Filum Terminale Arteriovenous Fistula: Importance of Clinical Judgment, Angioarchitexture and Technique of Spinal Angiogram to Decide Treatment Options

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Abstract

Background: Filum terminal arteriovenous fistula (FTAVF) is rare entity with varied and misleading presentation. Studying the angio-architecture is the key to better treatment outcomes.

Case presentation: A 56 year old policeman presented with slowly progressive walking difficulty, right lower limb weakness and paresthesia since last 1 year. On clinical suspicion and neuro-imaging using 3 Tesla MRI, flow voids and cord edema were noted from D8 - L3 level raising suspicion of vascular malformation. On spinal angiogram (DSA) FTAVF was noted at L3 - 4 level. It was fed by artery of filum terminal. Patient had undergone successful surgical excision of the FTAVF.

Conclusion: We need to have high index of suspicion for these vascular malformations in view of varied and misleading clinical presentation. Spinal angiogram is the gold standard in diagnosis and treatment planning for optimum outcome.

Keywords: Filum Terminale Arteriovenous Fistula (FTAVF); Angioarchitexture of Filum; Spinal Angiogram (DSA)

Introduction

Spinal vascular malformations with arteriovenous shunt represent rare and complex pathological entities characterized by considerable variation. Most widely used classification divides them into four categories: Type I, Dural arteriovenous fistulas (AVFs); Type II, intramedullary glomus AVMs; Type III, juvenile or combined AVMs; and Type IV, intradural peri medullary AVFs. For successful endovascular or surgical treatment of spinal AVF/AVM it is necessary to obtain data about their location, angio-architecture, and hemodynamics. Detailed angioarchitecture should include arterial feeders, site of fistula or nidus, draining vein, aneurysms on feeding artery, varix or stenosis/occlusion of draining vein. This information will facilitate the judicious use of endovascular and or surgical procedures. One should try to use the least invasive treatment modality as treatment related complications are higher as compared to brain AVM. To perform spinal AVF or AVM resection or occlusion one should use a targeted approach to the malformation, blocking only blood vessels supplying to the malformation and preserving the vessels feeding the spinal cord [1]. Vascular malformations in the filum are rarer compared with the spinal cord or Dural lesions, and only a few cases have been reported. Filum terminale arteriovenous fistula (FTAVF) were initially described by Djindjian., et al. in 1977 and were called “intradural extramedullary spinal AVFs." These were characterized by a direct arteriovenous shunting located on the ventral surface of the filum. Rodesch., et al. found a 3.2% prevalence of focal intradural terminal filum lesions (AVMs and AVFs) in their series of 155 patients with intradural arteriovenous shunts [2]. This case highlights efforts taken to diagnose rare filum terminal AVF(FTAVF) with discussion about angioarchitecture, technique of spinal angiogram and treatment options. We followed CARE guideline while reporting the case.
Case presentation

A 56 year old Policeman presented with complaint of slowly progressive walking difficulty since last one year. At the onset he noted heaviness of his right lower limb and felt that his right lower limb is not as strong as left. Initially he did not have much difficulty in his activities of daily living as well as work. Over next 3 - 4 months he observed progressive difficulty in getting up from sitting position, bearing weight on his right leg and limping while walking. He started requiring support while walking and had intermittent paresthesia's in right lower limb. He also complained of lower backache and constipation. There were no upper limb, urinary or cranio-bulbar complaints. On examination he had asymmetric weakness of both lower limbs with right side being affected more (right hip flexors 3/5, right knee extensors 4+/5 and at ankle 4+/5, left hip flexors 4+/5, left knee extensors 4+/5 and ankle 5/5), areflexia in lower limbs and planter extensor response on right side. Sensory examination revealed mildly decreased pain and temperature sensation in right lower limb and there was no sensory level. After detailed history and examination possibility of spinal cord lesion at dorso-lumbar level was suspected with differential diagnosis of Longitudinally Extensive Transverse Myelitis (LETM), neoplastic or vascular pathology. He was started on Injection methylprednisolone intravenously as pulse therapy and was investigated with nerve conduction studies and contrast MRI dorso-lumbar spine. Nerve conduction studies revealed evidence of right L2 - S1 and left L4-5 active motor axonal degeneration.

His previous MRI was performed at an outside center, on a low Tesla system which showed longitudinally extensive T2 hyperintense signal in the spinal cord from D7-8 disc level up to the conus. On review of this prior MRI and repeat study on a 3T system at our institute, prominent flow voids were seen around the cord including the conus and along the cauda equina nerve roots in the thecal sac. Diffuse persistent T2 hyperintensity was noted in the cord from D9 level up to the conus suggestive of edema. Based on these findings, a vascular malformation like dural arteriovenous fistula was suspected. Most common cause of these imaging features is (Type I) Spinal Dural arteriovenous fistula (SDAVF). On detailed Spinal DSA angiogram FTAVF was noted at L3-4 intervertebral disc level. It was fed by artery of filum terminal which was continuation of anterior spinal artery (ASA). This ASA has single hypertrophied supply by radiculomedullary artery arising from left D8 segmental intercostal artery. The fistula was drained by single vein from L3 - 4 level upwards emptying in to peri medullary venous plexus leading to venous congestive edema of spinal cord. Flow voids seen on MRI are due to these hypertrophied venous channels. In view of long and tortuous course of ASA it was decided to do surgical excision of FTAVF after multidisciplinary discussion. Patient had significant improvement in his clinical symptoms in the form of improved weakness of his right lower limb and gait in few weeks following surgery.

Patient’s perspective

Patient related outcome (PRO) measure on 100 point scale was 90 at the time of discharge.

Discussion

Anatomical consideration

The filum terminale is divided into two segments on the basis of anatomy. Filum terminal Internum is an intradural fibrovascular bundle measuring approximately 15 cm in length. It extends downward from the apex of the conus medullaris starting at the level of L1 - L2 up to S2 vertebra where dural sac ends. The filum terminal Internum pieces the inferior aspect of the arachnoid and dura and continues extradurally as the filum terminal extremum. After exiting the sacral hiatus, the filum terminale externum attaches to the posterior surface of the first coccygeal vertebral segment [3,4,5]. It consists mostly of bundles of collagen fibers and some bundles of neurogenic tissue and is covered by the pia mater, extending from the conus throughout the intradural course. Histologic study of the human filum terminale by Harmeier in 1933 demonstrated the presence of the central canal in the upper one-half of the filum, nerve fibers and all types of ganglion cells. Abundant connective tissues were observed. He concluded that the filum terminale internum had all the components of the spinal cord; therefore, pathologies in it should be similar to those in the spinal cord. Though it does not contribute to any neurological function, it is mainly said to fix, stabilize, and buffer the distal end of spinal cord from normal and abnormal cephalic and caudal traction [6,7].

Vascular anatomy of the filum terminale is unique. During embryonic development, the vascular supply of the spinal cord undergoes significant modification and pruning. The differential growth of the spinal cord and bony spinal column results in the descent of the spinal nerve roots and ascent of the spinal cord into its adult configuration by the end of first year of life. The blood supply to the spinal cord consists of an anterior spinal artery (ASA) and paired smaller posterior spinal arteries (PSAs). ASA is formed by two feeders from terminal vertebral arteries and runs caudally on ventral aspect of cord. PSA is relatively smaller caliber arise from respective posterior inferior cerebellar arteries and runs downwards on posterolater aspect of either side of spinal cord. Anterior spinal (ASA) and two posterior spinal arteries (PSA) are anastomosed to form a basket around the conus medullaris. This basket is formed by two branches of ASA which runs posterolaterally on either side to join respective PSA. From this basket ASA continues downward as single midline branch called artery of filum terminale which runs ventral to filum terminale. Artery of filum terminale rapidly diminishes in caliber, mostly nourishing the filum itself. It is fed by ASA and its radicular feeders. It is argued that there are no radicular nerves originating from the filum [8,9,10].

Radicular feeders of ASA (Radiculomedullary arteries) and PSA (radiculopial) arise from segmental arteries like intercostals, lumbar, lateral lumbar /median sacral, costocervical, vertebral and occipital and ascending pharyngeal artery branches of external carotid arteries. Segmental arteries at each level gives a radicular artery. At the surface of dura this gives radiculomeningeal artery counterpart of intracranial meningeal. After piercing dura it devides into anterior an posterior divison supplying respective nerve roots. The all level embryonic connections to ASA and PSA will regress leaving behind only 2 - 14 (average 6) connections to ASA which are called radiculomedullary arteries seen in a person. Similarly 11 - 16 connections to PSA which are called radiculopial arteries. These radiculomedullary arteries run along the corresponding radicular nerve to the point of its attachment with spinal cord and then turns downward forming typical hair pin bend there by continuing as ASA superiorly and inferiorly. One such dominant radiculomedullary artery in dorsal region is called Artery of Adamkiewicz. Similarly such dominant one in cervical region (lesser known)is called artery of Lazorthes The Level at which these feeders of ASA arise showed considerable variability. Number and size of radiculomedullary arteries are inversely proportional. These feeders can arise even at sacral or coccyegeal level from segmental branches of median sacral artery which continued downwards from aortic terminus, lateral sacral and coccyegeal arteries which arising from internal iliac artery. It is to be noted here that there are no direct arterial feeders to the artery of filum terminale. But it is always fed by ASA (as a continuation of anterior arterial axis) which in turn is fed by radiculomedullary arteries which can arise from any segmental levels from ponto-medullay junction (Intracranial) through cranio-vertebral junction to coccyx. There is a single vein of filum terminale which is also located on the ventral surface, but posterior to the ASA. Vein of the filum runs from its sacral end to the tip of the conus medullaris where it empties into perimedullary venous plexus. Unlike its arterial counterpart, the vein of the filum terminale connects the extradural venous plexus in the sacral canal to the anterior spinal vein (accompanying ASA) of the spinal cord. Venous flow can be either cephalad or caudad.

Technique of spinal angiogram

Spinal digital subtraction angiography is the gold standard and last resort for either confirming or ruling out vascular malformations of spine. It should ideally be done under general anesthesia to prevent motion artifact which can hinder the diagnosis of tiny malformations. All segmental arteries should be selectively cannulated unless one or few are pathologically occluded which can be known on aortogram done at the outset. It is pertinent to do selective angiograms of all segmental arteries systematically for confirmation or ruling out spinal vascular malformation. The entire length of ASA and basket at conus has to be visualized to be label it 'complete spinal angiogram'. All the radiculomedullary arteries, their levels of origin and hemodynamic flow are absolutely essential features to know to avoid devastating complications of treatment. The Flow voids and spinal cord edema seen on MRI gives clue about presence of vascular malformation but may not indicate site of malformation or arterial feeder of particular malformation. Hence complete Spinal Angiogram is absolutely essential. The strength of MRI magnet decides the resolution and sensitivity in depicting flow voids and cord edema. Ideally MRI should be done on 1.5 to 3T (tells) magnet. It is possible to miss flow voids and cord edema on lower strength magnets.

This fact must be kept in mind while evaluating the MRI and later doing spinal angiogram. As clinical presentations of different types of vascular malformation vary from acute myelopathy to progressive myelopathy and has significant overlap with non vascular pathologies of spine. Hence we should have high index of suspicion while evaluating these patients clinically and radiologically.

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Treatment options

Both endovascular or surgical options of treatment need complete understanding of angioarchitexture exact location and hemodynamics of the FTAVF. If length of feeding artery up to the site of fistula is short and less tortuous then endovascular embolization with N-butyl cyanoacrylate should be the first choice. However spinal surgery is also a good alternative with good results.

Conclusion

Learning Points from this case are

- We need to have high index of suspicion for these vascular malformation in view of varied and misleading clinical presentation.
- MRI will give hint about this lesion provided it is done on 1.5 to 3 T magnets. It is possible to miss flow voids and cord edema in lower strength magnets.
- Both endovascular and surgical treatment options give optimum and durable results without short term or long term complication provided it is selected judiciously.

Patient Consent

Patient has given written informed consent for the publication of this case. We are sure that at no point patient personal identity is disclosed.

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Conflict of Interest

Nil declared.

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Nil.

Bibliography


