.
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J. Neurosurg. Volume 64/June, 1986