Hemangioma Calcificans of the Spinal Cord

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ABSTRACT: A 27-year-old woman presented with hemotomyelia during pregnancy. Needle drainage of the spinal cord was followed by relief of symptoms. Symptoms recurred during a second pregnancy 4 years later and she presented with an avascular mass in the midcervical spinal cord. This lesion proved to be hemangioma calcificans, a densely calcified and ossified variant of cavernous angioma.

RÉSUMÉ: Hémangiome calcifié de la moelle épinière Une femme de 27 ans présente, lors d'une grossesse, une hématomyélie. Le drainage à l'aiguille de la moelle est suivi d'une amélioration des symptômes. Lors d'une seconde grossesse, 4 ans plus tard, on note à nouveau les même symptômes et on identifie une masse avasculaire mi-cervicale. Cette lésion fut confirmée comme étant un hémangiome calcifié, soit une variante calcifiée et ossifiée d'un angiome caverneux.

Can. J. Neurol. Sci. 1985; 12:321-322

Cavernous angiomas (cavernomas) account for 5 to 16% of spinal vascular malformations. Isolated intramedullary vascular malformations, including cavernomas, are rare.[1] While 14 cases of intracranial hemangioma calcificans, a densely calcified and ossified variant of cavernous angioma, have been reported in the literature,[2,3] we have found no previous reports of such lesions in the spinal cord.

Case Report

A 27-year-old woman was admitted during her first pregnancy in 1979. Two weeks earlier, after moving some furniture, she experienced aching pains in the neck and interscapular region. She then developed urinary hesitancy and frequency, a sense of a full bladder, obstipation, a sense of "walking on sponges" and unsteady station and gait.

She had taken oral contraceptives for seven years and had received injuries to tendons of the right arm and leg in a car accident 21 years before.

On examination, there was mild weakness of the interossei of the right hand, of finger, wrist and elbow extension and of hip flexion on the same side. Tone was increased in the legs and the deep tendon reflexes were brisk, especially those of the right arm. The left plantar response was extensor. Perception of pinprick was decreased posteriorly from C4 to the feet including the perianal area; anteriorly, the deficit was between T5 and T12. A small cutaneous nevus was present near the midback and a midsystolic click and murmur were heard medial to the cardiac apex.

Radiographs of the cervical spine were normal. Myelography showed an intramedullary lesion expanding the cord at the C6 level.

Laminectomy of the third to sixth cervical vertebrae was performed. On opening the dura, the cord was seen to bulge at C5. No other abnormalities were noted. A 25 gauge needle was inserted into this area and 2.2 ml of hemorrhagic fluid were withdrawn.

Post-operatively, the Babinski reflex had disappeared and, after a course of intermittent self-catheterization, normal micturition resumed. The rest of the pregnancy and delivery were normal. The patient became asymptomatic.

She was readmitted in 1984 with a six month history of painful dysesthesiae of the feet. A second pregnancy had been normal.

On examination, there was leg pain with neck flexion. There was decreased pinprick sensation from T4 to the left leg. The deep tendon reflexes were 3+ (4+ at the ankles) and the plantar reflexes were flexor.

Metrizamide CT scan from C2 to T1 and myelography showed a lesion expanding the spinal cord at C6 with atrophy above and below it. Spinal angiography revealed no abnormal vascularity.

Exposure of the cord at the same levels as before revealed cord dilatation at C5-C6. Old subpial blood was drained. There were no abnormal vessels on the surface of the cord. Myelotomy disclosed a lobulated mass of varix-like vascular spaces, the centre of which was stony-hard. It extended almost to the anterior surface, but could be separated from the surrounding gliotic cord and was completely removed, essentially splitting the cord longitudinally.

Except for a slightly spastic gait, full recovery was made.

The mass (Figure 1) consisted of a collection of closely contiguous, nonarterial vascular structures of varying sizes. The walls were composed of mostly hyalinized collagenous fibrous tissue and were lined by a single layer of endothelial cells. In many places the collagen merged with the calcified tissue and areas of bony metaplasia comprising the bulk of the lesion. The bony tissue was mature, lamellar bone containing hematopoietic marrow. Iron pigment and macrophage collections indicated prior hemorrhage.

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Received April 2, 1985. Accepted July 8, 1985
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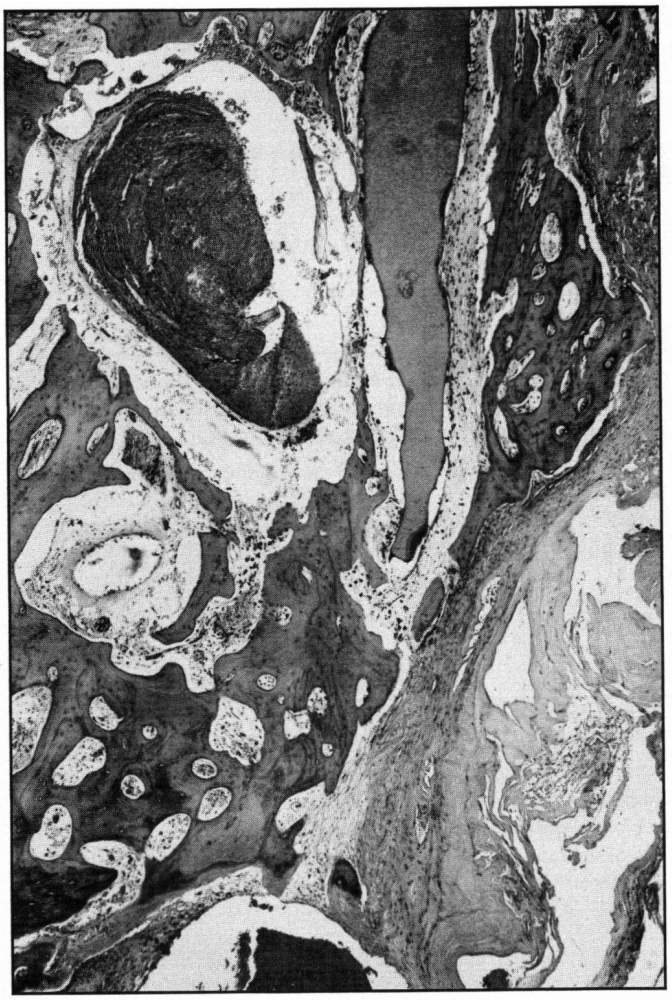

Figure 1 — Photomicrograph of the tumour. It is composed of non-arterial vascular channels with areas of dense calcification and bone formation. H&E X 32.

Discussion

Calcification and bone formation are known features of both intracranial and spinal cord cavernous hemangiomas.[45] However, it is only the massively calcified and ossified variety to which the term "hemangioma calcificans" is applied. Our case demonstrates the intramedullary location of such a lesion, previously only reported in intracranial sites.

Spinal vascular malformations in general may be complicated by subarachnoid hemorrhage, hematomyelia, compressive lesions of cord and roots and ischemic changes causing progressive radiculomyelopathy, the Foix-Alajouanine Syndrome.[6] The first two of these features are the most common. Chronic damage may result from pressure effects, thrombosis, abnormal venous drainage and "steal" phenomena.[1] Although surgical excision remains the preferred treatment, it was decided to treat by drainage alone owing to the risk of intramedullary surgery in an otherwise healthy, pregnant patient.[2] When the symptoms recurred, a definitive excision was made.

Our patient's symptoms began during her first pregnancy after she had been lifting heavy objects. Aggravation or precipitation of symptoms by exercise and by pregnancy have been documented,[7] but the actual risk that they will lead to symptoms is not known. It has been suggested that cerebral hemangioma calcificans represents a lower risk of hemorrhage than the usual kind of cavernoma.[8] Our case may not refute that claim, since the hemorrhage could have occurred into the common type of cavernoma, which calcified and ossified later. The bone may then have prevented hemorrhage during the second pregnancy, though enlargement could still have taken place.

Congenital anomalies such as segmental or nonsegmental cutaneous pigmented nevi have been reported in patients with spinal vascular malformation.[7] Our patient's nevus and mitral valve prolapse, being common findings, may simply be chance associations.

The intracranial forms of hemangioma calcificans appear avascular on angiography,[2] as did this spinal form, and their appearance on computerized tomography is not changed by contrast enhancement.[9] These radiographic features may aid in differential diagnosis.

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https://doi.org/10.1017/S0317167100035423 Published online by Cambridge University Press