Perimedullary arteriovenous fistula

Spinal cord vascular malformation, intradural located either on the surface of the cord or just under the pia without an intervening nidus.

There is a shunt between a radicular artery and intradural veins. The lack of a nidus differentiates these lesions from the type II and III spinal AVMs and the intradural location of the arteriovenous shunt with the involvement of arteries feeding the cord differentiate it from dural AVFs.

This type of spinal arteriovenous malformation was first described by Djindjian et al. in 1977, and subsequently classified as type IV spinal arteriovenous malformations (AVMs) by Heros et al.

Type 4 spinal vascular malformations of the American English French connection classification.

Epidemiology

They constitute 8-19% of spinal vascular malformations and are predominantly found in the thoracolumbar region, either on the anterior, lateral or posterior surface of the cord.

Classification

sub type I: single arterial supply (ASA), single small fistula, slow ascending perimedullary venous drainage

sub type II: multiple arterial supply (ASA and PSA), multiple medium fistulae, slow ascending perimedullary venous drainage

sub type III: multiple arterial supply (ASA and PSA), single giant fistula, large ectatic venous drainage

According to the generally accepted Anson and Spetzler classification, three subtypes are identified according to shunt flow and degree of vascular enlargement. In type IVa PMAVF, there is a slow-flow single shunt between a non-dilated anterior or posterolateral spinal artery and a spinal vein. Type IVb lesions show greater flow than in type IVa and an ampullary dilatation of the venous side of the shunt. Increased shunt flow causes dilated tortuous intradural veins. PMAVFs generally have more than one feeder, usually a dilated anterior spinal artery and one or two posterior spinal arteries. Type IVc PMAVFs, called giant perimedullary AVFs, have multiple high-flow dilated feeding arteries and gross dilatation of draining veins; varices or true venous aneurysms are encountered either near the shunt or more distally.

Sometimes there is difficulty in accurate labelling type IVa and IVb lesions. A revised classification by Rodesch et al. simplifies the task and separates PMAVFs into macro-AVF and micro-AVF. Macro-AVFs are high-flow direct shunts fed by one or more spinal cord arteries ending in a venous ectasia with secondary perimedullary venous drainage. Micro-AVFs are small lesions fed by one or more slightly enlarged arteries draining into veins that are not ectatic.

Clinical features

Due to progressive myelopathy, spastic paresis, sensory disturbance, and bowel and bladder
dysfunction can be the presenting symptoms. Cervical spinal arteriovenous fistulas (AVFs) are even rarer. These lesions differ from the ones in the thoracolumbar region and have an even wider mode of presentation.

**Differential diagnosis**

**Spinal dural arteriovenous fistula** (SDAVF) are considered to be acquired and should be distinguished from congenital intradural perimedullary arteriovenous fistulas (PMAVFs).

**Treatment**

The location and size of the fistula dictates the treatment strategy. The tiny filum terminale lesions appear to be a good indication for surgical treatment owing to accessibility problems with the endovascular approach. However, surgical treatment is no less challenging as sometimes a fistula embedded between congested veins is difficult to find intra-operatively and localization and eradication of the shunt may become very difficult.

**Case series**

2010

Spinal perimedullary arteriovenous fistula (AVF) or dural arteriovenous fistula (DAVF) presenting as intracranial subarachnoid haemorrhage (SAH) is uncommon. A total of 16 cases have been reported to date. A majority of the reports described cervical spinal DAVF, while two other case reports described intracranial SAH secondary to lumbar and thoracic DAVF, respectively. We report a 61-year-old Chinese man with intracranial SAH secondary to thoracic DAVF aneurysm, who presented with sudden, severe chest pain, initially suggestive of aortic dissection/acute myocardial infarction. However, a careful examination of the history and physical signs, followed by appropriate and timely investigations enabled effective treatment to be administered promptly with a good outcome. This serves to illustrate the importance of investigating the entire cerebrospinal system when neurological symptoms and clinical signs suggest extracranial primary pathology.

2005

Thirty-two SCAVFs (in 22 adults and 10 children) were treated between 1981 and 2000. These lesions were classified as microarteriovenous fistulas (mAVFs) or macroarteriovenous fistulas (MAVFs) according to shunt morphology. Location, architecture, presenting symptoms, and age group were detailed. The selection of patients for endovascular versus surgical treatments was analyzed, as were the anatomic and clinical results obtained by embolization with n-butylcyanoacrylate. Clinical status was evaluated according to the Karnofsky Performance Scale score.

Ten SCAVFs were found in the pediatric population (four mAVFs and six MAVFs). All four mAVFs presented with acute symptoms. Three mAVFs (two cervical and one thoracic) presented hematomyelia; in one patient with a thoracic AVF, subarachnoid hemorrhage was suspected. All six MAVFs were located in the thoracolumbar cord (five associated with hereditary hemorrhagic telangiectasias). Four of the six MAVFs presented with hemorrhage. In the adult population, there were 21 mAVFs (95%) and one MAVF (5%). Only two mAVFs were found in the cervical cord, all other shunts affecting the thoracolumbar region. Hemorrhage was present in 6 of the 22 cases seen in adults (27%). The symptoms of SCAVFs did not differ from those found in spinal cord arteriovenous shunts of nidus type. Pial venous reflux and congestion were the most frequently encountered features in both the adult and pediatric groups. Arterial aneurysms (different from false aneurysms)
Perimedullary arteriovenous fistula were not found in association with hemorrhagic presentation of SCAVFs. Mean follow-up in our series was 3.3 years. Of the MAVFs, 86% were embolized, with 67% cured. The others had more than 75% occlusion. All patients followed up improved significantly. Of the mAVFs, 48% were treated endovascularly. Successful embolization was performed in 75% of patients. One patient was not embolized because of vasospasm, whereas 67% percent of mAVFs were completely occluded, 22% were more than 90% occluded, and 11% were 75% occluded. Complementary surgery was deemed unnecessary. All patients with mAVFs improved significantly at follow-up. Transient complications occurred in 22% of all patients, with no permanent morbidity or mortality. No patient bled or rebled after embolization. Thirty-six percent of mAVFs were operated on because of anticipated technical difficulties for endovascular approach or distal localization of the shunt.

Endovascular treatment of SCAVFs stabilizes, normalizes, or improves neurological symptoms in all patients at long-term follow-up, with no bleeds or rebleeds. Embolization of SCAVFs with glue is a safe treatment that compares favorably with other approaches and significantly improves the poor natural history of the disease.

Nineteen patients with PMAVF (Type IVa in 9 patients, Type IVb in 6, and Type IVc in 4) were treated at Seoul National University Hospital between January 1988 and March 2001. Their mean age was 28 years (range, 6-52 yr), and the male-to-female ratio was 1.7:1. The mean follow-up period was 20 months (range, 2-55 mo). Most patients presented with symptoms of slowly progressive myelopathy (13 patients). On spinal angiography, all but 2 showed fistula at the level of the conus medullaris. The feeder was the anterior spinal artery and/or the posterior spinal artery in 14 patients and the posterior spinal artery in 5. All patients underwent endovascular or surgical treatment.

With endovascular treatment (11 patients; IVa, n = 5; IVb, n = 2; IVc, n = 4), complete angiographic obliteration of fistula was performed in 5 and partial obliteration in 4 (IVa, n = 1; IVb, n = 2; IVc, n = 1). Symptomatic improvement or arrest of progression was achieved in 5 of 9 patients with complete or partial occlusion. Embolization failed in two (IVa, n = 1; IVc, n = 1). With surgery (10 patients [IVa, n = 6; IVb, n = 4], including 2 patients with partial or failed embolization), most (9 of 10) were improved or stable.

Good results were achieved with surgery for Types IVa and IVb PMAVF located at the level of the conus medullaris. For Type IVc PMAVF, a fistula located on the ventral side of the spinal cord or above the conus medullaris, endovascular treatment might be considered. Because of rapidly evolving endovascular techniques, however, further studies are warranted.

1993

A series of 35 patients treated for an intradural perimedullary arteriovenous fistula (AVF) between 1970 and 1990 is reported. Angiography was performed on all of the patients, leading to the diagnosis. The patients were classified into Type I (4 patients), Type II (9 patients), and Type III (22 patients). One Type I patient was not treated, two others underwent surgery, and the last one was embolized. All of the Type II AVFs were treated, two by embolization, four by direct surgery, and three by surgery after incomplete embolization. All of the Type III AVFs were treated by endovascular detachable silicone balloon. Complete occlusion of the AVF was achieved in all treated cases of Types I and II AVF and in 15 cases of Type III AVF; for the 6 other cases of Type III AVF, incomplete occlusion was achieved. In the Types I and II AVFs, partial improvement was clinically observed in only half of the patients; the others remained unchanged. The 15 patients whose Type III AVF was completely embolized recovered completely, and four patients with Type III AVF who were incompletely embolized remained unchanged; 2 other patients with Type III AVF worsened after incomplete
occlusion, and 1 additional patient died a few hours after an attempt of endovascular occlusion of a cervical Type III AVF. The place of the perimedullary AVFs among the other vascular malformations involving the spinal cord is discussed according to this classification into three types. Their specific diagnostic and therapeutic difficulties are discussed, resulting in a simplified classification including two types of perimedullary AVF.

**Case reports**

Mühl-Benninghaus et al. report on a 9-year-old boy with a cervical PMAVF manifesting with headache and vertigo.


