Extensive Spinal Hemangioma Associated With Cutaneous Nevus in the Same Metamera: An Inusual Case of Paraplegia in the Peripartum

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Abstract

Cavernous hemangiomas most commonly occur in the cerebral hemispheres but can involve any part of the neuraxis including the spine. Very rare cases of spinal angiomas are associated with a skin lesion in the same metameric distribution in which case they constitute a segmental neurovascular syndrome, also known as Cobb Syndrome due to the description of a case in 1915. A case of a 37 year old woman with a cutaneous nevus from C7 dermatome to L3 dermatome which presented as pelvic limb paralysis 48 hours after giving birth to a healthy newborn by cesarean section is reported. The magnetic resonance demonstrated an enhancing extensive epidural mass from C7 to T7 and subsequently from T10 to L3. Histopathology revealed a spinal cavernous hemangioma. The aim of this study is to report a unique case of neurovascular segmental syndrome with extensive cervical, thoracic and lumbar involvement expressed as peripartum spinal cord compression syndrome. The clinical presentation, imaging findings, histopathology, differential diagnosis, surgical considerations and aggravating or precipitating situations included pregnancy are discussed. Although neurovascular segmental syndrome is a rare entity it should be considered if a patient with cutaneous angioma manifesting with radiculopathy or myelopathy signs, so that early diagnostic can lead to curative surgical treatment and more favorable outcomes.

Categories: Neurology, Neurosurgery, Obstetrics/Gynecology

Keywords: nevus flammeus, angiomatosis, compressive myelopathy, peripartum, spinal angioma, cavernous hemangioma

Introduction

Spinal angiomas represent 12% of all vascular pathologies of the spinal cord and four percent of lesions located in the epidural space [1]. Spinal cavernous hemangiomas most commonly arise from the vertebral body, being diagnosed as a finding or by the clinical expression of compressive myelopathy when the lesion extends to the epidural space and compresses the spinal cord. More commonly, the clinical presentation is slowly progressive in the form of spinal cord compressive syndrome or radiculopathy. There is occasional reports of late disturbance of sphincter function [1]. The sudden onset of symptoms is often secondary to enlargement of the lesion caused by intralesional hemorrhage, thrombosis, increased vascularization caused by hormonal effects, or mechanical venous occlusion [2]. Very rare cases of spinal angiomas are associated with a skin lesion in the same metameric distribution in which case they constitute a segmental neurovascular syndrome. Cobb syndrome is a rare non-hereditary neurocutaneous disease, characterized by spinal vascular malformations associated with a cutaneous vascular malformation located in the same metameric distribution. The skin lesions may occur anywhere in the dermatome, from the midline of the back to the abdomen [3]. Since Cobb’s original discussion, only 40 cases have been reported in the international literature; of those, 20 were children [3]. The presence of myelopathy or less commonly radiculopathy in patients with a birth nevus should make it necessary to perform a magnetic resonance to characterize the location and characteristics of a spinal vascular malformation that could be associated.

Case Presentation

A 37 year old woman in the postpartum period was referred to our hospital. Her history included the presence of paresthesias in the pelvic limbs for two months, with progression to paraparesis for one and a half months, which conditioned the use of a cane. 11 days prior to her admission, she underwent a cesarean section in another unit due to lack of labor progression. 48 hours after the cesarean section, the paresis worsens to paraplegia. Upon her admission, the presence of two monoform macula-shaped lesions with a purplish-red appearance located in the dorsal thoracic and lumbar region with well-defined borders, stands out. The gynecological examination revealed inadequate uterine involution, which is why it was evaluated by the gynecology service who found data suggestive of intrauterine hematoma vs hysterorrhaphy dehiscence. She was admitted to the operating room where a hysterectomy plus right salpingooophorectomy was performed without incident or complications. Four days after the gynecological procedure, she underwent laminoplasty of T12-L2 plus partial resection of the lesion where findings were reported as a highly vascularized extradural lesion in the right dorsolateral region of the spinal canal. Histopathology revealed a cavernous hemangioma.
The neurological examination reported muscle strength five out of five for C5-T1, zero out of five for L2-S1, deep bicipital tendon reflexes, supinator ++/++++, patellar reflex and bilateral Achilles reflex ++++/++++ accompanied of exhaustible clone, diminished tone. Sensitivity from C1 to T8 preserved in its protopathic and conscious proprioceptive modality, with decreased sensitivity from T9 towards the distal. Without presence of radicular or myelopathic pain, only local mechanical pain, without alteration in the bladder sphincter. Upon inspection, the presence of two monoform macula shaped lesions with a purplish red appearance located in the dorsal thoracic and lumbar region with well defined borders stands out (Figure 1). The cutaneous nevus extended from the C7 dermatome to the L3 dermatome as represented in the diagram in Figure 2.

**FIGURE 1:** Photographs showing the cutaneous nevus

Figure 1A) Monomorphic macules with well defined margins of a purplish-red appearance in the dorsal thoracic and lumbar region. Figure 1B) Cutaneous macule in the dorsal region of the chest

**FIGURE 2:** Diagram showing the extension of the cutaneous nevus

Figure 2A) Extension of the cutaneous nevus from the C7 dermatome to the L3 dermatome in a left lateral and posterior view. Figure 2B) Diagram of an axial section at the level of L2 representing the displacement of the conus medullaris (arrow), the involvement of the vertebral body (star) and the extension through the left neuro foramen to reach the ipsilateral psoas muscle (asterisk). Image Credits: Rogelio D. Flores Reyes Jr.

**Imaging findings**

A simple MRI with gadolinium of the cervicothoracolumbar spine was requested, in which an extraspinal and intraspinal lesion located extradurally was observed. MRI T1 weighted sequence showed a predominantly hyperintense extradural lesion of lobulated morphology with well-defined margins predominantly at the level of the posterior, left lateral margin and to a lesser extent anteriorly of the spinal canal with extension from C7 to T7 and subsequently from T10 to L3. It compresses the tectal sac and displaces the medullary cord. In addition, extension of the lesion was observed at the foraminal level from T11 to L2 on the left side and invasion of vertebral bodies from C8 to L4. Extension of the lesion of fatty tissue and left iliac psoas muscle was also observed. STIR-Weighted MRI image showed a hyperintense image
with respect to the spinal cord and hypointense with respect to the cerebrospinal fluid. (Figures 3-6).

**FIGURE 3: Sagittal MR images of the cervical and upper thoracic spinal cord**

Figure 3A) T1 weighted sequence showing a predominantly hyperintense extradural at the level of the posterior margin of the spinal cord from C7 towards caudal. Figure 3B) T1 with gadolinium where the same lesion is observed with intense homogeneous contrast enhancement. Involvement of the vertebral bodies from C8 is also shown. MR (magnetic resonance).
FIGURE 4: Saggital thoracolumbar MR images

Figure 4A) T2 weighted sequence showing a hyperintense epidural mass that extends from T10 to L3, which also invades the vertebral bodies from T9-L4, causing effacement of the tectal sac and compression of the spinal cord. Figure 4B) Hyperintense lesion in relation to the spinal cord on T1-weighted MR image. Figure 4C) STIR-Weighted MR image showing hyperintense image with respect to the spinal cord and hypointense with respect to the cerebrospinal fluid. MR (magnetic resonance)

FIGURE 5: Axial and Saggital MR images at L2 level

Figure 5A) Axial T1-weighted MR image with gadolinium where involvement of the vertebral body is observed, an epidural occupational lesion that causes displacement to the right of the cauda equina, extension to the left neuroforamen L2-L3 with involvement of ipsilateral psoas. Figure 5B) Saggital T1-weighted MR image with gadolinium where the extension of the lesion is observed in the craniocaudal axis from its lower portion that goes from T10 to L3 (arrow points a line at L2 level). MR (magnetic resonance).
FIGURE 6: MR images showing thoracolumbar extension

Figure 6A) Axial T2-weighted MR image at the level of L3 where displacement of the nerve roots of the cauda equina is shown. Figure 6B) Sagittal T1-weighted MR image where the hyperintense behavior of the lesion is observed. (arrow points a line to the L3 level). Figure 6C) Same level coronal T1-weighted with gadolinium where left predominance of the lesion is observed with extension towards ipsilateral neuroforamens. MR (magnetic resonance).

Surgical procedure and findings

A posterior approach guided by fluoroscopy was performed, an incision in the posterior midline, laminotomy with a T1-L2 drill. We proceeded to explore the epidural space, observing a dark purple mass on the dorsolateral side, predominantly on the left, vascularized with thinning of the dura mater.

Histopathological findings

Histopathology revealed cavernous hemangioma. In the histological section, the epidural lesion was composed of congested venous vessels in compact irregular groups that alternated with dystrophic calcifications. The vessel walls were of irregular thickness with anastomosed lumens without obvious elastic fibers. The endothelium showed no evidence of atypia (Figure 7).

FIGURE 7: Photomicrograph (haematoxylin and eosin).

Figure 7A) Original magnification X10. Figure 7B) Magnification x20; the venous vessels show simple flat endothelium without atypia. Figure 7C) Magnification 5X; nutrient arterial vessels are observed in the upper right corner and congestive venous vessels in irregular compact groups that alternate with dystrophic calcifications.

Discussion

Spinal cavernous haemangiomas are vascular malformations with a predilection for the thoracic spine. Magnetic resonance is diagnostic and total removal of the mass is the optimal treatment [1]. On average, two and a half vertebral segments are involved in each case, suggesting that epidural cavernous hemangiomas more likely tend to grow laterally than longitudinally [4]. This contrasts with the data presented in this case where both lateral and longitudinal extension were observed.

Epidural cavernous hemangiomas can develop anywhere along the spinal canal but predominantly develop at the thoracic levels with a predilection for the T2-T6 segment and multisegment involvement, followed by cervical, lumbar, and sacral levels in the order of reducing occurrence [2]. Pure epidural location, defined by the presence of 90% of the tumour volume in the epidural space is unusual [5]. Even more unusual is the cervical, thoracic, and lumbar extension described in this case.

Intraspinal vascular lesions can be divided into vascular tumors (hemangioblastoma and cavernous malformation) and arteriovenous malformations. They can also be divided into epidural and intradural...
lesions [4]. A key characteristic in magnetic resonance when arteriovenous malformations are suspected is that they usually present intensities of flow voids due to the high flow velocity of the vascular structures, which can be a key piece of information in the differential diagnosis between these two lesions.

Acute hemorrhage often leads to important neurologic deficit, requiring urgent decompressive surgery. However, the degree of functional recovery will depend to a considerable degree on whether the location of the bleeding is epidural or intramedullary. In most cases, an epidural injury has a better prognosis as it does not involve direct damage to neural tissue.

Since the description by Cobb in 1915 of a spinal hemangioma associated with paraplegia, isolated cases of spinal vascular malformations associated with cutaneous vacular lesions have been reported. This may be due in part to the lack of clinical expression of quiescent lesions. The vascular skin nevus found with Cobb syndrome is accompanied by a large variety of vascular pathologies. The intraspinal lesions are usually AVMs (high flow lesion) and rarely angiomias (low flow lesion) [6].

Although the location of spinal cavernous hemangiomas is usually limited to two or three vertebral segments at the thoracic level, in our case we observed an extensive longitudinal location at the cervical, thoracic and lumbar levels as well as a tendency to invade the vertebral bodies and paravertebral region that reached the left iliopsoas muscle. The extensive location of the lesion could raise suspicion of malignant behavior; however, cellular atypia was not observed in the histopathological study. The extensive location of the lesion seems to be better explained by its association with a cutaneous nevus, which suggests that the spinal lesion was present from birth, thus constituting a vascular segmental syndrome.

Spinal cavernous hemangiomas most often originate from the vertebral bodies, sometimes with secondary extension into the extradural space. The affected vertebra had coarsened trabeculae, which may be a critical finding for differentiating epidural cavernous hemangiomas of vertebral origin from foraminal nerve sheath tumors [7]. The pattern of involved vertebrae was described as vertical striations in sagittal reformatted images and as honeycomb pattern in transverse images [7].

Magnetic resonance image typically delineated a well circumscribed lobulated lesion. T1-weighted imaging demonstrated a homogeneously isointense lesion while T2-weighted imaging showed a hypointense mass slightly less intense than CSF. Uniform enhancement on T1 gadolinium enhanced [1]. Similar findings were observed in this case, unlike the fact that in the T1-weighted sequence a hypointense behavior was observed (relative to muscle and cord) (Figure 49).

Differential diagnosis of this entity should consider other epidural tumours, such as lymphomas, metastatic tumours, meningiomas, or neurinomas-neurofibromas [8]. When they do extend through the foramen as a dumbbell-shaped lesion, they do not enlarge the neuroforamen as much as a schwannoma or neurofibroma of similar size, and they often have an irregular or lobulated contour with strong enhancement [4].

Lymphoma usually appears isointense on T2-weighted images and exhibits less frequent paravertebral extension and intervertebral neural foraminal widening. An angiolioma is typically hypointense on T1-weighted images because of its fat content, while the fat in a cavernous hemangioma is usually absent [4]. An magnetic resonance scan with fat suppression sequences should therefore be performed in order to rule out this tumour type [8]. Extending through the intervertebral foramen region assume an “dumbbell” shape on axial magnetic resonance [9]. Which could confuse the diagnosis with lesions such as meningioma and schwannoma [2]. In neurogenic tumors, however, smooth contour instead of lobulated contour and frequent cystic change could be the clues to the differential diagnosis with cavernous hemangiomas [9].

Hemangiomas of intramedullary location always exhibit a mixed signal with a hyperintense hemosiderin ring on T2WI because of repeated bleeding, which is usually not observed in epidural cavernous hemangiomas [2]. This is probably due to the absence of a blood-spinal barrier in the epidural space, which favors faster removal of hemosiderin.

Estrogen has been well-recognized to play a crucial role in the development of these lesions by directly acting upon the endothelium of the vascular channels [2].

In relation to pregnancy has been proposed a mechanical theory: by obstruction of the blood flow from the paravertebral veins into the inferior vena cava by the gravid uterus. This increase in venous pressure may lead enlargement of epidural haemangiomas; hormonal theory: by overexpression of angiogenic factors during embryogenesis, such as Vascular Endothelial Growth Factor, basic Fibroblast Growth Factor and Placental Growth Factor, leading to a significant increase in malformations during this period of pregnancy [5]. Furthermore, the rapidly enlarging of the uterus in the third trimester may explain the relatively acute onset of symptoms [5]. In our case, the symptoms occurred and worsened in the last trimester of pregnancy, which may be related to the increase in estrogen at this stage; however, other processes that could explain an acute onset or worsening must be taken into account. Our theory also considers that this type of malformations may be susceptible to vascular steal phenomena that in turn can be precipitated by minor blood losses during the birth period.
In pregnancy, multiple mechanisms can promote the clinical expression of these lesions, either through direct stimulation of their growth through growth factors or hormones or through venous congestion explained by changes in the dynamics of venous flow linked to the growth of the gravid uterus.

The treatment of this type of lesions consists of complete surgical resection through a posterior spinal approach, in most cases a laminectomy. In this case, given the extension of the lesion from C7 to L3 and the need for a laminectomy of more than two levels, instrumentation of a large extension of the spine was necessary. The risk-benefit of surgery was evaluated, attempting a complete surgical resection in this hospitalization versus a three-level laminoplasty with the objective of relieving pressure in the site of greatest spinal cord compression and sending the specimen to pathology for histopathological identification of the lesion. Given the complications of the hysterotomy that our patient presented, performing additional complex surgery increased postoperative complications, such as delayed wound healing, pulmonary venous thromboembolism, etc., so a less aggressive surgery was performed in an attempt to alleviate spinal compression and thereby improve the strength of the lower extremities.

A careful dural opening must be carried out to avoid injuring vascular structures that could be adhered to the dural plane and that could lead to an increase in surgical time or injuries due to ischemia at the spinal cord level. Coagulation in the first instance of venous drainage can cause greater blood congestion and thus increase intraoperative bleeding, so nutrient arteries must first be identified with the aim of reducing the blood flow, subsequently proceeding to coagulate venous vessels.

**Conclusions**

Although the incidence of this type of entity is low, cavernous hemangioma should be included among the diagnostic possibilities when addressing an extradural soft tissue mass. Although extension to the neuroforamen is common, it is more common to find clinical symptoms consistent with myelopathy, probably due to better tolerance to chronic compression of the nerve roots compared to the spinal cord.

The surgical procedure as well as the ideal time for its performance depends on the patient’s clinical presentation. In a patient with compressive myelopathy with acute paraplegia, urgent decompression must be performed through a laminectomy.

**Additional Information**

**Disclosures**

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**Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following:  
**Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work.  
**Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.  
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