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Diagnosis and Treatment of Spinal Angiomas

By

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With 10 Figures

Summary

An account is given of 93 spinal angiomas found in 74 patients. 55 were solitary malformations and 38 were complex. In 5 patients angioma and angioblastoma were both present, and in 1 case an angioma was found in association with an aneurysm.

The main problems of morphology, diagnostic investigation, improvement of early diagnosis, complications as a result of bleeding and circulatory disturbances with arachnoiditis and myelo-malacia and the microsurgical treatment with radical removal and the palliative procedures are discussed.

Results depend on the situation of the angioma. After total extirpation of intradural angiomas in 34 cases, 14 patients recovered completely, 13 recovered partially, 5 were unchanged, and 2 deteriorated.

With the development of selective methods of angiography and improvements in operative technique, accurate diagnosis and operative treatment of spinal angiomas have become more practicable.

In contrast with cerebral angiomas there seems to be no agreement so far on operative methods and indications. The reasons for this seem to be scanty experience, diversity of clinical material, and differing presentations in relation to functional morphology and pathogenesis. In practice one of the most important problems is early diagnosis. I am reporting some of our own experiences in the hopes of answering some of these questions.

Morphology

Our observations deal with 74 patients with a total of 93 angiomas (Fig. 1). Of these 55 were *solitary* and 38 *complex*. In the latter group the vertebral-extradural type were more numerous than the extradural-intradural variety. Quite remarkable are those angiomas showing *involvement of all the contiguous layers*, apart from the skin and the dura,

	N Angiomas	Isolated	N	Combined Types		N Patients
Vertebral A.	13	5	8	Epidural	8	5
Epidural A.	30	15	15	Vertebral	8	
				Intradural	6	23
				Angioblastoma	1	
Intradural A.	42	32	10	Epidural	6	42
				Angioblastoma	4	
Angioblastoma	8	3	5	Epidural	1	4
				Intradural	4	
	93	55	38			74

Fig. 1. Spinal angiomas. Neurosurgical University Clinic Giessen 1954—1970

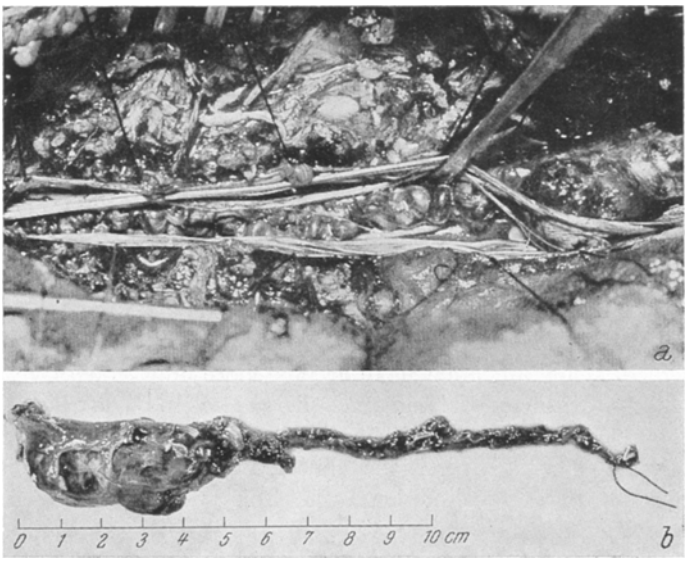

Fig. 2 a and b. Combination of spinal angioma and angioblastoma

from the subarachnoid space to the subcutaneous tissue. In one case an extradural lesion and a cutaneous angioma in the related ectodermal segment were both present.

The close relationship between *angioma and angioblastoma* was shown by the presence of both conditions in each of five patients (Fig. 2). In one case a secondary angioblastomatous malformation developed at the

site of a primary angioma. The combination of *spinal angioma and spinal aneurysm* is extremely rare (Fig. 3).

Epidural angiomas are in our experience more frequent than supposed, among these, cavernous and racemose angiomas predominated.

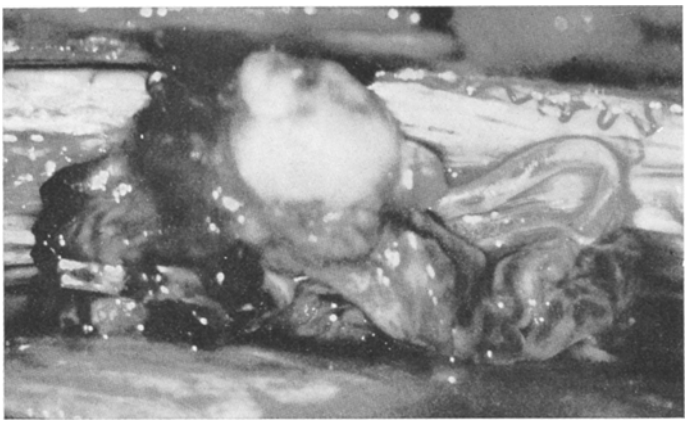

Fig. 3. Combination of spinal angioma and aneurysm

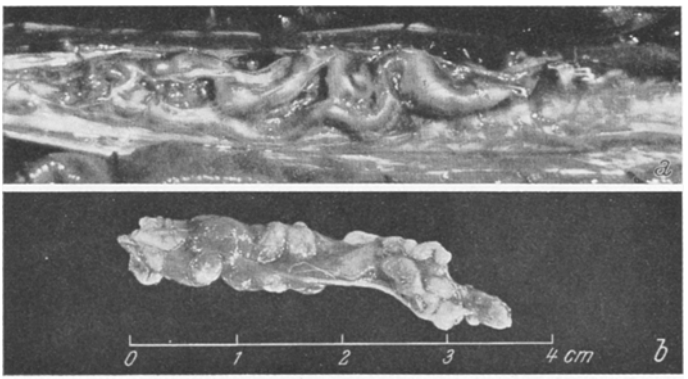

Fig. 4. Arterio-venous cirsoid angioma; a normal type, b solitary type

Another anomalous type is the lumbo-sacral vascular anomaly which presents with a chronic unilateral or bilateral sciatica and the rapid onset of footdrop. A recent investigation of 124 cases[3] including 48 without disc lesions showed varicose venous dilatation with but few definite signs of vascular malformation. The frequent combination of isolated vascular anomalies with lumbo-sacral dysraphism points to their congenital nature, so that possibly they also belong to the group of angiomas.

The most frequent and classical intradural angioma is the subarachnoid *arterio-venous cirsoid angioma* with one or more afferents ("feeding arteries") from the dorsal radicular arteries. As a rule it extends over four or five vertebral levels (Fig. 4 a); a lesion involving the whole length of the cord, or a solitary angioma, is less common (Fig. 4 b). One must emphasize the striking predilection for the dorsal aspect of the lower thoracic cord.

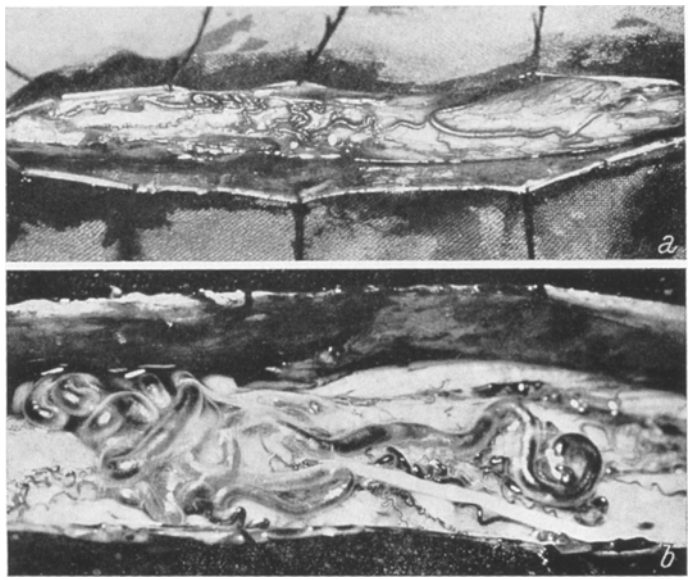

Fig. 5. Capillary angioma; a isolated, b combined

The *capillary angiomas* are quite distinct (Fig. 5 a). They lie subpially, communicating with the medullary arteries or emerging from them. I do not consider it justifiable to conclude from this that there is intramedullary involvement. Occasionally the large cirsoid angiomas are combined in their peripheral portions with capillary angiomas (Fig. 5 b). On only one occasion have we seen an *intradural cavernous angioma.*

It was not my intention to increase the vagueness of the classification by a further subdivision. My chief concern was to bring to light the *topographical relations.* Biopsy and angiography have removed to a considerable extent the uncertainty of histological classification and have also confirmed that the spinal angioma is predominantly an arterio-venous cirsoid angioma. It is doubtful if arterial and venous cirsoid angiomas can coexist (Fig. 6).

Slowing of circulation, darker coloured vessels, and partial thrombosis are explained by the specific pattern of the spinal circulation and

above all by the differing directions of the circulation. These appearances do not imply the presence of a venous angioma.

Diagnosis

Just as the *primary and secondary damage to the spinal cord* determine the symptomatology and course of the disease so also do they influence the treatment to be adopted. As in the case of the cerebral angiomas the most important and constant disturbance is the *local reduction of blood flow* on account of the *arterio-venous shunt, a spinal "steal syndrome"*.

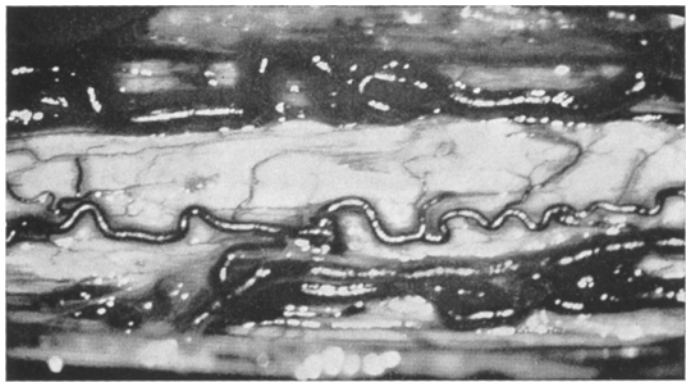

Fig. 6. Isolated arterial cirsoid angioma?

Even with large and elongated convoluted vessels intradurally, *primary compression* with total spinal block is unusual. This is substantiated not only by the Queckenstedt Test and myelography but also by the predominance of nuclear and pyramidal paraparesis and the relative rarity of posterior column defects.

In this connexion it should be pointed out that the so called Foix-Alajouanine syndrome is not a disease *sui generis* but is the typical sequel of the chronic circulatory disturbances of the cord caused by angiomas of various types.

Raised CSF protein and the partial or complete block which was present in almost 50% of our cases, are as a rule sequels of *adhesive and cystic* arachnoiditis. The most important cause of severe partial or complete adhesions is unsuspected subarachnoid haemorrhage (Fig. 7a, b, c). In some cases, an acute haemorrhagic space occupying lesion (extradural, subdural or intramedullary), or an angioma complicated by an angioblastoma, can lead to a spinal block. Early compression symptoms predominate in extradural angiomas, in contrast to the intradural lesions.

The main diagnostic problem is a *wrong diagnosis* (Fig. 8). In an earlier atlas of X-ray diagnosis it was reported that no extradural angioma was diagnosed[13, 14]. In four cases of intradural angioma the condition was suspected once, and three times the diagnosis was made after years of delay. The remaining cases were unrecognised for years

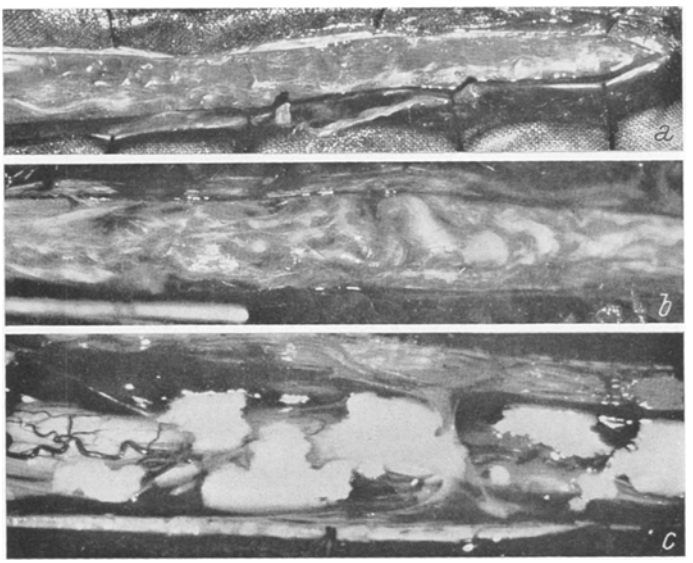

Fig. 7. Secondary arachnoiditis; a after fresh subarachnoid bleeding, b two years after subarachnoid bleeding, c calcified arachnoiditis in a case of capillary angioma and Bechterew disease

or even decades. A wide range of diagnoses had been suggested, led by *multiple sclerosis* in 15 and *disc lesions* in 14 cases. It is remarkable that on three occasions a diagnosis of juvenile kyphosis was made. Where a significant kyphosis occurs as the presenting symptom of a unilateral angioma it is the expression of muscular atrophy resulting from loss of anterior horn cells.

The *course of the disease* should give important clues to the possible diagnosis (Fig. 9). An *acute spinal apoplexy* and a *relapsing apoplectic course*—seen in seven out of eight extradural, and 13 out of 29 intradural angiomas—are always strongly suggestive of a spinal angioma. Whilst a *chronic progressive course* almost inevitably leads to the diagnosis of tumour, there were frequent cases with a remittent course—seven extradural and eleven intradural angiomas—which were regarded as having a solitary plaque of demyelination (*"monofocal" disseminated sclerosis*).

	Correct diagnosis	N	Diagnosis	N
Epidural angiomas n. 16	—	4	Cerv. spine syndrome	3
			Lumbar spine syndrome	4
			Intermittent clazdication	1
			DS (Multiple Sclerosis)	3
			Radiculo-myelitis	2
			Tumor	1
Intradural angiomas n. 28	after 2 yrs. DS	5	Cerv. spine syndrome	1
	after 4 yrs. Polyneuritis		Lumbar spine syndrome	6
	after 6 yrs. Cerebral-aneurysm		Juvenile kyphosis	3
	Suspected angioma 1		Tbc spondylitis	1
			Motor. Spinal claudication	1
			DS (Multiple Sclerosis)	11
			Myelitis	3
			Poliomyelitis	1
			Polyneuritis	2
			Subacute comb. degeneration of the spinal cord	1
			Syringomyelia	1
			Motor. neuron disease	1
			Tumor	5
			Cerebral SAH	5

Fig. 8. Diagnostic problems in spinal angiomas (Referral diagnosis) (1968)

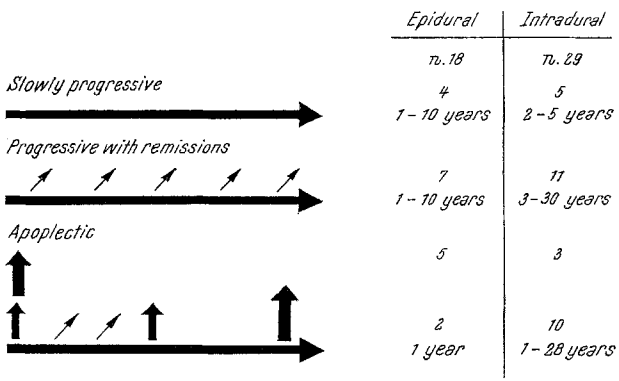

Fig. 9. Onset and course of spinal angiomas (1968)

Of more practical significance there are *certain precipitating factors* which have been noted in spinal exacerbations and apoplexies. Among our cases such factors included menstruation, pregnancy, bodily exertion, heat and cold, excitement, and irrelevant trauma. In the case of vertebral—extradural angiomas we have shown on three occasions

spontaneous fracture of a vertebra with extradural haematoma formation as a cause of sudden deterioration; with intradural angiomas we have seen one *subdural haematoma*, eighteen unrecognised *subarachnoid haemorrhages* in eight patients, and two *haematomyelias*.

The diversity of the types of angioma and their secondary variations demands the use of *special diagnostic procedures*. Investigations should start with examination of the CSF and Queckenstedt reaction, combined in non-acute cases with isotope myelography. *Spinal ossovenography*[12, 16] is recommended before considering myelography. This confirms or excludes an extradural angioma and a space occupying lesion. Enlarged and more numerous vessels, possibly paravertebral and in the soft tissue, confirm the presence of an angioma, whilst an interruption of the extradural venous plexus is due to an intraspinal space occupying lesion. Later on in acute cases and at one and the same session we undertake *selective angiography* according to the technique developed by Djindjian, Di Chiro et al.[2, 4, 5]. It is hoped that angiography (morphological and functional) of the spinal arteries will eventually be adopted as a routine method.

Up to the present one is unable to dispense with *myelography* as a means of demonstrating angiomas. Typical findings in 30% of angiomas are filling defects, loops of tortuous vessels, and arachnoiditis. Finally, the finding of a tumor in 10% of cases further underlines the great diagnostic value of myelography.

Operative Treatment

The most important therapeutic task in spinal angiomas, as with the comparable cerebral angiomas, is in accordance with most of the literature reports[2, 4, 7, 8, 12] the elimination of the impairment in blood flow produced by the arteriovenous shunt. The presence of compression by angioma, angioblastoma, or haematoma, influences the type of operative procedure adepted. In addition, a pre-operative study of the location, extent and type of the malformation and its feeding and draining vessels is desirable. Pre-operative angiographic demonstration and identification of feeding and draining vessels, particularly in the area of functionally vital arteries like the artery of Adamkiewicz, is still very far from being a matter of course and indeed in certain cases it is impossible.

Many problems of operative technique remain undecided. For this reason personal experience and examination of clinical material are important.

This is the reason why I want to draw attention exclusively to the technical problems of operation on intradural angiomas and angio-

blastomas. Of forty-six patients with this condition, thirty were operated on according to conventional practice and sixteen by micro-surgical methods. *These latter techniques*, generally presented in reports and films, as being the solution of therapeutic problems, should be critically assessed. Undoubtedly, the stereoscopic view with optimal illumination and the magnification of fine details like the small anastomoses between vessels of the angiomas and the sub-pial and cord vessels, are of great advantage. These, together with the possibility of dissection free from hazard, allow one to take care of big angiomatous vessels in every part of the malformation. *Bipolar coagulation* makes these angiomatous vessels shrink to thin strands and impresses us with its ease and elegance. In spite of the obvious fascination of this method such factors as the prolonged duration of the operation, an "over-cautious" dissection, and possibly danger to a cord already damaged by a too extensive use of bi-polar coagulation, must be taken into serious consideration. In one of our cases a deterioration of spastic paraparesis and sensory deficit resulted; in an earlier series operated on with the conventional method of uni-polar coagulation, using weak current and the finest hooks, such a worsening did not occur.

The extensive angiomas (one case of ours extended about 20 cm and another one of the totally extirpated angiomas about 26 cm) situated in the subarachnoid space adjoining the cord are supplied by several radicular arteries and very often by numerous tiny sub-pial vessels. The isolated coagulation of all feeding vessels is not possible and coagulation of single arteries is dangerous for the cord because the Steal Syndrome is increased. Ommaya et al. ("The Washington Group")[10] as well as Houdart et al.[4], advocate *selective intradural ligation of feeding vessels* which have been previously demonstrated by selective angiography. This procedure should lead to the removal of the compression which produced the cord symptoms. A. R. Taylor[15] advises "the occlusion of the supply in cases with a single vessel and unidirectional blood flow", but in all his four cases he removed those parts of the lesions compressing the cord, that is to say he carried out a subtotal removal of the angioma. All the same, it should be noted that by direct pressure measurements it can be demonstrated that compression of the feeding artery produces a drop in pressure from 70 mm Hg to 10–15 mm Hg. It is also worth while mentioning the similar endeavours of Chatterjee[1] and the interesting communication from Newton and Adams[9] about the *artificial embolization* of the afferent vessel to a ventrally situated angioma. In spite of modern techniques *radical operation* remains a problem in certain cases, particularly subpial angiomas with extensive adhesions and angiomas in older patients. It is not practicable in ventral angiomas. It has been claimed that one should begin the extirpation of the angioma with

ligation of the feeding vessels after these have been demonstrated by angiography or by inspection. While this may be highly desirable it is often not possible or necessary. One may encounter great difficulties resulting from earlier bleeding, with subdural and subarachnoid scars firmly adherent to angiomatous vessels, pia mater and cord (Fig. 10 a, b). In these cases, we start the extirpation at either the cranial or the caudal end after having removed the densest scars and then use continue with slight elevation of the loosened part of the angioma. After the identifi-

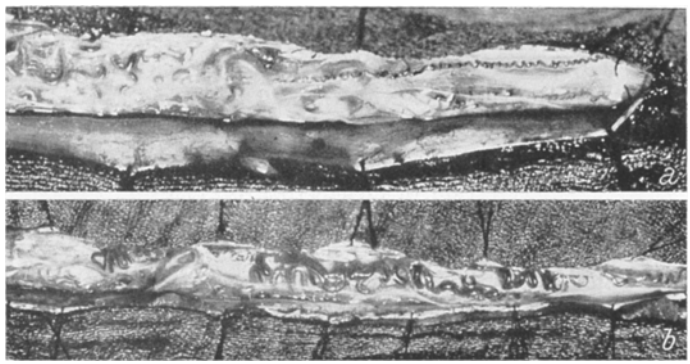

Fig. 10. Extensive (20 cm) subarachnoid angioma several years after at least two subarachnoid haemorrhages and nearly complete paraplegia. a Adhesive arachnoiditis with unusual vascularisation of the membranes, b after nearly complete removal of the adhesions which lasted more time than the following radical extirpation

cation of the lateral and dorsal feeding vessels, these are interrupted near the angioma away from possible medullary branches. In this way total extirpation becomes feasible without risk of cord damage, and mobilisation of the adhesions is easier.

Our own experiences confirm that the *uncomplicated extensive angiomas* which give the typical myelographic appearances are usually well demonstrated angiographically, even by the less experienced. They can generally be removed very easily. In individual cases, the angioma may be easily removed in continuity after dealing with the cranial and caudal main vessels, as in our case of an angioma combined with angioblastoma. As a rule, however, we find adhesions of different intensity and extent. In addition to the known difficulties of meningeal adhesions one must take note of the fragility of the angiomatous vessels and the ever present partial myelo-malacia. Our own experiences tend so much to justify an active attitude—because one half of our material corresponds to these cases—and yet, in the individual case there is so little precision in

assessing the indications for and against the type of procedure to be adopted. One third of our cases consisted of *capillary angiomas* either isolated or in combination with subarachnoid angiomas. These lie subpially. Even at higher magnification one is rarely able to decide to what extent, if at all, these vessels take part in the circulation and nutrition of the cord. Angiography is of no help because these malformations cannot be demonstrated. If these capillary angiomas are combined with large arterial malformations, the interruption of communication between the two kinds of angioma is not dangerous. Isolated capillary angiomas, which in the majority of cases are found in elderly patients with progressive functional cord deficiency, have been left alone. On myelography they show findings of atypical arachnoiditis.

Sustained functional improvement may be achieved by useful *palliative procedures*, such as partial extirpation, removal of portions of angiomas producing pressure, opening up of the cerebro-spinal fluid pathways in adhesive and cystic arachnoiditis, and widening of the dural sac by means of a graft of lyophilised human dura.

We are able to report practically no experiences in the treatment of *acute cases of spinal apoplexy*. In our own material we found one epidural haematoma, one subdural haematoma, two haematomyelias, and on twenty occasions subarachnoid haemorrhage. Four haematomas which showed a complete block were operated on within three days, but without success. Two elderly patients died weeks after the operation from pulmonary embolism. One patient with subdural and another with intra-medullary haematoma remained unchanged.

Prognosis

The prognosis depends almost exclusively on the pre-existing damage to the cord and the clinical symptoms. Out of *thirty-four patients with totally extirpated angiomas* fourteen recovered and returned to work. In thirteen cases functional improvement of varying extent facilitated rehabilitation up to the stage where they could walk. Five cases remained unchanged and two deteriorated.

We are in the beginning of a successful era in treating spinal angiomas. Now that the clinical and operative techniques have been established, with improved early diagnosis and more timely operation the number of cures will increase. I am sure that the radical operation will become the operative procedure of choice as was suggested by G. Perthes of Tübingen in 1926[11] after the first myelographically diagnosed and totally resected arteriovenous angioma. This was 18 years after Fedor Krause's first multiple ligature procedure[6] for spinal angioma.

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