Spinal cord vascular malformations with symptoms during menstruation

Report of two cases

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Two patients had the initial complaint of fluctuating paraparesis, which was most evident at menstruation. One patient had a semimonthly fluctuating deficit. Spinal cord compression and ischemia, secondary to the vascular mass, were considered the most likely mechanisms. Blood levels of estrogen and progesterone during the menstrual cycle may have had a contributory effect. Fluctuating spinal cord deficits associated with a consistent portion of the menstrual cycle should alert the physician to the possibility of an arteriovenous malformation of the spinal cord.

KEY WORDS • spinal cord vascular malformation • menstruation • fluctuating paraparesis • estrogen • progesterone • pregnancy

The clinical features of spinal cord vascular malformations have been well reviewed. Factors aggravating or precipitating symptoms include exercise, posture, injury, and pregnancy. During pregnancy, most patients become symptomatic in the third trimester, and although some neurological deficits may become permanent, most patients improve dramatically post partum.

Accepting that aggravation of symptoms and signs is a well recognized complication of pregnancy, we wish to call attention to two unusual cases in which the symptoms were related to the menstrual cycle itself.

Case Reports

Case 1

In 1959, during the third trimester of her second pregnancy, a 26-year-old woman experienced leg weakness characterized by difficulty in maintaining extension of the knees while climbing stairs. This resolved spontaneously, with a return to normal post partum. During the next 3 years, she noted intermittent weakness of her legs, but did not associate the weakness with menstruation until 1962. Before each menstrual period, she developed malaise, and during menstruation she had difficulty in climbing stairs, and her legs became weak. This pattern persisted with monthly recurrences of leg weakness during menstruation. She was first seen by one of us (C.R.S.) in 1966. At that time, examination during menstruation revealed mild symmetric weakness of the lower extremities, evidenced by impairment of heel and toe walking. She was unable to arise from a squatting position. There was a slight decrease in the quadriceps reflexes, and the Achilles reflexes were absent. Sensory ex-
amination revealed no abnormalities. A repeat examination later in the month revealed symptomatic improvement and resolution of all the previously demonstrated signs except for the Achilles reflexes, which remained absent. This pattern was confirmed on repeated examinations during the next 2 years.

In 1968, she was evaluated at another institution because of hypokalemia, but results of all studies were negative. During the next 2 years, the episodes of weakness lasted longer, beginning the week before the onset of menstruation, peaking in severity during menses, and gradually resolving the week after menses, during which time the patient’s strength returned to normal. Sensory abnormalities were confined to the lower extremities and consisted of a mild diminution of vibratory and joint position sensitivity as well as a mild decrease in pain and light touch sensitivity in a stocking distribution. Additional persistent deficits during this time included mildly impaired deep sensibility.

By 1970, 11 years after the onset of her symptoms, weakness was present for most of the month and she could walk only with great difficulty. However, mild objective improvement was noted at midcycle; this lasted 3 to 4 days.

Examination. Routine laboratory blood tests and X-ray films of the chest, thorax, and spinal column were normal. There were no fasciculations or fibrillation potentials. A Pan- topaque myelogram (Fig. 1) in May, 1970, revealed a partial block of the spinal subarachnoid space at L-1 by what appeared to be tortuous vessels adjacent to the posterior aspect of the cord and extending from L-1 to T-6. Above this level, the anterior spinal artery was large and tortuous. Spinal fluid examination revealed a protein level of 144 mg/dl but no pleocytosis. Femoral arteriography demonstrated an arteriovenous malformation (AVM) with a small tangle of vessels at the 11th thoracic interspace. Radicular venous runoff was observed under the right pedicles of T-10 and T-11.

Operation. The patient underwent bilateral laminectomy from T-9 to L-2, and a large AVM, approximately 14 cm in length, was resected with the aid of a dissecting microscope and bipolar coagulation.

Postoperative Course. The postoperative course was uneventful, with gradual improvement of strength in the lower extremities. One year after the operation, the patient could walk around the house without support, but she required a walker when out of doors. Mild leg weakness was present in the dorsiflexors of the foot. The quadriceps reflexes were symmetrically hyperactive, and the Achilles reflexes remained absent. There was a patchy hypalgesia in the L-5 and S-1 dermatomes.

The following case is presented because of a similar history, although there was no actual clinical documentation of neurological signs associated with menstruation.

Case 2

In 1965, 3 months after the delivery of her fourth child, a 39-year-old woman experienced intermittent episodes in which her right lower extremity seemed to “give way.” The weakness occurred semimonthly at midcycle and during the menses, and usually lasted 1 to 3 days before resolving. There was no pain, sensory loss, or bladder sphincter disturbance. The semimonthly weakness of

Fig. 1. Case 1. Myelogram showing low-grade obstruction of subarachnoid space by posterior arteriovenous malformation extending from L-1 to the midthoracic levels. Dilated and tortuous veins are evident (arrows).
FIG. 2. Case 2. Abdominal aortogram demon-
strates arteriovenous malformation (AVM) pro-
jected over L-1 interspace, just below hemostasis 
clip (HC). A = anterior radiculomedullary artery 
of Adamkiewicz; R = radicular artery of T-12; 
V = draining vein.

the right lower extremity became progres-
sively more severe during the next 4 years but 
fully resolved a few days after onset. She 
sought medical attention in 1969, and myel-
graphy revealed an AVM of the lower tho-
racic and upper lumbar cord. No specific 
treatment was recommended at that time. In 
1970, she underwent a hysterectomy and left 
ophorectomy at another institution. The 
neurological symptoms continued on a semi-
monthly basis and seemed to occur at the ex-
pected times of midcycle and menstruation.

Increasing disability led to excision of the 
AVM at another hospital in January, 1971. 
Postoperatively, she developed an asym-
metric paraparesis and sensory loss and com-
plete loss of bladder function. There was 
minimal improvement during the subsequent 
months, but the patient still complained of a 
semimonthly worsening of her conditioning in 
the form of increased weakness in the lower 
extremities lasting 1 to 3 days.

Examination. Neurological examination at 
our clinic at that time revealed a combined 
spastic and flaccid paraparesis with absent 
deep tendon reflexes in the legs. The anal 
sphincter tone was reduced, and there was 
moderate impairment of all sensory modal-
ties in the distal aspect of the lower ex-
tremities and perineum.

Results of routine laboratory studies were 
normal. An abdominal aortogram (Fig. 2) 
showed a small arteriovenous anomaly pro-
jected over the first lumbar interspace, just 
below the hemostasis clip placed at the time 
of the previous operation. The interspace 
filled by way of the anterior radiculome-
dullary artery of Adamkiewicz, which 
arose from the intercostal artery of T-12 on 
the left. A draining vein, ascending in the 
spinal canal, and multiple small venous 
channels in the low thoracic and upper lum-
bar regions also were demonstrated.

Course. It was decided that further sur-
gical intervention was not justified because of 
the existing permanent neurological deficit. 
Recent correspondence with this patient dis-
closed that her neurological status has 
remained unchanged and that the semi-
monthly fluctuations spontaneously ceased.

Discussion

Both of our patients complained of neuro-
logical symptoms in the lower extremities 
during menstruation, and neurological signs 
developed and resolved in association with the 
menses in Case 1. Our second patient also 
complained of similar symptoms at midcycle.

To our knowledge, previous reports con-
cerning spinal cord vascular malformations, 
in which symptoms and signs developed in 
relation to the menses, are few. Epstein, et 
al., described a patient with an arterial 
anomaly of the lower thoracic and upper lum-
bar spinal cord, whose lumbar pain was worse 
during menstruation. Pappenheim described 
a patient with a racemose angioma of the cer-
vical spinal cord in whom paresthesias 
developed in her legs and arms during 
menstruation. This association, as reflected in 
the literature, is exceedingly rare.

Although vascular malformations may 
produce local deficits by various mechanisms, 
including thrombosis, ischemia, and hemor-
rhage, Kaufman, et al., supported the con-
cept that symptoms and signs often resulted 
from compression of the spinal cord by the 
vascular mass. In both of our patients, fluc-
tuation in the size of the mass could have been 
one of the mechanisms whereby impairment 
of spinal cord function occurred. The size of 
the vascular malformation may vary in 
response to the circulating hormones estrogen 
and progesterone, the levels of which regulate
the menstrual cycle itself. These hormones have a number of physiological effects that could increase the size of the malformation, such as increasing venous distensibility during the second half of the menstrual cycle and causing increased blood flow, the latter effect being at a maximum at the time of ovulation. The exogenous administration of estrogen has been shown to dilate the ear veins of oophorectomized rabbits and to increase vascular ground substance, especially in smaller vessels. Estrogen also produces retention of total body sodium, chloride, and water, which secondarily increases the total blood volume.

All of these factors, alone or in concert, could theoretically increase the size of the vascular malformation and produce symptoms. If this happens, the maximum size should occur toward the middle of the menstrual cycle, near the time of ovulation. Such a mechanism could explain the symptoms at midcycle in Case 2 but not the changes in both patients during menstruation.

Blood flow through the endometrial spiral arteries closely parallels blood estrogen levels. During menses, when estrogen levels are low, there is a pronounced decrease in blood flow through these arteries, leading to ischemic conditions. Whether there is a similar effect of estrogen withdrawal on blood flow in the abnormal vessels composing the spinal vascular malformation is unknown, but mild ischemia could have produced the deficits in both patients at the time of menstruation.

Whether any of these mechanisms were responsible for the fluctuating neurological deficits in our patients is speculative at the present time. Hopefully, future studies in the areas of reproductive and vascular physiology will contribute more data on this interesting phenomenon.

Clinically, women of child-bearing age who have a history of a fluctuating paraparesis or lower extremity sensory symptoms, when associated with menstruation, should be suspected of having an occult spinal vascular malformation. Once this is considered, appropriate confirmatory diagnostic studies can be undertaken.

References


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