Vascular Malformations of the Spinal Cord
The Anatomic and Therapeutic Significance of Arteriography

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Ever since the studies of Virchow, a distinction has been made between tumors and vascular malformations in the central nervous system. Although described since the end of the 19th century, the study of vascular malformations of the spinal cord entered a new era with the advent of angiography. In fact, angiography not only shows spinal angiomas much better than was previously possible by surgical or anatomical examinations, but also encourages the hope of a logical treatment.

In 1956, Henson and Croft\(^1\) first reported a cervical arterio-venous aneurysm visualized with vertebral arteriography. In 1954, R. W. and C. W. Rand\(^2\) had succeeded in visualizing a malformation of the dorsal-lumbar junction by means of aortography. Subsequently, Höök and Lidwall\(^3\) and Morris\(^4\) blished 2 cases of cervical malformations injected by means of vertebral arteriography.

Since 1962, when Djindjian et al.\(^5\) reported their first case of opacification by aortography, we have been able to collect 15 cases of vascular malformations of the spinal cord studied by means of angiography. This number may seem surprising. We think that the frequency of spinal cord malformations will become more apparent with the increased use of the spinal angiography, just as has been the case with cerebral angiography. The diagnosis may also be made earlier, that is to say, with the first signs of the abnormality.

It is interesting to note that in the 15 cases of our series, the first symptoms appeared in 11 cases prior to the age of 12 (twice before 3 years, 4 times between 3 and 7 years, and 5 times between 7 and 12 years). However, in 4 cases the first symptoms appeared much later; we recall that in the case of cerebral arterio-venous aneurysms, although 70 per cent of the cases become symptomatic before the age of 30 years, 5 per cent appear after the age of 50.

Classical diagnostic procedures are undeniable helpful in identifying a vascular malformation of the spinal cord. However, the identification on plain x-rays or tomographs of the spine, of an enlargement of the central canal, or an erosion of its walls is only presumptive evidence. Positive contrast myelography (Lipiodol, Pantopaque) suggests the diagnosis of a vascular malformation in about 75 per cent of cases by characteristic images described in 1925 by Guillain and Alajouanine\(^6\), but, as Gross and Ralston\(^7\) pointed out, small malformations can probably escape this relatively elementary method of diagnostic study.

Like other authors such as Svien and Baker, and Tavera, we had expected big things from trans-osseous phlebography. But it is not reliable in demonstrating a dilatation of the venous plexus surrounding the spinal cord. It shows merely the increased pressure existing distal to the arterio-venous shunt which is characteristic of vascular malformations of the spinal cord as they appear with arteriography.

**Technique of Arteriography**

Arteriography is in fact the fundamental examination necessary to determine the existence of a vascular malformation, and precisely to define its position, its dimensions, and its afferent and efferent vessels. A rigorous technique is absolutely essential in order to avoid accidents, and to obtain clear x-rays. We therefore wish to emphasize certain technical points which seem to us important.

1. The use of general anesthesia.
2. The selection of the technique, vertebral or aortic, appropriate for the presumed site of the malformation.
3. The choice of a catheter of minimum length and maximum diameter in order to increase the concentration of the contrast medium.
4. Monitoring the position of the catheter on a television screen in order to avoid the introduction of the catheter into a major spinal artery.

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5. The use of a tri-ioded contrast medium the viscosity of which decreases when it is heated to a temperature of about 37°C.

6. Selection of the right quantity of medium injected, depending on the location of the injection.

7. The use of automatic injection under controlled pressure.

8. The use of seriographic technique in making the exposures.

9. The rigorous maintenance of apnoea while making the arteriographic exposures as well as while taking 2 plain x-rays prior to the injection, permitting one to obtain at different times in the seriography, the subtraction x-rays so important for the interpretation of these often complex studies.

Anatomical Study

Like malformations of the brain, spinal vascular malformations appear to be nothing more than arterio-venous aneurysms, their main characteristic being the existence of an arterio-venous communication without an intermediate capillary network. From one or several arteries, comes a group of vessels, more or less important, which open into the venous system by one or several veins, thus creating a malformation of variable size and volume, from the simple arterio-venous fistula, to the large cirsoid malformation.

1. The Malformation. This appears where the identification of artery and vein ends but it is supplied by a normal vascular system. Angiography studied in its time sequences show successively the opacification of one or more arterial pedicles supplying the malformation, the mass of abnormal vessels composing the arterio-venous shunt, and the venous pedicles draining the malformation.

A. The arteries supplying the malformation come from one or more pedicles and may be unilateral or bilateral. These arteries are often of important size and at times are really monstrous. On the other hand, although abnormal in appearance and size, they are of normal topography; that is, their origin, course and point of penetration of the vertebral canal is normal.

B. These arteries do not divide into a capillary network but terminate their course in the malformation as a mass of distended and sinuous vessels. This mass is probably nothing other than a vestigial vascular network. The malformation itself is difficult to isolate on the angiogram. The venous pedicles of drainage, in fact, are very quickly confounded with the malformation itself, and it is difficult if not impossible in this diffuse opacity, to differentiate which is related to the afferent pedicle, to the arterio-venous malformation, or to the pedicle of drainage.

C. Drainage veins are more numerous and larger when the malformation is larger and supplied by more pedicles. We believe, however, that here again it is not a question of topographically abnormal vessels, but of abnormally developed local veins.

One fact concerning these veins is very striking and should be emphasized. These veins may travel far from the arterio-venous shunt, upwards as well as downwards; this explains how some malformations seem to extend along the whole length of the spinal cord, and may form a mass much larger than the true malformation. This fact is of great practical value. These venous dilatations may produce a pressure syndrome at some distance from the malformation itself and myelograms may show, at this level, a picture of complete obstruction simulating that of a tumor.

Thus the malformation is really an arterio-venous shunt. In order to understand its importance, it would be necessary to study the blood flow in the malformation, just as has already been done in the brain, with the aid of gas measurements or by cine-angiography. Such a study would be difficult to accomplish since rates for normal blood flow in the spinal cord have not yet been established. Moreover arteriography of the spinal cord is not yet a simple, routine examination. However, we may say regarding cervical aneurysms injected by vertebral angiography, that their opacification appears more rapidly than the opacification of the vertebral system usually studied.

2. The Size of the Malformation. This is probably better defined by angiography than by anatomical examination. We have seen that drainage veins may extend some distance from the malformation and that they may at this point be mistaken for the main anomaly. The angiograms differentiates between the venous pedicles and the malformation itself, the size of which varies from a
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simple arterio-venous fistula to the enormous cirsoioid angioma. The less voluminous fistulae are difficult to distinguish by angiography, in spite of the special photographic process of subtraction, or the use of logeotron; it is probable that when spinal angiography improves in accuracy, it will be possible to see, as in the brain, very small fistulae now unrecognized.

The size of the malformation is only partially proportionate to the number of afferent pedicles, and it is true that the larger malformations are not necessarily the ones which have more afferent arteries. The size of the malformation, as well as the number of the afferent arteries, seems to be conditioned by the embryological evolution which modifies the initial metameric vascularization of the spinal cord; thus the vessels forming the malformation bear a relationship to the normal vascular system, which itself is not uniform throughout the full length of the spinal cord. One may distinguish the 3 areas, cervical, dorsal, and dorso-lumbar.

3. Localization by Level in the Cord. We are able to separate three topographic varieties of arterio-venous vascular malformations. Indeed, although our series of 15 cases is too small to be conclusive, we have been struck by the remarkable similarity in size of different malformations at the same general level.

A. Cervical malformations (3 cases). In our series these aneurysms are all of medium size, extending over 1, 2, or 3 spinal segments (Figs. 1–5). Their afferent arteries, which are numerous and bilateral, come from branches of the subclavian artery, namely, the vertebral artery, costo-cervical trunk, and the thyro-cervical trunk. Thus embryologically they originate from the first 7 somatic intersegmental arteries. One of our cases, however, showed a supplementary afferent artery arising from an aortic intercostal artery; it fed a malformation in the cervico-dorsal region (Figs. 6, 7 and 8).

B. Dorsal malformations (8 cases). Lesions in this area are the largest in our series, extending over 3, 4 or more segments (Figs. 9–17). However their afferent arteries are less numerous (2 or 3), are usually unilateral, and come from the intercostal branches of the aorta.

C. Dorso-lumbar malformations (4 cases). These malformations are remarkable for their small size, and their arterial source which is single, or more precisely, seems to be single (Fig. 18). The small size of malformations in this region explains the great difficulty in demonstrating them and the need for the most careful angiographic techniques. The origin, course and angiographic appearance of the single arterial source led us to believe that this pedicle is simply the artery of the lumbar enlargement or Adamkiewicz’ artery. This presents a very important problem when considering the possibility of ligating this feeding artery.

The cases we have considered are too few to give us much information. We need to look at many more angiograms in order to know whether these malformations are located at random or follow a more or less strict plan that makes them develop in one rather than another given area of the spinal cord. We are inclined to think the latter.

4. Width of lesions. The width of the malformation in relation to the spinal cord is difficult to ascertain precisely by angiography. However, according to the operative data and anatomical observations, these arterio-venous aneurysms always seem to be located on the posterior surface of the spinal cord. In only one of our cases did we find an artery on the anterior surface of the cord supplying a cervical aneurysm. It is impossible with present techniques to judge angiographically the penetration of abnormal vessels into the cord itself.

Pathogenic Origin

The analogy of arterio-venous aneurysms of the brain leads us to believe that the spinal malformations are also evolving congenital lesions with a fixed position based on normal anatomy. Thus they concern a delimited territory and are fed by a certain number of arteries. The territory involved, as well as the number of the vessels which form the malformation, does not increase with time. On the other hand the vessels, and particularly the drainage veins, do undergo dynamic modifications. Because of the lack of capillaries, the vessels are unable to adapt to general variations of circulation, and therefore the circulation inside the malformation is constantly being modified, setting the background for hemorrhage and thrombosis. Because of the histological anomalies of the vascular walls, the vessels undergo modifications and progressive distension that event-
Cervical Malformation. Case J.T., 22 years.

**Fig. 1.** Left subclavian arteriogram (subclavian puncture; subtraction x-ray). Malformation of oval form made up of fine entangled vessels, with 2 inferior pedicles issuing out of a large vessel that originates on the posterior surface of the vertebral artery and penetrates the spinal canal between C7 and C6. A lateral pedicle from the ascending cervical artery penetrates the spinal canal between C6 and C3.

**Fig. 2.** Left vertebral arteriogram (subtraction x-ray). Rectilinear anterior spinal artery, and a sinuous left posterior spinal artery, originating from the terminal vertebral artery and ending at the superior pole of the malformation.

**Fig. 3.** Profile arteriograph of the left subclavian artery (subtraction x-ray). Injected malformation extending from C4 to C6 vertically, and through the total diameter of the spinal cord in depth. The superior pedicles are supplied by the anterior spinal artery, the lateral pedicles by small branches issuing from the vertebral artery, and the inferior pedicle by a large curved branch of the vertebral artery. The thyroid gland is situated anteriorly to the vertebral artery.
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Cervical Malformation. Case J.P., 18 years.

Fig. 4. Spinal angiogram (by means of femoral aortography, catheter situated at the level of the aortic arch). The injected malformation appears along the C4–C5 vertebrae. The malformation is of circular form and made up of fine bundles of vessels. It is vascularized by a superior pedicle issuing from the right vertebral artery at the level of C3–C4 and of two inferior pedicles issuing from the ascending right cervical artery, penetrating the spinal cord at the level of C7–D1 and C6–C7. On the left side, there is an additional inferior pedicle issuing out of the ascending cervical artery and penetrating the spinal canal in the interval between C5–C6.

Fig. 5. Postoperative arteriogram (subclavian puncture). On the right, there is no filling of the malformation. On the left, a small portion of the malformation remains vascularized by the ascending cervical artery.

**Fig. 6.** Lipiodol myelogram (sub-occipital puncture). Image of blood vessels in the region of the cervical spinal cord; obstruction of the lipiodol at the level of D1.

**Fig. 7.** Spinal angiogram (subtraction). Femoral aortography, catheter in the left vertebral artery. Opacification of a large radiculo-spinal cervical artery issuing from the posterior surface of the vertebral artery at the level of C5, and descending along the cervical spine to the superior pole of the malformation situated at the level of D1-D2.

**Fig. 8.** Femoral aortogram (subtraction). Catheter tip at the level of D5. Opacification of a large branch of the 4th inter-costal artery, forming an arch, superimposed on the path of the aortic arch, and giving origin to two ascending arteries: the one on the left goes to the left inferior pole of the malformation; the other goes to a dilated vessel at the right inferior pole of the malformation. In addition, there are two lateral arterial sources coming from the right and left subclavian arteries.

Fig. 9. Femoral aortogram (subtraction). Catheter at the level of D6. Large feeding artery at the level of D6. Opacification of the malformation.

There are two arterial sources, the large inferior one arises from the right 6th intercostal artery and bends in order to penetrate the spinal canal. It passes through a loop and ascends to D3 and then descends to the superior pole of the malformation.

The superior pedicle is formed from the right 4th intercostal artery ascending up to D1, then descending to the superior pole of the malformation.

Note the dilated intercostal arteries (D7–D8) without clear evidence that they participate in the vascularization of the malformation.

Fig. 10. Aortogram (further filling and subtraction). Progressive opacification of the angiomatous mass.

Fig. 11. Aortogram (venous filling and subtraction). Appearance of large drainage veins, in a median position along the dorsal spine, extending upwards to the cervical level and downwards to the lumbar level looking like a very contorted spinal vein.

Filling of the large dorsal extraspinal plexuses which are very voluminous, and of the arch of azygous vein.
Dorsal Malformation. Case A.R., 3½ years.

Fig. 12. Spinal angiogram (aortogram through the femoral artery). Catheter at the level of D5. Two large afferent pedicles arising from the posterior surface of the aorta (4th and 6th left intercostal arteries). The antero-venous shunt is projected behind D4–D5–D6; it is a simple structure made up of a single large tortuous vessel.

Fig. 13. Spinal angiogram (subtraction). Late. The drainage veins are remarkable because of their size and because they extend upwards into the cervical region, and downwards to the conus medullaris.

Fig. 14. Profile spinal angiogram (subtraction). Early. One can see the two pedicles issuing from the posterior surface of the aorta.

Fig. 15. Profile spinal angiogram (subtraction). Late. Large spiralled vessel extending from the cervical level to the conus medullaris.
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Dorsal Malformation.
Case M.T., 25 years.

Fig. 16. Aortogram (subtraction). Opacification of enormous clusters of venous dilatations filling the spinal canal. Two radiculo-spinal pedicles; the large artery arises from the dilated 9th dorso-spinal branch; it forms a double bend, with the concavity on the left. The small superior arterial source comes from the 8th dorso-spinal branch of D8, ascends, then forms a loop with an inferior concavity which directs it to the superior pole of the malformation.

Fig. 17. Aortogram (subtraction). Venous filling. The inferior pole of the malformation has two large venous pedicles, one median and sinuous, the other lateralized, ending in the latero-vertebral venous plexus.

usually creates real masses that compress the spinal cord.

In addition, ischemic sclerotic changes may develop close to the malformation. This is the explanation for the clinical manifestations of arterio-venous malformations of the spinal cord.

Treatment

The development of a spinal cord vascular malformation can be very serious, principally in children or young adults, where it results, as a rule, in neurological deficiencies which are usually non-regressive.

The various accepted methods of surgical treatment, with a few exceptions, do not provide a really satisfying solution, since there seems to be no direct means of acting upon the arterio-venous shunts that are directly or indirectly responsible for the spinal cord lesions.

The total ablation of a vascular malformation of the spinal cord is not often possible. It can only be reasonably undertaken if the position of the abnormal vessels is purely extra-spinal and posterior. Often, due to the location of the extra-spinal malformation, there may be a pathological vascular penetration to the interior of the spinal cord itself. Because of the great functional density of the spinal cord, one cannot undertake dissection of the malformation without the risk of severe parenchymatous damage.

Ligation of the arterial pedicles of the malformation seems to us to be the logical solution. The precise anatomical picture provided by angiography eliminates much of the dangerous and blind character of partial exclusions or interruptions as they were formerly carried out.

However, from a theoretical point of view, the methods of arterial ligation, in order to be efficient, presuppose the total exclusion of the malformation. Experience gained through application of this technique to vascular abnormalities in the limbs or brain show that the persistence of a single supplying artery with its anastomotic circuits is enough to maintain the malformation and its injurious effects. This problem seems difficult to overcome in the spinal cord. Whatever the quality of the angiographic examina-
Practically speaking, 2 methods are available. The first is ligation of the afferent arteries at the point of their emergence from the big arterial trunks. The location of these arteries is not always easy, except at the dorsal level, where the intercostal arteries from which they originate present an obvious dilatation. This technique leaves great latitude for anastomotic circuits, and thus may reduce the risk of spinal ischemia. Unfortunately it also favors the revascularization of the malformation.

The second method is ligation of the arterial pedicles close to the malformation. This requires a laminectomy centered on the vessels concerned. Its principal advantage is the opportunity for selectivity which may reduce the possibility of anastomoses. Its principal difficulty rests in the identification of the arteries to be ligated. These are best found in the extradural space, as they enter the common dural sac. This penetration usually takes place near the nerve root; they also may enter through a separate orifice, different from the one used by the nerve root and situated on the anterior or antero-lateral surface of the dural sac. In the latter case it may be impossible to expose the desired artery.

When the arteries cannot be identified in the extradural space, separation of the afferent arteries on the interior of the dural sac remains the only solution. This is indeed delicate. The vascular confusion is in effect so pronounced that the isolation of the nourishing arteries is made extremely difficult. This is especially true in the dorsal region due to the narrowness of the vertebral canal, as well as the frequently monstrous size of the veins leaving the malformation in this region. In the cervical region on the other hand, where the vertebral canal is large, and the malformations of a moderate size, the intradural approach to the afferent arteries is more practicable.

We think that ligation of the arterial pedicle of vascular malformations of the spinal cord is the best treatment available and should be considered in relation to the site of the malformation:

1. For the cervical malformations which, you may recall, are of medium size supplied by numerous bilateral pedicles, we recom-
dread performing the ligation at the point of penetration into the dura mater and probably in 2 successive operations.

2. For the dorsal malformations, which are of a large size spread over several segments and supplied by a small number of arteries which are usually on one side, the best operation seems to be ligation at a distance from all these arteries outside the spinal column.

3. For the dorso-lumbar malformations which have a single arterial supply, probably the main lumbar artery, we do not feel that ligation is justifiable.

Clinical Cases

Eight patients in our series have been operated upon according to these principles. In 8 additional cases (2 dorsal malformations and one of the conus medullaris), explored by means of laminectomy we were unable to find any of the pedicles we had seen in the angiogram. In 3 of the successful operations (dorsal malformations) the ligation was done away from the malformation at the point where the intercostals leave the aorta. In one of these cases the 2 afferent arteries, which appeared to be isolated from the malformation, were ligated without any modification of the preoperative neurological condition (complete paraplegia of 3 month’s duration). In the 2 other cases of this type only one of the 2 principal afferent arteries was ligated. The postoperative angiographic control films showed a persistent supply to the malformation, but with a notable reduction of size as compared to the preoperative films, as well as significant slowing of the flow inside the malformation. The clinical state of these 2 patients who had suffered from paraplegia for several years was indeed much improved after the operation. There was a regression of the sensory deficit, diminution of the spasticity, and improvement of mobility and walking.

Finally, in 3 cases the afferent vessels were ligated either at the point where they penetrated the dural sac, or inside the dura itself. In one of these cases, a malformation of the cervico-dorsal junction, a superior afferent artery was ligated, and the 2 inferior arteries were left intact for later ligation if necessary. (The severe paraplegia remained unchanged.) In a second case of dorsal malformation, the superior afferent artery at D4 was ligated while the inferior afferent artery was left for ligation at a second stage. (The paraparesis was unchanged.) The last case in this group was a cervical malformation (Figs. 4 and 5). Three out of 4 afferent arteries were ligated at 2 successive operations. The remaining artery will be ligated at a third operation.

We have thought it wise not to ligate all the pedicles at a single operation. Experience will tell us whether this plan, which imposes repeated operations on the patient, should be followed.

It is hard to evaluate the therapeutic benefit of these operations because we have treated only a few cases and the period of follow-up has been short; moreover some of our patients are still under treatment. We are, everything considered, favorably impressed with our experience. Never, and this is important, never have we aggravated the neurological state of our patients.

Two incomplete ligations resulted in improvement which has been maintained for nearly a year.

We cannot actually demonstrate arteriographically any case that was totally cured by ligation. Fig. 5 shows, however, the anatomical effectiveness of the ligations. This patient had a cervical malformation and showed the clinical evidence of a Brown-Sequard syndrome with moderate hemiparesis; his neurological deficit is actually unchanged but has not gotten worse, whereas during the year preceding the operation, the symptoms had been steadily progressive. However, the patient’s cervico-brachial pain has completely disappeared.

Summary

Angiography (vertebral arteriography through the subclavian route or aortography) is a valuable method of studying vascular malformations of the spinal cord.

In our series of 15 cases we have never had regrettable complications attributable to the use of this technique. The diagnostic precision is infinitely better than that of the classical methods. In addition, arteriography provides an anatomical procedure rich in educational value.

All malformations that we have explored have been arterio-venous aneurysms. We think that in the spinal cord, as well as in the brain, the classical “arterial” or “venous”
angiommas are exceptional if they ever exist at all.

Our experience, although certainly modest, has been consistent and suggests that malformations with similar locations have similar morphology. This observation justifies an attempt at their classification in terms of their location; the confirmation of this classification still needs to be established through a larger series of cases.

Knowing the evolutionary gravity of vascular malformations in the spinal cord and the unsatisfactory character of the surgical solutions usually proposed, we have tried to use the anatomical data furnished by arteriography for a therapeutic purpose. In our hands ligation of the arterial pedicle has been a perfectly innocuous procedure. Moreover, we have reason to think that it is therapeutically efficacious. A larger number of cases and a longer follow-up period are necessary to confirm or disprove this opinion.

References