Spinal cord arteriovenous malformation with an associated lymphatic anomaly

Case report

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Spinal cord arteriovenous malformations (AVM's), like other vascular anomalies of the central nervous system, can be associated with similar vascular lesions of the skin and viscera. A 7-year-old girl, who presented with rapidly progressing paraplegia, was found to have a spinal cord AVM, cutaneous angioma, and a chylous malformation of the lymphatic system. She had previously undergone treatment for a posterior thoracic cutaneous angioma. At surgery, upon incision of the paravertebral muscle fascia, viscous pale fluid was encountered emanating from a foramen in the thoracic lamina. The spinal AVM was resected in spite of concern that the abnormality represented spinal osteomyelitis. Postoperatively, there was full return of function in the lower extremities, along with recurrent episodes of chylothorax, which slowly came under control with dietary manipulation. A review of the anatomy of the thoracic duct and nontraumatic causes of chylothorax is presented, and the association of cutaneous and central angiomas is discussed. Finally, the treatment of chylothorax is delineated.

KEY WORDS: spinal arteriovenous malformation • lymphatic system • chylothorax • angioma • phakomatosis

Case Report

This 7-year-old right-handed white girl was admitted to the Cardinal Glennon Children's Hospital of St. Louis with acute lower-extremity weakness. When she was 3 months old, a cutaneous vascular hemangioma over the right posterior thorax extending toward the midline was detected and treated with an argon laser. Before her acute illness, the patient had normal coordination, balance, and strength in the lower extremities. Twelve days prior to admission, she fell and struck the right posterior portion of her chest wall. Five days later she developed pain below the right scapula and experienced urinary retention and constipation. She subsequently developed severe right-sided back pain which radiated circumferentially to her sternum. Immediately prior to hospitalization she was unable to walk. Associated with this weakness were paresthesias, periumbilical pain, and a low-grade fever.

Examination. Physical examination on admission revealed a large hemangioma over the right posterior portion of the chest wall near the inferior aspect of the scapula, which extended to the posterior midline, and...
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FIG. 1. Left: Myelogram with water-soluble contrast medium, anteroposterior projection, revealing an enlarged thoracic cord (arrowheads) and erosions in the fifth through eighth ribs (arrows). Right: Cone-down view of the myelogram, showing filling defects (arrows) within the dye column.

A palpably enlarged urinary bladder. On neurological examination, no meningismus or spinal tenderness was present. The patient was alert and attentive. Cranial nerve function was normal. Motor strength in the upper extremities was normal, but varied in the lower extremities from 1 to 2/5 on the left and 2 to 3/5 on the right. Deep-tendon reflexes were hyperactive at the knees but normal at the ankles. Plantar reflexes were extensor bilaterally. Sensory examination was unreliable, with a suggestion of decreased pinprick, touch, and vibratory sensation in the lower extremities. There was a reduction of anal sphincter tone, and the bulbocavernous reflex was absent.

Thoracic and lumbar spine roentgenograms revealed no significant abnormalities, aside from erosion of the right fifth through eighth ribs posteriorly (Fig. 1 left). A myelogram performed via a lumbar puncture revealed a fusiform enlargement of the thoracic cord from T-4 to T-7, with serpiginous filling defects in the myelographic column at these levels (Fig. 1). Computerized tomography following myelography demonstrated a large right-sided intradural filling defect on the posterolateral aspect of the spinal cord extending from T-3 to T-6 (Fig. 2). The radiographic studies were interpreted as consistent with an AVM with possible intramedullary extension and erosion of the ribs secondary to enlargement of intercostal vessels. This impression was confirmed by spinal angiography. The AVM was supplied on the superior aspect primarily by a branch from the right bronchial artery (Fig. 3) and, on the inferior aspect, by a branch from the right sixth intercostal artery.

Operation. Although embolization therapy was considered, the patient deteriorated rapidly following admission and lost all motor and sensory function in the lower extremities on the 2nd hospital day. On the 3rd hospital day, a T1–5 laminectomy was performed to resect the spinal AVM. An attempt was made to monitor somatosensory evoked potentials intraoperatively, but no cortical potentials were ever obtained.

A midline skin incision was made, with care taken to avoid the cutaneous angioma during dissection of the subcutaneous tissue. The paravertebral fascia was incised in the midline and, as the insertion of the paraspinal muscles onto the spinous processes of T-3 and T-4 was cut, copious amounts of viscous white fluid filled the wound. The paravertebral muscles were elevated from the lamina and the fluid was found to
emanate from a fenestration in the lamina of T-3, near the pedicle. The bone itself was soft with multiple fenestrations on the right, and additional fluid came from the area of the transverse processes. The viscous fluid pulsed into the wound in rhythm with respiration. A Gram stain of the fluid revealed a moderate number of white and red blood cells and rare Gram-positive cocci. To the right of the T-4 pedicle a duct was discovered which communicated with the thoracic cavity. The appearance of the fenestrated lamina, coupled with the appearance of the fluid, suggested spinal osteomyelitis, and it was considered whether to terminate the procedure for fear of spreading infection intradurally. However, even though the fluid appeared purulent, on further review of the Gram stain only a few organisms were seen. As preoperative myelography did not show significant extradural compression, it was concluded that this extradural process could not be responsible for the patient’s neurological deterioration, and the procedure was continued. The AVM was identified on the dorsal surface of the cord, along with an intramedullary component penetrating 4 mm into the posterior columns. The AVM was resected in its entirety. During closure, air bubbles could be seen around the spinal column.

Pathological Examination. Histological examination of the tissues obtained at the initial operation confirmed the diagnosis of an AVM. The fenestrated lamina were normal, without evidence of osteomyelitis. Intraoperative cultures remained sterile.

Postoperative Course. In the recovery room, a rightsided chest tube was placed after a chest roentgenogram confirmed the presence of pneumothorax. There was immediate return of proprioception and touch sensation in the legs, followed by a slower return of motor function. Urinary function resumed on the 10th day. Aerobic cultures from the operating room showed no growth, and anaerobic cultures grew Propionibacterium acnes. Evaluation of the fluid draining from the chest revealed no red blood cells, 160 white blood cells/cc (96% lymphocytes and 4% polymorphonuclear leukocytes), a triglyceride level of 705 mg/dl (the serum level was 188 mg/dl), and a glucose content of 70 mg/dl, confirming the diagnosis of chylothorax. On the 5th postoperative day, because of persistent drainage from the chest tube, oral feedings were discontinued and parenteral nutrition was instituted. By the 15th day the patient was back on a routine diet and the chest tube was removed. A chylothorax redeveloped only to resolve spontaneously on subsequent serial chest roentgenograms.

Upon discharge, the patient had dramatically improved and used a back and leg brace for walking. She had no bowel or bladder dysfunction, and in the lower extremities her motor function was 4 to 5/5 bilaterally. Touch, pinprick, vibration, and position sensation was intact in the lower extremities with the exception of decreased position sense in the large toe of the right lower extremity. Bilaterally, knee jerks were hyperactive and Babinski responses were present.
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The patient was readmitted to the hospital 1½ months after operation for a recurrent right chylothorax (Fig. 4), and she was treated with 5 weeks of chest tube drainage, low-fat diet, and parenteral nutrition. Her chylothorax resolved and she was discharged in good condition. She was readmitted 1 year later for an expanding paraspinal cutaneous hemangioma. This was surgically excised after the intercostal arteries supplying it were divided through a thoracotomy. Multiple lymphatic leaks in the chest wall were noted intraoperatively, and the thoracic duct was ligated to decrease the risk of a recurrent chylothorax following surgery. Histological examination of the paraspinal soft tissue confirmed a capillary hemangioma. Neurological examination of the lower extremities was normal, with the exception of slight hyperreflexia.

Discussion

This patient underwent the successful resection of a thoracic subarachnoid and intramedullary spinal AVM, and experienced a gratifying recovery from complete paraplegia. Her postoperative course was complicated by chylothorax, which required protracted nutritional therapy and chest tube drainage. Although chylothorax can occur with transthoracic spinal procedures secondary to damage to the thoracic duct, this complication has not been seen with posterior procedures that do not extend anterior to the pedicles. This phenomenon has not been reported previously, and can increase the risk of mortality and morbidity of surgery if not recognized and treated promptly.

Embryology and Anatomy of the Lymphatic System

The lymphatic vascular system develops at the end of the 5th week in utero, 2 weeks after the cardiovascular system, arising as capillary offshoots from the endothelium of veins. There are six primary lymph sacs from which lymphatic vessels grow along main veins. The right and left thoracic ducts connect the jugular lymph sacs with the cisterna chyli (a lymph sac in the retroperitoneum), and an anastomosis forms between the two ducts. The adult thoracic duct develops from the caudal part of the right thoracic duct, the anastomosis of the two ducts, and the cranial part of the left thoracic duct, and drains into the venous system at the cranial part of the left thoracic duct, the anastomosis forms between the two ducts. The adult thoracic duct develops from the caudal part of the right thoracic duct, the anastomosis of the two ducts, and the cranial part of the left thoracic duct, and drains into the venous system at the angle between the internal jugular and subclavian veins. It begins in the abdomen, passes through or near the aortic opening of the diaphragm, ascends in the posterior mediastinum on the right side of the aorta, crosses obliquely at the T5–6 vertebras to the left behind the esophagus, and ascends to enter the left internal jugular vein. The thoracic duct receives lymph from the posterior mediastinal and upper intercostal nodes, in addition to the trunk and lower extremities.

The cisterna chyli and thoracic duct are notable for their variability. There may be multiple thoracic ducts or a single duct may end in multiple channels. The duct may be completely left- or right-sided. Two studies presenting the results of 1103 human cadaver thoracic duct dissections failed to demonstrate any portion of the thoracic duct traveling posterior to the spinal vertebral bodies, although many variations do exist anteriorly to the vertebra. There are frequent communications between the thoracic duct and veins during its course through the abdomen and thorax.

Causes of Chylothorax

Pressure in the thoracic duct is normally low. Thoracic duct lymphatic hypertension may develop when there is thrombosis of the great veins into which lymph flows, as well as occlusion of the thoracic duct due to hypoplasia or obstruction. This increase in pressure leads to lymphangiectasis (dilatation of the lymphatic vessels) of the tributary lymphatic ducts and small lymph vessels, which can rupture or leak chyle. Ligation of the thoracic duct results in hypertrophy of anastomotic channels with the intercostal veins, again occasionally with the development of chylothorax. Chylothorax has also been associated with lymphangiomas of the thoracic duct and bone.

Vascular Anomalies

There are several conditions in which vascular anomalies of the CNS are associated with similar lesions of the skin and viscera, as in Sturge-Kalischer-Weber syndrome (triaid of congenital cutaneous angiomas in the distribution of the trigeminal nerve, meningeal angiomas, and angiomas of the choroid) and von Hippel-Lindau disease (congenital angiomatosis of the retina and cerebellum, often with similar lesions of the spinal cord and cysts of the pancreas, kidneys, and other viscera).

The incidence of cutaneous angiomas with spinal vascular malformations is 8% to 26%, and numerous cases have been reported. Nevi (circumscribed new growths of skin of congenital origin, derived from neuroectoderm) and vascular cutaneous lesions (Wyburn-Mason syndrome) may be associated with spinal cord vascular anomalies at the same metamere (association of spinal cord level and peripheral tissue derived from that level), both arising from a developmental fault of the CNS. Spinal vascular anomalies may also be associated with angiomas of the omentum and of the lung, liver, and kidney.

Mixed combinations of hemangiomas and lymphangiomas are extremely rare. Large, incompetent, edema-producing lymphatic ducts have been found in the lower extremities in association with local arteriovenous shunts, perhaps due to a congenital malformation or secondary to hypertrophy resulting from increased tissue fluid production. One case has been reported in which a spinal AVM was associated with the development of compensating diaphragmatic lymphatic ducts, most likely due to the high pressure in the vascular compartment causing a disequilibrium of pressure between the venous and lymphatic systems.
ported associations between spinal cord vascular malformations and chylous abnormalities. In this patient, there appeared to be anomalous lymphatic channels located posteriorly, which were easily entered upon paraspinal dissection and led to an open communication between the wound and the right chest cavity. It must be stressed that no instrumentation occurred anterior to the transverse processes of the vertebrae.

There may be several explanations for the lymphatic anomaly we encountered. First, since cutaneous and visceral vascular anomalies have been described in association with spinal AVM’s in a metameric distribution, it is possible that lymphatic anomalies may also follow this pattern, since both the vascular and lymphatic systems are derived from lateral mesoderm. Second, it is possible that the AVM caused a chronic elevation in venous pressure with the subsequent hyperplasia of lymphatic ducts and development of collateral lymphatic vessels. Increased local tissue vascular circulation with corresponding increased lymphatic tissue fluid formation would also contribute to this. Dilated and collateral lymph channels are usually thin-walled and communicate with mediastinal and peritracheal lymphatic ducts. As the patient was in a prone position during surgery, increase in venous pressure may have distended the lymphatic system further, resulting in rupture. Extension of the rupture into the chest cavity may have been responsible for the chylothorax.

Both the spinal and cutaneous AVM of this patient may have played a role in the second postulated mechanism.

Treatment of Chylothorax

Prior to 1948, the mortality rate was 100% from nontraumatic chylothorax and almost 50% from traumatic chylothorax. Today, with the improvements in supportive and operative therapy available, the mortality rate is low. Diets consisting of medium-chain triglycerides or total parenteral hyperalimentation have been used successfully in selected patients. If drainage is still present, surgical treatment may be indicated. Home-based total parenteral nutrition is a therapeutic option while waiting for nonoperative spontaneous resolution.

Conclusions

Spinal AVM’s may be associated with peripheral vascular and chylous anomalies. The surgeon should be alert to the presence of thick fluids intraoperatively, and be prepared to initiate appropriate treatment for chylous drainage.

Acknowledgments

The authors thank Ms. Joan Bialon and Ms. Annette Reinarman for their assistance in preparing the manuscript, and Ms. Adele Moore for helping with foreign language translation.

References

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Manuscript received January 16, 1990. Accepted in final form April 2, 1990.
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