Management of juvenile spinal AVM's by embolization and operative excision

Case report

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A small group of spinal arteriovenous malformations (AVM's), most commonly present in children or young adults, are characterized by a large size, high flow, the presence of multiple feeders, and frequent extension to paraspinal structures. Cardiac output requirements may be significantly increased by these so-called "juvenile" malformations, and a bruit is commonly noted. This report describes the obliteration of a juvenile spinal AVM. Staging of embolization and operative procedures was used to obliterate the AVM successfully without morbidity.

KEY WORDS - spinal arteriovenous malformation - juvenile spinal malformation - operative technique - embolization

Spinal cord arteriovenous malformations (AVM's) are most commonly classified into one of four categories: Type I, long dorsal or dural spinal AVM's; Type II, compact or glomus lesions, usually intramedullary with multiple feeders; Type III, large juvenile malformations; and Type IV, direct arteriovenous fistulas without an intervening network of small vessels.

Type III or juvenile malformations are characterized by their large size, high flow, and frequent extension to paraspinal structures including bone (Fig. 1). Management of these formidable lesions usually consists of partial embolization and ligation of feeding vessels. Attempts at resection have, in general, been unsatisfactory. We report the complete obliteration of a Type III juvenile malformation using a combination of preoperative and intraoperative embolization with staged resection.

Case Report

This 33-year-old female physician presented with a history of a progressive upper-extremity deficit. She was well until approximately 24 months before admission, when she noticed increasing severe fatigue in the left upper extremity. Approximately 6 months before admission, during the 3rd month of her second gestation, she noted the onset of sharp radiating pain through the third, fourth, and fifth fingers of the left hand, associated with a constant burning sensation over the left medial forearm. The pain was aggravated by coughing or straining and became most severe during the final weeks of her pregnancy. After her baby was born via a normal vaginal delivery, the patient was aware of some improvement in pain; however, progressive weakness of her left upper extremity continued. She began dropping objects from the left hand and felt numbness in the first and second digits. Two weeks before admission, she noted the onset of pain and fatigue involving the right proximal upper extremity. She denied any symptoms of weakness or sensory changes involving the lower extremities or changes in bowel or bladder habits.

Examination. The left biceps, deltoids, and intrinsic muscles of the left hand were mildly atrophied. Muscle strength was graded 4/5 in all muscle groups of the left upper extremity, with no weakness apparent in the other extremities. Deep-tendon reflexes were 2+/5 on the right side. On the left the biceps and brachioradialis reflexes were absent, the triceps reflex was trace only, and the knee and ankle jerks were graded normal at 2+/5. Testing of light-touch and pin-prick sensation was remarkable only for a moderate hyperesthesia in the C-8 nerve root distribution of the left hand. There
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Fig. 1. Artist’s interpretation of a Type III “juvenile” spinal malformation. Note the presence of intra- and extramedullary components as well as extension of the arteriovenous malformation into the extraspinal soft tissues and bone.

Fig. 2. Axial computerized tomography section at the C5-6 vertebral level demonstrating uniform contrast enhancement of the arteriovenous malformation which extends from the spinal canal through the neural foramen. The carotid arteries and jugular veins are well visualized just anterior and lateral to the spine.

was no evidence of agnosias, apraxias, or extinctions. Ambulation was normal and cerebellar testing revealed no deficits. Auscultation over the chest and neck revealed a low-pitched Grade III/VI bruit over the suprascapular region bilaterally and over the left posterior neck. The remainder of the neurological and general physical examination was unremarkable.

Computerized tomography of the cervical spine revealed a vascular lesion involving the anterolateral aspect of the spinal canal, most prominent at the C-5 and C-6 levels (Fig. 2). Selective and subselective angiographic evaluation, using a combination of cut-film and digital subtraction techniques, demonstrated a large AVM with extra- and intraspinal components extending from C-5 to C-7 (Fig. 3). Major feeding vessels were supplied by branches from the right and left ascending cervical arteries, the right and left vertebral arteries, and the left thyrocervical trunk.

Preoperative Embolization. The initial approach to treatment in this patient consisted of selective transfemoral embolization of the arterial feeders to the AVM arising from the left thyrocervical trunk. The embolization procedure was performed using Ivalon particles ranging in size from 150 to 300 μ. Postembolization angiography demonstrated closure of the vascular channels to the malformation from this branch and a significant reduction in flow to the AVM from the left anterior cervical artery. The patient tolerated this procedure well without changes in neurological function.

Operation. Stage I. One day after embolization, the patient was taken to the operating room and a laminectomy was performed at the C4–6 levels. Bone dissection was extended laterally through the C5–6 nerve root foramen, exposing the dural root sleeve and sensory ganglion. Bleeding from the extensive vascular channels passing through the bone elements of the spine (Fig. 1) was controlled by packing with bone wax and Oxyel cotton. As predicted by angiography, inspection revealed grossly abnormal vasculature over the posterolateral aspects of the spinal dura and the C-6 dural root sleeve. A large arterial feeder to the AVM previously visualized by angiography was identified as it passed adjacent to the dural sleeve of the C-6 nerve root. With the aid of the operating microscope, this vessel was isolated, cannulated, and embolized to occlusion with Ivalon particles. The Ivalon particles were suspended in 30% Conray and injected under continuous fluoroscopic guidance. The progress of embolization was followed intraoperatively by periodic digital subtraction angiography.

The cervical dura and arachnoid were opened in the midline, exposing the intradural and intramedullary components of the malformation. The dentate ligaments on the left at C-4, C-5, and C-6 were divided, and the cord was mobilized medially and dorsally exposing a tangle of arterialized vessels ranging from 1 to 4 mm in diameter. These vessels were coagulated with the bipolar coagulator. A small intramedullary component of the malformation was resected from the dorsolateral surface of the cord at the C-6 level. The dural components of the malformation were obliterated with bipolar cautery, and attention was turned to the abnormal vasculature surrounding the C-6 nerve root sleeve and dorsal ganglion. Multiple small feeder vessels extending between the ganglion and adjacent left ver-
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FIG. 3. Selective subtraction views demonstrating the origin of abnormal vessels feeding the arteriovenous malformation. These vessels arise from the left (a) and right (b) vertebral arteries, and from multiple branches of the left (c) and right (d) ascending cervical arteries. Note the extremely rapid filling and marked dilatation of the draining veins, characteristic of these high-flow lesions.

tebral artery were coagulated and divided. The dura was opened over the root sleeve and ganglion and the sensory and motor fibers were dissected free; additional vascular components of the AVM beneath the dural surface were then obliterated with bipolar coagulation. The dural edges were brought into approximation, and the operative incision was closed in the usual manner.

Postoperatively, the patient did well with no major changes in neurological function. Angiography on the 5th postoperative day revealed a small residual portion of apparently extramedullary AVM ventrolaterally on the left side at the C-5 level. This residual AVM was supplied by radicular branches from the left and right vertebral arteries, as well as branches from the left anterior cervical artery.

Operation, Stage II. The day following angiography the patient was returned to the operating room. The previous incision was reopened, and the residual AVM was identified within the dura at the C-5 level. The dural component was obliterated. After a thorough inspection revealed no additional AVM within the operative field, the incision was closed in the usual manner. The patient was taken directly to angiography under general anesthesia, where evaluation demonstrated persistence of a small extraspinal component of the AVM at the C5–6 level fed by branches from the anterior cervical artery. This extraspinal portion was obliterated by selective embolization with Ivalon particles. A final angiogram demonstrated complete obliteration of the AVM (Fig. 4).

Postoperative Course. Following these procedures the patient suffered a transient increase in weakness in the deltoid, biceps, and triceps muscles on the left side. Grip strength, lower-extremity function, and bowel and bladder function remained unchanged. She rapidly improved and within 6 weeks returned to full strength with no residual deficits. The patient has since resumed her practice as a family physician and reports no limitation in activities.

Discussion

Type III spinal cord AVM's occur most frequently in adolescents and young adults, hence the term "juvenile." These lesions are characterized by their large size, rapid flow, and the presence of multiple feeders with extension to paraspinal structures. Large tangles of vessels in the mediastinum, pelvis, and retroperitoneal space are often associated with this type of spinal AVM. Cardiac output requirements may be significantly increased, usually as a result of the extraspinal component. A bruit is commonly heard on auscultation over the spine or, more rarely, is audible at some distance.19

Fortunately, these formidable lesions are rare. In our own series of 30 spinal cord AVM's, only one patient has fit into this category. Other authors have reported a similar low incidence. In a series reported by Om-maya, et al.,19 two of 26 patients had "juvenile" type malformations; Malis,14 in reviewing his personal experience with spinal AVM's, reported that three of 43 patients had Type III lesions. In general, the clinical presentation of patients with spinal AVM's may involve either progressive neurological deterioration or sudden (occasionally catastrophic) onset of neurological deficits.
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with or without subarachnoid hemorrhage. Symptoms are frequently aggravated by certain postures or activities such as coughing and straining or, as in the present case, by pregnancy.\(^1,4,10,17\)

With the exception of the direct trauma caused by subarachnoid hemorrhage, the pathophysiological basis for these neurological changes remains uncertain. At least four mechanisms have been proposed to explain these clinical features: 1) a vascular steal with resulting ischemia; 2) mass effect created by the distended tangle of abnormal blood vessels and draining veins, and the occasional venous aneurysms which are frequently thrombosed; 3) venous congestion secondary to the anomalous shunt, resulting in a reduced arteriovenous pressure gradient and decreased intramedullary blood flow; and 4) mild or subclinical hemorrhage with resulting arachnoiditis, scarring, and secondary ischemia. Some combination of these mechanisms is likely responsible for the clinical progression of symptoms in the patient with a Type III malformation, and can be expected to vary over time.

Whatever the mechanism, AVM's of the spinal cord have a poor prognosis if left untreated.\(^2,23\) In the classic report by Aminoff and Logue,\(^2\) only 9% of 60 patients were capable of unrestricted activity 3 years after the onset of symptoms, and 50% of patients became severely disabled during this short period.

Management of Type III juvenile spinal AVM's has usually consisted of partial embolization and ligation of the feeding vessels. Ommaya, \textit{et al.},\(^19\) reported marked clinical improvement of deficits in one patient with a Type III spinal AVM following embolization of a major feeding branch. Djindjian\(^4\) has also reported clinical improvement in patients treated with partial embolization. Attempts at complete resection of these lesions have usually been unsatisfactory. Malis,\(^14,16\) in describing his experience with the operative management of juvenile spinal AVM's in three patients, reported one death, no change in one patient who was severely debilitated prior to surgery, and a poor outcome (paraplegia) in the remaining case.

Based on our experience with the surgical management of large, complex, high-flow intracranial AVM's, a treatment protocol has been developed utilizing staged pre- and/or intraoperative embolizations combined with resection.\(^20,21\) This same approach was used to obliterate the juvenile spinal AVM presented in this case report. Staged transfemoral and intraoperative embolization of the AVM feeders to this spinal malformation was used to eliminate the extradural and much of the extramedullary component. Embolization was also helpful in reducing flow through the residual AVM and thus decreasing the risks of hemorrhage during surgery. The size and composition of the embolic material are critical to the safety of embolization of spinal AVM's, particularly if the AVM is supplied by branches that also feed the anterior spinal artery (as in this patient). In such cases, there are two requirements for safe embolization: 1) the arteries supplying the AVM must be larger than the anterior spinal artery; and 2) the particles chosen for embolization must be larger in diameter than the anterior spinal artery itself. We prefer to use Ivalon sponge (polyvinyl acetate) particles occasionally mixed with 1- to 2-mm Gelfoam cubes for the embolization of AVM's. Embolization with Ivalon particles has several advantages: these particles are permanent, are available in a range of sizes from 150 to 1000 \(\mu\)m in diameter, and are thrombogenic, so that over time thrombosis tends to extend within the AVM. The embolic material is mixed as a slurry with 30% Conray solution, and the progress of embolization is followed by fluoroscopy and digital subtraction angiography. While embolization may be crucial in the management of complex spinal AVM's, its use as the sole treatment for these lesions must be viewed with caution. Lasting benefit has not been established, and the incidence of recanalization, as occurs in cerebral AVM's after embolic therapy alone, remains unknown. Indeed, Hall, \textit{et al.},\(^8\) have reported delayed recanalization of spinal AVM's with the return of symptoms in five patients who had previously undergone complete transfemoral embolic obliteration of their lesions.

Monitoring the patients' neurological status is necessary to ensure safe embolization of spinal AVM's. During transfemoral embolization, the patient can be challenged by perfusion of Amytal (amobarbital) and/or lidocaine or by proximal and reversible occlusion of the vessel(s) to be embolized.\(^9\) During operative procedures, somatosensory evoked potentials are routinely monitored; however, they only permit evaluation of
dorsal column function. Experimental techniques such as recording motor evoked potentials will allow monitoring of the corticospinal tracts and should be useful for assessing anterior cord function.

Operative resection of intraspinal components is performed using standard microsurgical technique. Reports by multiple authors have demonstrated that intramedullary spinal AVM’s can be resected with good results. The techniques are essentially those used for resection of intramedullary cord tumors.

The importance of postoperative angiography cannot be overstressed. In this case, follow-up angiography demonstrated the presence of residual intraspinal AVM and permitted repeat surgery to be performed before the development of extensive scarring. The presence of scar tissue from previous surgery or subarachnoid hemorrhage can severely limit safe dissection in what would otherwise be a resectable spinal AVM. Repeat angiography also identified residual extraspinal components of the AVM which were obliterated by selective embolization.

References


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