Filum terminale arteriovenous fistulas: the role of endovascular treatment

Clinical article

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Object. The authors describe the clinical presentation, imaging features, and management of patients presenting with filum terminale arteriovenous fistulas (FTA VFs) and the role of transarterial treatment in their management.

Methods. The authors retrospectively reviewed data obtained in 10 patients with FTA VFs diagnosed between January 1990 and December 2011.

Results. Most patients (70%) were male, and the age of the population ranged from 31 to 72 years (mean 58.2 years). Clinical presentation was progressive paraparesis and sensory loss in the lower extremities in 9 cases, back pain in 7, radicular pain in 3, bowel/bladder disturbance in 5, and impotence in 1. The duration of symptoms varied between 2 and 24 months. Initial MRI studies showed intramedullary increased T2 signal, swollen cord, and dilated perimedullary veins in all patients. One patient had syringomyelia, presumably caused by venous hypertension transmitted by the perimedullary venous system. Embolization was attempted in 7 patients and was curative in 6 patients. Surgery was performed in the other 4 patients in whom embolization was unsuccessful or deemed not feasible. There was no treatment-related complication in either group. Symptoms, venous congestion in the cord, and syringomyelia improved on follow-up in all patients.

Conclusions. Embolization should be considered the treatment of choice for FTA VFs and can effectively treat the majority of patients presenting with an FTA VF. In a smaller group of patients in whom the angioarchitecture is unfavorable, open surgery is recommended.

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Key Words • filum terminale • spinal arteriovenous malformation • myelopathy • endovascular treatment • embolization • vascular disorder

Filum terminale arteriovenous fistula is an extremely rare type of SAVS that is defined as an abnormal communication of the artery of the filum terminale (the continuation of the anterior spinal artery) and the spinal vein (perimedullary vein). This direct communication leads to an increase in venous pressure in the perimedullary veins that may result in spinal cord congestion and edema. Clinical presentation is usually the result of progressive myelopathy or conus medullaris syndrome. Magnetic resonance imaging is the best modality by which to diagnose the radiological sequelae of the disease. Magnetic resonance angiography is an optional investigation that can differentiate other disorders from FTA VFs and other lesions that can mimic FTA VFs, such as SDAVs. Both endovascular treatment and surgery have been used to manage this condition. Endovascular treatment is usually technically difficult, due to the long and tortuous course of the anterior spinal artery and the artery of the filum terminale supplying the FTAVF.

This article, which combines the data from 3 large referral centers, describes the clinical presentation, MRI and angiographic findings, treatment, clinical outcomes, and follow-up MRI findings of FTA VFs and represents the largest reported series to date of patients with this condition.

Methods

We retrospectively reviewed all cases of FTAVF proven by spinal angiography between 1990 and 2011. Age,
sex, clinical presentations, MRI and angiographic findings, type and complication of treatment, and clinical outcomes were reviewed. In all cases the patients had been assessed clinically during admission and at 3–12 months after treatment. Follow-up MRI and/or spinal angiography were performed in most patients.

All patients underwent MRI examination and diagnostic spinal angiography while under general anesthesia. A diagnostic 5-F Cobra catheter (Merit Medical System) and a 0.038-in hydrophilic guidewire (Terumo) were inserted in each diagnostic angiography case to select potential arterial feeders—that is, intercostal, lumbar, and lateral sacral arteries (branches of internal iliac arteries).

At the time of embolization a 1.2-F Magic microcatheter (Balt) and 0.008-in Mirage microguidewire (ev3) were inserted into the 5-F Cobra catheter. The microcatheter was then advanced along the anterior spinal artery and the artery of the filum terminale until its tip was in the correct position. Liquid material, NBCA, was mixed with Lipiodol (NBCA concentration varied between 15% and 30%) and injected to obliterate the shunting zone. A control angiogram was obtained after embolization to assess the result of treatment.

In those cases in which embolization was unsuccessful or technically not feasible, surgery was performed; this involved clipping and disconnection of the proximal arterialized vein to cure the fistula.

### Results

A retrospective review of the neurovascular databases of 3 hospitals totaling 362 patients with SA VSs identified 10 patients with an FTAVF. Most of the patients were male (70%) and age of the cohort ranged from 31 to 72 years (mean age 58.2 years). Clinical presentation was back pain (7 patients; 70%), radicular pain (3 patients; 30%), both progressive paraparesis and sensory loss in the lower extremities (9 patients; 90%), bowel/bladder problems (5 patients; 50%), and impotence (1 patient; 10%). The duration of the symptoms ranged from 2 to 24 months (mean 10.8 months). Details are shown in Table 1.

All patients underwent MRI that demonstrated increased T2 signal within the lower thoracic cord and conus medullaris, enlargement of the affected cord segment, and dilated perimedullary veins (Figs. 1A and 2A), suggestive of an SA VS. Among SAVS diseases, SDAVF is often the first diagnosis to be considered because of its high prevalence.\(^1,2,4\) No evidence of hematomyelia in the spinal cord or conus medullaris was found. In 5 of 6 patients in whom gadolinium was administered, patchy intramedullary enhancement was shown (Figs. 1B and 2B). Three patients underwent contrast-enhanced time-resolved MR angiography, which revealed abnormal contrast filling of the perimedullary veins in the early arterial phase, confirming the diagnosis of SAVS; however, the exact location of the fistula could not be determined (Fig. 2C). In addition, one patient (Case 8) had syringomyelia from T11–12 to L2–3 and coexisting congenital anomalies such as a tethered cord with the tip of the conus medullaris at the inferior L-4 vertebral level and a hypoplastic lower sacrum.

One patient (Case 1) had a 4-year history of spinal stenosis and had undergone laminectomy and spinal fixation.
with metallic rods and screws. The orthopedic surgeon suspected that recurrent spinal stenosis was responsible for the recurrent symptoms. Lumbar myelography showed intradural, extramedullary serpentine filling defects along the lower thoracic cord and conus medullaris, but no significant spinal canal stenosis was seen (Fig. 3A). The result of this lumbar myelography suggested an intradural SAVS. Spinal MRI was performed to confirm the diagnosis (Fig. 3B).

Spinal angiography findings showed variable origins of the anterior spinal artery supplying the fistulas. The fistulas were supplied from the anterior spinal artery and had various locations between L2–3 and S2–3 (Figs. 1C, 2D, and 3C). There was a small venous ectasia of the proximal vein, just distal to the fistula in one patient (Case 4). A coexisting SDAVF on the right side of the sacrum was found in one patient (Case 2).

In 8 patients (Case 1–7 and 9), the multidisciplinary neurovascular team recommended endovascular treatment as the preferred management choice, but one patient (Case 3) declined embolization and elected to have surgery instead. In 6 of 7 patients a single session of embolization produced satisfactory results as demonstrated by the embolic material reaching the proximal aspect of the draining vein (Fig. 3D). In one patient (Case 9) embolization was recommended and attempted, but the tip of the microcatheter could not be placed in a suitable position and a surgical procedure was subsequently performed.

In the remaining 2 patients (Cases 8 and 10) surgery was recommended because of the extreme tortuosity of the anterior spinal artery made the success of a transarterial approach unlikely. In the end, successful disconnection of the fistulas by surgical means was achieved in 3 cases. The patient in Case 8 also underwent detethering of the cord during the same surgical session. There were no immediate or delayed postoperative complications.

The clinical assessment performed in all patients soon after treatment and again between 3 and 12 months after treatment classified patients into 1 of 4 categories: complete recovery, partial recovery, stable symptom, or worsening symptom. All patients who presented with back pain and/or radicular pain were classified as complete recovery. Patients who presented with paraparesis were classified as complete recovery in 2 cases and partial recovery in 7 cases. All patients who presented with sensory loss in the lower extremities were classified as partial recovery in 9 cases. Of the patients who presented with bowel or bladder dysfunction, 2 were classified as partial recovery and 3 were classified as stable. The patient who presented with sexual dysfunction (impotence) was stable. No clinical worsening or new symptoms were found during the follow-up in any patient.

Six of 10 patients underwent follow-up MRI, and 4 of 10 patients underwent follow-up spinal angiography. Three patients underwent neither MRI nor spinal angiography for logistical reasons. In 6 patients follow-up MRI performed between 3 and 24 months posttreatment demonstrated normal cord size and resolution of the increased intramedullary T2 signal and dilated perimedullary veins (Figs. 2E and 3E). Magnetic resonance imaging performed in the patient with syringomyelia (Case 8) on postoperative Day 6 revealed marked shrinkage of the syrinx (Fig. 1D).
Discussion

Spinal vascular arteriovenous shunts are generally divided into 4 groups according to their relationship to the dura: paraspinal, epidural, dural, and intradural shunts. The intradural arteriovenous shunts can be radicular, filum terminale, perimedullary, or spinal cord in location. The FTAVF is the least common location of these shunts, and only a few cases have been published. The filum terminale is normally supplied by the artery of the filum terminale, which arises from the termination of the anterior spinal artery at the inferior aspect of the conus medullaris—that is, it represents the continuation of the anterior spinal artery. The vein of the filum terminale travels ventral to the filum terminale (but dorsal to the artery) and runs cephalad to join the anterior spinal venous system. When there is an abnormal communication between the artery and the vein of the filum terminale, an AVF, both vessels are enlarged. The fistula will then cause an increase of intravascular pressure in the veins around the spinal cord, resulting in a disturbance of spinal cord drainage. Consequently, the spinal cord becomes congested, and venous ischemia and infarction will occur if no treatment is provided.

Clinical presentations of FTA VFs consist of gradual onset of back pain, radicular pain, motor weakness, decreased sensation of the lower extremities, and bowel and bladder dysfunction (urinary incontinence, difficulty in urination and defecation, and sexual disturbance) resulting from thoracic cord and conus medullaris venous congestion. These symptoms and the pathophysiology may mimic those of other SAVSs such as the more frequently encountered SDAVFs. Our patients had symptoms similar to those reported in previous studies. Interestingly, one patient (Case 2) had no neurological deficit despite obvious increased T2 signal within the lower thoracic cord on MRI. His symptoms (back and radicular pain) and abnormal MRI findings resolved after complete closure of the fistula.

Magnetic resonance imaging of the spine is the modality of choice with which to evaluate spinal cord disease including SAVSs. The most important sequence to diag-
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nose SAVSs, in our experience, is the sagittal T2-weighted fast spin echo sequence, which may show hyperintensity within the cord and intradural tortuous vascular flow voids at the cord surface. However, the MRI findings of FTAVFs are nonspecific and can mimic SDAVFs and perimedullary AVFs that also demonstrate dilated perimedullary veins, increased intramedullary T2 signal, and cord swelling. Although MRI differentiation of these arteriovenous shunts is sometimes difficult, there are some clinical and imaging clues by which to differentiate these entities—for instance, the age of the patient and size of the venous ectasia. Perimedullary AVFs usually occur in young age groups (childhood, teenage, or young adult) and may contain a giant venous ectasia as has been reported in one case by Matushita et al. On the other hand, SDAVFs and FTAVFs usually occur in patients over 60 years old and of male sex, as was the case in the majority of cases in our study. However, we had 2 patients who were younger in age (31 and 39 years), which is uncommon in this disease. A venous ectasia in FTAVFs, if present, is usually small in size, similar to those in our study and as reported by Lim et al., but has never been reported in SDAVFs. In addition, contrast-enhanced MR angiography of the spine is very helpful in distinguishing FTAVFs and SDAVFs by showing different locations of the fistulas and arterial feeders. Demonstration of enlarged anterior spinal artery and artery of filum terminale suggests FTAVFs, whereas there is no such finding in SDAVFs.

However, spinal angiography has remained the gold standard and is still required in patients with SAVSs to precisely identify the location of the arteriovenous shunt and the supply to the lesion, as well as the supply to the spinal cord, and to exclude metachronous lesions. The radiculomedullary artery that contributes to the anterior spinal artery and continues as the artery of the filum terminale represents the arterial supply in FTAVFs. Dual blood supply to FTAVFs—that is, the contribution of the anterior spinal artery from two segmental arteries—has been described in previous studies but was not demonstrated in our series.

Treatment of FTAVFs can be either by endovascular or surgical means, and the choice of treatment depends on the experience of neurointerventionalists and neurosurgeons in each institute. The goal of treatment in FTAVFs is to obliterate the fistulous point in one of two ways: 1) by applying glue (NBCA) to the beginning of the draining vein during the endovascular procedure or 2) by clipping of the proximal arterialized vein during a surgical procedure. For embolization, a 1.2-F Magic microcatheter was inserted into the arterial feeder and navigated toward the distal part of the artery of the filum terminale, just proximal to the fistula, and a liquid glue mixture (15%–30% glue) was injected until the beginning of the vein was reached. Of the 7 cases in which the embolization was considered likely to be curative, this goal was achieved in 6 (85%). The overall success rate of embolization in our study was 60% (6 of 10 cases), which represents a higher success rate than previously reported. A very important factor that will enable superselective distal catheterization of the feeding vessel is for the guiding catheter to be in

![Fig. 3. Case 1. A: Lumbar myelogram showing intradural, extramedullary serpentine filling defects (arrows) along the thoracolumbar region. B: Sagittal T2-weighted MR image demonstrating increased T2 signal in the lower thoracic cord and conus medullaris (single arrow) and flow voids around the thoracolumbar area (double arrow). C: Selective angiogram of the left T-12 intercostal artery demonstrating an FTAVF at L-5 (arrow) that is supplied from the anterior spinal artery. The FTAVF drains to the perimedullary veins. D: Subtraction image showing the glue cast (arrow) occluding the fistula and proximal draining vein. E: Sagittal T2-weighted MR image at 1-year follow-up revealing resolution of the venous congestion of the spinal cord and dilated flow voids (perimedullary veins).]
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Duration of FU (mos)</th>
<th>Clinical Symptoms</th>
<th>Type of FU Investigation</th>
<th>MRI</th>
<th>Duration of FU</th>
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<td>CR</td>
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<td>resolution of increased T2 SI &amp; reduced size of PMV</td>
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<td>6</td>
<td>CR</td>
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<tr>
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<td>6</td>
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<tr>
<td>6</td>
<td>12</td>
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<td>9</td>
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<td>10</td>
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<td>PR</td>
<td>PR</td>
<td>12 mos</td>
<td>closure of fistula</td>
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* CR = complete recovery; FU = follow-up; PR = partial recovery.
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a stable position. The tip of the 5-F Cobra should be advanced and positioned distally in the segmental artery as close as possible to the origin of the arterial feeder (radiculo-medullary artery) to stabilize the microcatheter system. The anatomical and technical factors that may affect the stability of the guiding catheter have been noted by Fanning et al. From a point of view of risk, the filum terminale does not carry any neurological function, closure of the artery of the fistula does not result in neurologological complications, and the procedure thus should be safe unless there is reflux of the embolic material upstream to the anterior spinal arterial system. This is the reason why we favor injection of NBCA rather than Onyx (ev3) in this particular situation.

Surgery is recommended when embolization is technically not feasible or if the embolic material did not reach the beginning of the draining vein. When the fistula is obliterated, regardless of the type of treatment, the pressure in the perimedullary veins will decrease, as noted by Hassler et al. in the setting of post-treatment SDAVFs. According to this pathomechanism, the symptoms will gradually improve or at least stabilize. Evidence of improvement on follow-up MR images demonstrating a reduced size of the dilated perimedullary veins, decreased cord swelling, and decreased intramedullary T2 signal is also expected.

Understanding the cause of syringomyelia is helpful in explaining why this MRI finding improved in Case 8 (Fig. 1) after treatment. Syringomyelia is usually a secondary condition caused by other abnormalities such as a Chiari I malformation, posterior fossa tumor, or spinal cord tumor, with different mechanisms of syrinx formation such as blockage of the CSF flow pathway. The association of syringomyelia and SAVSs has rarely been reported. In their cases of spinal arteriovenous malformation, Srivatanakul et al. proposed a hypothesis of syrinx formation in which venous hypertension causes exudation and collection of fluid inside the spinal cord due to disruption of the blood-brain barrier. When the circulation and absorption of the CSF are unable to compensate for this overproduction of fluid, a syrinx is formed. This hypothesis can explain the significant improvement in the syringomyelia in our patient (Case 8) after disconnection of the fistula (Fig. 1D), resulting from a decrease of pressure in the perimedullary veins.

Limitations of this study are the small number of cases owing to rarity of the disease, short follow-up duration of clinical outcomes, lack of follow-up spinal angiography studies, and lack of follow-up MRI studies in some patients. Long-term MRI follow-up is recommended to assess spinal cord status. Incomplete cord recovery documented on MRI should warrant repeat spinal angiography to rule out residual, recurrent, or metachronous disease.

Conclusions

Endovascular embolization should be considered the treatment of choice for FTAVFs unless extreme tortuosity of the anterior spinal arterial feeder predictably precludes arterial access depending on local expertise.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Chanthanaphak, TerBrugge. Acquisition of data: all authors. Analysis and interpretation of data: Chanthanaphak, Pongpech. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Chanthanaphak. Statistical analysis: Chanthanaphak. Study supervision: TerBrugge.

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