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Dural arteriovenous malformations of the spine

Clinical features and surgical results in 55 cases

LINDSAY SYMON, T.D., F.R.C.S., HIDEYUKI KUYAMA, M.D., AND BRIAN KENDALL, F.R.C.P., F.R.C.R.

Gough Cooper Department of Neurological Surgery, Institute of Neurology, and Department of Neuroradiology, The National Hospital, Queen Square, London, England

✓ The clinical and angiographic findings of 55 patients with a spinal dural arteriovenous malformation (AVM) are reviewed, and the results of surgery assessed. The symptoms of dural AVM are usually gradual in onset, and hemorrhage from this type of AVM is less common than in true spinal cord angiomas. Other clinical features and the myelographic findings are similar to those of spinal cord angiomas. On angiography, the nidus of dural AVM's usually projected lateral to the spinal cord. Clipping of communicating vessels between the AVM and the coronal plexus was carried out in 50 patients, and decompressive laminectomy only in five cases. Surgery led to improvement of disturbed gait or arrest of a previously progressive course in 85% of those managed by clipping communicating vessels. The pathophysiology and surgical treatment of dural AVM's are discussed.

KEY WORDS • **dural arteriovenous malformation** • **pathophysiology** • **spinal angiography** • **selective vein clipping** • **dura mater** • **spinal vein**

THE topographical classification of spinal angiomas has hitherto been based upon the relationship of the lesion to the various compartments of the spinal canal. Most authors have classified spinal angiomas as intradural (intramedullary, extramedullary), extradural, or vertebral. However, the recognition of the angioma itself is occasionally difficult since it is associated with enlarged arterialized veins which can be mistaken for an angioma. The development of angiography and microsurgery have facilitated the recognition of the nidus, which is the arteriovenous shunt — the basic vascular lesion. An angiographic review of spinal angiomas has shown that many of these lesions have a nidus of abnormal vessels.[5]

Spinal dural arteriovenous malformations (AVM's) in which the nidus lies on the dura mater are generally considered uncommon, and most of these have been reported as epidural angiomas associated with intradural or vertebral angiomas in the same patient.[12,14] In 1977, Kendall and Logue[7] presented 10 cases of spinal dural AVM. Later, Merland, *et al.*,[11] reported these lesions as "radiculomeningeal arteriovenous fistulae" and suggested that anteriovenous (AV) fistulas constitute the majority of retromedullary malformations. Our experience suggests that, in adults, the dural type of lesion is the commonest spinal AVM, with a fairly characteristic clinical picture and a simple method of treatment. This paper reports the clinical and angiographic findings in cases of spinal dural AVM, and assesses the results of surgical treatment.

Clinical Material and Methods

Seventy-eight patients with a spinal AVM attended either the National Hospital for Nervous Diseases, Queen Square (NHQS) or the Maida Vale Hospital, between 1952 and 1982. In 60 cases, the diagnosis was established myelographically and later confirmed angiographically or at operation. The other 18 patients, whose diagnosis was made on clinical or myelographic findings, were not operated on. The former group, of which eight cases have been reported by Kendall and Logue,[7] make up this study. Of this group, 59 were operated on in the National Hospitals (Mr. Valentine Logue, and L.S.) and one at the University of Wales (Mr. R. D. Weeks).

In 55 patients, the AVM was situated on or adjacent to the surface of the dura mater. Definitive surgical treatment was achieved in 50 of these patients (Group

TABLE 1

Age and sex in 60 patients with confirmed spinal arteriovenous malformations

Sex	Location of Lesion	0–9 Yrs	10–19 Yrs	20–29 Yrs	30–39 Yrs	40–49 Yrs	50–59 Yrs	60–69 Yrs	70–79 Yrs	Total
female	dura					2	1	2	1	6
	spinal cord				1					1
male	dura			1	1	7	19	19	2	49
	spinal cord		1	1			1	1		4

TABLE 2

Initial symptoms of dural arteriovenous malformations

Initial Symptoms	Cases	
	No.	Percent
remote pain	8	15
local back pain	7	13
root pain	6	11
leg weakness	16	29
sensory disturbance	13	24
disturbed defecation	2	4
disturbed micturition	2	4
disturbed sexual function	1	2
total cases	55	

TABLE 3

Symptoms at diagnosis of dural arteriovenous malformation

Symptoms at Diagnosis	Cases	
	No.	Percent
leg weakness	52	95
sensory disturbance	49	89
disturbed micturition	49	89
disturbed defecation	47	85
back pain	19	35
disturbed sexual function	17	31
root pain	15	27
remote pain	13	24
total cases	55	

I) and laminectomy for decompression in five (Group II). Since we would now regard Group II as only partially treated, their subsequent course could be regarded as some form of control. In the remaining five cases, the angioma lay in the substance of the spinal cord. Complete excision of the angioma was carried out in one case, incomplete excision in three cases, and decompressive laminectomy in one case.

The present study is based on hospital records and outpatient review. Symptoms and signs were analyzed in detail. Response to the operation has been assessed by reference to the functional capacity of patients, estimated according to the study of Aminoff, *et al.*[3,10] Thus, disturbances of gait have been graded as follows:

Grade 1: occurrence of leg weakness, abnormal stance or gait, no restriction of activity;
Grade 2: restricted activity;
Grade 3: requires one stick or some similar support for walking;
Grade 4: requires crutches or two sticks for walking;
Grade 5: unable to stand, confined to bed or wheelchair.

Disturbances of micturition have been classified as follows:

Grade 1: hesitancy, urgency, or frequency;
Grade 2: occasional urinary incontinence or retention;
Grade 3: total urinary incontinence or persistent retention.

Subsequent to surgery, many of these patients continued to attend the NHQS regularly for follow-up review. Some patients attended for follow-up observation at another hospital, and their notes were kindly sent to us.

TABLE 4

Duration of symptoms before diagnosis of dural arteriovenous malformation

Duration of Symptoms	Cases	
	No.	Percent
< 1 wk	0	
1 wk to < 1 mo	0	
1 to < 6 mos	6	11
6 mos to < 1 yr	12	22
1 to < 2 yrs	12	22
2 to < 3 yrs	6	11
3 to < 10 yrs	14	25
≥ 10 yrs	5	9
total cases	55	

Summary of Cases

Clinical Data

Age and sex distribution are shown in Table 1, there were seven females and 53 males. The majority of patients with dural AVM's were middle-aged (average 57 years, range 29 to 75 years). By contrast, three of the five patients with spinal cord angioma were under 40 years of age, one of them being only 14 years old.

The most frequent initial symptom of dural AVM was pain (39%) (Table 2). In eight patients pain was referred remotely in a nonspecific manner and in seven it was localized in the back. In six patients pain was radicular in distribution. Weakness of the legs occurred in 16 patients (29%), and sensory symptoms other than pain in 13 patients (24%). Disturbance of micturition, defecation, or sexual function was the initial complaint in five cases (10%). Symptoms by the time of diagnosis are summarized in Table 3. The most common symp-

TABLE 5

*Clinical and angiographic findings in 55 cases of spinal dural AVM**

Case No.	Age (yrs), Sex	Symptoms†	Muscle Wasting	Deep-Tendon Reflexes in Legs	Fasciculation	Abdominal Reflexes	Sensory Disturbance			Site of Shunt	Feeding Arteries	Draining Veins	Anterior Rad-Med Origin
							Superficial	JPS	Vib				
1	63, M	M, S, Sp, I	−	increased			T-10	+	+	rt intervertebral foramen at T6–7	rt 6th IA	2 veins into CP	lt 9th IA
2	63, M	Sp, S, M, I	+	decreased		−	T-11	+	+				
3	60, M	S, M, Sp	+	increased	+	+	T-12	−	+	lt posterolateral at T12–L1	lt subcostal artery	1 vein into CP	lt 10th IA
4	58, M	S, M, Sp, I	+	decreased	+	−	L-2	+	+	rt posterolateral at T6–7	rt 6th IA		
5	64, M	S, I, Sp, M	−	increased		−	T-6	+	+	posterior at T6–7	rt 6th & 7th IA's	2 veins to intervertebral foramen	lt 9th IA
6	58, M	S, M, Sp, I	+	normal	+	+	L-5	−	+	rt posterolateral at T9–10	rt 9th IA	1 vein into CP	lt 11th IA
7	55, M	S, M, Sp, I	−	increased		−	T-8	+	+	posterior at T4–5	rt 4th IA	1 vein into CP	rt 8th IA
8	63, M	S, M, Sp	−	increased		−	T-6	+	+	lt posterolateral at T-6 nerve root	lt 6th IA	1 vein into CP	lt L-1 artery
9	73, F	M, Sp, S	+	increased		−	T-9	+	+	rt posterolateral at T-9	rt 8th & 9th IA's	2 veins to CP	lt 7th IA
10	65, M	M, S, Sp	−	normal		−	T-11	+	+	rt lateral at L-1	rt L-1 artery	1 vein to CP	lt 8th IA
11	55, M	S, Sp, M	+	decreased		−	T-11	+	+	lt lateral at T-9	lt 9th IA	1 vein to CP	lt 9th IA
12	72, M	S, M, Sp	+	decreased	+	−	T-10	+	+	posterior at T11–12	lt 11th IA	1 vein to CP	lt 9th IA
13	53, M	S, Sp	+	increased		−	T-9	+	+	lt posterolateral at T-5	lt 6th IA	2 veins to CP	lt 8th IA
14	66, M	Sp, M, S	+	normal		−	T-6	+	+	lt posterolateral at T-7	lt 7th IA	1 vein to proximal	lt 9th IA
15	57, M	S, Sp, M	+	normal		−	L-2	+	+	rt intervertebral foramen at T3–4	rt 3rd & 4th IA's		
16	51, M	S, M, Sp	−	normal		+	S-2	+	+	rt intervertebral foramen at T10–11	rt 10th IA	2 veins to CP	lt 9th IA
17	55, M	M, Sp, S	+	increased		−	L-2	+	+	posterior at T9–10	rt 9th IA	1 vein to CP	
18	53, M	S, Sp, M	+	normal	+	−	S-1	−	+		aortogram		
19	44, M	S, I, M, Sp	−	increased		+	S-1	+	+		aortogram		
20	65, M	S, M, Sp	+	increased		+	T-9	+	+	lt posterolateral at T9–10	lt 10th IA	1 vein to CP	lt 10th IA
21	50, M	S, M, Sp	+	decreased		+	T-10	+	+	lt intervertebral foramen at T11–12	lt 11th IA	1 vein to CP	lt 10th IA
22	51, M	S, M, Sp, I	−	decreased		−	T-12	+	+	rt posterolateral at T-9	rt 9th & 8th IA's	1 vein to CP	lt 10th IA
23	54, F	S, M, Sp	−	decreased		−	T-10	+	+		rt S-1 artery		
24	62, F	M, S, Sp	+	normal		+	T-10	+	+		lt 8th IA		lt 11th IA
25	61, M	M, Sp	−	increased		−	intact	−	−	rt anterolateral at T-6	rt 5th & 6th IA's	1 vein to CP	rt 8th IA
26	63, M	M, S, Sp, I	+	increased		−	L-1	+	+	rt lateral at S-1	S-1 & S-2 artery	1 vein to CP	lt 9th IA

* Cases 1 to 50 underwent laminectomy and clipping of the feeding vessel, and Cases 51 to 55 underwent laminectomy only. Abbreviations: AVM = arteriovenous malformation; M = motor disturbance; S = sensory disturbance; Sp = sphincter disturbance; I = impotence; JPS = joint position sense; Vib = vibration sense; Rad-Med = radiculomedullary; IA = intercostal artery; CP = coronal plexus; VA = vertebral artery; FT = filum terminale. + = present; − = absent.

† Symptoms in order of onset.

TABLE 5 (*continued*)

Case No.	Age (yrs), Sex	Symptoms†	Muscle Wasting	Deep-Tendon Reflexes in Legs	Fasciculation	Abdominal Reflexes	Sensory Disturbance			Site of Shunt	Feeding Arteries	Draining Veins	Anterior Rad-Med Origin
							Superficial	JPS	Vib				
27	69, M	S, M, Sp	+	increased		–	T-11	+	+	lt posterolateral at T-12	lt subcostal artery	1 vein to CP	lt 9th IA
28	37, M	S, M, Sp	–	increased		±	T-12	–	–	posterior at T6–7	rt 8th IA	1 vein to CP	
29	56, M	M, Sp, I, S	+	increased		+	L-1	+	+	lt posterolateral at L-2	lt L-2 artery		
30	44, M	S, M, Sp	–	increased		–	T-10	+	+	lt intervertebral foramen at T10–11	lt 10th IA	1 vein to CP	
31	75, M	M, I, S, Sp	+	decreased		+	L-3	+	+	lt intervertebral foramen at T4–5	lt L-3 artery	1 vein to CP	rt 10th IA
32	54, M	S, M, Sp, I	–	increased		–	T-6	–	–	rt posterolateral at T-8	rt 8th IA	1 vein to CP	rt 9th IA
33	59, M	S, M, Sp, I	+	decreased		+	L-5	+	+	rt posterolateral at L-2	rt L-1 & L-2 artery	1 vein to CP	rt 11th IA
34	47, M	S, M, I, Sp	–	increased		–	L-1	–	–	posterior at T-6	rt 6th & 7th IA's		
35	69, F	M, S, Sp		increased		–	L-4	+	+	posterior at T12–L1	rt subcostal artery		lt 10th IA
36	64, M	S, M, Sp	+	increased		–	T-12	+	+	lt posterolateral at T-5	lt 4th, 5th & 6th IA's	1 vein	lt 9th IA
37	48, M	S, M	+	increased		–	L-5	+	+	lt intervertebral foramen at L1–2	lt L-1 artery	1 vein to CP	lt 10th IA
38	60, M	S, M, Sp	+	increased		–	L-4	+	+	posterolateral margin of foramen magnum	lateral branch of VA	1 vein to CP	
39	52, M	S, M	–	normal	–	–	L-5	+	+	lt intervertebral foramen at T11–12	lt 11th IA	1 vein to CP	lt 9th & rt 10th IA's
40	69, M	S, M, Sp	+	increased	+	–	L-4	+	+	rt intervertebral foramen at T7–8	rt 7th IA	1 vein to CP	lt 12th IA
41	67, M	S, M	–	increased		–	L-2	+	+	rt posterolateral at T-8	rt 7th & 8th IA's	2 veins to CP	lt 2nd & 10th IA's
42	54, M	S, M, Sp, I	–	increased	–	+	intact	–	–	rt intervertebral foramen at T11–12	rt 11th IA	1 vein to CP	
43	58, M	M, S, Sp	–	decreased		–	L-4	+	+	rt posterolateral at T-7	rt 7th IA		rt 9th IA
44	65, M	S, M, Sp	+	normal		+	T-11	+	+	rt posterolateral at T-8	rt 8th IA	2 veins to CP	lt 8th IA
45	67, M	I, Sp, M, S		increased		+	L-4	–	+	lt posterolateral at T-6	lt 6th IA		lt subcostal artery
46	51, M	Sp, S, M	–	normal	–	+	T-10	–	+	rt posterolateral at T5–6	rt 5th & 6th IA's	1 vein to CP	lt 10th IA
47	47, M	M, Sp, S	+	decreased	–	–	L-2	+	+	rt posterolateral at T7–8	rt 8th IA	1 vein to CP	rt 10th IA
48	64, M	S, M, I, Sp	–	increased	–	+	L-4	+	+	rt posterolateral at T-11	rt subcostal artery		lt L-2 artery
49	63, M	S, Sp, I, M	+	decreased		–	T-2	+	+	lt intervertebral foramen at T12–L1	lt subcostal & L-1 arteries	2 veins to CP	lt 9th IA
50	46, F	Sp, M, S	+	increased		+	S-3	+	+	central pelvis	lt 4th lateral sacral artery	sacral on to FT	lt subcostal artery
51	42, F	S, M, Sp	–	increased		–	T-11	+	+		no angiography		
52	56, M	M, S, Sp, I	+	increased	+	–	T-9	+	+		aortogram		
53	61, M	M, S	+	increased		–	L-4	+	+		no angiography		
54	41, M	S, M, Sp	–	decreased		–	T-10	+	+		no angiography		
55	29, M	S, I, Sp, M	+	normal		–	L-5	+	+		no angiography		

* Cases 1 to 50 underwent laminectomy and clipping of the feeding vessel, and Cases 51 to 55 underwent laminectomy only. Abbreviations: AVM = arteriovenous malformation; M = motor disturbance; S = sensory disturbance; Sp = sphincter disturbance; I = impotence; JPS = joint position sense; Vib = vibration sense; Rad-Med = radiculomedullary; IA = intercostal artery; CP = coronal plexus; VA = vertebral artery; FT = filum terminale. + = present; and – = absent.

† Symptoms in order of onset.

tom was weakness of the legs (95%), accompanying impairment of bladder function in 49 patients (89%), and impairment of bowel control in 47 (85%). Most patients had a combination of motor, sensory, and sphincter disturbance.

The clinical history of the dural AVM was slowly progressive in 43 patients (78%), progressive with remission in six (11%), and acute in onset in another six (11%). The duration of symptoms before diagnosis was less than 1 year in 33% and less than 3 years in 66% (Table 4).

A summary of clinical and angiographic findings in the 55 cases of dural AVM are shown in Table 5. Some factors were found to influence the production of symptoms. In 23 patients (42%), the symptoms were aggravated by exercise or by certain postures. In two cases, the symptoms were related to preceding trauma. In one case, a spinal bruit was recorded. Examination showed that there were 31 patients (56%) with muscle wasting, and the same number had pathologically increased tendon reflexes in the legs. In 13 cases, the tendon reflexes in the legs were absent or obscure. Fasciculation of involved muscles was recorded in seven patients (13%), and loss of abdominal reflexes in 37 (67%). All except two patients had sensory signs that could be related to a lesion of the posterior column and spinothalamic tract.

Myelography demonstrated dilated and tortuous vessels in 51 (93%) of the 55 patients and a complete block in one case (Table 6). In three cases myelography had been carried out elsewhere and the films were not available to us for review. Selective angiography was carried out in 51 cases (47 cases of dural AVM, four cases of spinal cord angioma), and aortography in three cases of dural AVM (Tables 5 and 7). The site of the shunt was evident in 45 cases of dural AVM (90%) and in four cases of spinal cord angioma. In 37 (82%) of 45 dural AVM's, the nidus was projected lateral to the spinal cord and 11 (30%) of these AVM's were situated in the intervertebral foramen (Fig. 1). Most of the draining veins extended medially to join the coronal venous plexus.

In all spinal cord angiomas the shunts were situated within the cord substance or on the surface of the cord. One angioma was fed by the anterior spinal artery, the others by a variety of radicular vessels. Figure 2 shows the site of the shunt related to the age of the patients with spinal AVM's. Most dural AVM's were located between the T-5 and L-3 vertebrae, although one was found on the dura of the foramen magnum, and several on the dura of the sacral sac.

TABLE 6

Summary of myelographic findings in 55 cases of dural arteriovenous malformation

Myelographic Findings	Cases	
	No.	Percent
typical angioma picture	51	93
complete block	1	2
partial block	0	
films no longer available	3	5

Surgical Techniques

The management of these cases in our clinic has evolved from the long laminectomy suggested by Shep-

TABLE 7

Clinical and angiographic findings in five cases of spinal cord angioma

Case No.	Age (yrs), Sex	Symptoms†	Muscle Wasting	Deep-Tendon Reflexes in Legs	Fasciculation	Abdominal Reflexes	Sensory Disturbance			Site of Shunt	Feeding Arteries	Draining Veins	Anterior Rad-Med Origin
							Superficial	JPS	Vib				
56	35, F	S, M, Sp	–	increased		–	S-1	–	–	lt intradural at T12–L1	rt L-2 artery	1 vein to CP	lt subcostal artery
57	24, M	S, M, Sp	+	increased		–	T-2	+	–	intradural posterior at T2–3	lt 6th IA		
58	14, M	M, S, Sp	–	increased	–	+	T-11	+	+	intradural rt posterolateral at T-11	rt L-1 & L-2 artery		lt 9th IA
59	53, M	Sp, M, S, I	–	decreased	+	–	T-11	+	+	no angiography			
60	60, M	S, M	+	decreased		–	L-3	–	–	intradural anterior & posterior at T-12	anterior spinal artery		lt subcostal artery

* Abbreviations: M = motor disturbance; S = sensory disturbance; Sp = sphincter disturbance; I = impotence; JPS = joint position sense; Vib = vibration sense; IA = intercostal artery; CP = coronal plexus; Rad-Med = radiculomedullary. + = present; – = absent.

† Symptoms in order of onset.

hard,[18] with excision of portions of the coronal plexus. We now regard this approach as unsound. Our current procedure involves identification of the communication between the dural AVM and the coronal plexus; this channel is usually a single arterialized vein, which may be exposed by limited laminectomy and doubly clipped and excised. The nidus itself, if accessible on the dura, may be coagulated or excised at this time, but if, as is not infrequently the case, it is separated from its points of communication with the coronal plexus by several segments, then the nidus itself is left undisturbed. In some instances, for example, the nidus may be related to the sacral sac, draining into the coronal plexus through a large arterialized vein lying between the roots of the cauda equina (Fig. 3), where it is easily identified and occluded. The advantage of this form of surgery is obvious. Disruption of the coronal plexus is minimal, and postoperative deterioration is now unknown.

In nine cases the dural AVM was excised. Histological examination showed the nidus on the dura mater to be the AVM (Fig. 4) and intradural dilated vessels to be abnormal veins with a greatly thickened wall containing elastic tissue.

Results of surgical treatment are indicated in Figs. 5, 6, and 7. The majority of patients were moderately or severely disabled before surgery using the criteria of Aminoff and Logue.[3] Forty-six patients of Group I were available for follow-up review. In 39 (85%) of these 46 patients a previously progressive disturbance of gait was arrested by operation. Improvement of disturbed gait was noted in 20 of 31 severely disabled patients (Grades 3, 4, and 5) and in 12 of 15 moderately disabled patients (Grades 1 and 2). No severely disabled patients regained a normal gait. The disturbance of gait improved by two grades in seven patients and by one grade in 25 patients. Control of micturition improved in 26 Group I patients, returning to normal in 11 of these. In Group II there was no improvement of disturbed gait, or micturition.

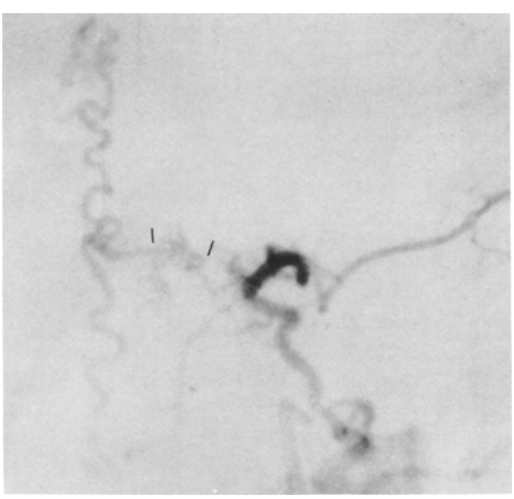

FIG. 1. Case 8. Left sixth intercostal angiogram showing a dural arteriovenous malformation (between *markers*) situated laterally in the spinal canal and in the left T6–7 intervertebral foramen. It is supplied by two small branches from the posterior division of the artery and drained by a single vein into the coronal venous plexus.

Discussion

The incidence of spinal angioma is between 3.4% and 11.5% of spinal cord tumors.[8,16] Angiomas are topographically distributed in the various portions of the spinal canal as well as along the longitudinal axis of the spinal column.[21] Umbach[20] analyzed intra- and extradural lesions separately and found that eight of 24 spinal vascular lesions were epidural, two involved both the extra- and intradural spaces, and 14 were situated intradurally, forming the largest group. In Pia's series,[15] however, 108 of 168 cases of spinal angioma were extradural angiomas, and 25 of these epidural angiomas were complex angiomas associated with intradural or vertebral angioma. Fifty-three of his patients had intradural angiomas, and another seven had vertebral angiomas. At the lumbosacral level, the majority of the angiomas lay in the epidural space, and Pia suggested that epidural angiomas were more frequent than was supposed. This large difference in the frequency of the distribution of angiomas cannot be a coincidence but may reflect the difficulty of topographical classification,

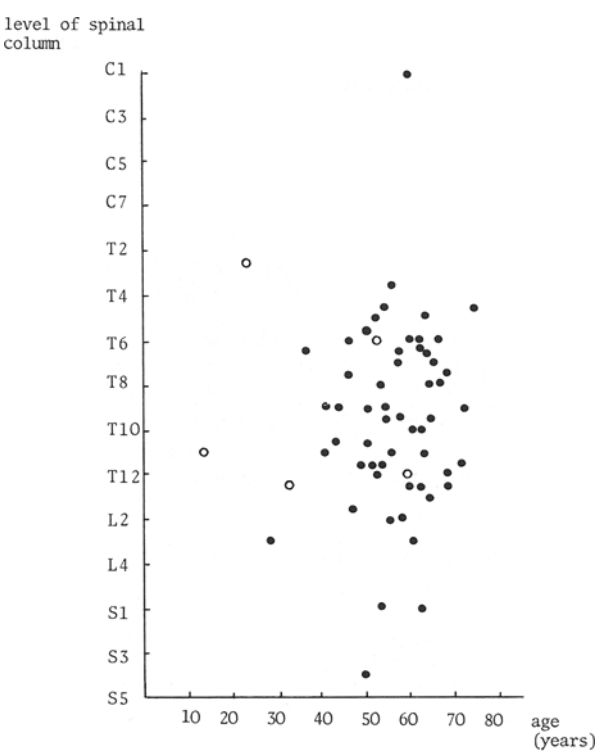

FIG. 2. Site of shunt related to age. Spinal dural arteriovenous malformations are shown by *black circles* and spinal cord angiomas by *open circles*.

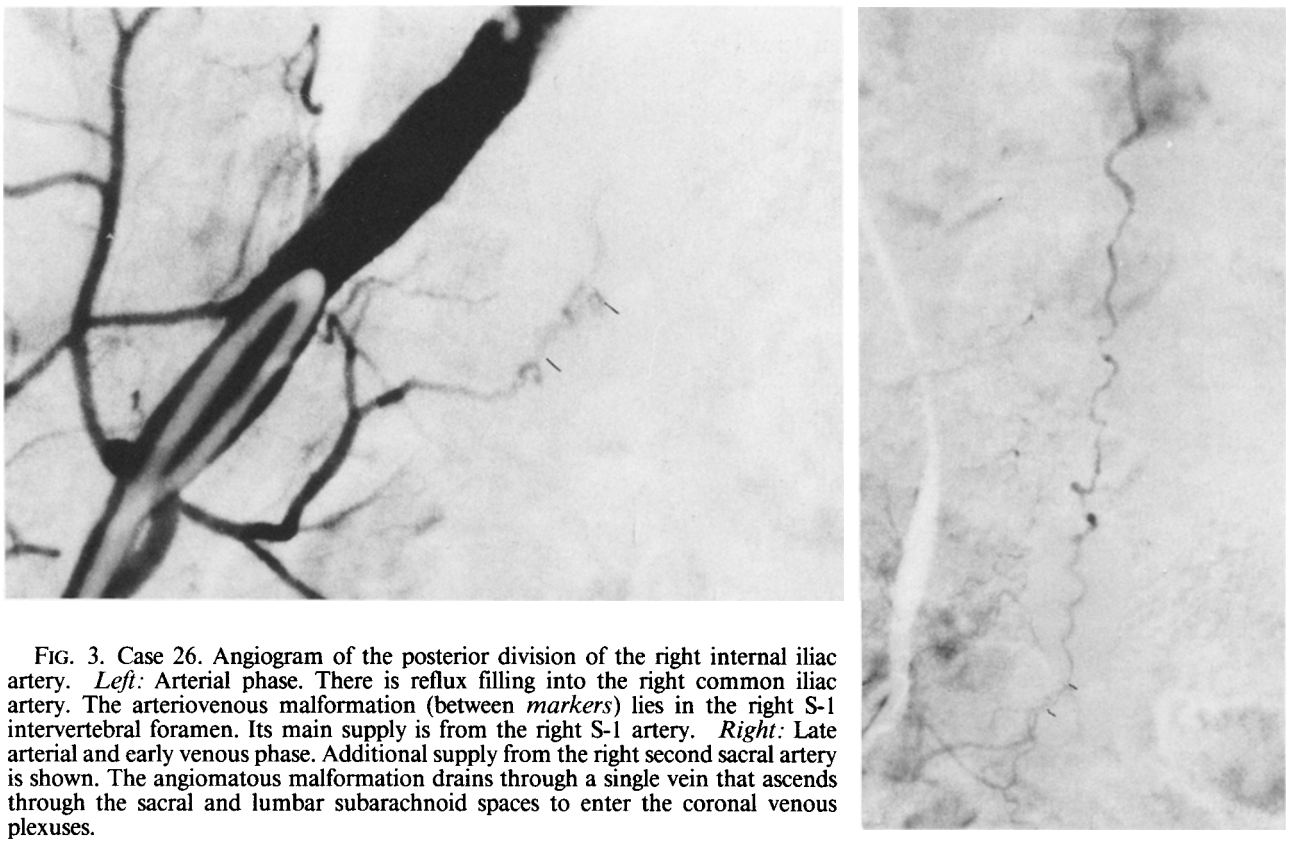

FIG. 3. Case 26. Angiogram of the posterior division of the right internal iliac artery. *Left:* Arterial phase. There is reflux filling into the right common iliac artery. The arteriovenous malformation (between *markers*) lies in the right S-1 intervertebral foramen. Its main supply is from the right S-1 artery. *Right:* Late arterial and early venous phase. Additional supply from the right second sacral artery is shown. The angiomatous malformation drains through a single vein that ascends through the sacral and lumbar subarachnoid spaces to enter the coronal venous plexuses.

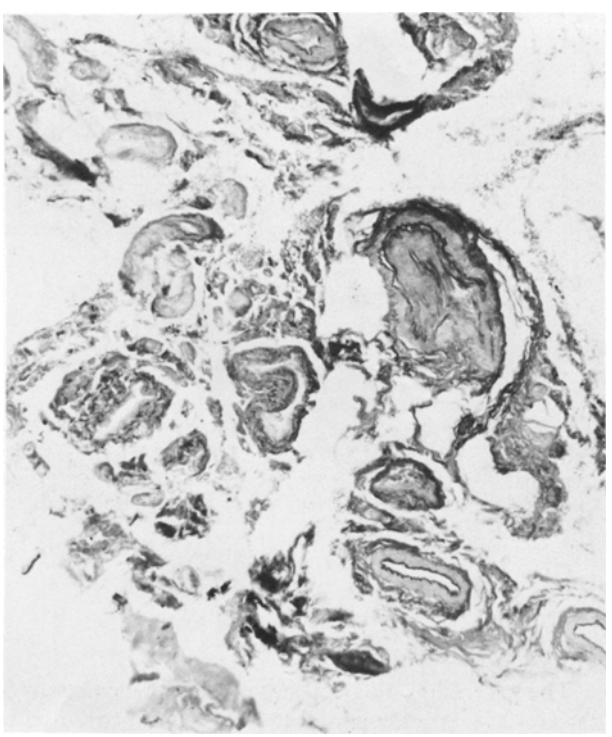

FIG. 4. Case 34. Photomicrograph of a surgical specimen showing numerous blood vessels. Some of them are arteries with well-formed internal elastic lamina, others are veins. Elastica van Gieson, × 105.

especially in series preceding the widespread use of selective spinal angiography.

At present, the incidence of spinal dural AVM's is not known accurately. In 55 of our own 60 patients, the AVM was situated extradurally in the dura mater. The remaining five angiomas involved the spinal cord. None of our patients had a so-called "retromedullary angioma." Merland, *et al.*,[11] suggested that the majority of retromedullary angiomas are radicomeningeal AV fistulas, which are the same type of spinal dural AVM's as we describe here. The incidence of dural AVM's was thus extremely high and that of spinal cord angiomas was low. There are two reasons for this. First, our classification of spinal angioma is different from that of other authors. The classification of our series is based upon accurate identification of the nidus, mainly by angiography and partly by surgical findings. In most previous reports these dural AVM's have not been recognized, since arterialized draining veins passing from the dura to the cord have been mistaken for an artery running up to a fistula on or within the cord. Such arterialized draining veins are not, in fact, arteries of supply, since they join and run into the coronal venous plexus; moreover, no shunt can be found in relation to such a vessel. The second reason for the low incidence of spinal cord angiomas, predominantly found in children and young adults, may be case selection, for a general pediatric service is not provided at our institution.

Clinical Findings

Males were affected more often than females in a ratio of 9:1. The onset of symptoms can be at any age but is most common in the sixth or seventh decades. The average age of 57 years for dural AVM's is higher than that for spinal cord angiomas (37 years), even when we take into account case selection differences.

Our study shows that the initial symptoms are not specific for dural AVM's, and that they are similar to those of cord compression from any cause. A disturbance of micturition or defecation was the initial symptom in only four patients. However, this was one of the most common symptoms at the time of diagnosis, by which time most of the patients had a highly suggestive combination of motor, sensory, and sphincter disturbances. The sphincter disorder is similar to that occurring in cases of intradural angiomas, in which it develops soon after the onset of symptoms and much earlier than with either extramedullary benign neoplasms or intrinsic gliomas.[2]

The clinical history of dural AVM's differs from that of spinal cord and extradural angiomas. Pia[14] indicated that an apoplectic course, found in 39% of cases of extradural and 45% of intradural angiomas, is always strongly suggestive of a spinal angioma, and that a chronic progressive course usually suggests the presence of a spinal tumor. In his series, 17% of epidural angioma patients had extradural hematoma, and 38% of intradural angiomas were associated with subarachnoid hemorrhage (SAH), subdural hematoma, or hematomyelia. In other series,[13,22] the symptoms of spinal cord angioma were often of sudden onset, and SAH was common. The symptoms of dural AVM, however, are usually gradual in onset. None of our patients with dural AVM showed either extradural or intradural hematoma at surgical exploration. Arachnoiditis, which was possibly due to an earlier episode of hemorrhage,[14] was shown in six of our 55 cases of dural AVM (11%) and three of the five spinal cord angiomas. Each patient with arachnoiditis showed acute onset or sudden aggravation of symptoms during the slowly progressive course. The results suggest that the sudden aggravation of neurological deficit is due to an episode of hemorrhage and that bleeding from dural AVM's, which is usually mild but leads to neurological deficits, is less common than in cases of spinal cord or epidural angioma.

One of the most common signs is sensory disturbance relating to the lesion of the posterior column and spinothalamic tract. Muscle wasting and fasciculation of involved muscles were demonstrated in 56% and 13% of cases of dural AVM, respectively. The common sign is a combined upper and lower motor neuron deficit in the legs.

The duration of symptoms of dural AVM was less than 3 years in 66% of cases. Symptoms were often exacerbated by straining, by certain postures, or by exercises, which are the same precipitating factors as in cases of spinal cord angioma.[14]

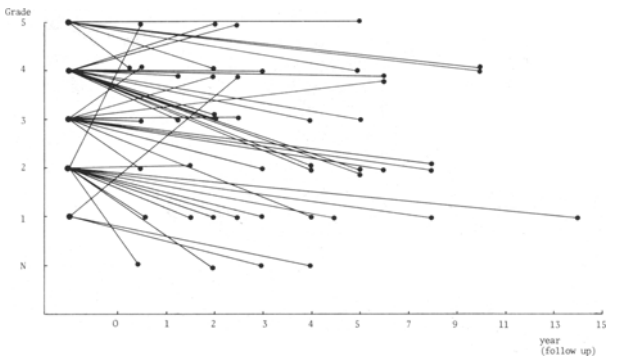

FIG. 5. Changes of grade of gait disturbance at latest follow-up examination after operation for spinal arteriovenous malformation. For definition of grading see text.

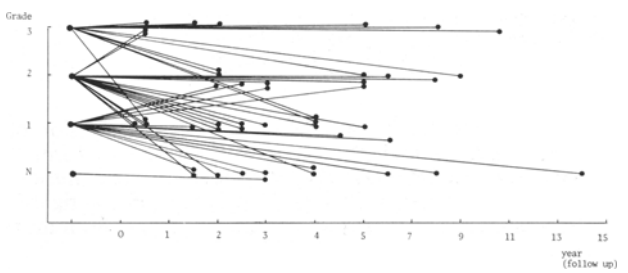

FIG. 6. Changes of grade of micturition disturbance at latest follow-up examination after operation for spinal dural arteriovenous malformation. For definition of grading see text.

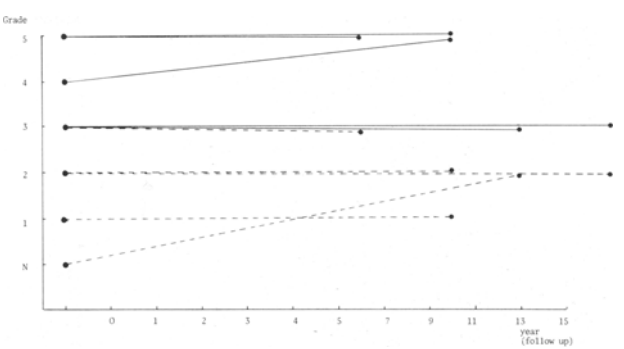

FIG. 7. Changes of grade of gait disturbance *(solid line)* and micturition disturbance *(broken line)* at latest follow-up examination after decompressive laminectomy for spinal dural arteriovenous malformation. For definition of grading see text.

Pathophysiology

The pathophysiology of these neurological deficits of dural AVM is uncertain, but it has been attributed to compression of the spinal cord by the venous bulk,[17] to arachnoiditis due to SAH,[14] or to ischemia resulting from raised venous pressure and the steal phenomenon.[9,14,17]

Compression of the cord by enlarged draining veins seems improbable to us, since myelographic evidence of obstruction in the subarachnoid space was rare, and there was no evidence of cord compression at surgery. Moreover, decompressive laminectomy was ineffective in all five cases in which it was the sole treatment (Group II). Arachnoiditis was not common in our series, and therefore seems unlikely to be the cause of symptom production.

Increased venous pressure is the most important factor in the production of neurological deficit.[1,7] The shunt itself is situated on the dura mater and its venous drainage is, at least in part, to the coronal venous plexuses which lie longitudinally over the posterior and lateral surface of the cord. Much of the confusion relating to so-called "spinal angiomas" has arisen from the failure to recognize the dural site of the shunt. The disordered physiology depends upon a high-pressure draining vein leading from the region of the shunt and communicating with the coronal plexus on the cord itself. This vein may have a very long intradural course, as, for example, from a shunt in the sacral sac. Distension of the coronal plexus may extend from the lumbar enlargement as high as the cervical region. The erroneous impression that these dilated veins were all angiomas led to preliminary attempts to excise them over great lengths. In fact, division of the communicating vein between the shunt itself and the coronal plexus resolves the cause of interference with function of the spinal cord. It is interesting also that in one of our cases where the shunt was on the dura at the foramen magnum (Case 38, Fig. 8) the symptom complex followed a pattern very similar to that in dural AVM's of the most common thoracolumbar region, suggesting that the principal site of the interference with cord function from high-pressure coronal venous plexus blood is in the region of the lumbar enlargement, whatever the site of the dural AVM. This presumably is strongly dependent upon posture.[6] The coronal plexuses receive blood from radial veins draining the intramedullary circulation. The shunt in the dura leads to an increased pressure not only in the epidural veins, but also in the coronal plexus. The AV pressure gradient and spinal blood flow are decreased as a result of increased intramedullary venous pressure. The reduced AV pressure gradient in turn leads to intramedullary vasodilation and to the development of increased tissue pressure possibly associated with progressive exhaustion of the autoregulatory capacity.[19] The intravascular pressure changes are transmitted directly to the adjacent cord substance, and progressive formation of edema may result from this undamped pulsation as well as from the underlying ischemic process. In the most advanced cases, extensive ischemic loss of cord tissue may result.

Investigations

Cerebrospinal fluid analysis was often abnormal, with an elevated protein content. Myelography of dural AVM's usually demonstrated dilated tortuous vessels, and this finding is similar to cases of spinal cord angioma.

Angiography has demonstrated that many spinal cord angiomas have a nidus of abnormal vessels.[4,5] The nidus of dural AVM's is also usually visible on angiog-

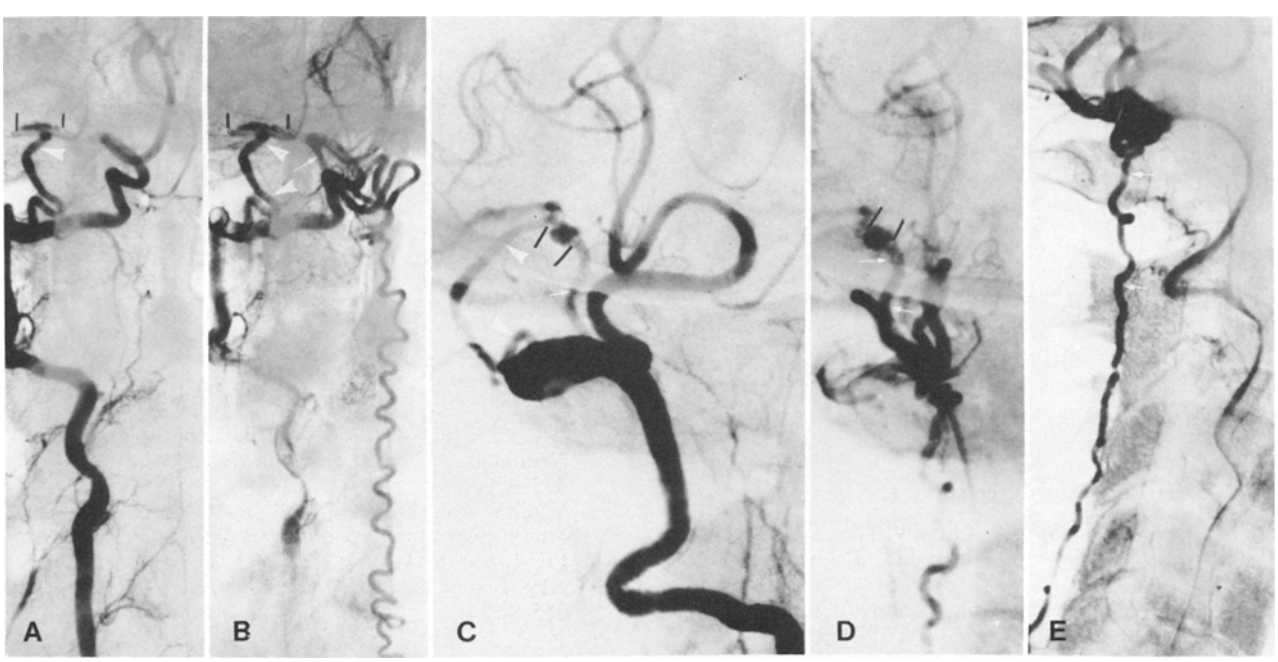

FIG. 8. Case 38. Right vertebral angiograms, anteroposterior (*A* and *B*), and lateral projections (*C*, *D*, and *E*). The dural arteriovenous malformation (between *markers*) is on the right side of the foramen magnum. It is supplied by an enlarged posterior meningeal branch *(arrowheads)* of the vertebral artery and drains medially through a single large vein *(arrows)* into the posterior coronal venous plexus, which fills caudally.

raphy as an irregular conglomeration of small vessels, because the feeding arteries converging on the shunt divide into small branches. Drainage from the shunt is usually via one vein, sometimes several. These veins are usually wider than the shunt vessels.[7] These angiographic findings are similar to those in cases of spinal cord angioma.[5] Merland, *et al.*,[11] suggested that the lesion is not an angiomatous network but a single AV fistula, based on the angiographic findings; however, we believe it is an AVM rather than an AV fistula, because histological examination has shown numerous vessels with an admixture of arteries and veins (Fig. 4). The location of the dural AVM is characteristic. In 82% of dural AVM's the shunt was projected lateral to the spinal cord. In 30% of these instances, the nidus encroached into the intervertebral foramen. In each case, one or sometimes two veins emerged to penetrate the theca and enter the tortuous and dilated veins of the coronal venous plexus on the cord.

Results of Surgical Treatment

The operative results of treatment of spinal angioma depend not only on the location of the shunt but also on the type of operation.[15,22] Decompressive surgery has had no effect in any of our patients in whom it was the sole method of treatment. Dural AVM's should be treated by excision of the shunt where possible, or disconnection of the shunt from the coronal plexus. Total excision of all dilated veins on the coronal plexus is unnecessary and may be damaging.[5] Compression of the cord by the enlarged veins is improbable.

Most of these lesions can be treated without increasing the neurological deficit. In 20 (65%) of 31 severely disabled patients and 12 (80%) of 15 moderately disabled patients, surgical treatment led to improvement of disturbed gait. These results are better than has been reported in cases of intradural angiomas,[15] and suggest that good results are most likely in those patients who have mild or moderate neurological deficit preoperatively. The duration of symptoms is long, but neurological deficits develop progressively and finally become irreversible. It seems to us, therefore, that a progressive neurological deficit, especially in its early stages, is a good indication for operation. The simple disconnection of the nidus of the shunt from the coronal venous plexus is effective in most cases, apparently permanently, and is substantially without risk.

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Address reprint requests to: Lindsay Symon, T.D., F.R.C.S., Gough Cooper Department of Neurological Surgery, Institute of Neurology, The National Hospital, Queen Square, London WC1N 3BG, England.