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Spinal dural arteriovenous fistulas – An underdiagnosed disease

A review of patients admitted to the spinal unit of a rehabilitation center

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Abstract *Background* Spinal dural arteriovenous fistulas (SDAVF) are rare and present with non-specific symptoms. The diagnosis is difficult and it is therefore conceivable that patients may not be recognized. *Methods* We reviewed the intake forms of patients who had been admitted to the spinal cord injury ward of a rehabilitation center in the period 1980–2004 to identify possible patients with an undiagnosed SDAVF. Clinical and radiological data were evaluated in selected cases. *Results* In 20 of 1429 newly admitted patients to the rehabilitation center (in 614 of whom trauma was not the cause), we restudied the CT myelograms, MRI scans or spinal angiograms and in two of these we found an undiagnosed SDAVF, and one cerebral dural arteriovenous fistula. One of these three was diagnosed with SDAVF 8 years after the admission to the rehabilitation center; the other two patients had never been diagnosed with SDAVF. In 9 patients a diagnosis of SDAVF had already been established by the time they were admitted to the spinal cord unit. In 20 other patients the admission diagnosis was a vascular lesion or 'progressive myelopathy' but appropriate radiological studies had been destroyed or had never been performed. *Conclusion* Our results suggest that spinal dural arteriovenous fistulas are an underdiagnosed condition.

Key words spinal dural arteriovenous fistulas · undiagnosed · rehabilitation center

Introduction

Spinal dural arteriovenous fistulas (SDAVF) are rare lesions occurring predominantly in middle aged men (Gilbertson et al. 1995). They consist of an arteriovenous shunt between a branch of a dural artery and a medullary vein, which leads to increased pressure in the venous system. This in turn results in a reduced pressure gradient in the spinal cord and in a slowly progressive myelopathy (Kendall, Logue 1977).

The symptoms of the disease initially consist of gait disturbances, numbness, and paresthesiae (Jellema et al. 2003). Because these symptoms are non-specific and the disease is rare, a spinal fistula is often not suspected and the time to diagnosis is long (Jellema et al. 2003). This is the more regrettable since the condition can be treated by operation or endovascular embolisation (Jellema et al. 2004). It is possible that there are patients in whom the diagnosis is never recognised. To identify such patients we studied the records of a rehabilitation center with a special department dedicated to the care of patients with spinal cord lesions.

Materials and methods

The intake forms of 1429 patients who were newly admitted to the spinal cord injury ward of 'de Hoogstraat' rehabilitation center (Utrecht, The Netherlands) between 1980 and June 2004 were studied. This form consists of a single page containing the reason for admission, diagnosis, date of admission and summary of clinical history. Re-admitted patients were not included in this study.

When there was any suspicion of SDAVF, or if the diagnosis was not fully clear, the medical records (which also contains the medical correspondence) and the imaging studies were reviewed (when available) with the help of an experienced neuroradiologist (MS). Trauma cases were excluded from further analysis.

Results

Between 1980 and June 2004 1429 patients were newly admitted to the spinal cord injury ward (about 58 patients a year). Of these, 950 patients were men (66 %). In most patients (815 patients, 57 %) the spinal cord injury had been caused by an accident (Table 1). There were 8 patients who had been previously diagnosed with SDAVF (0.5 %) and one patient who had a cerebral fistula with spinal drainage.

Apart from the 9 patients with a proven cerebral or spinal DAVF, there were 40 others in whom a vascular cause of the spinal cord lesion had been diagnosed or in whom SDAVF was at least a possibility, i. e. patients with a syndromal diagnosis of 'progressive myelopathy' or 'transverse myelitis' (Table 2).

CT-myelography had been performed in 12 of these 40 patients; in one of these (see below, patient No3) an enlarged vein was suspected (written report, the films had been destroyed). MRI of the spinal cord had been performed in 23 patients. Catheter angiography with digital subtraction had been performed in 4 patients, in none of whom had evidence for a spinal fistula been found. In 6 patients none of these three investigations had been performed.

The radiograph films had been destroyed in 14 patients (which is possible in the Netherlands, if ten years have elapsed after the last imaging study).

Of the remaining 20 patients imaging files were re-evaluated. A spinal or cerebral dural arteriovenous fistula was suspected on restudying the films in two patients, and one patient was diagnosed with SDAVF at post mortem examination.

Table 1 Diagnoses according to the admission form in 1429 patients

Diagnosis	Number of patients (%)
Trauma	815 (57 %)
Tumor	117 (8.2 %)
Spina bifida	63 (4.4 %)
Iatrogenic (during spinal or aortic surgery)	49 (3.4 %)
Multiple sclerosis	38 (2.7 %)
Cauda equina syndrome	31 (2.2 %)
Spinal stenosis	25 (1.7 %)
Transverse myelitis	23 (1.6 %)
Epidural hematoma	20 (1.4 %)
Spinal artery syndrome	19 (1.3 %)
Aortic aneurysm/dissection	18 (1.3 %)
Guillain-Barré syndrome	16 (1.1 %)
Spinal tuberculosis	14 (1 %)
Brain stem infarction	12 (0.8 %)
'Progressive myelopathy'	12 (0.8 %)
Poliomyelitis anterior acuta	10 (0.7 %)
DAVF	9 (0.6 %)
spinal	8
cerebral fistula with spinal drainage	1
Angioma	7 (0.5 %)
Arteriovenous malformation	6 (0.4 %)
Other specific diagnoses	136 (9.5 %)
Total	1429 (100 %)

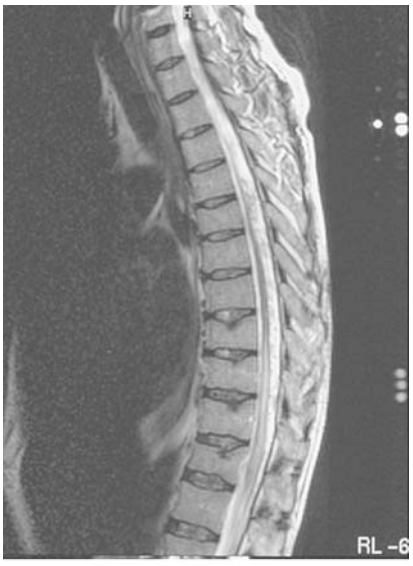
Case reports

The first patient was admitted to the spinal cord injury ward at 42 years of age in 1984, with a medical history of a suspected anterior spinal artery syndrome at 30 years of age. A year before he had developed slowly progressive micturition difficulties and gait disturbances. MRI of the spinal cord was performed in a neurological department, eight years after he had been admitted to the rehabilitation center (Fig. 1a and 1b). At that time he was diagnosed with SDAVF and was subsequently treated by embolisation. After this treatment this patient was able to walk without aid.

The second patient was a 43 year old man; he developed progressive weakness of his arms and legs in 1992. He also developed voiding difficulties. MRI showed an enlarged cervical cord with hyper-intense signal changes from the medulla oblongata to the thoracic spinal cord and flow voids at the dorsal aspect of the

Table 2 Diagnoses according to the medical records in 49 patients with a proven SDAVF or with a suspected SDAVF on the basis of the clinical information. If there was no doubt in diagnosis of patients listed in Table 1, they were not mentioned in Table 2. (SDAVF = spinal dural arteriovenous fistula)

Diagnosis	Number of patients
Anterior spinal artery syndrome	18
Progressive myelopathy	9
SDAVF	9
Angioma	5
Arteriovenous malformation	3
Transverse myelitis	3
Other	2



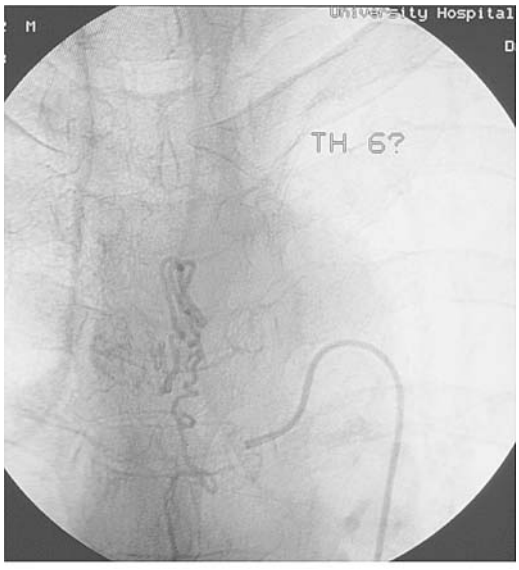


Fig. 1a MRI of the spine of a 50 year old patient with a proven left sided SDAVF at T6. Eight years before he had been admitted to the spinal unit of the rehabilitation hospital. The T2 weighted image shows signal changes from T8 to the conus medullaris and flow void signals dorsal from the spinal cord, representing an engorged perimedullary vein.

Fig. 1b Selective catheterisation of the left T6 intercostal artery. Spinal angiogram shows an enlarged and tortuous medullary vein, which is fed by a dural branch. Same patient as in Fig. 1a

cord (Fig. 2). Spinal angiography was performed, but cranial vessels were not visualised and no SDAVF was found. This patient may well have had a cerebral dural arteriovenous fistula (CDAVF), resulting in venous congestion of the cord (Asakawa et al. 2002). This patient became wheelchair bound and died of recurrent pneumonia and respiratory insufficiency.

The third patient was 69 years old when in 1983 he first complained of pins and needles in both feet. He also developed gait disturbances and micturition difficulties. A myelogram in 1984 showed a possibly enlarged perimedullary vein, but no action was taken upon this finding. His deficits evolved to a complete transverse lesion of the spinal cord and he died a few years later. Post mortem examination revealed tortuous and enlarged perimedullary veins, arteriolisation of medullary veins and virtual disappearance of the gray matter in the lumbar and lower thoracic cord, all consistent with SDAVF.

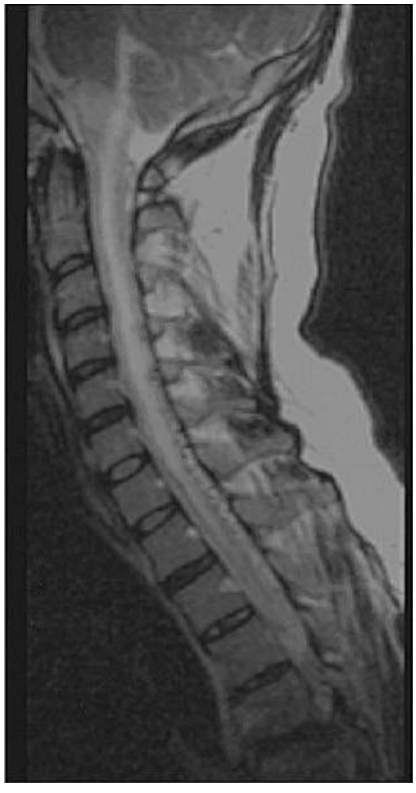


Fig. 2 MRI of a 43 year old man with a suspected cerebral dural arteriovenous fistula (CDAVF). The T2 weighted image shows hyperintense signal changes from the medulla oblongata to the thoracic spinal cord and flow void signals at the dorsal aspect of the spinal cord

Discussion

In this study we found two patients who were admitted to the rehabilitation center with spinal cord dysfunction probably caused by a spinal dural fistula that was unknown at the time and a third patient probably had a cerebral dural fistula causing venous hypertension in the spinal cord. Although these three patients represent 0.2 % of the patients with spinal cord lesions admitted to the rehabilitation center, they represent 0.5 % of patients with a non-traumatic spinal cord lesion and even 8 % of those in whom the diagnosis was a vascular lesion or a syndromal diagnosis. It is possible that even this is an underestimate of the true proportion of undiagnosed patients with an SDAVF, because in half the patients with a syndromal diagnosis the films had been destroyed (14/40) or appropriate radiological studies had never been performed (6/40).

The first patient was diagnosed with SDAVF eight years after he had been admitted to the rehabilitation ward. During this period he slowly deteriorated. The second patient underwent spinal angiography, but intracranial vessels were not visualised. This is a well-known pitfall in the diagnosis of SDAVF. All intercostal

and lumbar arteries should be catheterised, and if no fistula is found, then the deep cervical arteries and iliac arteries should be visualised. Cerebral angiography is indicated if the fistula is not found at any of these levels (Gobin et al. 1992).

All three patients had been seen by neurologists in the 1980s or early 1990s when diagnosis was more difficult than nowadays, because MRI was less widely available. Furthermore, the disease has received more attention in the last few years, at least in the Netherlands. Nevertheless, because of the rarity of SDAVF and the non-specific symptoms especially at the onset, it is still possible that patients are undiagnosed at the time they are transferred to a rehabilitation center. This is particularly worrisome because SDAVF is a treatable cause of myelopathy. Treatment can stop or even reverse symptom progression (especially with regard to muscle strength and gait disability) in about two thirds of patients (Jellema et al. 2004).

This is the first systematic study addressing the question of underdiagnosis of SDAVF in patients treated at a spinal unit. In one previous report two patients were described in whom the diagnosis of SDAVF was not made until a late stage(Grandin et al. 1997), but larger series show long delays in the diagnosis is far from uncommon (Jellema et al. 2003). It is a matter of speculation whether our findings apply to spinal units in other regions or countries. In the initial phase of the disease physicians often think a polyneuropathy is responsible for the symptoms because the first symptoms of SDAVF often consist of paresthesias in the feet or legs and progressive gait disturbances (Jellema et al. 2003). The diagnosis should be strongly suspected when bladder symptoms, sexual dysfunction or sensory symptoms in the perineal region supervene (Jellema et al. 2003). Then MRI of the spinal cord should be performed. MRI almost invariably shows swelling of the spinal cord and T2 weighted signal abnormalities, in combination with flow void signals representing enlarged tortuous veins on the dorsal aspect of the spinal cord. Even then these MRI images may be misinterpreted as inflammatory transverse myelitis, demyelinating disease or a tumor, such as astrocytoma. This should nevertheless prompt further investigation, which should consist of intra-arterial angiography (DSA), even in the absence of flow voids (Gilbertson et al. 1995). Catheterisation should be performed of the radicular arteries which are fed by the vertebral, deep cervical, ascending cervical, posterior intercostal, lumbar and lateral sacral arteries. If no fistula is found the procedure should be extended to the cerebral arteries, including ascending pharyngeal artery, meningohypophyseal trunk, middle meningeal artery and occipital artery. It should be recognised that cord edema may be distant from the site of the fistula. Even patients with intracranial fistulas may present with lower extremity weakness and sensory disturbances caused by edema of the lower cord (Brunereau et al. 1996). In those cases venous congestions may start in the lowest part of the spinal cord.

In conclusion, our review of records at a spinal rehabilitation unit shows that SDAVF may be an underdiagnosed entity because of the rarity of the disease and the non specific symptoms at onset. In any patient with a progressive and especially with an ascending myelopathy SDAVF should be included in the differential diagnosis.

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