VASCULAR MALFORMATION OF THE SPINAL CORD

Report of a Case

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The occurrence of vascular malformations and vascular tumors of the spinal cord is perhaps sufficiently infrequent and interesting to justify the recording and discussion of a case which is unique in the experience of the writer. Most neurologists, through the writings of Lindau, Cushing, Bailey, Dandy, Olivecrona, and others, have become familiar with these conditions in the brain. However, except for single case reports or reports of small groups of cases, there has been no attempt until recent years to make a comprehensive study and classification of these vascular abnormalities of the spinal cord.

In 1941, O. A. Turner and J. W. Kernohan¹ reported a pathologic study of 46 cases of vascular malformations and tumors of the cord. In 1944 Wyburn-Mason² published a monograph entitled *The Vascular Abnormalities and Tumors of the Spinal Cord and Its Membranes* in which he presented a classification of these conditions, thoroughly discussed each type, and added 57 cases to the literature. A very complete bibliography has been included in his book. Prior to 1941 he had found only 140 cases in the literature. His classification is as follows:

(A) Abnormalities

(I) Venous abnormalities

- (a) Secondary venous abnormalities, that is, those occurring below a tumor of the cord or associated with arachnoiditis or calcification of the cord.
- (b) Angioma racemosum venosum, that is, extensive venous varicosities affecting the pia and central regions of the cord.
- (II) Arterio-venous angioma
- (III) Arterial anomalies
 - (a) Associated with congenital heart disease.
 - (b) Alone.
- (IV) Syphilitic aneurysm of the spinal arteries.
- (V) Telangiectases, including so-called cavernomata or cavernous angiomata.

(B) True Tumors

(VI) Hemangioblastoma or hemangio-endothelioma.

- (a) Angioreticuloma, or Lindau's tumor, occurs in the cord or on a nerve root and may be associated with syringomyelia, or with similar tumors elsewhere in the nervous system and cysts in other organs.
- (b) Extradural hemangioblastoma.

(VII) Lymphangioma.

Arterial anomalies in the form of large tortuous collateral arteries, with or without the formation of a local aneurysm, may cause spinal cord compression in cases of coarctation of the aorta or patent ductus arteriosus. The case to be reported here, however, showed no evidence of congenital heart disease, and in view of the condition exposed at operation, it probably should be considered in the above classification as an arterial anomaly without congenital heart disease. Unlike 3 cases reported by Wyburn-Mason,² this case did not exhibit symptoms and signs of cord compression, but only those of nerve-root irritation, probably because of the low location of the lesion in the vicinity of the conus medullaris and cauda equina. The arterial anomaly appeared to be entirely extramedullary, and none of its constituent vessels was seen to penetrate the cord.

A word of caution regarding the surgical treatment of these vascular abnormalities of the cord may be worth-while. Radical attempts to excise, ligate, or coagulate these lesions should be avoided in most instances because of the danger of resultant degenerative changes in the cord due to alterations in blood supply.

Roentgen therapy is of little avail in the presence of vascular abnormalities which are composed of large vessels.

Case Report

A white man, aged 46, was first examined at the Clinic on May 11, 1946.

History. About eighteen months previously the patient had first experienced sharp pains in the calves of both legs. The pain was like a toothache and lasted three or four hours. A week later similar pain was experienced and lasted for two weeks. It was exaggerated by coughing and straining to move the bowels. Six weeks later pain recurred in the calves and also in the buttocks and lower back. The patient found it more comfortable to sleep while sitting up in a chair. Since that time he had had intermittent pain occurring in either buttock or leg. Two weeks before admission the patient had experienced severe pain in the left buttock, left popliteal area, left calf, and lower anterior surface of the left thigh. At the same time there was less severe pain of similar distribu-

tion on the right side. There had been no numbress, no loss of strength in the legs, and no loss of sphincter control.

During the eighteen months of pain experienced by this patient the following diagnoses had been made: sacro-iliac strain, neuritis, arthritis, male menopause, muscular sprain, and sciatica. The following forms of treatment had been carried out without relief: osteopathic and chiropractic adjustments, novocain injections in the sacro-iliac area, strapping of the back, and short-wave therapy.

General examination revealed a strong, healthy looking, middle-aged man, not acutely ill. There was no evidence of pulmonary, cardiac, or intra-abdominal disease. Rectal examination showed no masses, and there was no palpable evidence of prostatic neoplasm. Temperature was 98.1°, pulse 96, blood pressure 160/100. There was no evidence of zonal vascular abnormality or pigmentation of the skin.

Neurologic examination showed limited motion of the lumbar spine in all directions but no flattening of the normal lumbar curve and no list. The patellar and Achilles reflexes were very sluggish and equal on both sides. Motor function of the legs was normal. Sensation was normal throughout. There was no spasticity, no Babinski sign, and no clonus.

Roentgen examination of the lumbosacral region of the spine showed no increase in the width of the canal, no erosion of vertebral bodies or pedicles, and no other abnormalities.

Laboratory studies of the blood and urine, including the blood Wassermann reaction, were all normal.



FIGURE. Vascular malformation of spinal cord.

Lumbar puncture between the second and third lumbar vertebrae on May 21, 1946, showed clear yellow cerebrospinal fluid at 170 mm. H_2O pressure. There was a complete spinal subarachnoid block by the Queckenstedt test. Only 1.5 cc. of fluid could be obtained for examination. This showed no cells, a trace of globulin, 4000 mg. total protein per 100 cc., and a negative Wassermann reaction.

Roentgen myelography was carried out after the introduction of 3 cc. of pantopaque into the lumbar subarachnoid space between the fourth and fifth lumbar vertebrae. The opaque medium failed to ascend in the spinal canal beyond the level of the third lumbar vertebra when the head of the patient was lowered on the fluoroscopic table.

As a result of the above clinical and laboratory data a diagnosis of intraspinal tumor was made. It seemed evident that its lower end did not extend below the level of the third lumbar vertebra. Laminectomy was advised and carried out on June 13, 1946. The spinous processes and laminae of the twelfth thoracic vertebra and of the upper three lumbar vertebrae were removed. The dura was opened in the midline throughout the length of the exposure, and through the transparent arachnoid large tortuous red blood vessels could be seen overlying the posterior aspect of the conus medullaris and extending downward among the strands of the cauda equina. When the arachnoid was opened, the serpentine mass of vessels was clearly visualized. The vessels were quite red in color and appeared to be slightly elevated above the surface of the cord. The largest vessels in the mass appeared to be about 1.5 mm, or 2.0 mm, in diameter. They did not appear to plunge into the substance of the cord, and several tortuous vessels extended downward below the lower limit of the exposure in close proximity to the strands of the cauda equina, especially on the left side of the spinal canal (figure). The tortuous mass of vessels overlying the conus medullaris appeared to extend upward beyond the upper limit of the exposure, but no attempt was made to enlarge the bony opening. It was impossible to determine from gross observation whether the vessels were arteries or veins, but it was the operator's impression that they were probably arteries. No aneurysmal dilatations were observed, and there was no evidence of any neoplasm such as a neurofibroma, meningioma, or ependymoma. It was considered inadvisable to attempt any radical removal of this vascular malformation. Ligation or electrocoagulation of the vessels in the mass might have resulted in serious neurologic dysfunction which was not present prior to operation. The dura was closed, and the soft parts were united in layers in the usual manner.

Following operation the patient received a course of roentgen therapy to the spinal cord and cauda in an effort to sclerose the vascular mass. He was discharged from the hospital on the seventeenth postoperative day without complications. He was last observed on August 31, 1946, at which time he stated that his pain was less severe than prior to operation and that he had returned to work on July 15. He did experience some pain on walking, but on the whole he felt definitely improved. His motor, sensory, and sphincteric functions were still normal, and his patellar and Achilles reflexes could not be elicited.

References

- Turner, O. A., and Kernohan, J. W.: Vascular malformations and vascular tumors involving spinal cord; pathologic study of 46 cases. Arch. Neurol. & Psychiat. 46:444-463 (Sept.) 1941.
- 2. Wyburn-Mason, R.: The Vascular Abnormalities and Tumours of the Spinal Cord and Its Membranes (St. Louis: The C. V. Mosby Co., 1944).