Perimedullary arteriovenous fistula

- Progressive quadriparesis in a young woman due to a spinal perimedullary arteriovenous fistula (PMAVF type IVa) successfully treated with endovascular therapy: A case report
- Endovascular treatment of a craniocervical junction dural arteriovenous fistula associated with lateral medullary syndrome: A case report
- Intradural venous engorgement of CSF-venous fistula mimics spinal dural arteriovenous fistula on MRI: A novel case report and review of literature
- Treatment of 23 spinal perimedullary arteriovenous fistulas in a single center: A simple and practical treatment strategy
- Rare and Easily Misdiagnosed Intracranial and Craniocervical Junction Dural Arteriovenous Fistulas With Spinal Perimedullary Drainage
- Evaluating the Role of Onyx Embolization in the Management of Spinal Dural Arteriovenous Fistulas: A 20-Year Single-Center Experience
- Successful endovascular treatment for pediatric ruptured cervical spinal perimedullary arteriovenous fistula: A case report
- Surgical Management of Spinal Dural Arteriovenous Fistula: A Case Report

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Spinal dural arteriovenous fistula (SDAVF) and perimedullary arteriovenous fistula (PAVF) are two distinct types of spinal vascular malformations within the spinal cord and its surrounding structures. They differ in their location, anatomy, and clinical presentation.

This spinal cord vascular malformation, is intradural located either on the surface of the cord or just under the pia without an intervening nidus [1].

There is a shunt between a radicular artery and intradural veins [2].

The lack of a nidus differentiates these lesions from the type II and III spinal arteriovenous malformations and the intradural location of the arteriovenous shunt with the involvement of arteries feeding the cord differentiates it from dural arteriovenous fistulas.

This type of spinal arteriovenous malformation was first described by Djindjian et al. [3] in 1977, and subsequently classified as type IV spinal arteriovenous malformations (AVMs) by Heros et al [4].

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 [5].

Classification

Perimedullary Arteriovenous Fistula Classification.

Clinical features

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).

Complications

Spinal arteriovenous fistulas (SAVFs) are underdiagnosed entities that can lead to severe morbidity from spinal cord dysfunction or hemorrhage

Treatment

Perimedullary arteriovenous fistula treatment.

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 [6)].

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) 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 [7)].

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 [8].

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 [9].

Case reports

Lara-Reyna et al. from the Department of Neurosurgery, OSF Saint Francis Medical Center, Peoria, Illinois present a case of intraluminal microsurgical access for occlusion with a hemostatic agent of a type IV SAVF near the conus medullaris as an alternative to clip occlusion to avoid nerve root compromise.

Temporary microsurgical clipping of the SAVF led to nerve root compromise detected via intraoperative monitoring. Instead, the authors advanced elongated pieces of a hemostatic agent directly into the arterial lumen via arteriotomy to create direct obliteration of the fistula without intraoperative monitoring changes.

Lessons: In patients unable to tolerate clipping of the SAVF because of nerve root involvement and neurophysiological signal decline, open access of the vessels and direct intraluminal obliteration using a hemostatic agent should be considered as an alternative method of fistula occlusion [10].

A case of microsurgical disconnection of a PMAVF supplied by the artery of Adamkiewicz with fistulation at the ventral spinal cord [11].

Mühl-Benninghaus et al. report on a 9-year-old boy with a cervical PMAVF manifesting with headache and vertigo [12].

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