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CASE REPORT

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Spinal epidural hemangioma related to pregnancy

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Abstract We report the case of a 39-year-old woman with adolescent idiopathic scoliosis presenting with myelopathy secondary to a spinal epidural hemangioma. MRI showed an epidural soft tissue mass within the spinal canal between T5 and T9 with severe spinal cord compression. Symptoms had a temporal relationship to her pregnancy. Surgical removal of the epidural hemangioma rapidly relieved her symptoms and neurologic deficits. Follow-up examination 2 years later demonstrated normal motor and sensory function, without any neurologic sequelae or progression of deformity.

Keywords Spinal epidural hemangioma · Pregnancy · Scoliosis · Myelopathy · Radiography · CT · MRI

Introduction

Spinal hemangiomas were first described in 1867 by Virchow [1], and usually involve the vertebral body with occasional epidural extension [2]. They are found in approximately 10% of all people [3], but are rarely symptomatic or of clinical importance. Less commonly, they may occupy the epidural space without bony involvement. Epidural lesions are frequently symptomatic [4, 5].

Hemangiomas, also called cavernous angiomas, cavernous malformations or cavernomas, are vascular hamartomas that, by definition, do not grow with mitotic activity [6]. Most evidence supports hemorrhage and thrombosis with organization and recanalization as the mechanisms of growth [7], with additional support for hormonal control [8]. Epidural hemangiomas without vertebral involvement are rare, with 56 cases reported [4, 7, 10, 11, 12, 13, 14, 15, 16, 17]. We report here on a thoracic spinal epidural hemangioma causing myelopathy in a 39-year-old woman. To our knowledge, this is the fourth report of an epidural hemangioma related to pregnancy [8]. Angiolipomas during pregnancy have been reported, and are also rare [17, 18].

Case report

A 39-year-old woman presented with complaints of intermittent decreased sensation in her lower extremities with paresthesias in her toes and occasional clumsiness. She denied any bowel or bladder symptoms. She de-

livered a healthy child 2 months prior to onset of her current symptoms. She had similar, though less severe, symptoms which occurred in her third trimester of pregnancy. An investigation by a neurologist at that time did not include imaging studies. The symptoms had completely resolved without intervention.

Her past medical history was significant for adolescent idiopathic scoliosis, diagnosed at age 12 years, and successfully treated with bracing in her teenage years. There was no curve progression in her adulthood. Her obstetric and gynecologic history was notable for multiple miscarriages, fibroids and endometriosis, for which she underwent laparoscopy as well as laser myomectomy. She had received several cycles of hormonal therapy for infertility including Clomid and Pergonal prior to becoming pregnant. She denied any symptoms associated with hormonal therapy.

Her initial physical examination demonstrated 5/5 motor strength throughout both upper and lower extremities. She was hyperreflexic in her lower extremities with 2 beats of clonus and bilateral positive Babinski signs. The patient had poorly defined sensation to pin prick and light touch in the thoracic area: T8 on the left, T11 on the right. Laboratory data were normal. Anteroposterior and lateral films of the thoracolumbar spine showed a right thoracic curve from T5 to T12 measuring 44°, which was unchanged from previous films (Fig. 1). The patient was started on oral steroids at the time of the office visit due to myelopathic symptoms.

A CT myelogram and an MRI examination of the cervical and thoracic spine were obtained, which revealed an extradural soft tissue mass within the spinal canal extending from T5 to T9. There was severe spinal cord compression. Other than indolent erosion, there were no abnormalities noted in the vertebral bodies or posterior elements (Figs. 2, 3).

Differential diagnosis for the extradural tumor based on age, location and MRI appearance included epidural lipomatosis, epidural hemangioma and lymphoma. The two other common extradural tumors, neurofibromatosis, a relatively common spinal cord tumor, and extradural meningioma, a relatively uncommon tumor, were excluded by the MR appearance.

The patient was admitted to the hospital and underwent urgent spinal cord decompression. A T5–T9 laminectomy was performed with excision of the tumor. A firm epidural mass was found completely filling the epidural space and compressing the cord. The dura was found to be normal with no evidence of infiltration from the tumor. Feeding and draining vessels were controlled with electrocautery. Spinal fusion was not performed given the limited laminectomy and relative stability of the spine [19]. Both frozen and permanent sections revealed a vascular tumor consistent with a hemangioma in an adipose tissue stroma. The specimen was reddish-brown and measured 5×2×1 cm.

Microscopic examination of the excised tissue showed a lesion composed of blood vessels within a generally mature fatty background stroma (Fig. 4). In some regions, the blood vessels consisted of disorganized and compressed small arterioles and arteries to large tortuous arteries. Cavernous regions were present focally (Fig. 5). Veins of variable diameter were also present in some regions. All the vascular elements were suspended in the adipose tissue with only a small amount of fibrous connective tissue. Cytologic atypia of the vascular endothelial linings was not present. Estrogen receptor sensitivity was not examined on the specimen. On the basis of pathologic analysis, a diagnosis of spinal epidural hemangioma was made.

Postoperatively, the patient's neurologic examination returned to normal. An MRI examination was performed 6 weeks postoperatively,

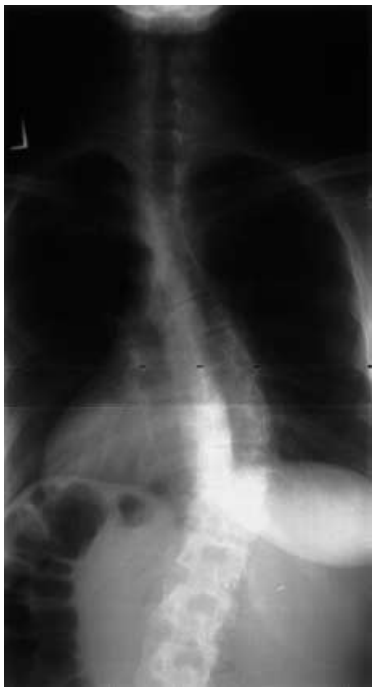

Fig. 1 Posteroanterior view of the spine demonstrates idiopathic dextroscoliosis of 44°

which documented the complete excision of the tumor. Eighteen months after resection of the tumor the patient remained asymptomatic, with no clinical or radiographic evidence of recurrence on MRI. She continued to do well and returned to recreational athletics. Furthermore, there was no progression of her scoliosis on radiographs.

Discussion

Hemangiomas are benign, slow-growing cavernous or capillary tumors [13]. They are characterized histologically by abnormally dilated blood vessels lined by a thin endothelium, closely clustered together. No neural tissue is found between blood vessels in a hemangioma. Elastic fibers and smooth muscle fibers within the walls are scant if not absent [6]. Hemangiomas occur more commonly with increasing age and in women [20]. Epidural hemangiomas represent approximately 4%

Fig. 2 **A** Axial T1-weighted SE sequence (TR/TE=500/14) at the T9–10 level demonstrating the lesion (*arrow*) to be hyperintense compared with the bone, spinal cord and muscle, and only slightly hypointense compared with fat. **B** Sagittal T2-weighted FSE image [TR/TE=2750/76 (Ef.)] shows a heterogeneous lesion, hyperintense to all surrounding structures except cerebrospinal fluid. It is posterior to and compresses the spinal cord in the mid-thoracic region. **C** Axial T2-weighted FSE image [TR/TE=2450/110 (Ef.)] at the T6–7 level shows heterogeneous high signal (*arrow*) signal causing displacement of the spinal cord to the left anterior aspect of the spinal canal. No foci of low signal in the mass are present to suggest calcification. **D** Axial T2-weighted FSE image [TR/TE=2450/110 (Ef.)] at the T9–10 level demonstrating the spinal cord (*straight arrow*) and the dural sac (*curved arrow*) with the right canal high signal lesion at this level. Asymmetry of the cord within the canal is partly related to the patient's underlying scoliosis

Fig. 3 Axial image from CT myelogram at the T7 level demonstrating cord compression by posterior right soft tissue. *V* vertebral body, *straight arrow* spinal cord, *curved arrow* compressed dural sac, *open arrow* extradural mass. There is no bony destruction. There is generalized widening of the canal by the mass from indolent erosion

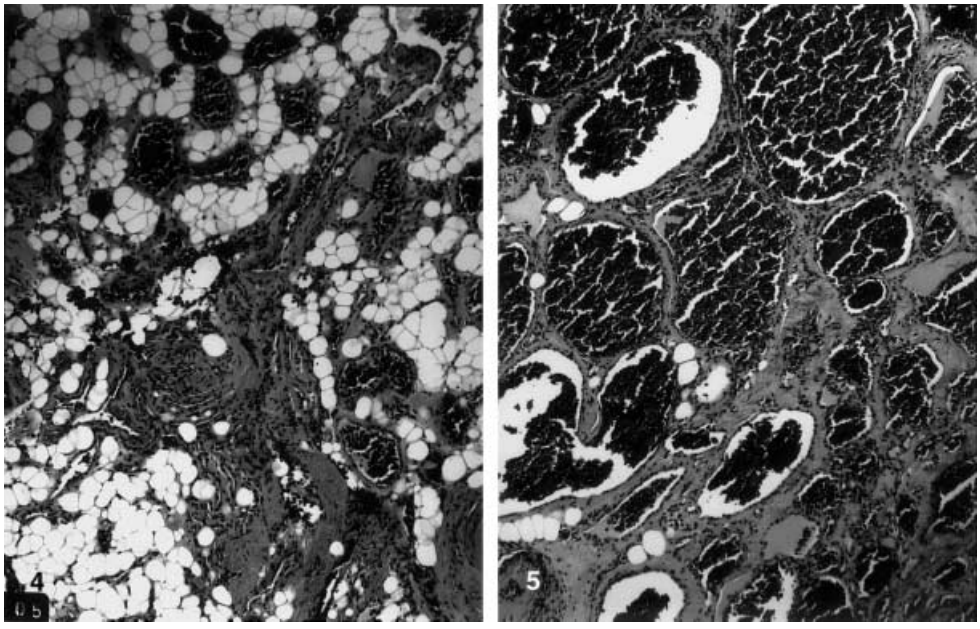

Fig. 4 Photomicrograph showing a mixture of small, capillary-like vessels around an irregular larger vessel with vessels of variable diameter in the surrounding adipose tissue (H&E, ×10)

Fig. 5 Photomicrograph showing greatly dilated blood-filled vascular spaces in a cavernous region (H&E, ×10)

of all epidural tumors and 12% of all intraspinal hemangiomas [5]. Any part of the spine may be affected, but the thoracic spine is the most common symptomatic location [21].

Although histologically and immunohistochemically identical, these vascular anomalies may exhibit different clinical behavior and radiologic features depending on location. Therapy, therefore, is dictated not only by the histologic appearance of the lesion, but also by its location. Treatment options for the asymptomatic patient with a vertebral hemangioma include observation, embolization, radiotherapy, and surgical resection alone or in combination. All modalities are effective but must be tailored to the specific needs and condition of the patient.

As opposed to vertebral lesions, most cases of epidural hemangioma are characterized by the insidious onset of symptoms over several months with recurrent symptomatology and symptom-free intervals. Symptoms may include local pain, tenderness, radiculopathy and myelopathy, and can be aggravated by trauma, exercise, pregnancy and straining, secondary to physiologic or pathologic variations in blood flow [22]. Vascular skin dysplasias, as described by Johnston, may also be present [22].

During pregnancy, two mechanisms may account for the symptomatology. The first is an increase in venous pressure resulting from mechanical obstruction by the pregnant uterus of the blood flow from the paravertebral veins into the inferior vena cava. The second theory is based on the effect of the hormonal alterations associated with pregnancy on a pre-existing vascular lesion. Increased blood volume may lead to distention of the vascular channels in these lesions [8].

Estrogen levels increase markedly during pregnancy and fall abruptly following parturition. They begin to rise once again when the normal menstrual cycle is re-established. In non-nursing mothers, this may be expected to occur 6–8 weeks following delivery. In nursing mothers, this may not occur for 3–5 months. The hormonal action may act directly upon the endothelium of the vascular channels.

Radiographs usually are not diagnostic, but may show evidence of erosion of the pedicles, the laminae, the vertebral bodies, and of enlargement of the intervertebral foramina. Myelograms can show compression, as in this case. Spinal angiography is usually negative, in contrast to vertebral body lesions [16]. MRI is the diagnostic modality of choice.

The signal characteristics of this mass are high, compared with muscle and spinal cord, on T1-weighted and fast spin echo T2-weighted images. This suggests the presence of fat or acute hemorrhage. Rarely, soft tissue tumors, such as melanoma and alveolar soft part sarcoma, can have these characteristics [23, 24, 25]. Focal areas of high signal can be seen with intratumoral hemorrhage [26]. The presence of indolent erosion and widening of the central canal excludes acute hematoma from the differential diagnosis. The heterogeneity of the mass on the T1-weighted images goes against lipomatosis. Also, lipomatosis does not generally cause bony erosion. Although focal calcifications are often seen in hemangiomas, the finding is not always present. Finally, the mass on the CT scan is variable in density, with areas as low as –20 Hounsfield units and as high as 30. This also argues against lipomatosis, which should have homogeneous low density.

Total or partial excision is indicated in symptomatic cases. Graziani et al. [27] reported on 11 cases of symptomatic hemangiomas of the spinal epidural space. Four thoracic cases in their series were all posterior with foraminal extension, with excellent results after surgical excision. Intraoperative findings typically show a posterior dark epidural lesion in fat tissue with foraminal extension easily dissectible from the dura. It is sometimes difficult to differentiate cavernous angiolipomas from hemangiomas because the admixture of fat tissue and vessels can vary. Specimens in acute neurologic deterioration show varying degrees of hemosiderin, indicating an intra- or extralesional hemorrhage. Postoperative radiation therapy is advocated for unresectable lesions, postoperative residuals, and medically unstable patients [28]. Embolization plays a small role in purely epidural lesions, because they are often angiographically silent.

The mixture of the various components, including venous and arterial elements, with a wide range in luminal diameters suggests that the lesion is a hamartomatous process rather than a specific neoplastic vascular lesion. Given the occasional inability to fully characterize the behavior of these and similar lesions, the lesion presented here has been referred to as "arteriovenous hemangioma." The presence of the adipose tissue component in this lesion might lead one mistakenly to the diagnosis of angiolipoma. However, in this case, the adipose tissue component represents the matrix in which the vascular components grow, and by which they are supported.

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