A PATIENT THAT CHANGED MY PRACTICE

Practical Neurology 2006; 6: 48-51

# An elderly man with slowly ascending numbness of the legs, followed by incontinence and paraplegia

**J van Gijn**

Professor and Chairman,
Department of Neurology,
University Medical Centre,
Heidelberglaan 100,
3581 CX Utrecht, Netherlands; J.vanGijn@umcutrecht.nl

**Figure 1**
Inner surface of the spinal dura mater (dorsal surface, seen from the ventral side), at the thoracic (left) and lumbar (right) levels. Thickened and tortuous veins run along the length of the dura at the level of the thoracic and lumbar cord.

10.1136/jnnp.2006.088294

*It was 1984 and George Orwell's dire predictions had not become true. No Thought Police were watching me, but perhaps I was a bit too much out on my own in the management of difficult patients. Past publications had not been rewritten by a Ministry of Truth, but were patiently waiting to be discovered. I was not bound by party slogans such as "Ignorance is Strength", but there were many things I did not know, even as a newly appointed professor of neurology in Utrecht, less than a year before. It was time for my first lesson in humility*

The patient was a modest 69 year old man, the retired director of a large firm. He consulted me in May 1984 because of trouble with his legs. This had started in December of the previous year, with altered sensation in his feet; at first they felt "itchy", then gradually "leathery". In February the back of his thighs had started to feel numb, later his left buttock as well. In the past month his sexual potency had decreased and he had twice been incontinent of faeces. On examination muscle power was completely normal. I confirmed decreased skin sensation in the feet, the back of the left thigh, and in the perineal region. Both ankle jerks were absent and the left knee jerk was decreased; the plantar responses were normal.

Given the combination of sensory deficits in sacral dermatomes and disturbances of voiding and erection, I suspected a problem in the conus medullaris or (despite the absence of pain) in the cauda equina—perhaps a tumour. For the benefit of the younger readers I should clarify that at this time MRI was not yet anywhere in sight on the medical scene. The patient was admitted for a myelogram of the lumbar region, which showed only a dural cyst at the level of the S2 root on one side. The CSF showed one white cell/$mm^3$ and a slightly increased protein level. After some hesitation—on my part as well as that of the neurosurgeons—this abnormality was surgically explored but nothing other than a cerebrospinal fluid filled cyst was found. A subsequent EMG showed positive waves and fibrillations in the quadriceps, gastrocnemius, and erector spinae, at several levels.

These EMG findings should have alerted me to a disorder of the spinal cord extending above the conus medullaris, despite the absence of weakness, but apparently I was discouraged by the fruitless operation and the patient was followed as an outpatient. In October 1984 the motor problems became clinically manifest and within a few days his left leg became weak. On examination the weakness proved to be in a "pyramidal" distribution (most marked on hip flexion, knee flexion, and foot dorsiflexion) and he now had bilateral Babinski signs. The deficits of skin sensation had extended over both legs and buttocks. He was readmitted for a descending myelogram (with contrast agent being injected at the cervical level), which again showed no abnormalities except a suggestion of a dilated vein dorsal to the cord. Spinal angiography was considered but not pursued because our radiologists had little experience with the procedure and the gains seemed uncertain.

My diagnosis at this stage was some kind of degenerative disease of blood vessels in the spinal cord; some similar cases had been described in the neurological literature as subacute "necrotic" or "angiodysgenetic" myelopathy.[1,2] I had nothing to offer except my support when the right leg also became weak, and by April 1985 he was no longer able to stand and became wheelchair bound. Fortunately he could remain at home, thanks to the utter dedication of his wife; nevertheless bedsores necessitated several admissions to a spinal rehabilitation centre. In 1990 he was operated for prostatic carcinoma and when, in January 1994, a supraclavicular metastasis was detected he decided to have no further treatment. He died in March 1994 and donated his body to science, which in practical terms meant that after the post-mortem his body was transferred to the department of anatomy for the instruction of medical students.

On inspection, the spinal cord was flattened, from the low dorsal level downwards. Dilated and thickened veins ran along the length of the inner surface of the dura, on the dorsal side

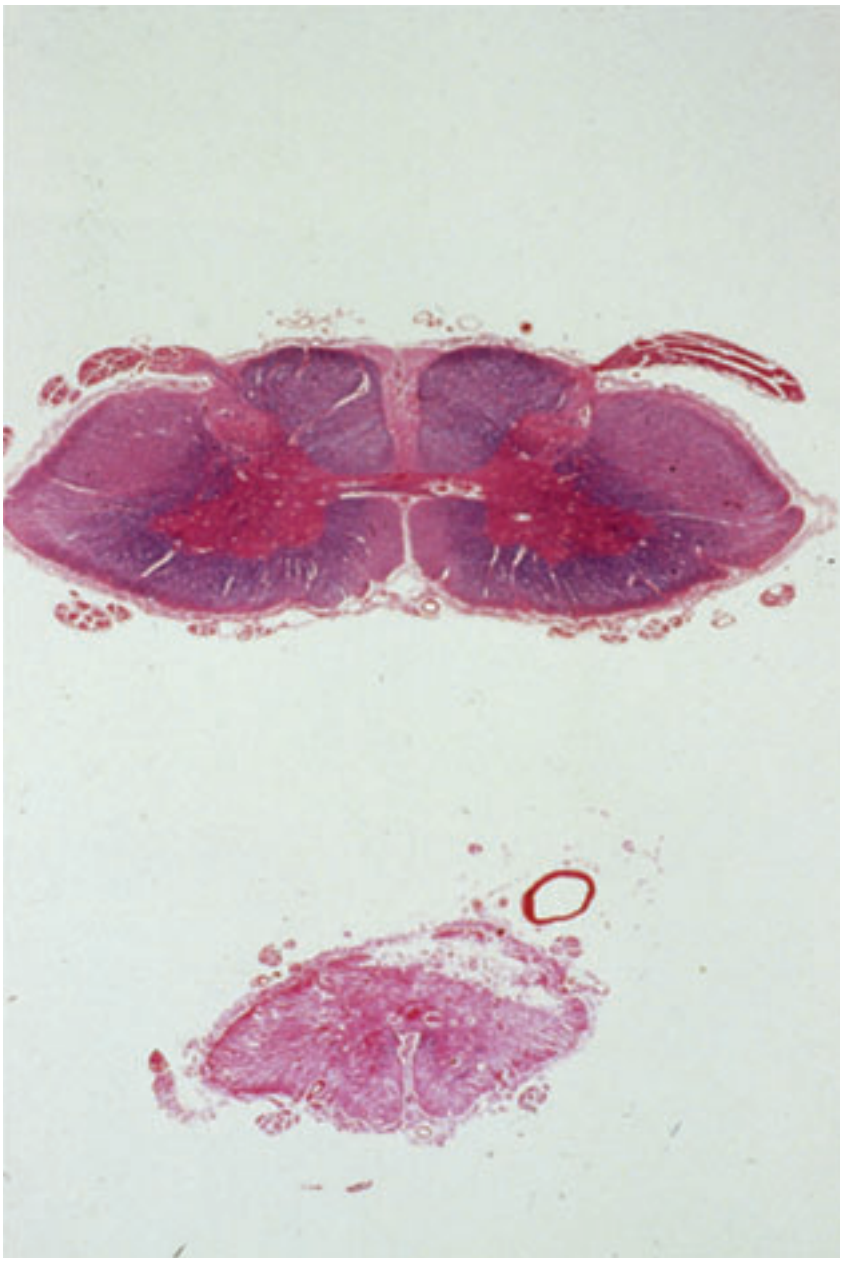


**Figure 2**
Transverse section (haematoxilin-eosin) of the lumbar spinal cord of the patient (bottom) and a control subject (top). Extensive gliosis and atrophy, with disappearance of the gray and white matter structures. Also note the large vein adjacent to the dorsal aspect of the cord.

(fig 1); these abnormal vessels also covered some posterior roots at the thoracic level. On microscopical examination there was almost complete gliosis of the cord at the lumbosacral level (fig 2). The thoracic and lumbar cord contained numerous thickened, sclerotic vascular structures (fig 3).

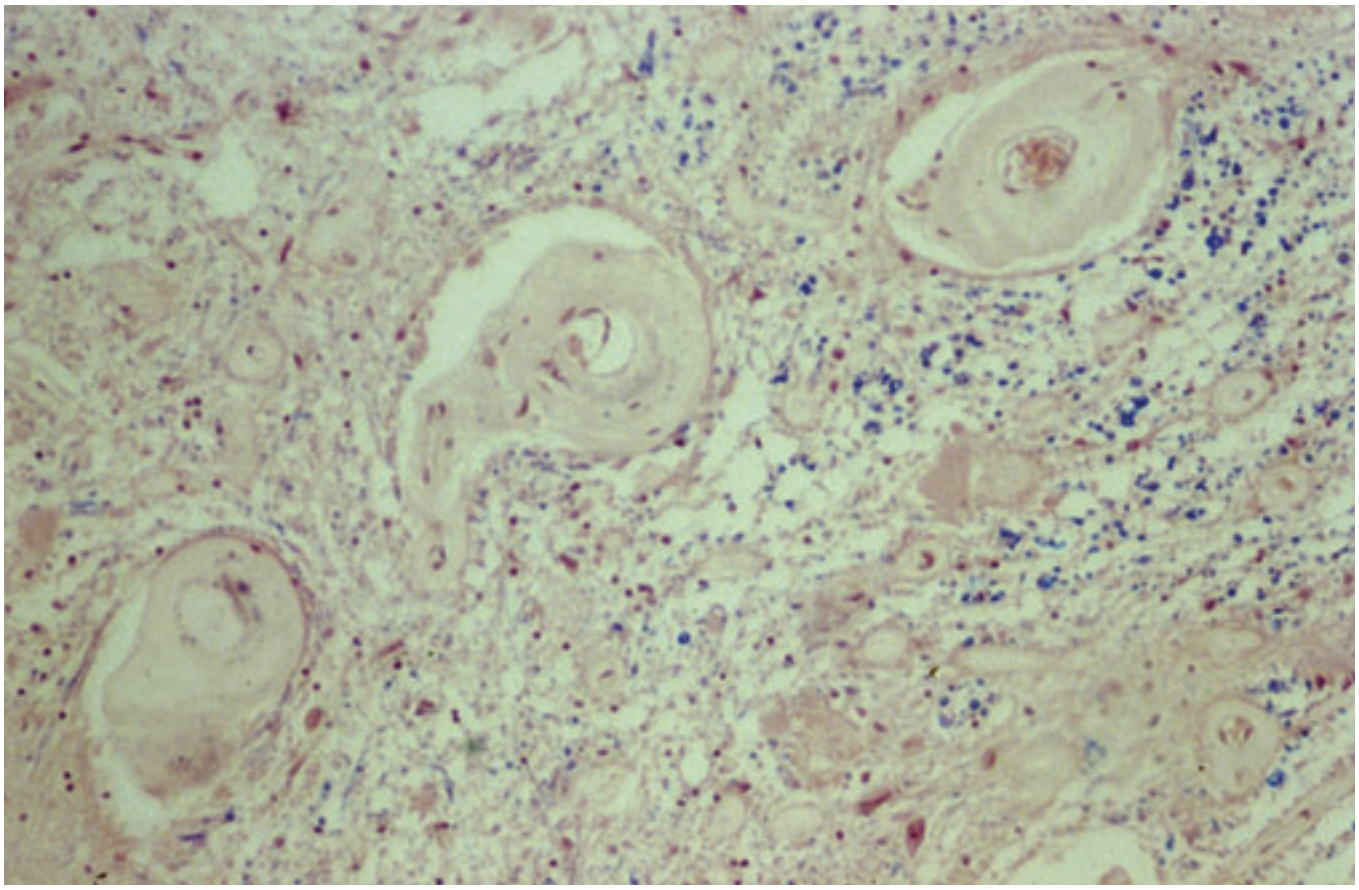
Had this patient been admitted to the National Hospital, Queen Square, London, even 10 years before, the correct diagnosis would have been made without much delay. The publications from this institution in 1974 about "arteriovenous malformations" had still left some uncertainty about the pathogenesis of the abnormality, but they contained several important observations[3,4]: that the essential feature was an arteriovenous shunt, with venous congestion and ischaemia of the spinal cord; that the clinical features could be progressive over months and even years; and, most importantly, that the deterioration could be halted and sometimes even reversed by surgical treatment (in which, we now know, closure of the shunt was the essential feature—not removal of the dilated veins).

Today, with improved imaging techniques and extended surgical observations, it has become clear that arteriovenous abnormalities of the spinal cord are more diverse.[5] An important distinction is between arteriovenous malformations (congenital) and arteriovenous fistulas (acquired). My patient had the most common form, an (intra)dural arteriovenous fistula.[6] For mysterious reasons that mainly affects middle aged men and the cause is unknown. The fistula develops in the intervertebral foramen, between a radicular artery and vein, usually at the thoracic level. Because the radicular vein is connected with the venous outflow of the spinal cord, the cord becomes increasingly congested and eventually ischaemic, usually first at the caudal end (the conus medullaris) and subsequently at higher levels. All the venous structures become thickened and tortuous as a result of the increased pressure. The clinical features parallel this ascending course, spreading in the course of weeks, months, or years from the feet to the legs and buttocks, followed by incontinence and sexual dysfunction, and finally by upper motor neuron features.[7] Initially the clinical features may suggest a polyneuropathy or, if sensory symptoms appear only later, spinal muscular atrophy.[8] For closure of the fistula, surgery has been largely replaced by endovascular embolisation.[9]

This patient changed my practice in several ways. First, I learned how important it is to interact regularly with neurosurgeons and to keep abreast of their literature. Second, although in distinguishing nosological entities we often have to rely on similarity with some previous observations, I now mistrust the notion of "obscure degenerative conditions". Third, if this patient had not himself insisted on a postmortem I would have felt less guilty about missing the diagnosis and I would have been less active in teaching and writing about this treatable condition, in order to prevent others missing the diagnosis.

Since then, my colleagues and I have diagnosed and treated scores of patients with

10.1136/jnnp.2006.088294



*Today, with improved imaging techniques and extended surgical observations, it has become clear that arteriovenous abnormalities of the spinal cord are more diverse*

a spinal dural arteriovenous fistula. On the other hand, it goes almost without saying that I now think of the diagnosis too often. In Orwellian terms, "doublethink" is still difficult. The latest example was an 84 year old woman who, one evening, while sitting in an easy chair, had sudden weakness of both calf muscles, a few hours apart. Eventually the cause turned out to be spontaneous rupture of both Achilles tendons, precipitated by a course of steroids for her asthma—but that is another story.

## ACKNOWLEDGEMENTS

Dr Bart van der Worp and Dr Korné Jellema critically commented on a previous version of this essay.

## REFERENCES

1. Foix Ch, Alajouanine Th. La myélite nécrotique subaigüe. *Rev Neurol* (Paris) 1926;**46**:1–42.
2. Jellinger K, Minauf M, Garzuly F, *et al.* [Angiodysgenetic necrotizing myelopathy (report on 7 cases)] Angiodysgenetische nekrotisierende Myelopathie (bericht uber 7 Fälle). *Arch Psychiatr Nervenkr* 1968;**211**:377–404.
3. Aminoff MJ, Logue V. Clinical features of spinal vascular malformations. *Brain* 1974;**97**:197–210.
4. Aminoff MJ, Barnard RO, Logue V. The pathophysiology of spinal vascular malformations. *J Neurol Sci* 1974;**23**:255–63.
5. Spetzler RF, Detwiler PW, Riina HA, *et al.* Modified classification of spinal cord vascular lesions. *J Neurosurg* 2002;**96**:145–56.
6. Criscuolo GR, Oldfield EH, Doppman JL. Reversible acute and subacute myelopathy in patients with dural arteriovenous fistulas. Foix-Alajouanine syndrome reconsidered. *J Neurosurg* 1989;**70**:354–59.
7. Jellema K, Canta LR, Tijssen CC, *et al.* Spinal dural arteriovenous fistulas: clinical features in 80 patients. *J Neurol Neurosurg Psychiatry* 2003;**74**:1438–40.
8. van der Meulen MFG, Rinkel GJE, Witkamp TD, *et al.* A man with progressive weakness in his legs. *Lancet* 1999;**354**:830.
9. Jellema K, Sluzewski M, Van Rooij WJ, *et al.* Embolization of spinal dural arteriovenous fistulas: importance of occlusion of the draining vein. *J Neurosurg* (Spine) 2005;**2**:580–3.

**Figure 3**
Thickened, 'arterialised' veins in the thoracic spinal cord (haematoxilin-eosin).