Congestive myelopathy (Foix-Alajouanine Syndrome) due to intradural arteriovenous fistula of the filum terminale fed by anterior spinal artery: Case report and review of literature

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Abstract

Spinal arteriovenous fistulas are rare entities. They often present with congestive myelopathy but are infrequently diagnosed as the cause of the patients' symptoms. Only one such case has been described previously in Indian literature. We describe one such case who presented to us after a gap of 3 years since symptom onset and following a failed laminectomy where the cause was later diagnosed to be an intradural fistula in the filum terminale fed by the anterior spinal artery and review the available literature.

Key Words

Anterior spinal artery, congestive myelopathy, filum terminale, intradural arteriovenous fistula

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Ann Indian Acad Neurol 2013;16:436-40

Introduction

Spinal arteriovenous fistulas are rare entities. They may be extradural or intradural. The intradural variants may have feeders from the radicular arteries [called dorsal type] or anterior spinal artery [ventral type]. The artery of the filum terminale is a continuation of the anterior spinal artery and contributed to the fistula formation in our case. The high flow in these fistulas caused intramedullary venous hypertension and the entity of congestive myelopathy.

Case Report

A 54-year-old male presented with gradually progressive history of walking difficulty, weakness, and decreased sensation in lower limbs, diffuse gluteal, and leg pain and bowel and bladder incontinence of 3 years duration. He had undergone L4-L5 fenestration and microdiscectomy (in another hospital) 2 years previously for a disc bulge that was thought to be the causative pathology. As there was steady deterioration even after this procedure, he underwent nerve conduction studies that were inconclusive and was on various types of medication and physiotherapy. A repeat magnetic resonance imaging (MRI) of the whole spine was now carried out, which showed multiple flow voids in the dorsal sub arachnoid space (from C3 to L4) and T_2 hyperintense signal change in the dorsal cord from D6 body to the conus [Figure 1]. At current presentation, he was unable to stand, even with support. Bilateral foot drop was present. Right lower limb power was grade 3/5 in hip flexors and knee extensors and 2/5 in knee flexors, ankle plantar flexors, and hip extensors. Left lower limb power was grade 4/5 in hip and knee flexors and extensors and 3/5 in ankle plantar flexors. Power in bilateral ankle dorsiflexors, toe flexors, and extensors was grade 0/5. There was wasting of the muscles of the lower limb and foot. Bilateral knee and ankle jerks were absent. All modalities of sensation were decreased from L3 dermatome downwards bilaterally with complete perineal anesthesia.

Spinal digital subtraction angiography (DSA) showed an arteriovenous fistula (AVF) of the filum terminale at L4 level fed by a single mid-line descending anterior spinal artery (ASA) that was augmented by a radicular feeder from D12 level. A tortuous vein was looping back from L4 and running cranially [Figures 2 and 3].

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	DOI: 10.4103/0972-2327.116931

The patient underwent L3 laminectomy and disconnection of the AVF by coagulating and cutting the feeding artery and vein after application of separate "Liga" clips proximally and distally on both. Intra operatively, the vein was tortuous and thickened. There was no evidence of any filling of the vein after the artery had been initially disconnected. The arachnoid was clear and thin and had no stigma of prior subarachnoid hemorrhage. The patient had slow recovery of motor and sensory functions and also regained reasonable sphincter control in the following 6 months. His dysesthesias and leg and gluteal pain also disappeared. A repeat MRI done at 5 months follow-up showed disappearance of flow voids as well as cord T_2 hyperintensity [Figure 4]. Currently at 14 months follow-up, he is independently ambulant, continent and has full power in both lower limbs.

Discussion

Congestive myelopathy due to spinal arteriovenous fistulas (AVF's) was first described by Foix and Alajouanine in 1926 who called it necrotizing myelopathy[1,2] and subsequently by Lhermitte in 1931.[1] In 1974 Aminoff and Logue hypothesized that in the presence of an AVF there is increased intramedullary venous pressure with resultant decrease of the arteriovenous pressure gradient and concomitant decrease of cord perfusion.[1,3] Irrespective of the location of the fistula it is the lower part of the cord that is affected commonly and shows signal changes on MRI. Two reasons are put forward to this phenomenon– (a) the intraspinal venous system is valveless and due to gravity the lower part of the cord is more congested and (b) the collateral outflow from the lower part of the dorsal cord is less even in normal subjects[1] and this leads to impaired dissipation of venous hypertension found in patients with AVF's.

This affliction of the lower part of the cord leads to the commonly observed symptoms of dysesthesias, non-dermatomal sensory loss, walking difficulty, wasting, weakness of lower limbs, buttock pain, and sphincter dysfunction.[1-3]

Often this condition is misdiagnosed as polyradiculopathy, polyneuropathy, intramedullary tumor, etc., and the same has been highlighted by various authors.[1,3] Another cause of the misdiagnosis is the rarity of the condition[2] and has led one study (in the Netherlands) to speculate that a neurologist can

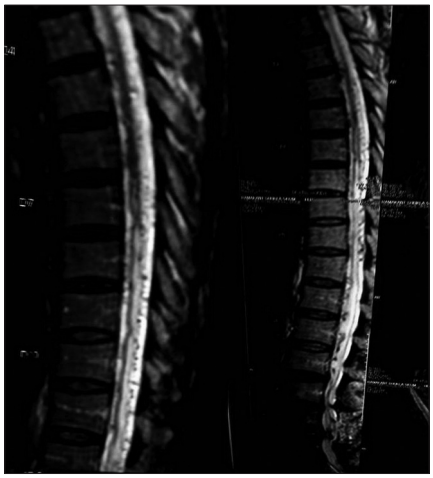

Figure 1: Magnetic resonance imaging spine (sagittal view) showing multiple flow voids in the dorsal sub arachnoid space (from C3 to L4) and hyperintense signal change in the dorsal cord from D6 body to the conus

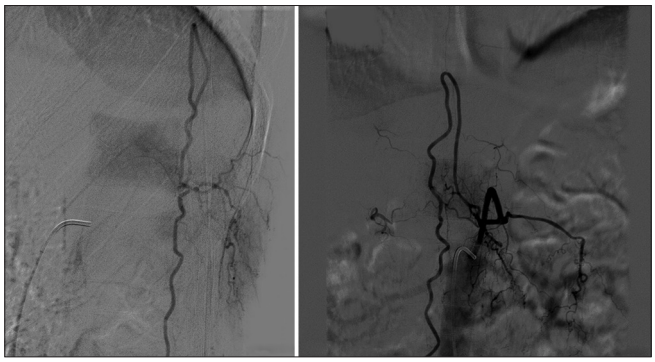

Figure 3: Anterior spinal artery is augmented by a radicular feeder from left D12 level that takes a hairpin bend as it descends. Digital subtraction angiography showing the arteriovenous fistula of the filum terminale with the enlarged, tortuous vein looping back from L4 and running back cranially

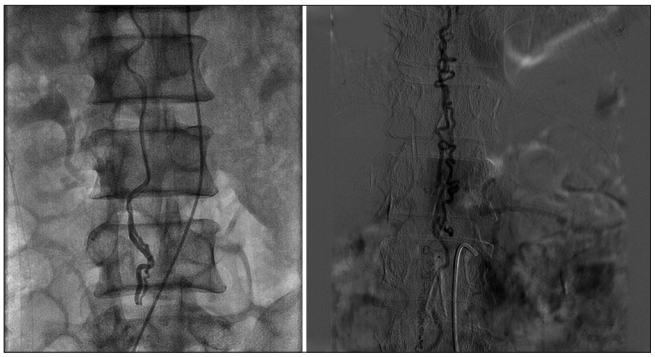

Figure 2: DSA showing an arteriovenous fistula of the filum terminale at L4 level fed by a single midline descending ASA. DSA (lateral view) showing that the arterial feeder (ASA) abutting the posterior cortex of the vertebral body

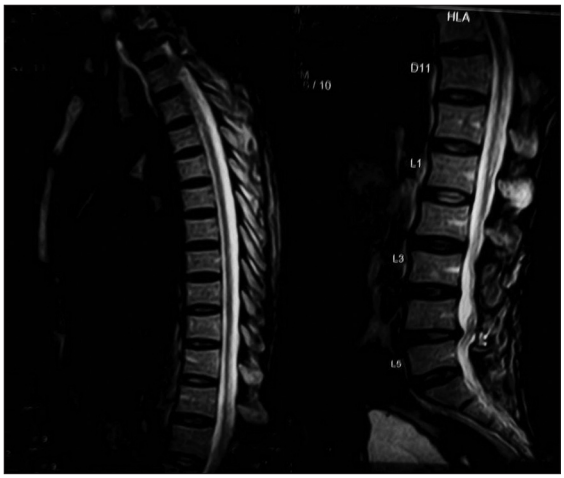

Figure 4: Magnetic resonance imaging done at 5 months follow-up showing disappearance of flow voids as well as cord hyperintensity

expect to see one such case only every 4-8 years.[1] The result is that 40-63% of patients have disease of 1-3 years duration and 10-34% of patients have disease for greater than 3 years by the time they are diagnosed.[1] Our patient too gave a history of more than 3 years duration in the course of which he had been subjected to lumbar laminectomy and multiple nerve conduction studies to no avail.

There are several classifications of spinal vascular lesions. Spetzler *et al*.[4] grouped them into three major classes [Table 1]. Of these spinal AVF's are further sub classified depending on their vascular supply [Table 2].

Intradural (ventral or perimedullary) AVF's as a separate entity were first described by Djindjian in 1977[1,5] and are said to account for 20% of all spinal vascular malformations.[3] Filum terminale AVF's is a rare variant[5,6] of intradural AVF's. The filum terminale is supplied by a single feeder that is a continuation of the ASA or a branch descending from the "vascular basket "at the conus.[5,7] There is no radicular arterial supply as there are no rootlets that exit the filum.[5] The vein of the filum terminale drains the blood either cranially into the ventral vein of the spinal cord or caudally into the sacrum extra dural plexus.[5,7] Hence any AVF below the conus that has supply only from the ASA with no lumbar radicular augmentation can be considered to be a filum terminale AVF.

Rodesch *et al*. found 3.1% prevalence of filum terminale AVF's in patients with intradural AVF's.[5] We performed a PubMed search using the terms 'congestive myelopathy' and

Table 1: Classification of spinal vascular lesions (Spetzler 2002)

Neoplastic vascular lesions
Hemangioblastomas
Cavernous malformations
Aneurysms
Arteriovenous lesions
Arteriovenous malformations (nidus present)
Arterivenous fistulas (nidus absent)

Table 2: Classification of spinal AVF's (Spetzler 2002)

Extradural
Intradural
Ventral (shunt between the anterior spinal artery and a draining vein also called Type IV lesions, intradural or perimedullary AVF's; usually high flow)
Small
Medium
Large
Dorsal (shunt between artery and vein at the level of the dural root sleeve also called spinal dorsal arteriovenous fistulas or SDAVF's; usually low flow)
Single feeder
Multiple feeders

AVF's=Arteriovenous fistulas, SDAVF's=Spinal dural arteriovenous fistulas

Table 3: Summary of cases of filum terminale AV fistulas

Name of the author	No. of cases	Age and sex	Duration of symptoms	Clinical features	Imaging findings	Treatment modality	Outcome
Meisel	1	30/M	Not available	S1 radicular pain on left side	Angiogram showed arterial feeder of filum arising from medullary basket	Surgery	Cured
Tender	2	70/M 58/M	7 years 18 years	Low back pain and lower extremity weakness. Underwent laminectomy for spinal stenosis. Worsened 1 year later with further weakness, decreased sensation, urinary retention and constipation	Lumbar myelography showed serpentine filling defects at thoraco-lumbar junction. Angiogram showed ASA augmented by left 11th intercostal artery. Fistula at L4 level	Surgery Surgery	Walking with a cane 1 year post-surgery Walking with a cane 1 year post-surgery
				Bilateral leg weakness and paresthesias. Operated for spinal stenosis 3 times. 1 year before diagnosis wheelchair bound, urinary retention and constipation	Hyperintense cord signal from mid-thoracic to conus and flow voids from T12 to L3 in subarachnoid space. Angiogram shows feeder from ASA augmented by 9th left intercostal artery		
Jin	1	61/M	10 years	Leg pain	Multiple perimedullary signal voids from D10 to L3. Fistula fed by ASA augmented by right 9th intercostal artery and 2 branches of left lateral sacral artery. Fistula at L5 level	Endovascular followed by surgery	Complete resolution of symptoms
Lim	4	60/M 48/M 53/F 63/F	Not available	Gluteal pain (2 patients), motor weakness (3 patients), sensory changes (4 patients), sphincter dysfunction (3 patients)	Flow voids in all 4 patients. Cord signal change in first 3 patients (Case 1 and 2 in conus, case 3 up to D9). Angiogram showed ASA feeder augmented from left L1, left D10, right L1 respectively in the first three cases and right L4 and left lateral sacral artery in the last case. Fistula at L3, L4-L5, L4-L5 and L3-L4 respectively	2 surgery 2 endovascular	Follow up period 2 to 6 months. All patients with motor weakness improved, 2 completely and 1 partially. Sphincter dysfunction improved in 2 out of 3 patients

Contd...

Table 3: Contd...

Name of the author	No. of cases	Age and sex	Duration of symptoms	Clinical features	Imaging findings	Treatment modality	Outcome
Trinh	2	57/M 63/M	2 years 3 months 4 years	Back pain, leg weakness, numbness. Underwent lumbar fusion 2 years before presentation and recent onset (3 months) worsening of power and sphincter dysfunction Back pain, numbness and weakness right foot. Weakness worsened after surgery for lumbar fusion	ASA fed by left D10 intercostal artery fistulating at L5 level Flow voids in subarachnoid space and increased signal in dorsal cord and conus.Fistula located at L4-L5 level. Precise location of artery to vein transition not apparent	Both surgery	Both patients improved in motor power and lower limb sensations and became independently ambulant. Case 1 had a pseudomeningocele which was reoperated 1 month after surgery. Intra operatively there was cauterisation injury to S3 and S4 nerve roots. However, his sphincter functions improved
Witiw	1	62/M	6 months	Lower leg paresthesias, weakness, incontinence and impotence	Hyperintense cord signal from mid-thoracic to conus and flow voids in subarachnoid space. Angiogram showed enlarged ASA augmented by right 8th intercostal artery. Fistula at S2-S3 level	Surgery	At 10 months follow up regained continence, ambulant with assistance of cane, improvement of paresthesias, sexual dysfunction persisted
Kumar	1	44/M	8 months	Walking difficulty, paresthesias, bladder complaints (urgency and poor flow). Had undergone L4-L5 microdiscectomy few months previously	Hyperintense cord signal in dorsal cord with flow voids that enhanced post-contrast. Angiogram showed enlarged ASA augmented by right 9th intercostal artery. Fistula at L4-L5 level	Surgery	Independently ambulant
Present case	1	54/M	3 years	Diffuse gluteal and leg pain, weakness of lower limbs, wasting, walking difficulty, sensory loss and urinary and fecal incontinence	Multiple flow voids in the dorsal sub arachnoid space (from C3 to L4) and hyperintense signal change in the dorsal cord from D6 body to the conus. ASA is augmented by a radicular feeder from left D12 level. Fistula at L4	Surgery	Independently ambulant and continent. Residual urinary volume in bladder 45 ml. No fecal incontinence

ASA=Anterior spinal artery

'AVF' and retrieved only 29 articles. A search with the terms 'filum terminale' and 'AVF' yielded 28 results. However, after removing cases associated with extradural fistulas, diastematomyelia, schwannomas, hemangioblastomas and aortocaval fistulas, we found only 12 cases of filum terminale AVF's fed by the ASA causing congestive myelopathy [Table 3].

Both surgical and endovascular treatment options are mentioned in literature. Lin *et al*.[5] conclude that if there is a large distance from the origin of the artery to the fistula site, clipping would be the safer option than embolization. Others[3] too have emphasized on the need for a high index of suspicion to establish diagnosis and hold that surgery is safer with decreased risk of spinal cord ischemia as compared to embolization.[3,8] Trinh *et al*.[6] have also preferred surgery rather than endovascular treatment in their two cases. Witiw *et al*.[9] have reported indirect treatment of the AVF by transection of the filum as have Tender *et al*. in their two cases.[10]

In our case, the hairpin loop cranially at the point where the radicular artery augmented the ASA precluded any attempt at embolization and hence, surgery was carried out.

All cases described in literature have had a satisfactory recovery. Factors determining prognosis are said to be duration of illness, sphincter involvement at presentation and location of fistula however, there is no consensus on any of these.[1] Before our case, only one such has been published in Indian literature.[3] Five of these 12 patients had undergone surgical procedures previously in the lumbar spine before a correct diagnosis was made.

To conclude, diagnosis of congestive myelopathy due to a spinal vascular malformation requires a high index of suspicion. Any patient with slowly progressive paraparesis and T_2 hyperintensity in cord with flow voids in the spinal subarachnoid space must be investigated carefully with a spinal DSA. Filum terminale AVF's, if found can be easily treated with simple surgical disconnection that has gratifying results with clinical and radiological improvement seen in a short time.

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How to cite this article: Krishnan P, Banerjee TK, Saha M. Congestive myelopathy (Foix-Alajouanine Syndrome) due to intradural arteriovenous fistula of the filum terminale fed by anterior spinal artery: Case report and review of literature. Ann Indian Acad Neurol 2013;16:432-6.
Received: 26-01-13, **Revised:** 10-03-13, **Accepted:** 28-02-13

Source of Support: Nil, **Conflict of Interest:** Nil