Spinal cord compression due to Masson’s vegetant intravascular hemangioendothelioma

Case report

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The authors present the case of spinal cord compression in a 16-year-old boy due to the rare vascular lesion, Masson’s vegetant hemangioendothelioma.

KEY WORDS  • spinal cord compression  • Masson’s tumor  •  hemangioendothelioma  • differential diagnosis

In 1923 Pierre Masson described a tumorlike papillary proliferation of the endothelium within an infected hemorrhoidal vein. Since then this unusual lesion has been found in a variety of other locations, and its benign nature has been well established. Only a handful of the reported cases have involved the neuraxis,1,5,11,14 and in all of these instances the tumor was found in the intracranial compartment. Here we present what we believe to be the first report of spinal cord compression due to Masson’s vegetant hemangioendothelioma.

Case Report

This 16-year-old Caucasian boy presented with a 1-month history of progressive weakness in both legs and a 1-week history of midthoracic back pain with radicular radiation that was severe enough to wake him at night. These symptoms had become rapidly worse in the few days prior to presentation, and he had experienced some hesitancy of micturition for 24 hours. He had previously been healthy and had not suffered any injury to his back. The family history was unremarkable, and he had never been exposed to carcinogens or ionizing radiation.

Examination. On general examination, the skin, mucous membranes, and cardiovascular, respiratory, and gastrointestinal systems were entirely normal. Neurological examination revealed intact cranial nerves and arm function. In the legs there was a symmetrical spastic increase in tone, Grade 4/5 weakness in an upper motor neuron pattern, and hyperreflexia with bilateral extensor plantar responses. There was a partial sensory level response to pinprick at T-8 and a moderate reduction in joint position sense in the toes. Anal sphincter tone was reduced on digital examination. Routine blood tests and chest x-ray film results were normal. A computerized tomographic myelogram demonstrated marked cord compression at the T-6 level due to a posterior extradural mass with an abnormal texture to the overlying lamina (Fig. 1). The cerebrospinal fluid had an elevated protein content of 0.8 mg/L but was otherwise normal. Magnetic resonance imaging was not available at the time, and in light of the patient’s rapid deterioration, he was immediately taken to surgery.

Operation. At operation, the T-6 lamina was found to be abnormal, with large vascular spaces on its deep surface. The tumor lay within attenuated extradural fat and com-
prised a red–black nodular mass associated with dilated extradural veins. It could be separated cleanly from the dura throughout, and a complete resection was achieved with sacrifice of part of the right T5–6 facet joint.

Pathological Examination. The surgical specimen measured $4 \times 2 \times 1$ cm. Microscopically, it consisted of organized thrombus separated into numerous pseudovascular channels that were lined by a single layer of endothelium. Papillary projections were seen on the luminal side, and there were no mitoses or other evidence of malignancy (Figs. 2 and 3). Residual elastic fibers were demonstrated with a van Gieson stain in the main vessel walls. The appearance was thus typical of Masson’s vegetant hemangioendothelioma.

Postoperative Course. The patient’s leg power and bladder function improved markedly within 24 hours, and he was discharged after 10 days, at which time he could walk with a cane and was fully continent. At 6-month follow-up examination, he was entirely asymptomatic and had returned to playing football.

Discussion

Masson’s vegetant hemangioendothelioma, sometimes also known as “intravascular papillary endothelial hyperplasia,” is an unusual benign growth. Since it was first described, the lesion has been found in a variety of locations, including the skin and subcutaneous tissues, muscle, pelvis, liver, pharynx, and large neck veins. The age distribution for Masson’s vegetant hemangioendothelioma is wide, with a slight female preponderance noted in most series. Its importance lies mainly in the fact that the histological appearance may be mistaken for that of a malignant vascular tumor such as angiosarcoma or Kaposi’s sarcoma. This distinction must be made to avoid unduly aggressive treatment, because Masson’s vegetant hemangioendothelioma is invariably cured by adequate resection.

Historically, the lesion is confined to the vascular lumen. It consists of blood spaces and thrombotic material that are fragmented by florid papillary projections of fibrous tissue-bearing hyperplastic endothelial cells. Features that might suggest malignancy such as solid cellular areas, necrosis, mitotic figures, pleomorphism, endothelial layering, and an inflammatory cellular infiltrate, are all uncommon.

Masson’s vegetant hemangioendothelioma may arise apparently de novo or within an identified preexisting vascular abnormality such as a hemangioma, pyogenic granuloma, or cavernous angioma. Its lymphatic counterpart has been described within cystic hygroma. The question of pathogenesis remains unanswered. Masson and several subsequent researchers have suggested that it is a benign but truly neoplastic process of the endothelium. However, similar histological appearances may be found within ordinary venous thrombi or arterial thromboemboli and so other researchers prefer to regard it as an unusually exuberant form of the normal granulation response within thrombus. This theory does not discount its ability to
form an expansive and compressive mass as the initial growth could cause vascular occlusion, leading in turn to stasis, further thrombosis, and recruitment of contiguous vessels. There has been no suggestion that Masson’s vegetant hemangioendothelioma is associated with the various congenital and acquired causes of blood hypercoagulability, and our patient had no known risk factors for thrombosis.

Instances of Masson’s vegetant hemangioendothelioma that involved the neuraxis as noted in previously reported cases have all been present in the intracranial compartment. These cases presented variously as a giant tumor in an infant, as an intracerebral hemorrhage from a presumed cavernous hemangioma containing focal Masson’s vegetant hemangioendothelioma change, with raised intracranial pressure due to venous sinus obstruction, and with epilepsy in a case of multiple neurocutaneous Masson’s vegetant hemangioendothelioma lesions. Ours is the first reported case to present with spinal cord compression, and we believe that this lesion had probably arisen within an extradural cavernous hemangioma. There was nothing remarkable about our patient’s clinical course and the lesion’s appearance on computerized tomographic myelogram was not unusual. However, our patient made an excellent recovery after a complete resection and the many reports of successful surgical treatment of Masson’s vegetant hemangioendothelioma at other sites would therefore suggest that his prognosis is favorable. Spinal Masson’s vegetant hemangioendothelioma is clearly very unusual, but we would recommend that the diagnosis be considered in cases of spinal cord compression due to vascular tumors to prevent needless adjuvant therapy.

References


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