Treatment of Spinal Cord Vascular Malformations by Surgical Excision

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Recent developments have now made direct surgical attack the treatment of choice for spinal cord vascular malformations. We are reporting 17 cases treated with surgical excision, the last 11 of which were operated on under the operating microscope.

There is much confusion in the literature concerning the histological nomenclature used to describe varieties of spinal vascular malformations. This confusion is partly the result of the lack of opportunity for adequate microscopic study of the entire lesion. We prefer to follow the classification of Bergstrand, et al., who divided these malformations into: 1) angioma cavernosum, 2) angioma racemosum, and 3) angioreticularoma. Some vascular malformations will show characteristics of more than one group, making classification difficult. From the surgical standpoint the extent of the lesion and the amount, if any, of intramedullary involvement are of much greater importance than the specific type of malformation present. The 17 cases we are reporting are primarily of the angioma racemose type, some largely venous, others predominantly arterial.

It has been shown that spinal cord vascular malformations are much more likely to occur in certain areas of the cord. In only one of our cases was the lesion confined to the cervical area. The remainder were located in the thoracic or lumbosacral region, with the latter site the most common location. Often the malformation was so extensive as to involve the entire thoracic and lumbar cord with even occasional extension into the cauda equina. Only two of our cases were females, although an exacerbation of symptoms in women during menstruation has been reported, and several authors have called attention to an increase in symptoms during pregnancy with subsidence after delivery. Newman has stated that he believes the increase in symptoms in such cases may be due to “venous congestion” from the distended uterus and interestingly suggests the possibility of some “hormonal factor acting on the vessel walls.” Although none of our cases was a child, several authors have reported the occurrence in children and even in infants.

Clinical Picture

History. The clinical history is usually one of three types. There can be 1) a slow progression of neurological symptoms and signs, 2) progression followed with regression or a stationary period, or 3) a sudden apoplectic onset.

Neurological Examination. This may range from an entirely normal neurological examination up to that of a complete transverse myelitis. It is interesting that a rather high percentage of patients have some impairment in bladder control. In many cases, the complaints and neurological findings are such as to suggest the diagnosis of herniated disc, poliomyelitis, spinal cord tumor, multiple sclerosis, or even hysteria. The correct diagnosis is much more apt to be made in those cases having a sudden onset, bloody spinal fluid, and neurological findings indicating some sort of myelopathy. Matthews reports the occasional presence of a spinal bruit as a diagnostic sign of spinal vascular malformation.

Spinal Fluid. Nearly all writers on the subject agree that in the presence of spinal vascular malformations the spinal fluid protein is almost always increased. There may be a slight increase in the spinal fluid white cell count. The Queckenstedt test is usually normal except in the occasional case having a “spinal block” from a very large malformation.

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Radiological Findings

*Plain X-Ray Films of the Spine.* These will seldom show anything to indicate the vascular nature of the lesion. Rarely there may be widening of the pedicles at the site of a large malformation. Even more rarely an associated vascular involvement of the spinal vertebrae may be seen, but this is more often present in association with an extradural vascular lesion than with an intradural lesion. Peculiarly, a high percentage of patients with spinal cord vascular malformations show considerable osteoarthritic vertebral changes.

*Myelography.* The very typical myelographic picture of the “worm-like” mass of vessels is well known (Fig. 1). Whenever a complete block is demonstrated on myelography and the cause is obscure, contrast medium should then be instilled from the opposite end of the vertebral column in order to delineate the extent of the block. Occasionally this will demonstrate the cause of the block to be a vascular malformation. Most all vascular malformations of the cord are located on the posterior rather than the ventral surface. It is for this reason that these lesions are better demonstrated myelographically with the patient lying in the supine position. In searching for such vascular lesions it is essential that at least 10 cc of contrast medium be employed. It would seem that the value of air myelography is very limited in diagnosing this particular disease.

*Spinal Angiography.* Angiographic demonstration of the abnormal vasculature of the spinal cord represents a great advance in not only the diagnostic area but also in planning the surgical attack. In recent years several authors have shown how well these vascular malformations can be demonstrated with one of several angiographic techniques. Retrograde femoral catheterization, brachial catheterization, and vertebral angiography are being utilized. The use of multiple cassette changers and subtraction technique is essential. We wonder if, with further perfection in angiographic technique and the use of stereoscopy in subtraction, it may not even become possible to reveal the presence and extent of any significant intramedullary extension of these vascular lesions? Our own experience with spinal angiography is limited, but we well recognize the value of the procedure (Fig. 2). Detailed preoperative spinal angiographic studies offer the best information as to the extent of the vascular malformation, and postoperative angiographic studies will reveal the “completeness” of removal. In contrast to myelography, angiography can give precise information as to the nature and location of the “feeder” arteries, which can be of great help in the surgical removal.

**Treatment**

In years past the treatment of these vascular spinal malformations has, for the most part, consisted of decompressive laminectomy and deep x-ray therapy. Decompression is of little or no value and then only in
the presence of a "spinal block." Some cases are actually worse after decompression. We agree with Odom, et al., who state that x-ray therapy has no place in the treatment of this lesion. Section of the dentate ligaments has been recommended by Teng and Shapiro in reporting six cases of "arterial anomaly" of the cord. Direct surgical attack on these formidable lesions has, generally, met with little enthusiasm, although occasionally very good results have been reported. The first reported removal of a spinal vascular malformation was by Perthes in 1927 who diagnosed the lesion with Lipiodol, surgically excised it, and reported the patient showed a gratifying degree of recovery. More recently a number of authors have reported limited experience and success with surgical removal. Very recently, Houdart, et al., have encouraged ligation of only the "feeder" vessels, as have Baker, et al. At surgery most of our cases, particularly those done under the operating microscope, have shown such a multitude of anastomosing branches that it is difficult for us to conceive that "feeder" ligation alone will produce satisfactory long-term results when employed routinely.

There have been 17 cases of spinal vascular malformation with surgical removal in this clinic. Total removal has been attempted in each case; failure to accomplish this was usually due to significant intramedullary extension or to excessive length of the malformation. While some of the very large malformations that produce a spinal block may have some of their symptoms relative to cord compression, we believe that all spinal vascular malformations cause most, if not all, of their symptoms by syphoning off important blood supply from the cord. This is a type of "spinal steal syndrome." Shephard has likewise suggested they act as a shunt and thereby produce symptoms. If there is suf-
sufficient impairment in the spinal cord circulation, then actual pathological changes occur within the cord; many of these are irreversible. Antoni has reported three cases of spinal vascular malformation showing varying degrees of myelomalacia at autopsy. Removal of these vascular lesions must therefore be done before irreversible changes occur.

Surgical excision is best accomplished in those cases that do not have a significant degree of intramedullary extension. At present it is impossible to determine preoperatively whether intramedullary extension exists. It is well known that intramedullary spinal tumors are sometimes associated with dilated vessels on the surface of the cord, and Krishnan and Smith report such a case with vessels "resembling a racemose angioma." Dilated vessels on the cord surface in conjunction with an intramedullary tumor should not be confused with a vascular malformation.

Technique. The following surgical technique is employed in the removal of the spinal vascular malformation. The entire length of the malformation is exposed by laminectomy. This often requires a laminectomy extending over six or more segments. If detailed spinal angiographic studies have not been done, it frequently will be necessary to extend the skin incision and laminectomy, for the malformation is often more extensive than suspected from the myelographic studies. The dura should be opened very carefully in order to avoid tearing one of the huge, fragile surface vessels. Any major arterial feeding vessels are then sought and will usually be found along the lateral edges of the cord and malformation. They can often be identified due to their rather large size, arterial pulsation, and by a more reddish color because of their high content of arterial blood. Not only are these vessels more easily identified under the operating microscope but the whole removal of the malformation is greatly facilitated with magnification (Fig. 3). These arterial feeders must first be clipped. We have found many of these feeding arteries to be vestigial in character and to have a very thin wall so that any clip applied to them must close very tightly or the vessel will continue to bleed. These clipped feeders are then each coagulated and severed between the clip and malformation. Preoperative spinal angiography will often greatly aid in the identification of these feeding vessels.

The actual excision of the mass of coiled vessels is then begun, usually starting at one end of the lesion. The malformation will be lying beneath the arachnoid, which often is thickened. The arachnoid is first cut along the edges of the vessels so that these can be mobilized. Under magnification many numerous small anastomosing vessels will be seen running from the malformation to the underlying normal nutrient vessels of the cord. All of these anastomosing connections must be coagulated with the bipolar electrode and then cut. This very careful dissection of the malformation is continued until the entire mass can then be lifted from the cord (Fig. 4). Very frequently the entire malformation will be found on removal to consist of primarily one large vestigial vessel with numerous coils (Fig. 5). If intramed-

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ullary extension does occur the malformation can be severed from that portion, or if the extension is not large it can sometimes be removed with very careful dissection under magnification.

Following removal the dura is closed tightly. Occasionally these malformations may be so long that removal of the entire lesion in one operative session is not deemed prudent. Three of our cases each underwent two separate operative procedures.

In evaluating the results of treatment it is necessary to bear in mind the natural course of the disease if untreated. Once a patient has begun to have symptoms from a spinal cord vascular malformation, either slow progression or repeated episodes with increasing deficit can be expected. In those patients with a subarachnoid hemorrhage from the lesion who were fortunate enough not to have suffered a severe transverse myelitis, the threat of such a catastrophe at some future time was very real.

With this in mind, we have attempted to classify our operative results into one of several broad, somewhat arbitrary, groups (Table 1). Such a classification is only approximate for the extremes of any one group at some point merge imperceptibly into those of the succeeding group. We have divided our results into "excellent," "good," "fair,"
<table>
<thead>
<tr>
<th>Case No. (yrs)</th>
<th>Operation</th>
<th>Preoperative Findings</th>
<th>Postoperative Findings</th>
<th>Results</th>
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<tbody>
<tr>
<td>1 (38)</td>
<td>T11-L1 Localized knotty mass—total excision, Had 4 mm feeding artery</td>
<td>Flaccid weakness both legs, rt. &gt; left. Unable to support weight. Hypalgesia, both legs. Peri-anal analgesia. Sphincter difficulties.</td>
<td>No weakness in legs. No sensory loss in legs. Still has urinary retention. Bowel control is good. No apparent difficulty in erection or ejaculation.</td>
<td>Good</td>
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<td>3 (64)</td>
<td>T7-L1 Total removal</td>
<td>Bilateral spasticity legs with ataxia in walking. No discernible weakness. All sensory modalities decreased below D-12. Urinary incontinence.</td>
<td>Now walks well. Spasticity and ataxia not apparent. Vibration decreased below knees. No urinary incontinence.</td>
<td>Excellent</td>
</tr>
<tr>
<td>4 (59)</td>
<td>T8-L1 Total removal</td>
<td>Weakness both legs—marked on rt. Paresthesia in legs but objective sensory examination normal. Has pain in legs.</td>
<td>No weakness. Walks without difficulty. Paresthesia only in rt. buttock area. No pain.</td>
<td>Excellent</td>
</tr>
<tr>
<td>5 (25)</td>
<td>1st Operation T2-T7 only decompression done.</td>
<td>Spastic paraparesis legs. Walks only with help and cane. Marked decrease in sensation below D6. Impairment in urinary sphincter control.</td>
<td>As above</td>
<td>Poor</td>
</tr>
<tr>
<td></td>
<td>2nd Operation T10-T12 Partial removal</td>
<td></td>
<td></td>
<td>Poor</td>
</tr>
<tr>
<td>7 (43)</td>
<td>T6-L1 Total removal</td>
<td>Severe spastic weakness legs, rt. &gt; left. Able to walk only with great difficulty. 'Sacral syndrome' including urinary incontinence.</td>
<td>Some improvement in spasticity and ataxia. Walking improved—estimated 50% Vibratory sense decreased in legs.</td>
<td>Fair</td>
</tr>
<tr>
<td>11 (37)</td>
<td>C1-C4 Total removal</td>
<td>Subarachnoid hemorrhage. No neurological deficit.</td>
<td>No neurological deficit.</td>
<td>Excellent</td>
</tr>
<tr>
<td>Case Age No.</td>
<td>Operation Details</td>
<td>Preoperative Findings</td>
<td>Postoperative Findings</td>
<td>Results</td>
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<tr>
<td>13 (55)</td>
<td>Cerebellar angiomata removed'50 T2-T6 Partial removal 2nd Operation T6-L2 '67 Remainder removed</td>
<td>Moderate weakness legs. In walking, circumducts left leg and uses cane. Decreased sensation legs. Partial urinary incontinence. Severe weakness both legs. Unable to walk. Sphincter difficulties.</td>
<td>Improvement in weakness for 6-8 months and then progression to where unable to walk.</td>
<td>Fair</td>
</tr>
<tr>
<td>14 (46)</td>
<td>1st Operation elsewhere Decompression T8-T9 2nd Operation T7-L1 Partial removal with lysis arachnoid at site of decompression.</td>
<td>Severe spastic weakness both legs. Able to walk with difficulty. Decreased sensation in legs. Severe weakness both legs. Unable to walk. Analgesia below D11.</td>
<td>No improvement following surgery. Unable to walk. For first 48 hrs. no spasticity and good motor function. Then severe motor weakness followed with gradual improvement. Can now walk but only slowly and with help. Patient happy to have no pain or paresthesia. Normal micturition.</td>
<td>Poor</td>
</tr>
<tr>
<td>15 (27)</td>
<td>1st Operation T1-T6 '64 Partial removal 2nd Operation T7-T12 '67</td>
<td>Severe weakness in legs. Walks only with cane. Marked sensory decrease below T6. Partial urinary incontinence and impotent. Complete paraplegia.</td>
<td>Improvement for 8 months and then progression to complete paraplegia. Complete paraplegia unchanged.</td>
<td>Fair</td>
</tr>
<tr>
<td>16 (61)</td>
<td>T5-L1 Total removal at these levels but has extension above T3 and not removed.</td>
<td>Moderate weakness and spasticity of rt. leg. Decreased sensory modalities below D12. Left &gt; rt. Sacral anesthesia, left. Urinary incontinence—needs catheter.</td>
<td>Rt. leg now normal strength. Left leg weak but improving. Continued sensory level at D12 but loss is still improving. Left sacral anesthesia and left penile anesthesia remains. Has only 100 cc residual urine and improving.</td>
<td>Good</td>
</tr>
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and “poor” groups. If the patient had little or no neurological deficit preoperatively and the malformation was removed without incurring any significant increase in deficit, we considered this an “excellent” result. In addition any patient who had a significant deficit preoperatively which cleared after surgery was likewise classified as an “excellent” result. A “good” result was recorded when moderate, but incomplete, improvement in the neurological deficit occurred and the patient was able to walk without help. A “fair” result indicated some improvement in the neurological picture but that the patient was still left with considerable deficit. A “poor” result indicated no improvement or increased deficit following surgery.

Seventeen patients have had a total of 20 operations in this clinic with either total or subtotal excision of their spinal vascular
malformations. An “excellent” result occurred in 5 cases, a “good” result in 5, a “fair” result in 3, and a “poor” result in 3. Insufficient time has elapsed since the operation on our last case to judge the operative result. Evaluation of these results demonstrates in Cases 5, 14, and 15 that spinal decompression and partial removal of the malformation may have an ill effect on the patient whose condition is getting worse. We presume that these procedures enhance the “spinal steal syndrome” and thus create an additional impairment of the vascular supply of the cord.

Such generalized terms are useful for roughly indicating results but a better estimation of operative results can be obtained by looking at Table 1 and comparing the preoperative neurological abnormalities with those present postoperatively in each of our cases.

We believe that the treatment of spinal vascular malformations should be by surgical excision. Most of the lesions can be totally removed without increasing the neurological deficit. The best results are obtained in those patients who have little or no neurological deficit preoperatively. We therefore select those cases for excision which either have had an acute episode of subarachnoid hemorrhage with no neurological deficit, or show a slow progression of neurological symptoms and signs. Cases with complete long-standing paraplegia in flexion are excluded. The use of the operating microscope is a great adjunct and is strongly recommended for those experienced in its use.

Summary

Spinal vascular malformations may be divided histologically into one of three groups: angioma cavernosum, angioma racemosum, and angioreticuloma. The 17 operative cases we have reported were all of the angioma racemosum type. These malformations tend to occur in the thoracic or thoraco-lumbar-sacral areas of the cord. The clinical picture may be one of slow progression, or progression with regression, or of a sudden apoplectic onset. The neurological examination may range from one that is entirely normal to one showing a complete transverse myelitis. Spinal fluid examination will usually show an increase in spinal fluid protein. Routine x-rays of the spine will rarely suggest the diagnosis. Myelography will usually show the mass of “worm-like” vessels commonly associated with the diagnosis of vascular malformation. Myelography in this disease is best carried out with the patient lying on his back rather than prone, and it is important to use at least 10 cc of contrast medium. Spinal angiography gives the most accurate information as to the size and extent of the malformation and also the precise location of the “feeder” vessels. Total excision of these lesions is the treatment of choice and can often be done without appreciable decrease in the neurological function. Operation is best done under the operating microscope and use of the bipolar coagulator is absolutely essential. We have reported the technical details in the surgical excision of these lesions and also our operative results in 17 patients.

References


