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Case Report

Filar arteriovenous fistula mimicking upper motor neuron palsy: A case report with review of the literature

Anand Kumar Das[1], Saraj Kumar Singh[1], Subhash Kumar[2]

Departments of [1]Neurosurgery, [2]Radiodiagnosis, All India Institute of Medical Sciences, Patna, Bihar, India.

E-mail: Anand Kumar Das - dranand0822@gmail.com; *Saraj Kumar Singh - dr.sarajkumarsingh@gmail.com; Subhash Kumar - drsubhashk@aiimspatna.org

***Corresponding author:**
Saraj Kumar Singh,
Department of Neurosurgery,
All India Institute of Medical Sciences, Patna, Bihar, India.

dr.sarajkumarsingh@gmail.com

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ABSTRACT

Background: Filar A-V fistula is a rare entity. It requires a high degree of suspicion to diagnose. Magnetic resonance imaging (MRI) findings are often nonspecific and spinal angiogram is required to diagnose it.

Case Description: A 63-year-old male patient presented with Grade 4 spastic paraplegia and significant sensory disturbance below D8 level along with severe vesicorectal dysfunction. On imaging flow voids were present at lower dorsal and lumbar level in MRI (T2 sequence). Patient underwent spinal digital substraction angiography (DSA) which was suggestive of filar fistula at L4-L5 level. Patient underwent surgical exploration with L4-5 laminectomy. Feeding artery was identified using indigocyanine green (ICG) dye and excised along with filum and dilated vessels. Patient recovered symptomatically in postoperative period.

Conclusion: Filar fistula is a rare lesion and it presents with long standing progressive congestive myelopathy. It requires a high degree of suspicion to diagnose it. DSA is the gold standard for diagnosis and management planning. Surgical approach utilizing the ICG dye is best treatment options in such cases.

Keywords: Filar arteriovenous fistula, Myelopathy, Spinal angiogram, Venous hypertension

INTRODUCTION

Filar arteriovenous (A-V) fistula is a rare type of A-V shunt. It is a direct communication of artery of filum terminale and a single draining vein. These types of fistulas are 3 times more common in males than females.[2] Patients usually present in late stage with severe neurological dysfunction.[3] It requires a high degree of suspicion to diagnose a spinal A-V fistula. Magnetic resonance imaging (MRI) findings are often nonspecific and spinal angiogram is required to diagnose it.[8] Here, we are discussing a case that presented to us with severe neurological dysfunction. Initially, there was a diagnostic dilemma but it was diagnosed as filar fistula after spinal angiography and later on treated successfully using indigocyanine green dye.

CASE REPORT

A 63-year-old male patient presented with insidious onset gradually progressive difficulty in walking, decreased sensation below chest, and bladder bowel dysfunction for the past 11 years. The patient was operated for lumbar spondylolisthesis 11 years back. Multiple MRIs (whole spine) were done at different hospitals but no definite diagnosis was reached. His neurological

deficit continued to worsen and he became paraplegic. On examination, he was found to have spastic paraplegia (power – 0/5 in both lower limb, tone – spasticity, and bulk reduced in both lower limbs). There was significant sensory disturbance below D8 level along with severe vesicorectal dysfunction. Deep tendon reflexes were absent in the lower limb but normal in the upper limb. Plantars were extensor on both sides. Cerebral MRI was normal. MRI spine revealed multiple flow voids at lumbosacral level in T2 with cord signal changes at dorsolumbar level [Figure 1a]. On spinal digital substraction angiography (DSA), filar fistula at L4-L5 level was found [Figure 1b] with feeder from the right medullary artery arising from the left D11 level [Figure 1c]. It was draining into ascending vein which was dilated, tortuous, and extending up to mid dorsal level.

Patient underwent surgery with L4-5 laminectomy. Thecal sac was opened over virgin area at L4 level. On opening dura, dilated veins were visualized posterior to the filum. Indigocyanine green (ICG) was injected to identify feeding and draining vessels [Figure 2a]. Feeder from the artery identified. Vein and fistula were also localized which were running parallel to the artery [Figure 2b]. All the three with filum was coagulated and excised. After excision, repeat ICG injection was done, showing no fistulous tract or venous filling from the site. Postoperatively, patient showed very mild improvement with power in both lower limbs. His bowel, bladder disturbance was same as preoperative.

DISCUSSION

Filum terminale AV fistula (AVF) is a rare variety of spinal AV shunt.[3] Artery of filum is caudal extension of anterior spinal artery which is present anteriorly. Vein of filum is a single venous structure which runs posteriorly from filum to conus and ascending toward medullary vein. Pathogenesis of AVF between these two vessels can be congenital or acquired. Congenital ones are associated with tethered cord, diastematomyelia, spina bifida, and syringomyelia. Acquired ones present at later stage of life.[6] Underlying pathogenesis is same as of an AVF causing steal phenomenon. In filar AVF, there is increase venous flow and venous pressure. It is transmitted as increased intramedullary pressure to conus and spinal cord leading to congestive myelopathy.[3] 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 and this leads to impaired dissipation of venous hypertension found in patients with AVF's.

Patients usually present with dysesthesias, nondermatomal sensory loss, walking difficulty, wasting, and weakness of lower limbs, buttock pain, and sphincter dysfunction. Due to nonspecific clinical manifestations and rarity, this condition is usually misdiagnosed as polyneuropathy, polyradiculopathy, or intramedullary tumor.[4] Our patient presented with slowly progressive myelopathy and became paraplegic. He was diagnosed and operated for spine degenerative disease but could not recover.

MRI is first diagnostic step for such patients of progressive myelopathy. Prominent vascular flow voids and vascular enhancement around conus and lower dorsal spinal cord are common diagnostic findings in MRI T2 sequence. However, to differentiate between perimedullary fistula and filar fistula, spinal angiogram is necessary. DSA is must for characterization of the angioarchitecture of the lesion. It

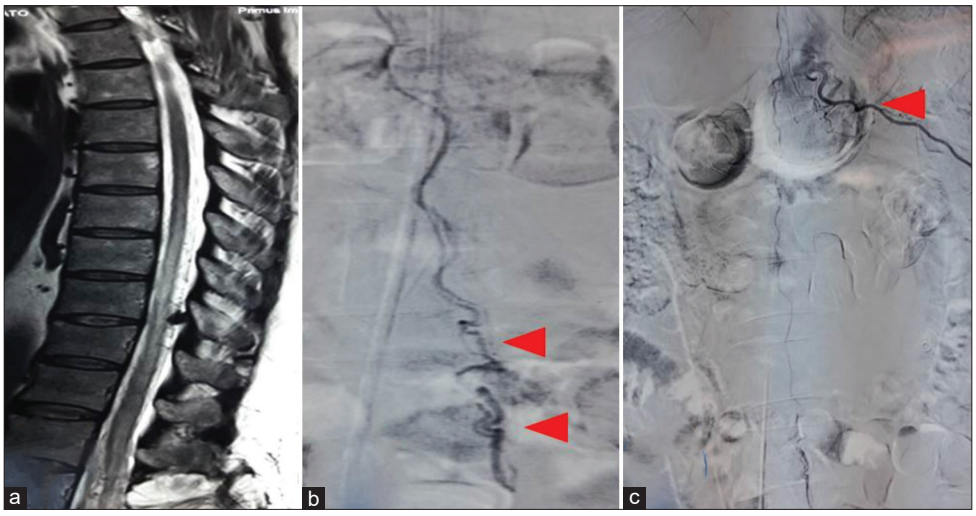

Figure 1: (a) Sagittal T2 sequence showing multiple flow voids. (b) Digital substraction angiography (DSA) showing filar artery and dilated vein running together (red arrowheads). (c) DSA showing feeder from the right medullary artery (red arrowhead).

Table 1: Summary of previous case reports with present one.

Case reports	Year	Age/(years)	Sex	Duration (Months)	Symptoms				Feeders	Location	Treatment	Outcome
					S	M	BB	P				
Kumar *et al.*[5]	2011	44	M	8	+	+	+	+	ASA (T9)	L4-5	Surgery	Imp
Trinh *et al.*[6]	2011	57	M	24	+	+	+	+	ASA (T9)	L4-S1	Surgery	Imp
		63	M	48	+	+	+	+	ASA (T9)	L4-5	Surgery	Imp
Fischer *et al.*[1]	2012	69	M	>12	+	+	+	+	ASA (T9)	L4	Surgery	Imp
Wajima *et al.*[7]	2016	78	M	12	+	+	+		ASA (T8/9)+LSA	S1/2	surgery	Imp
Author case	2022	63	M	132	+	+	+	+	ASA (T11)	L4-5	Surgery	same

M: Motor, S: Sensory, P: Pain, B/B: Bladder and bowel, ASA: Anterior spinal artery, LSA: Lateral sacral artery, Imp: Improvement

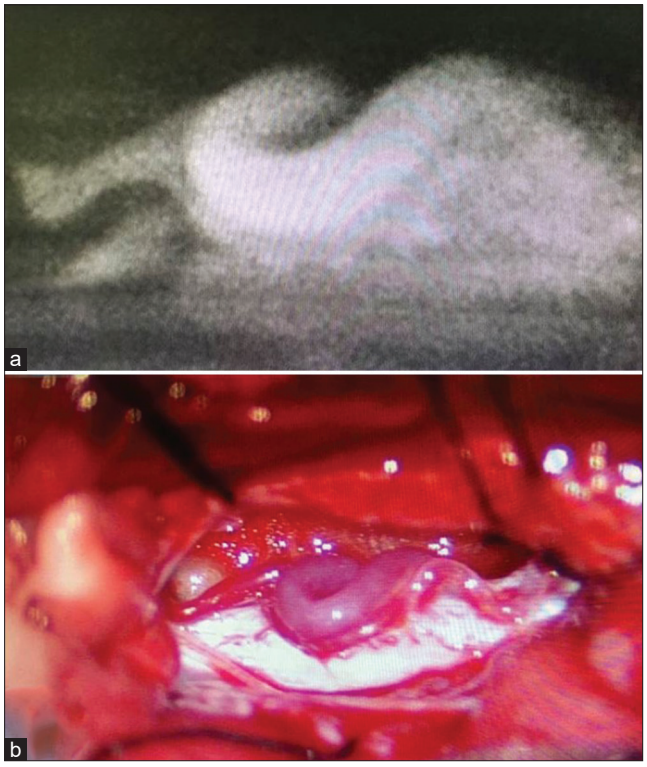

Figure 2: Intraoperative visualization of dilated vein under indigocyanine green dye (a) and under microscope (b).

identifies all arterial feeders, draining vein, and fistulous connection. It also detects any additional associated spinal vascular malformations. On DSA, filar fistula is typically characterized by a focal, single fistula located well below the conus in the lower lumbar spine. The fistulous point is usually defined by the transition from the smaller artery to the larger proximal draining vein.[1] Most filar fistulas are located at the lower lumbar level or at lumbosacral level and in majority of cases, it is supplied by one feeder, the artery of the filum terminale. On MRI, our patient was having flow voids from conus to dorsal spinal level. In DSA, it came out as filar fistula at L4-5 level.

Main therapeutic target is complete obliteration of fistula with preservation of spinal cord circulation. Surgical intervention is the most favored treatment for filar fistula in expert hands.

Surgery is advocated when filar fistula has a single feeder with long distance to the AVF and is safer than endovascular treatment. The overall success rate is close to 100%, and complication rates are low (<5%).[6] For surgical obliteration, precise localization and intraoperative view of feeding artery and fistulous point is necessary. Feeding artery is smaller than the other parallel draining vein. Intraoperative ICG fluorescein angiography is very useful in identifying the feeding artery, draining vein, and exact shunt location. Tightly adherent artery and the vein along the filum are separated and connection is identified. Temporary clip is placed over the feeding artery. Before sectioning of fistulous connection, ICG angiography is repeated and nonfilling of fistulous connection and draining vein is noted. As filum terminale is not having neurologic property, it can be sectioned if needed or if vessels are tightly adherent.[5,9] In our case, we occlude the fistulous connection and also sectioned the filum terminale. However, we highly recommend sectioning the actual identified fistula and sparing the filum. Neurophysiology monitoring is also very helpful in neurologically intact patients. Overall surgical intervention is safe and compatible and approach is also frequently utilized for other procedure.

Endovascular procedure and embolization requires selection of microcatheter for arterial feeder and navigation at the most distal part of the artery, just proximal to the fistula. Risk of anterior spinal artery (ASA) dissection, thrombosis, or vasospasm is very high while manipulation. High flow fistulae can cause dilatation of ASA making manipulation with microcatheter safer. However, still surgical management is recommended due to low morbidity and high occlusion rate.[7] Table 1 compares various case reports with our case presented here.

CONCLUSION

Filar fistula is a rare lesion and it presents with long standing progressive congestive myelopathy. DSA is the gold standard

for diagnosis and management planning. Surgical approach utilizing the ICG dye is best treatment options in such cases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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