Direct spinal arteriovenous fistula: a new type of spinal AVM

Case report

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A patient presenting with progressive paraparesis was found to have a spinal arteriovenous fistula at the T3-4 vertebral level. The lesion consisted of a direct communication of the anterior spinal artery with a very distended venous varix that drained mostly superiorly to the posterior fossa and simulated a posterior fossa arteriovenous malformation (AVM) on vertebral angiography. The patient was treated by surgical ligation of the fistula through an anterior transthoracic approach. He deteriorated abruptly on the 4th postoperative day, probably because of retrograde thrombosis of the enlarged anterior spinal artery. Over the next few months, he improved to the point of being able to walk with crutches. He has also regained sphincter control.

The different types of spinal AVM's are reviewed. Our case does not fit into any of these groups. A new category, Type IV, is proposed to designate direct arteriovenous fistulas involving the intrinsic arterial supply of the spinal cord.

KEY WORDS: spinal cord, arteriovenous malformation, arteriovenous fistula

Spinal cord arteriovenous malformations (AVM's) can be classified into one of three categories: Type I, the dorsal extramedullary AVM; Type II, the compact, usually intramedullary AVM with multiple feeders; and Type III, the extensive juvenile malformation. This report describes a patient with a direct arteriovenous fistula involving the anterior spinal artery, which cannot be classified under any of these categories. The literature is reviewed and it is proposed that a new category — Type IV, a direct arteriovenous fistula involving the normal arterial supply of the cord — be added to the classification of spinal AVM's. An unusual surgical approach to this lesion is described.

Case Report

This 31-year-old man had suffered from progressive paraparesis of insidious onset for approximately 2 years before admission to the Massachusetts General Hospital. By the time of admission he was essentially wheelchair-bound. Vertebral arteriography, performed in Belgium, had initially revealed what appeared to be an AVM of the posterior fossa. The patient was referred to our institution for possible proton-beam treatment of the lesion.

Examination. Selective vertebral and spinal arteriography performed in Boston and Paris revealed a direct arteriovenous fistula formed by large descending and ascending anterior spinal arteries that communicated directly with a distended vein that drained mostly upward to the posterior fossa but also, to a lesser degree, inferiorly. The fistula was located at the T3-4 vertebral level (Figs. 1, 2, and 4). Dynamic computerized tomography (CT) with contrast enhancement and metrizamide indicated that the lesion was extramedullary and located anterior to the spinal cord (Fig. 3). An attempt to treat the lesion by balloon occlusion was undertaken. It was possible to navigate a detachable balloon successfully through the right vertebral artery and down the descending anterior spinal artery which could be temporarily occluded, but the fistula was kept open by the flow from the ascending spinal artery. It was felt that it would be futile to occlude the descending branch while leaving the ascending branch open (Fig. 4), and it was decided to approach the lesion surgically.
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Operation. After careful review of the anatomic location of the fistula, an anterior transthoracic approach to directly expose the lesion was planned. The fistula was located in the midline, but the arteries entered the fistula to the right of the midline and the distended venous aneurysmal pouch projected to the left. We therefore chose to approach the lesion from the right side. The anterior vertebral column from T-2 through T-5 was exposed anterolaterally through a right-sided thoracotomy incision, after resection of the head of the fourth rib. Using the high-speed air drill, we resected a wedge of the T-3 and the T-4 vertebral bodies to expose the dura over the anterolateral aspect of the spinal cord. The dura mater was then opened linearly for a length of about 3 to 4 cm just to the right of the midline. The distended venous pouch was well visualized through the dura mater using this approach.

The anatomy seen at surgery was exactly that predicted by angiography. Some care had to be exercised in developing, under high magnification with the microscope, a plane between the ascending and descending arterial branches and the vein; the vein was relatively thin-walled, and the arteries were considerably thicker although much smaller in diameter. After a plane was developed, it was possible to clip and divide the two arterial feeders which entered the distended venous pouch through a common “hilum” as depicted in Fig. 4. After clipping both feeders, the venous pouch became much softer and the blood within it became blue (“venous”), as compared to the previous red color. We carefully inspected the now softer venous pouch for other feeders and could find none. The dura was closed in a watertight fashion without fear of compression since the venous varix was now partially collapsed. Because only the anterolateral portion of the vertebral bodies was removed, we did not feel that bone grafting was necessary. The thoracic incision was closed in the usual manner.

Postoperative Course. The patient’s condition remained unchanged until the 4th postoperative day, when his legs suddenly became weaker and he was barely able to move his feet. In addition, he lost the partial control of bladder function that he had had preoperatively. We obtained an immediate metrizamide CT scan which showed no block or evidence of extrinsic compression of the spinal cord. The venous varix now appeared to be thrombosed. At this point we considered that the deterioration was probably related to thrombosis of the anterior spinal artery and that surgical intervention would not be beneficial. About 1 week later we obtained repeat arteriograms. The vertebral arteriogram showed no filling of the descending branch of the anterior spinal artery, which suggested ascending retrograde thrombosis from the point of surgical occlusion. Spinal angiography showed no filling of the ascending branch of the anterior spinal artery when its previous main feeder, the left L-3 vertebral
artery, was injected. This again suggested the possibility of descending retrograde thrombosis. There was no opacification of the draining venous complex with either vertebral or selective spinal angiography.

A partial dehiscence of the thoracic incision occurred and was treated by resuturing in the operating room. The patient returned home to Belgium about 3 weeks after surgery with only minimal improvement from his deteriorated neurological state. Since then, however, he has gradually made a very rewarding recovery to the point that, at about 6 months after the operation, he was able to walk with crutches and he had regained useful control of his sphincter muscles. He is continuing to improve at the time of this report.

Discussion

Almost all the reported spinal AVM’s can be categorized under one of three major groups. The first group (the most common type of spinal AVM) is the long single coiled vessel malformation located almost invariably in the dorsal aspect of the lower thoracic cord and the conus. This lesion was called “angioma racemosum venosum” by Wyburn-Mason in his classic monograph.38 Djindjian and his colleagues10,14,15 referred to this lesion as “malformation rétro-médullaire.” Di Chiro, et al.,8,17 called them “Type I” spinal AVM’s and Malis26 prefers to call them “long dorsal arteriovenous malformations.” These lesions are almost always extramedullary and are diagnosed most frequently in males between the ages of 40 and 70 years. They usually have one, occasionally two, and rarely three or more large feeders which enter the dura dorsolaterally and drain into the long coiled vessel which frequently extends for many segments above and below the region of the “feeders.” The long coiled vessels are usually under low pressure, but contain arterialized blood. These are usually “low flow” malformations which arteriographically fill relatively slowly. Clinically, they usually present with a syndrome of progressive neurological deterioration with occasional abrupt exacerbations, and only rarely are there remissions. The symptoms can be temporarily intensified by exercise, which mimics the syndrome of neurogenic claudication. Pregnancy and Valsalva maneuvers can also ex-
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FIG. 3. Computerized tomography scans, horizontal section of the cervical spine. Upper: Scan after cerebral angiography. The huge vein (arrow) is filled with contrast medium in the middle of the cervical canal. The cord cannot be visualized but it must be severely compressed by the vein which occupies most of the canal. Lower: Scan after subarachnoid injection of metrizamide. The huge vein (long arrow) is in the middle of the canal, surrounded by metrizamide anteriorly and laterally. The dilated anterior spinal artery (short arrow) is also in the subarachnoid space, surrounded by metrizamide on the left side of the vein. The cord (curved arrow) is a crescent, in the anterior concavity of which the huge vein is embedded.

acerbate the symptoms, probably by increasing venous pressure. 

Presently, it is believed that the likely pathophysiology accounting for the progressive myelopathy in these patients is chronic venous hypertension. At surgical exploration it is rare to find evidence of recent hemorrhage or of spinal cord compression to account for the symptoms, and there is usually no surgical or pathological evidence of extensive thrombosis. A “steal” phenomenon has been implicated, but this is unlikely since the arterial supply to the lesion is usually separate from the intrinsic cord supply, and most patients treated only by ligation of feeders improve, at least temporarily, which would be the opposite effect expected if a steal phenomenon were the main pathophysiological mechanism. It appears that the distended coils seen myelographically and at surgery in these patients are indeed the dilated and arterialized coronal veins which normally drain the cord. Venous hypertension then results in pathologically demonstrable chronic intramedullary ischemic changes. Subarachnoid and, rarely, intramedullary hemorrhages do occasionally occur in these patients accounting for the abrupt onset of symptoms, but the frequency of bleeding is less than in the higher-pressure lesions discussed below.

Evidence has been accumulating to indicate that, in most if not all of the Type I spinal AVM’s, the actual malformation or shunt is extradural, usually related to the dural sleeve of a dorsal root. The lesion then drains intrathecally through one or two efferent vessels which penetrate the dura, usually at the point adjacent to a dorsal root. The efferent vessels become the “feeders” bringing arterialized blood into the coronal venous plexus which, with time, becomes dilated and tortuous for a variable distance. This concept, which has now been proven repeatedly by sophisticated selective angiography and also by direct surgical observation, explains why excision of the dural fistula without opening the dura or embolization of the fistula usually results in lasting and frequently dramatic improvement. The improvement seen with these simpler procedures supports the hypothesis that venous hypertension is indeed the important pathophysiological mechanism, and suggests that the traditional radical excision of all the dilated abnormal venous coils is unnecessary in these patients.

The Type II spinal AVM is a true compact angioma fed frequently by multiple feeders and draining into the coronal venous plexus that surrounds the cord and becomes tortuous and distended with arterialized blood for a variable distance up and down from the lesion. These lesions have also been called “angioma racemosum arteriovenosum” or simply “angioma arteriovenosum” and “glomerus malformation.” The majority of these lesions are intramedullary; only a few are
extramedullary. They occur most frequently in the cervical region, but can also occur throughout the thoracic and lumbar areas. This type is as common in males as in females and presents with equal frequency throughout adulthood. They are high-flow high-pressure lesions which opacify rapidly and drain early angiographically. They are mostly fed by branches from the anterior spinal artery, but those located in the cervicomedullary junction are usually located dorsally and are fed by branches of the vertebral artery which reach the lesion from posteriorly, thus making these particular lesions more susceptible to surgical resection.

Type II lesions have a less predictable natural history. Patients with these lesions frequently present with an acute neurological catastrophe, usually from intramedullary hemorrhage. They can also present with progressive myelopathy or with fluctuating neurological symptoms. The lesions are frequently associated with large aneurysmal varicosities which can produce significant cord compression. They are difficult to treat because of their usual intramedullary location and the fact that the feeders are often multiple, arising directly from the anterior spinal arterial system. However, successful treatment by embolization by feeder ligation, or by microsurgical resection has been reported.

Type III lesions are the juvenile malformation. Fortunately, these awesome lesions are uncommon. They occur most frequently in adolescents and young adults, are mostly intramedullary, and have multiple feeders involving sometimes several vertebral segments. Occasionally they have extramedullary, spinal, and paraspinous extensions. Their natural history is not well known, but most patients with these AVM's do poorly. The only available treatment is a partial resection in the volume of the lesion by embolization, feeder ligation, or partial surgical excision.

Clearly, our case does not fit into any of the above categories. There was no "glomus" or true AVM in our patient; rather, the anterior spinal artery communicated directly with a vein without an intervening small vessel network. In other words, our patient had a true arteriovenous fistula. He had no history of significant trauma and we must presume that the fistula was spontaneous. We were unable to find an identical case in the literature. We found two examples of patients with Rendu-Osler syndrome who had a cervical AVM draining through massively distended veins up to the posterior fossa simulating a posterior fossa AVM on vertebral angiography. In both of these patients, however, there was a compact nidus between the afferent arteries and the draining veins. It is likely that cases similar to ours have indeed occurred, but have been interpreted as unusual Type I or Type II malformations.

In our patient, the presenting progressive myelopathy could have been due partially to the mass effect from the distended venous varicosity, which clearly deformed the cord, although the preoperative metrizamide study did not show a complete block (Fig. 3). Venous hypertension, as previously discussed, could also have played a role. In addition, a steal phenomenon may have indeed been possible in our case, in view of the fact that the anterior spinal artery itself was involved in the fistula. The postoperative deterioration was most likely due to retrograde thrombosis in the anterior spinal artery above and below the level of ligation at the point of entry into the fistula. An alternative explanation would be thrombosis and swelling of the venous varicosity; however, a metrizamide CT scan performed at the time of deterioration did not show a significant block.

We felt justified to treat this patient surgically in view of the poor prognosis of progressive paraparesis in cases of spinal AVM's. Aminoff and Logue found that, of 60 patients presenting with leg weakness, 10 became unable to walk within 6 months and 28 within 3 years. Most of the remaining patients were significantly incapacitated within 5 years of onset of leg weakness. The logical treatment for our patient appeared to be direct ligation of the fistula. Although we had no previous experience with intradural exposure of the ventral aspect of the thoracic cord, we thought that this could be achieved through a standard transthoracic anterior approach by drilling the central aspect of two vertebral bodies. We were concerned about the effects of ligating the anterior spinal artery. We could demonstrate no other major intrinsic arterial supply to the thoracic cord, and feared that ligation of the spinal artery above and below the fistula could lead to retrograde thrombosis and result in cord ischemia, as indeed probably happened. However, we saw no alternative way to eliminate the fistula. Since performing this operation, we have read a report of a patient with an intramedullary spinal AVM who was treated by ligation of the anterior spinal artery feeding the lesion. This was accomplished by a transthoracic approach with resection of the T-8 vertebral body. Postoperative deterioration did occur, but the patient gradually improved over the ensuing months. Postoperative angiography showed residual filling of part of the lesion.

On the basis of this case, we propose a new category of spinal AVM, Type IV, to refer to direct arteriovenous fistulas involving the intrinsic arterial supply of the cord. In this type, a very distended venous outflow is to be expected. In our case, the massive venous drainage into the posterior fossa simulated a posterior fossa AVM on vertebral angiography. Symptoms can result from spinal cord compression by the distended veins, from venous hypertension, and from a true steal phenomenon, since the fistula involves the intrinsic arterial supply to the cord. When the exact location of the fistula is demonstrated, a relatively simple surgical approach with ligation of the fistula can be considered.

Acknowledgment

The authors wish to thank Dr. Raymond Kjellberg for kindly referring the patient.
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References


Manuscript received April 19, 1985.
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J. Neurosurg. / Volume 64 / January, 1986