Lobular capillary hemangioma of the cauda equina

Case report

ROBERT N. N. HOLTZMAN, M.D., PAUL M. BRISSON M.D., RICHARD E. PEARL, M.D., AND MICHAEL L. GRUBER, M.D.

Divisions of Neurosurgery, Orthopedics, and Neurology, Beth Israel Medical Center and Cabrini Medical Center, New York, New York

This 56-year-old woman presented with a 1-year history of low-back pain, sciatica, and paresthesias in the right S-1 dermatome. On examination the patient was shown to have a right-sided Lasègue’s sign, normal strength, hypalgesia in the right S-1 dermatome, and a slight diminution of the right Achilles tendon reflex. Magnetic resonance imaging revealed a 2-cm intradural enhancing lesion at the level of the L-4 vertebra. Laminectomy of L3–L5 vertebrae was performed, and intradural exploration disclosed a blueberry-appearing tumor that was surrounded by an intense arachnoiditis and attached to the right S-1 nerve root. A cystic collection of cerebrospinal fluid was seen caudal to the tumor. Complete removal required transection of the adherent nerve root fascicles. Histological analyses indicate that the lesion was a lobular capillary hemangioma, which, to the authors’ knowledge, appears to be one of the first recorded examples of such a case.

KEY WORDS • hemangioma • lobular capillary hemangioma • pyogenic granuloma • cauda equina

Capillary hemangiomas are typically cutaneous or subcutaneous lesions\(^4,10\) that may also arise in mucosal tissues.\(^3\) They most frequently occur during childhood and are characterized by elevated, reddish-purple lesions that are composed of capillary-sized vessels lined by a flattened epithelium. In this context they are commonly referred to as “strawberry nevus” and “cherry angioma.”\(^4\) The term “lobular capillary hemangioma” is applied in a generic sense to a number of hemangiomas, the cytoarchitecture of which is described as “nodules of small capillary sized vessels, each of which is subserved by a ‘feeder’ vessel.”\(^4\) These include capillary hemangioma of infancy, epithelioid hemangioma, and pyogenic granuloma.\(^2\) (The term pyogenic granuloma has been considered by some authors to be a misnomer.\(^6\))

The present case of a lobular capillary hemangioma that is associated with the S-1 root of the cauda equina without dural attachment and that simulated a neurofibroma is distinct, unusual, and appears to be one of the first recorded examples of such a case.

Case Report

History. This 55-year-old woman suffered nocturnal leg cramps for more than 20 years. One year prior to admission, after an emergency requiring that she descend 50 flights of stairs, she developed right-sided sciatica and low-back pain. These symptoms progressively worsened. Lumbar spinal stenosis was diagnosed on computerized tomography scanning. Treatment with epidural steroid injections produced significant relief for several months before the symptoms returned and worsened.

Examination. The patient was moderately obese. No cutaneous lesions were noted. Neurological deficits included hypalgesia in the right S-1 dermatome and a decreased right ankle jerk reflex.

Gadolinium-enhanced magnetic resonance (MR) imaging of the lumbar spine revealed a 2-cm homogeneously enhancing intradural lesion opposite the L-4 vertebral body (Fig. 1).

Operation. Laminectomy of L3–L5 vertebrae was performed, and intradural exploration revealed a blueberry-colored lesion that arose from the roots of the cauda equina in a manner characteristic of a neurofibroma and derived its blood supply from radicular vessels. It was completely covered by matted, adherent nerve roots and ensconced in a bed of dense arachnoidal adhesions. A loculated cyst of arachnoid tissue that contained cerebrospinal fluid was present caudal to the tumor. Complete excision was accomplished by individually separating the matted nerve roots overlying the tumor, cautery shrinkage without violation of the capsule, microdissection of the surrounding arachnoidal adhesions, and transection of several densely adherent nerve trunks and fascicles.
Postoperative Course. The patient’s postoperative recovery was uneventful. Neurological examination immediately after surgery and at 6 months disclosed a residual right S-1 radiculopathy from which the patient rapidly recovered such that she was able to stand on the toes of both feet together but only briefly on the tiptoes of the right foot. She was ambulating independently, with the intermittent assistance of a cane, and returned to work at 2 months. Postoperative MR imaging of the brain and the cervical, thoracic, and lumbar spine revealed no residual tumor at the operative site and no other tumors.

Pathological Examination. Grossly, the tumor consisted of a round, well-circumscribed rubbery tan-red nodule that measured 1.9 \times 1.5 \times 1.1 \text{ cm}. Three white nerve bundles were present on its surface. Serial sectioning revealed a variegated tan-red glossy multiloculated surface.

Microscopically the tumor was characterized by a well-circumscribed nodule that was partially surrounded by nerve trunks (Fig. 2 left). It was divided into lobules that were separated by a myxoid, occasionally hyalinized, stroma. The lobules were composed of tightly packed blood-filled, variably dilated, well-formed capillaries that were lined by plump but bland-appearing endothelial cells. Larger feeding and draining vessels were present. Stromal cells were not seen (Fig. 2 right). Histologically the tumor was characteristic of lobular capillary hemangioma as seen in other sites.

Discussion

Lobular capillary hemangiomas are characterized by the presence of lobules of normal, capillary-sized channels that are tightly aggregated into nodules, each of which is nur-
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ished by a feeding vessel. They are commonly seen in cutaneous and subcutaneous locations but have been rarely encountered in the neuraxis. Reports of lobular capillary hemangiomas attached to the dura, peripheral nerves, muscles, skin, and mucous membranes are cited, but no examples of attachment to the roots of the cauda equina have been reported. It remains possible that similar lesions have been reported using other terms such as “benign hemangioendotheliomas,” however, more recent classification suggests that all hemangioendotheliomas should be considered tumors of intermediate malignancy.

Other vascular lesions such as cavernous angiomas within the spinal cord parenchyma and cauda equina are distinct from lobular capillary hemangioma both clinically in their propensity for hemorrhage and cytoarchitecturally in their microscopic appearance of having larger, dilated vascular channels arranged in diffuse and haphazard patterns with inflammatory cells scattered throughout the stroma. Hemangioblastomas are distinguished by the presence of stromal cells and by their largely intramedullary and parenchymal sites of origin.

The tumor’s origin in the cauda equina in the present case raises the possibility of a congenital formation or the spontaneous development of a vascular tumor in relation to capillary structures along or within the cauda equina roots. It is presumed that the nerve root capillary is the site of origin of this lesion and that the adherence of the other roots was a reflection of its associated arachnoiditis. The intense arachnoiditis that was noted to surround the tumor may have resulted from minute bleeding or the diapedesis of erythrocytes. It also contributed to the formation of the caudally placed loculation of cerebrospinal fluid.

The absence of any tumors elsewhere in the neuraxis in our patient and the tumor’s benign pathological appearance, combined with the knowledge that these tumors are unlikely to bleed spontaneously, suggest that the prognosis for the patient who undergoes surgical removal is good.

However, reports of the spontaneous eruption of lobular capillary hemangiomas with widespread dissemination on the skin as well as the appearance of multiple skin satellite lesions after surgical removal of a solitary lobular capillary hemangioma indicate that close postoperative surveillance of patients with these tumors in the neuraxis is necessary.

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References


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