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TITLE OF CASE

Unusual Case of Intracranial Dural AV Fistula presenting with acute myelopathy

AUTHORS OF CASE ***Please indicate corresponding author by *(after the author's name)***

Mohamed Abdelsadg, Avinash Kumar Kanodia*, Peter Keston, James Galea

SUMMARY ***Up to 150 words summarising the case presentation and outcome***

We present a case of Intracranial arterio-venous fistula with perimedullary venous drainage presenting with acute myelopathy, which is an unusual presentation of this uncommon condition. Subsequent catheter angiogram defined the arterial feeders from meningohypophyseal trunk and petrosal branch of middle meningeal artery. The patient was successfully embolised which resulted in complete obliteration of fistula and significant resolution of brainstem and cervical cord changes along with clinical improvement.

BACKGROUND ***Why you think this case is important – why you decided to write it up***

Dural arteriovenous fistulae (DAVFs) are heterogeneous collection of conditions, involving presence of shunts between dural arteries and dural venous drainage, they can occur either intracranially or in the spinal dura. [1] Intracranial DAVFs account for 15% of all cerebrovascular malformations. Cranial DAVF with perimedullary venous drainage are uncommon amongst these, but are more likely to have an aggressive neurological course and present with myelopathy and brainstem changes. Only a few isolated case reports and small case series have been described, with the majority of those cases involving the dura within the posterior cranial fossa or at the foramen magnum. [2] We present a case of DAVF with perimedullary venous drainage, clinically presenting as rapid onset myelopathy and discuss some of the characteristics and issues involved with pathophysiology and management.

CASE PRESENTATION ***Presenting features, medical/social/family history***

A 65 year old female, while on cruise in the Mediterranean, developed weakness in her right hand grip that soon progressed to involve the rest of the right sided upper and lower limbs. Within next 3 hours, she developed dizziness, unsteadiness of gait, frequency of urine, vomiting and fluctuating consciousness. She was attended to by the ship's doctor; her symptoms were attributed to sea-sickness and low serum potassium for which she was started on replacement therapy. She had a background history of hypertension, stage II chronic kidney disease (ex-kidney donor), and no previous history of trauma or craniotomy. Her symptoms persisted and on the following day, she was noted to have developed weakness on the left lower limb. She was therefore transferred to the nearest hospital in Puglia, Italy.

INVESTIGATIONS ***If relevant***

Patient had a CT scan of brain which was reported to be normal. Subsequent MRI brain showed evidence of brainstem swelling, and high signal in T2 images was detected in the lower medulla oblongata and upper cervical cord (Figure 1). There was a thin rim of peripheral low T2 signal around upper cervical cord. On diffusion imaging, there was predominantly high ADC signal suggesting vasogenic oedema, although there was suggestion of a small area of restricted diffusion in medulla on left side, suggesting a small infarct (Figure 2). On intravenous contrast, there was patchy enhancement and vascular congestion in lower brainstem and upper cervical cord (Figure 2). She underwent cerebral digital subtraction angiography (DSA) which revealed a DAVF in posterior fossa on left side (Figure 3). The arterial supply was from the tentorial branch of the meningohypophyseal trunk (MHT), with a very small feeder arising from the petrosal branch of the middle meningeal artery (MMA). The venous drainage was to an isolated segment of the left superior petrosal sinus (SPS) and from there via a communicating superior petrosal vein to the pontomedullary venous plexus and upper cervical perimedullary plexus. Small radiculomedullary communicating veins draining to the paravertebral venous plexus were also noted. The vein of Rosenthal was noted to be absent. There was a sluggish flow in the vein of Galen, and a stenosis of a short segment of the left transverse sinus.

DIFFERENTIAL DIAGNOSIS ***If relevant***

Based on MRI findings, there can be a potential differential of encephalitis, neuromyelitis optica (NMO), tumor, sarcoid and vascular aetiologies. There were no febrile changes nor was the enhancement pattern on MRI particularly suggestive of encephalitis. Overall clinical picture and MRI enhancement pattern was also inconsistent with NMO, sarcoid or tumour. The large area of abnormality was difficult to attribute entirely to acute infarct either on clinical basis or diffusion MRI. Due to vascular congestion (Figure 2d) and changes in the upper cervical cord (Figure 1f), a possibility of a DAVF was raised, although the site was difficult to exactly guess on MRI, but possibly posterior fossa. On subsequent DSA, the diagnosis was confirmed and no differential was considered.

TREATMENT ***If relevant***

It was decided to embolise the DAVF. A microcatheter was fed via the middle meningeal artery feeder and the tip was positioned within the nidal network. PHIL^{TM} 25% (precipitating hydrophobic injectable liquid) was used to occlude the nidus and the arterialised segment of the superior petrosal sinus. Complete angiographic occlusion at completion was obtained (Figure 4).

OUTCOME AND FOLLOW-UP

The intra-and post operative course was uneventful. The patient made a very good recovery and was discharged in good condition 2 weeks later. After 6 week of discharge the patient was free of symptoms a part from minimal remaining weakness (MRC grade 4+) in the right lower limb. She was followed up in the neurovascular outpatient clinic 3 months following discharge, she was walking independently, with complete resolution of her symptoms. Her repeat MRI showed significant resolution of brainstem and upper cervical cord changes with minor residual signal

changes (Figure 5). However, there was an infarct on inferior aspect of left cerebellar hemisphere.

DISCUSSION ***including very brief review of similar published cases (how many similar cases have been published?)***

Majority of DAVFs are idiopathic, while a minority are associated with preceded events like trauma, cranial surgery, or dural sinus thrombosis. [3] Intracranial DAVFs represent about 6% of supratentorial and 35% of infratentorial vascular malformations. [4] On review of literature, we could identify 54 cases of DAVF with perimedullary venous drainage. In the majority of the reported cases, the fistula was located on the dura mater of the posterior fossa, out of which, 18 were situated in the tentorium cerebilli, 11 in petrosal sinus and petrous apex, 9 in foramen magnum, 4 at torcula, 7 at transverse/sigmoid sinuses, 1 each described in occipital region and base of posterior fossa while no exact description of the site of the fistula was given in 3 cases. [2,5,6] The only reported site outside the posterior cranial fossa was in the cavernous sinus. [2] With regards to the laterality of the shunt site, out of the 54 cases reported only 38 cases described the laterality, 76% were left sided , 21% were right sided and 2.6% (single case) the fistula was situated in the torcula and received bilateral blood supply from both occipital arteries. [7] There was a clear male preponderance with 66% males and 34% females (where gender was identifiable). [2] Mean age was 52.8 and 55.4 years for females and males respectively.

DAVFs may present incidentally or with symptoms related to the location and pattern of venous drainage [8]. In 40 out of the reported 54 cases, there were symptoms and/or signs of progressive myelopathy during initial presentation (74%) while 8 patients had had no symptoms or signs of myelopathy. They presented with meningeal irritation and subarachnoid haemorrhage (SAH)- (4), aphasia (1), coma secondary to intraventricular haemorrhage associated with SAH (2) and headache, ataxia and cranial nerve palsies (1). [7,9] In those presenting with myelopathy, most commonly, the symptoms were slowly progressive, starting with sensory disturbance and motor loss in the lower limbs extending to the upper limb. Sphincter function was usually impaired, while the brainstem symptoms were the last to appear. [10] Acute onset of symptoms has been reported in 25%. [2]

The pathophysiology of myelopathy has been a matter of debate. [1] There are 3 hypothesis described; venous hypertension, arterial steal and direct compression by enlarged veins, clot or varicose vessels. Venous hypertension is more widely accepted; the fistula causes increased pressure in the perimedullary venous system with stagnation of the blood flow in intramedullary veins, leading to reduction in perfusion in microcirculation and vasogenic oedema. [11] However, there has been occasional case of high-flow caroticocavernous fistula presenting with cervical myelopathy due to compression of the cervical spinal cord by dilated perimedullary veins. [12] We wonder if there are altered patterns of venous drainage that can predispose a patient to develop myelopathy in these patients. Interestingly, in our case the vein of Rosenthal was absent, with a sluggish flow in the vein of Galen, and stenosis of a short segment of the left transverse sinus. While some of these could be related to high flow venopathy from DAVF, it is possible that absence or paucity of alternate venous drainage pathways may result in predominant drainage through perimedullary venous plexus, predisposing to myelopathy and other complications such as venous infarcts. Rarely, these patients can present exclusively with cerebral signs and symptoms without myelopathy. In such patients, a working drainage for the venous blood from the congested perimedullary vessels into the epidural plexus via a medullary-radicular vein has been described, that would explain the missing myelopathy. [7]

The cause of rapid progression is not always certain, but generally, it is believed to be presence of thrombosis in vascular channels, infarct or haemorrhage. In our case, the patient presented first with right sided rapidly progressive weakness, associated with some brainstem symptoms and later the left lower limb weakness that rapidly evolved within 24 hours. There was suggestion of a small recent infarct in left side of medulla that may have caused sudden onset and deterioration.

The initial imaging diagnosis of such patients presenting with myelopathy is often challenging, since presenting symptoms are related to spine and they usually have spinal imaging in the first instance. It is important to identify the MRI changes that primarily tend to be oedema in brainstem and cervical cord. A low peripheral signal on T2 weighted images has been described. [13] On DWI, there is usually high ADC signal due to vasogenic oedema, although it can vary if

it is complicated by infarcts. Presence of abnormal vascular channels in subarachnoid space is helpful and points towards a vascular aetiology. As expected, no parenchymal nidus is seen. Complications can include venous infarction, parenchymal and subdural hematomas, and cerebral vasogenic oedema. [14]

The definite diagnosis is by catheter digital subtraction angiography (DSA), which would include all 6 vessels. All of the reported cases in literature had more than single arterial feeder, the reported main arterial suppliers however were: middle meningeal artery, occipital artery, ascending pharyngeal artery, meningo-hypophyseal trunk (MHT), vertebral artery, posterior meningeal artery, Artery of foramen rotundum, and meningeal branches of the internal and external carotid arteries. Based on pattern of venous drainage, DAVFs draining to perimedullary veins are classified as type V based on Cognard classification, which signifies quite aggressive clinical course. [15]

In the management of cranial DAVF the pattern of venous drainage is determinant. As in all dural fistulas, treatment is directed toward closure of the draining vein, either by surgical or endovascular techniques (trans-venous or trans arterial). [1] In the literature, the management was declared in 34 out of 54 cases, embolization (15), surgery (8) and embolization followed by surgery (11). In view of potential serious complications, the goal of the treatment is to obtain a rapid, complete anatomical cure of the lesion. Generally, all treatment options (embolization, surgery or embolization followed by surgery) remain justifiable, there is no evidence to superiorise any, yet it depends on the patient presentation, the lesion characteristics, the surgeon preference and the availability of equipment and expertise. Complications can happen, including irreversible brainstem damage. [16] Cortical venous reflux may persist in some patients after treatment, which is associated with a mortality of 10.4%. [17]

We have presented an uncommon case of DAVF with perimedullary venous drainage, who presented with progressive myelopathy, which was uncommon as it was rapid in onset, probably due to acute infarct. The patient was treated with transarterial embolisation, that completely obliterated the fistula and improvement in imaging and clinical picture. This condition being rare, can be difficult to clinically suspect and diagnose quickly and will often need specialised centres for final diagnosis. These can be treated with vascular intervention or surgery, depending upon available expertise and overall anatomy and accessibility of the fistula.

LEARNING POINTS/TAKE HOME MESSAGES ***3 to 5 bullet points***

1. Cranial DAVF is an important although rare cause of acute myelopathy.
2. Diagnosis is often difficult on cross sectional imaging and DSA is essential for final diagnosis.
3. Depending upon anatomy, treatment is often successful and complete
4. The clinical outcome is good, if complete obliteration of DAVF is achieved.

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Figure captions

Figure 1. Initial MRI. T2 sagittal (a), FLAIR sagittal (b), T2 axials (c,d) show presence of oedema in medulla and upper cervical cord (white arrow). ADC map (e) show high ADC value suggestive of vasogenic oedema (black arrow). T2 axial (f) though upper cervical cord showing low peripheral T2 signal (white arrow).

Figure 2. DWI (a) and ADC map (b), showing a probable small area of restricted diffusion (arrows) suggesting small recent infarct. T1 axial (c) and T1 axial post contrast (d) showing vascular congestion and enhancement (white arrow).

Figure 3. DSA. Left ICA injection. Lateral views (a,d) and AP views (b,e) showing MHT (broken arrow in a), site of fistula/nidus (black arrows) and perimedullary venous drainage (white arrows). Left ECA injection, lateral view (c), showing nidus (black arrow). Superselective MMA injection (f) showing site of fistula/nidus (black arrow) and perimedullary venous drainage (white arrow).

Figure 4. Embolisation. PHIL^{TM} cast seen (black arrows in a,b). Post embolisation left ICA injection, lateral view (c), shows absence of fistula.

Figure 5. Follow up MRI. FLAIR sagittal (a), T2 axials (b,c) show significant resolution of oedema with some residual changes (black arrow) and left cerebellar infarct (white arrow in c).

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