A Rare Case of Spinalduralarterio-Venous Fistula (SDAVF)

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The authors report a rare case of spinal duralarterio-venous fistula (SDAVF), supplied by radicular artery, in a young female. SDAVF usually occurs(at 5-10/million) in a middle aged(avg 55-60 years) males. The condition has never been reported under the age of 20years. A 15-year-old female presented with progressive subjective ascending weakness of lower limbs associated with tingling numbness since 6 months. Spinal magnetic resonance imaging suggested a possible SDAVF confirmed on Spinal MRAngiography and CT angiography. The patient was advised Embolisation but subsequently treated conservatively on request with expectant management and followed over a period of 1year . At present ,patient has subjective improvement of symptoms with no other complaints .

Keywords: Spinal duralarterio-venous fistula (SDAVF), SpinalM Rangiography, Embolization, Radicular artery.

Introduction :

Spinal duralarterio-venous fistula (SDAVF) is a rare(annual incidence of 5-10 cases per million) and enigmatic disease entity[4]. The clinical features andstructural changes have been recognized since 1926, and the pathophysiology and the essentials of treatment since 1974, but up to the present day it is unknown why these fistulas develop. The fistula is between a radicular artery and the corresponding radicular vein within the dural root sleeve leading to congestion of the venous outflow of the spinal cord and eventually ischaemia.

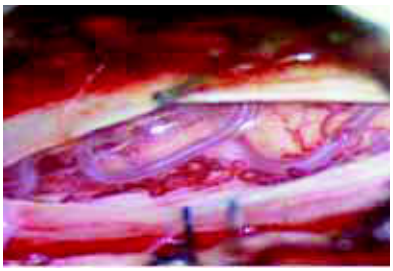

Figure 1 : Clinical picture.

The clinical signs and symptoms as well as the course of disease are sometimes disguising, so the final diagnosis is established relatively later the initiation of diagnostic evaluation[24]. However, recent advances in selective angiography and embolization techniques made an earlydiagnosisand treatment of this disease possible preventing severe neurologic deficits. The pathophysiologic mechanism in SDAVF is a nonhaemorrhagic venous hypertensive myelopathy, which leads to venous congestion and chronic hypoxia, and potentially to venous infarction and irreversible spinal cord lesion[25,26,27].

In any middle aged patient with ascending motor or sensory deficits in the legs, SDAVF should be considered in order to prevent irreversible handicap.

Case report :

A 15year old girl presented with history of, gradual onset ascending tingling numbness over left lower limb(below knee) since 6 months, gradual onset subjective weakness of left lower limb which increased on exertion and decreased at rest, progressively increasing and ascending in nature since 6 months. Patient did not have any radicular/radiating pain or backache.

Examination did not reveal any local spinal tenderness. Neurological evaluation revealed hypoesthesia below L3 dermatome, normal power, reflexes and SLR-full and free.

Plain radiographs(fig.2 and fig.3) showed no remarkable findings

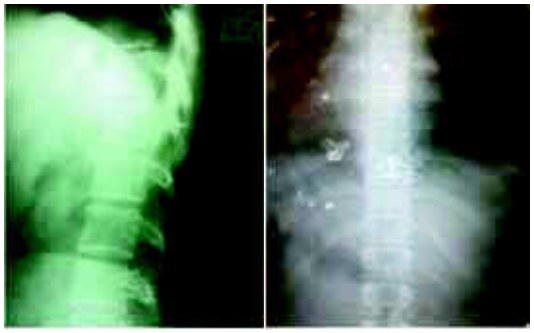

Figure 2 : Lateral view. Figure 3 : AP view.

Doppler -

Arterial Doppler of both lower limbs was normal, thus ruling out local vascular pathology

MRI showed, a 12x9 mm intramedullary heterogenous lesion at D11 with subtle peripheral enhancement, prominent vascular flow voids in extradural space, posterior to cord, diffuse cord hyperintensity from D5 to L1 s/o cord edema.

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Figure 4 : MRI T2W sagittal.

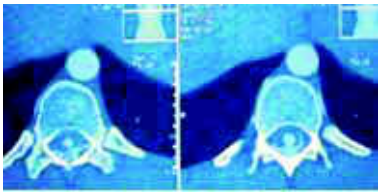

Figure 5 : CT Angio– axial.

Contrast enhanced CT Angiography study was suggestive of, a bunch of abnormal pial vessels on posteriorsurface of spinal cord, malformation supplied by feeding radiculararteries at D10 to L1 on right side and L1-L2 on left side.

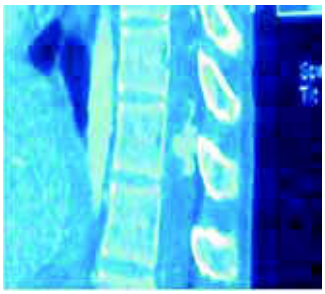

Figure 6 CT Angio– sagittal.

A diagnosis of spinal-dural AV fistula was made from above radiological findings.

Patient was advised endovascular embolization using N-butyl 2-cyanoacrylate (NBCA), but patient refused for the same and opted for conservative management.

A follow up after 10 months revealed subjective improvement in neurological symptoms, no additional neurological deficit.

Discussion :

Spinal duralarterio-venous fistulas are underdiagnosed despite being the most common spinal vascular malformation accounting for about 70% of all spinal vascular malformations[5,6,7]. A major German referral center for spinal vascular disease arrived at incidence of 5-10\million cases\per year in the general population[4]. The exact etiology is unknown. A strong male predominance is noted with males being affected five times more often than women. Mean age at diagnosis of 55 to 60 years. No patient under the age of 20 has ever been reported.[7,8,9,10,11,12,13,14,15] .Most fistulas are in the thoracolumbar region (most commonly between T6-L2) and solitary. SDAVFs are supplied by radiculomeningeal arteries. The AV shunt is located inside the dura mater near the nerve root. Increased pressure within the venous system because of arterialization leads to venous congestion causing chronic hypoxia and progressive myelopathy.[16]

SDAVFs are organized into three groups based on the embryologic development of their venous drainage: ventral, dorsal and lateral epidural (most common type). The shunts develop in the lateral epidural space at the junction of the radicular veins to the epidural venous system and due to outflow obstruction blood flow is shunted into the perimedullary veins.

SDAVFs often go underrecognized because the symptoms can be subtle and non localizing and can be mistakenly attributed to coexisting abnormalities, such as lumbar stenosis, disc herniation, or vertebral anterolisthesis, that appear in imaging studies Patients, who are mostly middle-aged , develop a progressive myelopathy, which at the early stages of the disease often mimics a polyradiculopathy or anterior horn cell disorder.Initial clinical symptomatology vary and could include gait disturbance and sensory symptoms such as paraesthesias, sensory loss and also lower extremity radicular pain.[1]. Classically, neurologic symptoms progress and can often be ascending. In the more latter stages of the disease, bladder and bowel incontinence, erectile dysfunction and urinary retention can be seen.[23]. Upper motor neuron disease showing clonus and positive Babinski sign can be seen usually following lower motor signs.[17,18,23]

Diagnosis can be aided by MRI and MRA with but Catheter angiography is still the gold standard in the diagnosis of SDAVF.MRI shows swelling of the spinal cord, with a centrally located hyperintense signal on T2-weighted images, and with hypointense'flow void' phenomena dorsal to the cord, representing enlarged and tortuous veins.[2,19,20,21,22].Cord enhancement can be seen due to blood-spinal cord barrier breakdown with chronic venous congestion. As the disease progresses, the spinal cord can atrophy.

First-pass enhanced MRA can show venous filling and confirm the shunt and may be able to better define the level of the shunt and in effect decrease the radiation dose the patient will need for subsequent traditional angiography. Traditional selective angiography can show stasis of contrast within the radiculomedullary arteries with delayed venous return suggesting congestion. Normal venous return after injection of the anterior spinal artery can exclude the possibility of a SDAVF.

Treatment is centered around occluding the most distal arterial supply with the most proximal draining vein. Options include surgical occlusion or endovascular treatment. The primary goal is to halt the progression of pathology. Only about 67% of patients have regression of motor symptoms with only about 33% having

improvement in sensory disturbances. Impotence and sphincter disturbance rarely resolve. Deterioration of symptoms after initial improvement should suggest recanalization of shunt or development of another shunt.

Conclusion :

In a middle age patient with ascending sensory or lower extremity weakness on exertion, AV malformation, particularly SDAVF should be among the differential considerations. With significant neurological symptoms SDAVF requires definitive management (embolization / operative).

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