Section of Neurology

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Meeting February 3 1966

Vascular Disorders of the Spinal Cord

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Angiomas of the Spinal Cord

Arteriography of angiomas of the spinal cord is a delicate technique, but it allows a study of these malformations which is impossible by other conventional methods of diagnosis.

Plain pictures and tomography of the spine may, in showing enlargement or erosion of the spinal canal, or in demonstrating a scoliosis, provide presumptive evidence of an angioma. Even more, myelography may show a characteristic picture in about three out of four cases; finally, intraosseous venography may show a dilatation of the epidural venous plexus, even if it does not fill the angioma itself.

Only arteriography can really demonstrate the angioma, and outline its position, extent, afferent pedicles and draining veins. A rigorous technique is essential if one is to avoid complications as well as obtain accurate pictures.

Points which appear essential to us are use of a general anæsthetic; choice of a technique adapted to the presumed site of the angioma (subclavian arteriography for the cervical region, aortography for the dorsal and lumbar regions); increased concentration of the iodized dye in the vessels filled by using a catheter of the shortest length and the largest calibre; adjustment of the position of the catheter by television screening; automatic injection under controlled pressure; and the use of serial pictures at the rate of three or four per second.

As with cerebral angiomas, spinal vascular malformations seem to be nothing more than arteriovenous aneurysms, of which the essential feature is an arteriovenous communication without any intermediate capillary. One or more of the arteries forms a cluster of vessels which discharge into one or more draining veins, producing a malformation of variable size, from a simple arteriovenous fistula to a cirsoid malformation.

The malformation: The normal arrangement of artery and vein disappears at the level of the malformation, which is itself connected to a normal vascular system. Arteriograms, studied at different phases, show successive delineation of nourishing arterial pedicles of the mass of abnormal vessels, constituting the arteriovenous shunt, and the draining pedicles.

(1) The feeding arteries of the malformation come from one or several pedicles and in this case may be unilateral or bilateral. These arteries are often of large size, occasionally being really enormous, abnormal in their appearance or their pattern; they are, however, normal in their origin, course and penetration of the dural canal.

(2) These arteries do not end in a network of capillaries, but lead to the malformation which comprises a cluster of dilated vessels and sinuses. This is really nothing more than the vestigial vascular network. This malformation is difficult to show on arteriograms. The draining venous pedicles quickly become superimposed on the malformation itself and it is a delicate problem, in this diffuse opacity, to distinguish between afferent pedicle, arteriovenous malformation and draining pedicle.

(3) The draining veins are more numerous and important with a more extensive malformation, fed from very numerous pedicles, but they are almost always impossible to classify. We think, nevertheless, that it is not so much a question of abnormal topography as of regional veins which are abnormally developed. The veins may travel a long way from the arteriovenous shunt, upwards and downwards, and this explains how certain malformations seem to spread over a great length of the cord; they can assume a size much larger than the malformation itself, and this has great practical importance, because the venous dilatations produce a compression syndrome at a distance from the so-called malformation and intrathecal lipiodol may show a complete block at this level, suggestive of a tumour.

The importance of the extent of the malformation is probably better appreciated by arteriography than by anatomical examination. Arteriography allows of the differentiation of the various vessels of the malformation itself, of which the extent varies from a simple fistula to a huge cirsoid mass.

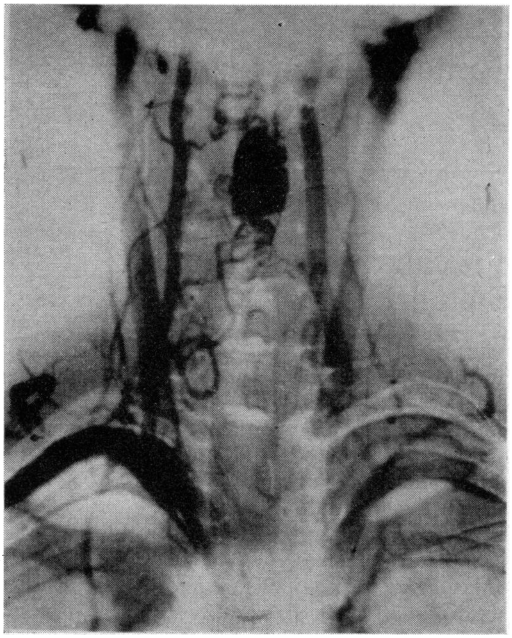

Fig 1 J P, aged 18 years. Spinal angiogram (by means of femoral aortography; catheter situated at level of aortic arch). The angioma lies over the level of C4/5; it is of circular form, consisting of fine vessels fed by several pedicles. A superior one is seen arising from the right vertebral artery at the level of C3/4. Two inferior pedicles are seen arising from the ascending cervical artery on the right side, penetrating the spinal canal at the level of C7/D1 and C6/7. On the left side, there is another inferior pedicle arising from the ascending cervical artery and penetrating the spinal canal at C5/6

The small fistulæ are difficult to see arteriographically, in spite of the photographic procedures of subtraction and the logetron, but it is reasonable to suppose that when the techniques of arteriography of the spinal cord are improved, it will be possible to demonstrate, as in the brain, really small fistulæ, at present unrecognized.

Regional localization allows the topographical distinction of three varieties of arteriovenous aneurysms (angiomas).

(1) Cervical angiomas are of medium size, extending over one, two or three spinal segments. Their afferents, which are multiple and bilateral, obtain all their branches from the subclavian artery: vertebral artery cervico-intercostal trunk, thyro-cervico-scapular trunk (Figs 1 and 2). One of our cases, however, had a supplementary afferent arising from an intercostal branch of the aorta. It was an angioma at the cervicodorsal junction.

(2) The dorsal angiomas are the largest and cover three or four segments or more; however, their afferents are less numerous (two or three) and, as a rule, unilateral. They come from the aortic intercostal vessels (Figs 3 and 4).

(3) The dorsolumbar angiomas are quite unusual in their small size and in their arterial afferent supply, which is or appears to be single. The small size of the angiomas of this region make them the most difficult to demonstrate and necessitate rigorous photographic procedures. The nourishing pedicle appears single; its origin, its course and arteriographic appearance suggest that it is often no more than the artery supplying the lumbar enlargement or the artery of Adamkiewicz (Fig 5); and this is a problem when considering the possibility of ligating the pedicle. Certain angiomas are, on the other hand, nourished essentially by the posterolateral artery of the spinal cord, and this differentiation is essential because it permits ligature of the nourishing pedicle.

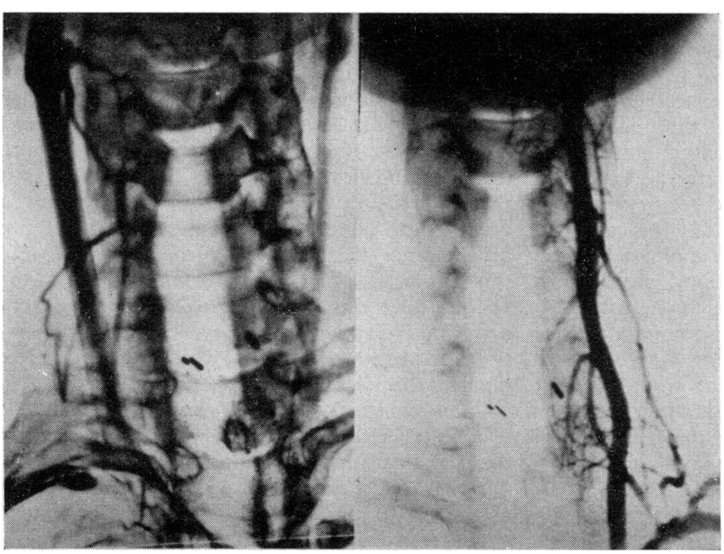

Fig 2 J P, post-operative arteriogram (subclavian puncture). Left, a small portion of the angioma is filled from the ascending cervical artery. Right, no filling of the angioma

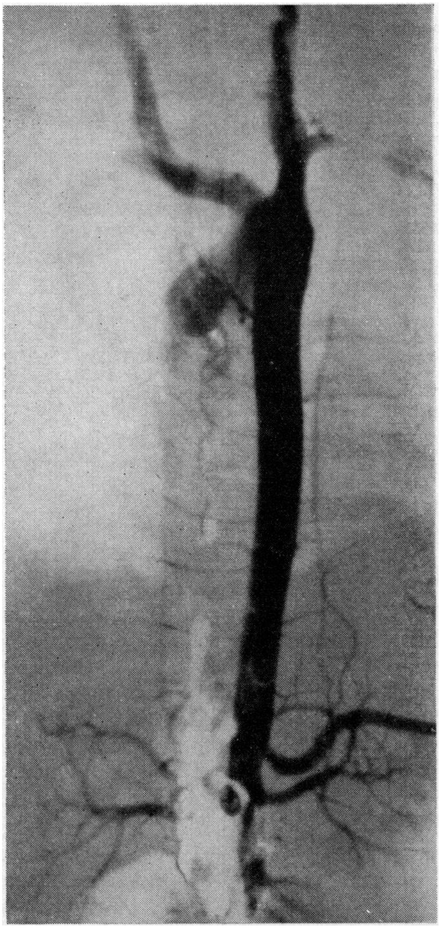

Fig 3 *F B, aged 4 years. Aortography (subtraction): two pedicles are visible. The upper one comes from the fourth intercostal artery on the right, and curves upwards before it joins the upper pole of the angioma. The inferior pedicle arises from the ninth intercostal artery and joins the lower pole of the angioma*

The topographical classification of our series, which is based on 23 cases, will only be of proven value in the light of further experience. It will be necessary to carry out a great number of arteriographic observations to know if these malformations are formed at random, by an arrest in capillary maturation, or obey the more or less strict rules of development, depending on the region of the cord, as we are inclined to think.

Treatment

Except for some cases of unusually pure extramedullary pattern, it seems impossible, in the majority of cases of angiomas, to contemplate excision. The extramedullary angiomatous mass is often more important and larger than the intramedullary portion, but the penetration of abnormal vessels within the cord precludes removal by a dissection along a plane of cleavage at the limits of the pathological vessel, as can be achieved in the cerebral hemispheres, without considerable risk to the patient.

For this reason the only possible therapy seems to us to be ligature of the afferent arterial pedicles; this can be achieved either at a distance from the cord at the level of the afferent vessels, or within the spinal canal where the vessels penetrate the dura mater. However deficient they are, these ligatures at a distance are not ineffective, and in some cases they have been followed by improvement in the paraplegic condition. It is quite probable that these ligations lessen the pressure within the malformation and the draining veins, perhaps allowing some improvement in vascularization of the cord tissue.

We think that ligation of the arterial pedicle of spinal angiomas should be considered in relation to the site of the malformation; for the cervical angiomas which are irrigated by numerous and bilateral pedicles the ligature must be at the penetration of the dura and probably performed at successive operations; for the dorsal angiomas

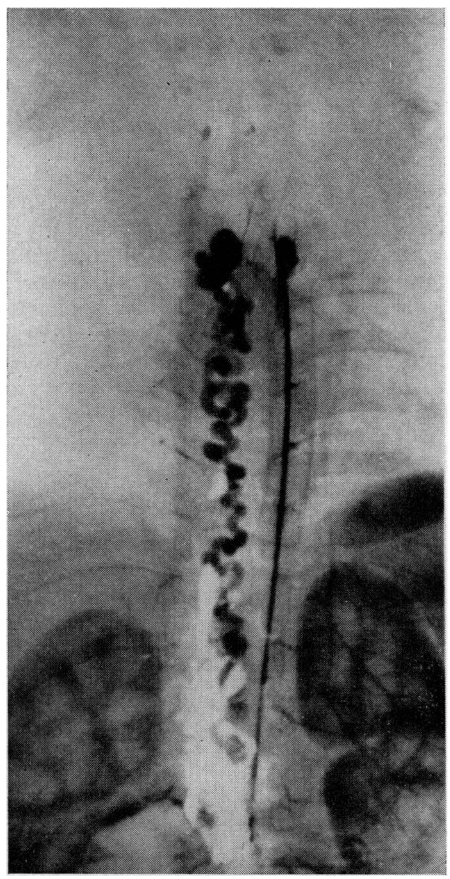

Fig 4 *F B. Aortography (subtraction): shows the large, tortuous inferior efferent vein*

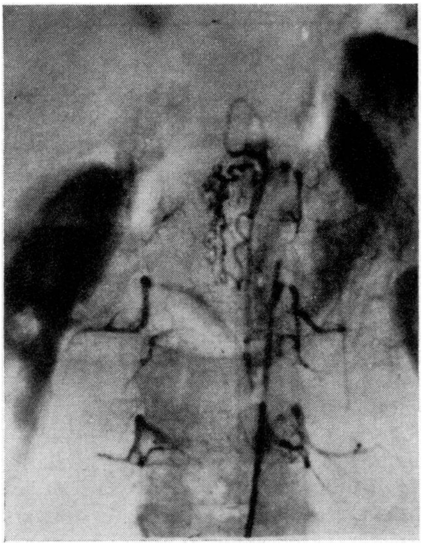

Fig 5 M D, aged 32. Aortography via the femoral artery (subtraction): increase in size of the third lumbar artery on the left. The spinal branch of this vessel enters the spine between L3 and L4, ascends to the lower margin of T12 and then descends again to a point opposite the disc space L1/2, where it divides into two branches. One goes to the upper pole and the other to the lower pole of a fine angiomatous network lying in front of the first and second lumbar vertebral bodies. This arterial pedicle is the Adamkiewicz artery

which are large malformations spread over several segments and which are fed by a few pedicles, usually from one side only, the better procedure seems to be ligation at a distance, outside the spine; for the dorsolumbar angiomas, if the afferent supply is provided by the artery of Adamkiewicz, the condition cannot be treated surgically. If the pedicle is provided by a posterolateral network surgery is practicable.

Conclusion

An arteriographic study of vascular malformations of the spinal cord suggests that these malformations are arteriovenous aneurysms. Arteriography allows a clear picture of the afferent arterial supply of the malformation, of the arteriovenous shunt and of the draining veins, and indicates the morphological similarity of angiomas at identical segmental levels of the cord, allowing, therefore, an attempt at topographical classification of these malformations. These anatomical facts offer the new possibility of planned surgical treatment which attempts to obliterate the circulation of the angioma by ligature of its afferent arterial supply.

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Clinical Aspects of Spinovascular Disease

The study of spinovascular disease began with an experiment, remarkable in the history of medicine for having come before clinical observation. Three hundred years ago the Danish scientist Niels Stensen, or Steno, in his treatise on the action of muscles (1667) mentioned the reversible paralysis of a dogfish's tail which he had induced whenever he tightened a ligature around its descending aorta:

'... cum viderim ligata aorta descendente sine praevia sectione, partium posteriorum omnium motum voluntarium toties cessare, quoties vinculum stringebam, iterumq; tot vicibus redire, quot vicibus nodum laxabam; ... sublato vinculo supervixit canis sine ullo motus incommodo.'

According to Haller (1762) this experiment, priority for which he wrongly accorded to Swammerdam (1667), was repeated many times by later investigators. Steno may have thought of the dogfish, *piscis ex canum genere*, as an experimental animal after he had examined the head of the Mediterranean monster, *Canis carcharia* (Fig 1), which his patron the Grand Duke of Tuscany, Ferdinand II, had obtained for him. Noticing that the shark's teeth resembled local stones he began a study which led to the foundation of petrology.

The paraplegia of aortic occlusion may be spinal or peripheral; but two centuries passed after Steno's experiment before physiologists learned that the type of paralysis depended on the site of the ligature, on the degree and duration of compression and, above all, on the pattern of the individual animal's circulation (Schiffer 1869, Hoche 1899). The lesson had to be learned again with the advent of aortic surgery (Crafoord & Nylin 1945, Gross 1945) and the hazard of