SPINAL CORD ANGIOMA—NON-PROGRESSIVE DISEASE?

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Abstract. In an analysis of 214 patients admitted to the Midland Spinal Injuries Unit with non-traumatic cord damage 18 patients had arteriovenous malformations (AVM) of the cord. This paper describes the experience gained by this unit in the rehabilitation of these patients. A brief review of literature is included, essentially to highlight the relentless progress of the disease. The overall rehabilitation potential of these patients was poor. Fifty per cent of patients had further neurological loss during the follow-up period after the onset of paraplegia.

Key words: Spinal cord angioma; Rehabilitation potential; Diagnosis; Therapy.

Introduction

SINCE its first description by Hebold (1885) and Gaupp (1888) several terms have been employed by various authors to describe the histological appearance of spinal cord AVM. The most recent classification by Bergstrand *et al.* (1964) divides them into three groups: (1) Angioma cavernosum, (2) Angioma racemosum and (3) Angioreticuloma (haemangioblastoma). This classification is now more commonly recognised. By 1971 there were nearly 500 cases of spinal cord AVM reported in the literature (Yasargil 1971). The age at onset in the majority of these cases was between 30 and 50 years with a distinct male predominance. Lower dorsal and upper lumbar cords were most frequently involved. Rarely they may present as Lindau syndrome which comprises of haemangioblastomas at various sites in the central nervous system and tumours and cysts of visceral organs, transmitted as a dominant trait. Spinal angioma may also be associated with retinal or cerebellar haemangioblastoma. Cases have been reported of spinal AVM co-existing with hereditary haemorrhagic telangiectasia (Rendu-Osler-Weber syndrome) (Merry 1976). Bony anomalies of the spine, scoliosis, congenital heart diseases, mesenchymal or ectodermal abnormalities, polycythemia have all been reported in association with spinal AVM.

Materials and Methods

Case records of 18 patients who were diagnosed elsewhere but who were admitted for rehabilitation to the Midland Spinal Injuries Unit between 1956 and 1975 were studied. Thirteen of these patients spent their primary rehabilitation at this unit, five patients were initially treated elsewhere but followed-up at the Midland Spinal Injuries Unit, for further rehabilitation or complications of paraplegia. Three were lost to follow-up, and two were discharged from follow-up because of good functional recovery.

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Results

Age at onset of symptoms of the 18 patients varied between 18 and 69 years. Age and sex distribution is shown in Figure 1. There were four females and 14 males. Duration of symptoms before diagnosis varied from 2 days to 8 years (Table I).

FIG. 1

TABLE I

Duration of symptoms before diagnosis	
0-1 month	5
6 months	3
1 year	1
2 years	3
4 years	3
8 years	1
Unknown	2

Disturbed micturition was the most common presenting symptom. Sensory disturbance was difficult to ascertain and in several cases motor loss was vague and diffuse, sometimes transient. In two patients the initial symptom complex could not be confirmed (Table II).

Initial neurological level at the time of admission is shown in Table III.

The clinical findings altered during the hospital stay and over the follow-up period. Five patients improved during the rehabilitation and follow-up. In nine

TABLE II

Initial symptoms	
Radicular pain	8
Back pain	3
Calf pain—cramps	5
Paresthesia	3
Girdle pain	2
Subarachnoid haemorrhage	3
Disturbed micturition	10
Premature ejaculation	1
Unsteady gait	3
Weakness of the legs	9

TABLE III

Neurological level on admission to rehabilitation centre	
Cervical	1
Upper Dorsal (D1-4)	1
Mid Dorsal (D5-8)	4
Lower Dorsal (D9-12)	4
Lumbar	6
Unknown	2
Total	18

patients there was definite deterioration of neurological deficit, three patients had no detectable change, while one patient made marginal neurological gains though without much functional alteration (Table IV).

Disability developed gradually in seven patients. Acute onset of neurological deficit occurred in six patients, including one whose paraplegia followed childbirth and another following varicose vein surgery (Table V).

Myelogram was known to have been carried out in 15 patients while the records were incomplete in three. There was one false positive and two false negative myelograms and 12 were diagnostic. The false positive myelogram led to discectomy at L5/S1, but the diagnosis was subsequently confirmed by cisternal myelogram. One of the false negative reports was later corrected following a repeat myelogram. Thus only in one of the 15 patients myelogram failed to be diagnostic. Spinal angiography was done in only three of the 15 patients and was diagnostic in two.

Sixteen patients had laminectomies while two had no surgery. One of the 16 had a myelotomy. Angioma was excised in only two patients and partially excised in two others. One patient had two laminectomies. None of the patients were treated by selective angiographic embolisation. Two patients were found to have thrombosed AVM during surgery.

Of the 13 patients for whom the records were complete (three patients were

TABLE IV

No.	Initial	Sex	Neurological level on first admission	Neurological level on final follow-up	Follow-up since onset
1	PJ	M	C2 Brown Sequard	Complete recovery	3 months
2	CB	M	D4 paraplegia	D4 paraplegia	9 years
3	EH	F	D6 paraplegia	D3 paraplegia	20 years
4	FG	M	D6 subcomplete	D4 complete	1 year
5	ER	M	D6-7 subcomplete	D3 complete	6 years
				C7 sensory	
6	MS	F	D7 paraparesis	Walking with sticks	1 year
7	DD	F	D10 Brown Sequard	D7 complete hydrocephalus, optic atrophy	4 years
8	SK	M	D10 paraparesis	Walking	4 years
9	HA	M	D10 paraplegia	D8 paraplegia	21 years
10	MF	M	D10 paraplegia	L2 paraparesis	4 years
11	PD	M	L1-2 paraparesis	L1-2 paraparesis	4 years
12	PT	M	L1 paraparesis	D8 paraplegia	3 years
13	AJ	M	L1 paraplegia	L1 paraplegia	4 years
14	DH	M	L2 subcomplete	D11 complete	5 years
15	JW	F	L4 subcomplete	L1 complete	12 years
16	BC	M	L4 paraparesis	D10 paraplegia	3 years
17	HB	M	No definite level	Walking with sticks	4 years
18	WM	M	No definite level	Walking with sticks	3 years

TABLE V

Development of disability	
Acute onset (2 days-3 months)	4
Gradual onset with slow progression	7
Slowly progressive with step-wise worsening	2*
Postpartum	1
Following unrelated surgery	2
Following minor trauma	1
Unknown	1
Total	18

* The initial neurological deficit in one of the two patients in this group followed a minor trauma.

lost to follow-up and two were discharged because of good functional recovery), five patients died of causes listed in Table VI.

Two patients were above 65 years at onset of symptoms. Of the remaining 16 patients one who had complete recovery and three paraplegics, are employed. One patient died after being a typist for a short time. Two others, a secretary and a housewife, were lost to follow-up. Nine patients never managed to secure a job.

Thirteen patients in this series had their primary rehabilitation in this unit and spent an average of 5.2 months on their initial admission (range 2-11 months).

TABLE VI
The cause of death

Cause	No.	Duration of survival since paralysis
Uraemia with pressure sores	1	4 years
Uraemia with nephrolithiasis	1	9 years
Cerebralvascular accident	1	21 years
Cerebral angioma blocked Spitz Holtzer valve	1	4 years
Massive subarachnoid haemorrhage	1	1 year

Pressure sores have been a major problem in at least four patients and have been a contributory factor to the death of two patients. Cerebral haemangioma, associated with optic atrophy from increased intracranial pressure was found in one patient. One patient had diabetes, one a malrotated large bowel and another mitral stenosis.

Review of Literature and Discussion

The incidence of spinal cord AVM, according to Béraud (1972) is between 3.3 and 11 per cent of all spinal cord tumours. Yasargil (1971) in a review of 500 cases puts the incidence at 4.3 per cent. Aminoff and Logue (1974) reported a 4 : 1 male to female ratio, while Yasargil 3.3 : 1. In the present series the ratio is 14 : 4. Even though all the patients in this series were adults, the youngest case reported in literature was 1 year old and the oldest 72. Fifty per cent of the patients in the present series were aged between 35 and 65. Aminoff *et al.* (1976) recorded that 46 of their 60 patients were between 40 and 70 years. In accord with other series, a majority of patients in the present series had lower dorsal or dorso-lumbar lesions. Only 14 of the 60 patients reported by Aminoff *et al.* (1974) had radicular pains, compared to 8 of 18 patients in this series. In common with Tobin *et al.* (1976), Yasargil (1971) and Béraud (1972) weakness was the most common presenting symptom in our series. Gregorius *et al.* (1970) in a review of patients with spinal cord AVM record radicular pain as the most common symptom. Aggravating factors such as menses, pregnancy (Newman, 1958), parturation, injury and exercise have all been well documented. In our series two patients had some form of trauma precipitating paraplegia and one patient had signs of intrathecal haemorrhage 15 hours after normal delivery. Unrelated surgery has been the aggravating factor in two patients.

Yasargil (1971), Béraud (1972), Aminoff *et al.* (1974) and Tobin *et al.* (1976) reported that most of the patients developed signs and symptoms gradually. In the present series 50 per cent had major paralysis within 1 year of onset of initial symptoms, six of the 17 patients whose records were complete had acute onset of paralysis in less than 3 months (Table I). On the other hand 48 of 60 patients reviewed by Aminoff *et al.* (1974) had progressive deterioration of neurological disability. Similarly in the report by Tobin *et al.* (1976) 51 of the 71 patients reviewed were slowly progressive. This apparent difference in the course of disability is likely to be due to the fact that rehabilitation units receive most of the acutely paralysed patients.

Even after paralysis there is a relentless deterioration of neurological disability. As shown in Table IV 50 per cent of patients deteriorated during the follow-up period and only three of the 18 paralysed patients had nearly unchanged neurological status. Pathogenesis of progressive neurological loss is uncertain. There is evidence to believe that haemangioblastoma are mesenchymal rest tumours (Lindau, 1926) and haemangioblastomatous Syringomyelia is probably due to transudation of fluid from the tumour (Lindu 1931). In a review of 85 cases of spinal haemangioblastoma Browne *et al.* (1976) noted that 43 per cent had syringomyelia. Spinal cord AVM have been shown to have several rapid arteriovenous shunts (Bergstrand *et al.*, 1964). Arteriovenous shunting has been demonstrated in other spinal cord tumours as well (Yuhl *et al.*, 1976). Progressive cord ischaemia results from a 'steal phenomenon' (Yasargil 1971, Béraud 1972), and this may contribute to progressive neurological loss. In our series only two patients had excision of the AVM, one was a cavernous haemangioma while the other was a haemangioblastoma.

Even though only four of the 16 patients eligible to work were employed, three more patients were employed at least for a period after rehabilitation. However 50 per cent of patients never returned to work, partly due to the fact that a good number of them were above 40 years of age at onset and partly due to continuing neurological signs and symptoms. Adequate rehabilitation in younger patients should improve this poor employment prospect of these patients.

Radiotherapy was not employed in any of the patients in this series and has no effect whatever on vascular malformations of the cord (Béraud 1972). Laminectomy alone or in conjunction with coagulation of AVM has not been beneficial in most of our patients. Yasargil (1971) noted that 42.7 per cent of patients were worse off following decompressive surgery, while only 18.7 per cent had some improvement of subjective complaints. The finding of thrombosed veins at laminectomy in two patients in our series probably represents Foix-Alajouanine disease (1926). Yasargil (1971) believes that spinal cord AVM are best treated by surgical excision, possibly employing microsurgical techniques and throws doubt on the efficacy of ligation of feeding vessels. Kasdon *et al.* (1976) emphasised that surgical exploration of all spinal AVM should be performed despite angiographic appearance. Spinal angiography first reported by Höök and Lindvall (1958) has now become a very useful diagnostic tool. In our series three patients had spinal angiography, only two were technically satisfactory, even though myelograms were diagnostic in all three. Newton *et al.* (1968) reported successful obliteration of the artery of Adamkiewicz in a case of spinal cord angioma leading to neurological recovery. Lead pellet embolisation employed in the early days has now been replaced by crushed muscle, Gelfoam (Decker *et al.*, 1975) and more recently by expandable compressed Ivalon sponge (Ausman *et al.*, 1977). Djindjian (1975) reviewed 34 cases treated by selective embolisation and recorded four cases in whom there was temporary deterioration of neurology with complete subsequent recovery. He believes selective embolisation to be the best available treatment of spinal cord AVM. Extensive excision of angioma using microsurgical techniques appeared to be superior to earlier forms of surgery but selective spinal arterial embolisation perhaps holds a promise for the future.

Conclusion

This study shows that neurological deficit continues to deteriorate in at least 50 per cent of patients who had paralysis from spinal cord arteriovenous mal-

formation. Half the patients in this series did not return to their work even after good rehabilitation. A greater proportion of patients had acute onset of paralysis in this series compared to other reports. Five of the 13 patients died as a direct sequela of AVM and paraplegia.

RÉSUMÉ

Une analyse de 204 malades avec lésions de la colonne vertébrale non-traumatiques révéla 18 malades avec angioma de la colonne vertébrale. Divergence de la littérature en ce qui concerne la manière dont la perte neurologique commence est notée. La perte continua pendant les soins post-hospitaliers dans 50% des cas, et contribua à la mort de 5 malades. Dans l'ensemble le potentiel pour la réhabilitation de ces malades est pauvre. La valeur de divers procédés diagnostiques et thérapeutiques est discutée.

ZUSAMMENFASSUNG

Bei Untersuchung von 204 Patienten mit nichttraumatischen Rückenmarksschädigungen zeigte es sich, dass bei 18 der Patienten ein Rückenmarksangioma bestand. Eine Abweichung von der in der Literatur beschriebenen Art und Weise des Auftretens der neurologischen Symptome wurde festgestellt. Ein permanenter neurologischer Verlust wurde bei 50% der Fälle beobachtet, und dieser trug dazu bei, dass 5 der Patienten starben. Allgemein besteht bei diesen Patienten nur eine geringe Aussicht auf Genesung. Der Wert verschiedener diagnostischer und therapeutischer Massnahmen wird besprochen.

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