Extensive dural arteriovenous malformation

Case report

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 \checkmark A child with an extensive dural arteriovenous malformation (AVM) of the sigmoid sinus and bilateral occlusion of the transverse sinus is reported. His intracranial venous flow had become rerouted through the right cavernous sinus and then into the right ophthalmic veins. The AVM was associated with severe hydrocephalus and a progressive devastating neurological deficit. The hemodynamic and intracranial pressure changes as well as the mechanism of sinus thrombosis are discussed. The role of aggressive surgical treatment is emphasized.

KEY WORDS • arteriovenous malformation • dura mater • sigmoid sinus • hydrocephalus • thrombosis

URAL arteriovenous malformations (AVM's) in the region of the transverse sigmoid sinus, although rare, represent a well-defined entity from both clinical and angiographic aspects.^{1,7-9,13,14} The etiology and pathology of these anomalies remain controversial.^{1,7,12-14,24,25} The clinical findings are determined by the volume, direction, and route of the venous drainage. The disease often progresses, leading to increasing neurological deficits, elevation of intracranial pressure, hydrocephalus, seizures, and hemorrhage.^{2,7,8,13,24} Unilateral thrombosis of the sigmoid sinus is recognized to be associated with these lesions.^{4,10,11,18,24} In rare cases, the thrombosis is bilateral.^{2,4,5,20,24}

We report a case that exemplifies the devastating neurological deficit that results from intracranial hemodynamic and pressure changes associated with this type of AVM. The patient made a remarkable recovery after radical surgical treatment.

Case Report

This 13-year-old Syrian boy was admitted to the King Faisal Specialist Hospital on October 30, 1984. His medical history started 6 years before, when his right cheek was struck by a stone. The injury was mild. Several months later a gradually increasing proptosis of the right eye and swelling of the forehead and nose was noticed. One year prior to admission, the patient had increasingly severe headaches and deteriorating consciousness. He had been seen in several medical centers prior to his present referral.

Examination. The child was severely cachectic, had several decubitus ulcers, and was debilitated, with flexion contractures of all four limbs. He had proptosis of the right eye with marked dilatation of the right superior ophthalmic and facial veins. A loud bruit was heard over the right side of the head. He was awake, but did not follow commands and spoke only a few words. Bilateral papilledema was present. The ocular movements were intact.

Pre- and postcontrast computerized tomography (CT) scans demonstrated hydrocephalus and extensive dilatation of vascular structures within the cranium and right orbit. Peripheral rim calcifications of these vascular structures were present, suggesting dystrophic changes in the walls of arterialized veins (Fig. 1). The initial arteriograms showed a direct dural arteriovenous fistula (Fig. 2 left). The major feeding vessel was a markedly hypertrophied right middle meningeal artery communicating with the dural venous sac, which drained into the sigmoid sinus. Additional communications with this dural venous sac were via some unnamed branches originating from the right internal carotid artery in the petrous bone and also from a large meningo-orbital branch (also known as "the superior orbital branch of the middle meningeal artery") emanating from the lacrimal division of the ophthalmic artery (Fig. 2 *right*). The venous phase of the arteriogram demonstrated bilateral transverse sinus occlusions, and aneurysmal dilatation of the sigmoid sinus and torcula Herophili (Fig. 3). Extracranial drainage was entirely via the right cavernous sinus and then into the superior and inferior ophthalmic veins.

Operations. At the first operation, the right middle meningeal artery was occluded by a Fogarty catheter balloon. A similar balloon was lodged more proximally in the internal maxillary artery, after which the external carotid artery was ligated. A few days later a small right frontotemporal craniotomy was performed with clipping of the meningo-orbital artery. Angiography then

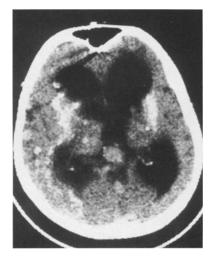


FIG. 1. Plain computerized tomography scan at the level of the lateral ventricles demonstrating hydrocephalus and calcifications.

demonstrated that the fistula was filled from hypertrophied branches of the intrapetrous portion of the internal carotid artery (Fig. 4 *left*).

At the third operation, temporal craniotomy was performed with clipping of the internal carotid artery feeders behind the petrous pyramid and total excision of the aneurysmal sac contained within the two leaves of the dura mater. Postoperative angiography confirmed complete obliteration of the AVM (Fig. 4 *right*). The route for cerebral venous drainage via the ophthalmic veins remained, but the pressure and size of the dilated vascular structures were much reduced. The ophthalmic veins collapsed with the patient in an upright position.

Postoperative Course. The patient underwent grafting of the decubitus ulcers, a ventriculoperitoneal shunt was placed, and the flexion contractures of his limbs were treated orthopedically. After 6 months of rehabilitation, he was able to walk and converse with continually improving mental and physical skills. One year after surgery, he has nearly normal mentation and neurological function. He is being treated for hip dislocation.

Discussion

The cause-and-effect relationship between the presence of the dural AVM and the thrombosis of the sinus is controversial. Sundt and Piepgras²⁴ have suggested that the dural AVM is an acquired abnormality evolving from revascularization of the previously thrombosed sinus — the sinus thrombosis being the primary event. Contrary to that, the sinus thrombosis may develop as a result of the hemodynamic and morphological changes caused by the turbulence of the AVM. Over a century ago, Virchow postulated that the path-

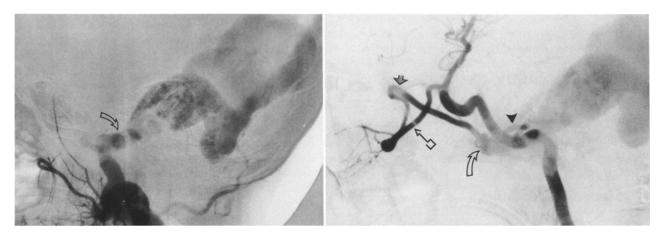


FIG. 2. Left: Right external carotid arteriogram, lateral view, showing the middle meningeal artery markedly hypertrophied and connecting directly to the dural venous fistula (arrow). Right: Right internal carotid arteriogram, lateral view, depicting the enlarged right meningo-orbital artery (closed arrow) arising from the lacrimal division of the ophthalmic artery (straight open arrow). The curved open arrow shows the point of arteriovenous connection, and the arrowhead the meningeal feeding vessel arising from the internal carotid artery in the petrous bone.

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ogenic events that led to thrombus formation included: 1) stasis; 2) changes in blood coagulability; and 3) alterations in the vessel wall.³ Such phenomena certainly may occur in dural AVM's. Seeger, *et al.*,¹⁷ demonstrated radiographic signs of thrombus formation in the cavernous sinuses in eight consecutive patients with carotid-cavernous fistulas. In five of these patients, progression of thrombosis was revealed on follow-up angiography. They hypothesized that the arterial shunt into the venous sinus elevates the venous pressure to a critical point at which the flow in the tributaries is decreased, resulting in blood stasis within the sinus.

Many patients with sigmoid sinus AVM's present

with elevated intracranial pressure,¹⁰ which, if it is of sufficient magnitude, may compress and collapse the dural sinus wall.^{15,19} Osterholm¹⁵ demonstrated that this collapse of the dural sinus occurs in the transverse sinus area. Stasis within the sinus would then follow.

When these dynamic and morphological changes in the sinus occur, endothelial and subendothelial changes can ensue, leading to thrombosis. Stewart, *et al.*,²² presented experimental data showing leukocytic invasion into a venous wall following stasis in the vein associated with surgical trauma to adjacent tissue. The leukocytic invasion caused venous endothelial damage and desquamation. Endothelial desquamation exposes subendothelial collagen, initiating platelet adhesion and

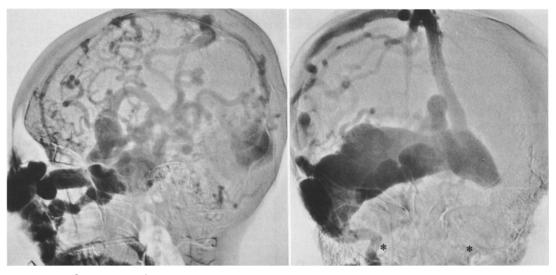


FIG. 3. Left: Right carotid angiogram, venous phase lateral view, demonstrating the markedly dilated cortical veins, bilateral occlusion of the sigmoid sinuses, and venous drainage exiting entirely through the right superior and inferior ophthalmic veins. *Right:* Anteroposterior view showing the dilated dural venous sinus draining superiorly. There is bilateral occlusion of the sigmoid sinuses. *Asterisks* mark the location of the nonvisualized jugular bulbs.

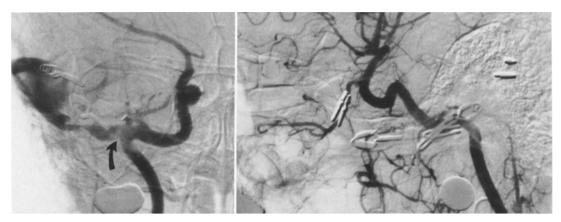


FIG. 4. Left: Right carotid arteriogram, frontal view, after clipping of the meningo-orbital artery. The arrow indicates the petrous branches arising from the internal carotid artery and communicating with the dural sinus. *Right*: Right common carotid arteriogram, lateral view, after clipping of the internal carotid artery feeders, demonstrating complete obliteration of the dural arteriovenous malformation.

aggregation as well as activation of the intrinsic clotting system leading to thrombosis.²¹

The injection of contrast medium may further contribute to the process of thrombosis. Ritchie, et al.,¹⁶ showed experimentally that leukocyte accumulation in the venous wall may be exaggerated by contrast medium. Clagett and colleagues³ stated that a direct effect of the contrast medium on the vascular endothelium and on red blood cells will contribute to thrombus formation. This finding is supported by the occurrence of thrombosis at angiography in dural AVM's, as reported by Kühner, et al. (their Case 2),9 and Storrs and King (their Case 4).²³ and as reported in several cases of carotid-cavernous fistula thrombosis at or soon after angiography.¹⁷ Finally, the turbulence caused by the shunt may have further contributed to the thrombotic process. The occurrence of thrombosis of the sinus would explain the abrupt neurological deterioration in our own and other reported cases.

Although spontaneous closure of a dural AVM has been reported,^{9,11,23} complete obliteration of the fistula is recommended.²⁴ Simple ligation of the arterial feeder is rarely successful. Recent techniques of arterial occlusion by embolization have merit,^{6,26} and surgical intervention may be curative if the embolization fails to completely obliterate the fistula. Serial stages of management were recommended by Storrs and King.²³ Embolization and/or ligation of the major feeding vessels, followed by repeat angiography and then total surgical excision, minimizes blood loss, decreases the shunt pressure, may allow total excision, and permits gradual correction of the disturbed hemodynamics of the cerebral circulation.

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