CASE REPORT
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Solitary Hemangioblastoma at the Filum Terminale: A Case Report and Review of Literature

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Spinal hemangioblastomas at the filum terminale are rare. Only a few reports present immunohistopathological features of spinal hemangioblastomas. A 55-year-old woman presented with sustained low back pain and intermittent numbness in both lower extremities. She already had a decompressive surgery for a left L5-S1 extraforaminal lesion 7 months ago. Her incidental mass lesion had been slightly enlarged for 7 months and concordant symptoms had also been prolonged. Her tumor was removed en bloc with a unilateral hemilaminotomy and bilateral decompression technique. The patient's symptoms improved immediately after the surgery. In immunohistochemical stains, the stromal cells were positive for inhibin-alpha, neuron specific enolase (NSE), and the endothelial cells were positive for CD31, CD34, and factor VIII related antigen in confirming the diagnosis of hemangioblastoma. We report a rare case of intradural extramedullary spinal hemangioblastoma at the filum terminale. Complete en bloc resection was achieved with no morbidity. The immunohistopathological staining include inhibin-alpha confirmed an accurate diagnosis of the hemangioblastoma distinguishing from other vascular tumors.

Key Words: Spinal hemangioblastoma · Filum terminale · Immunohistopathological stain

INTRODUCTION

Tumors located around filum terminale are rare. Tumors commonly found in this area include neurogenic tumors such as ependymomas and schwannomas. Spinal hemangioblastomas are rare tumors comprising 1.6 to 2.1% of all spinal cord tumors[2,13]. The most reported hemangioblastomas present in spinal nerve roots are associated with von Hippel-Lindau (VHL) syndrome[8]. Hemangioblastomas located at the filum terminale intradurally and extramedullarily, with no related condition or VHL syndrome, are even rarer. These tumors' morphologic and radiologic features are confusing and specified immunohistopathological stains are essential to confirm the diagnosis. In this report, we present a rare case of a hemangioblastoma located at the filum terminale, which was diagnosed after immunohistopathological stains.

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CASE REPORT

A 55-year-old woman presented with sustained low back pain and intermittent numbness in both lower extremities for about a year. She already had undergone on a decompressive surgery on the left L5-S1 extraforaminal disc 7 months before. She had been suffering from a gradual worsening of low back pain, which radiated to her left lower extremity, for 6 months at her initial presentation. The neurologic examination at her first hospital visit revealed symptoms and signs that were caused by her left L5-S1 extraforaminal disc. Motor and sensory distributions were intact and we identified no pathologic reflex. Deep tendon reflexes in both knees and ankles were normal.

Lumbosacral magnetic resonance images (MRI) showed a left L5-S1 extraforaminal disc with a small incidental intradural mass at the L3 level. T1-weighted enhanced MRI demonstrated a homogenous enhancement of the mass (Fig. 1). We regarded the mass as an incidental asymptomatic neurogenic tumor, so we performed the decompressive surgery only on the left L5-S1 extraforaminal lesion. The left lower extremity symptoms improved completely after the surgery. However, her low back pain still remained and was slightly aggravated for 7 months after the surgery. We performed follow-up enhan-

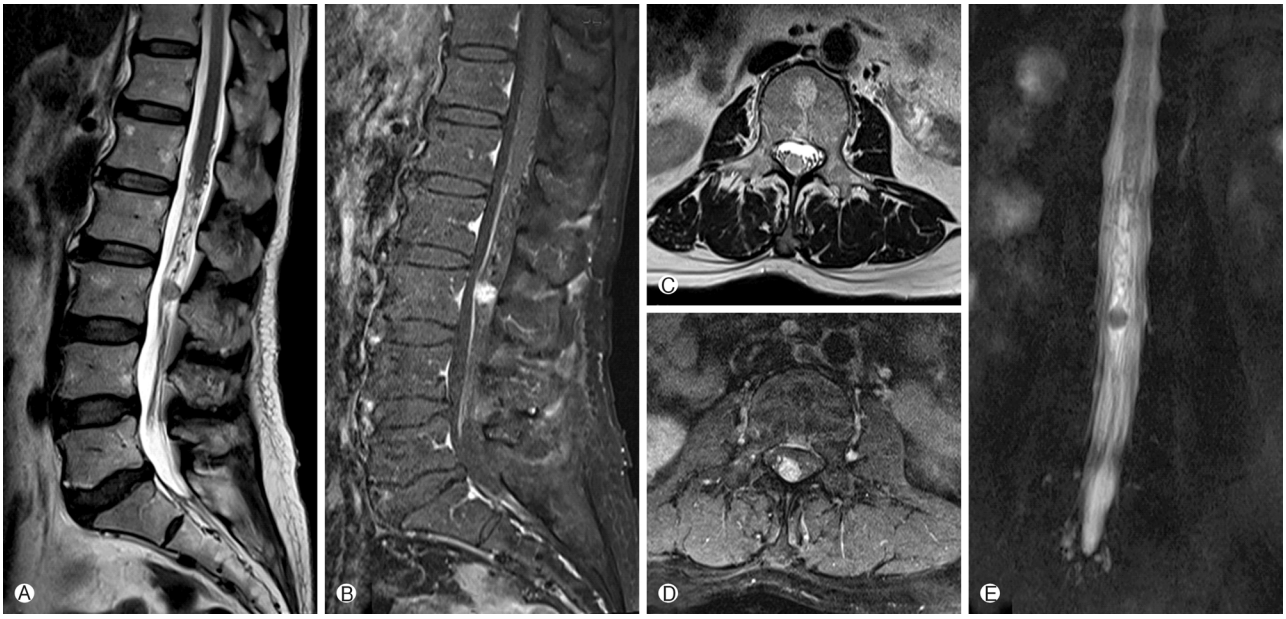

Fig. 1. T2-weighted sagittal (**A**) and axial (**C**), and T1-weighted gadolinium-enhanced sagittal (**B**) and axial (**D**), images show a slightly enlarged intradural mass with engorged vessels above the lesion. MR myelogram (**E**) presents a filling defect at the L3 level with a mass shadow.

ced lumbosacral MRI, which revealed that the mass had grown slightly. An MR myelogram showed a filling defect at the L3 level with a mass shadow (Fig. 1). The mass was located in the posterior two-thirds of the spinal canal and the nerve rootlets were displaced anteriorly. We noticed engorged intradural vessels above the mass, which were regarded retrospectively as a clue for a diagnosis of hemangioblastoma. An additional neurologic examination, family history, laboratory findings, and radiologic studies demonstrated no sign of VHL syndrome.

The preoperative diagnosis was a neurogenic tumor that had arisen from the spinal rootlets, such as a schwannoma or an ependymoma. We performed a hemilaminotomy on the right L3 and a partial hemilaminotomy on the right L2 and L4. We exposed the thecal sac's midline by undercutting the spinous processes, like the unilateral laminotomy with bilateral decompression (ULBD) technique. We performed a midline myelotomy and exposed the dark red colored tumoral mass, which was smooth and lobulated with dilated vessels on its surface and was attached firmly to three rootlets. We carefully dissected these adhesions and removed the mass en bloc. We encountered abundant bleeding at the mass' surface, so we had to meticulously control the bleeding during the tumor dissection.

Postoperatively the patient was free of pain and showed no neurological deficits, except for the slight numbness in both lower extremities that subsided after two hours. Postoperative MRI showed complete evacuation of the tumor mass.

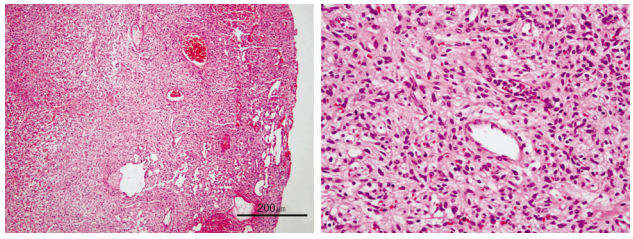

Fig. 2. Histopathology of the tumoral mass with HE staining x 100 (left) and x 400 (right); note the fine vascular capsule and the rich capillary network. Prominent capillary vasculatures and stromal cells with clear cytoplasm were apparently presented.

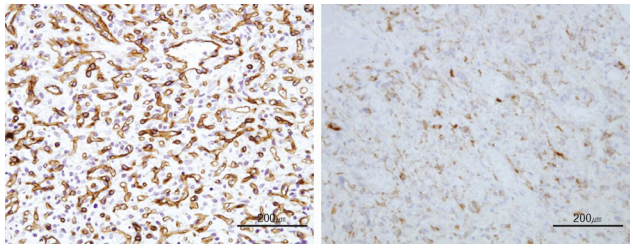

Fig. 3. Specified immunohistopathological staining of the tumor. The tumor was immunopositive for CD34 (left) in endothelial cells, and inhibin-alpha (right) in stromal cells.

The first histopathological diagnosis was a capillary hemangioma. There were prominent capillary vasculatures and stromal cells with clear cytoplasm after hematoxylin and eosin (HE) staining (Fig. 2). However, microscopically the tumor

was composed of two components such as prominent small blood vessels and stromal cells. The immunohistochemical stain showed positive for CD31, CD34, and Factor VIII related antigen in endothelial cells, and for inhibin-alpha, NSE, and S-100 protein in stromal cells. Inhibin-alpha is helpful for differentiating hemangioblastoma and other vascular tumors such as capillary hemangioma (Fig. 3).

DISCUSSION

The medical literatures show that hemangioblastomas comprise between 1.6% and 2.1% of all spinal cord tumors[2,13]. Reported incidences show that intradural extramedullary hemangioblastomas represent about one-fifth of all spinal hemangioblastomas. The affected level varies from cervical to cauda equine, and all spinal levels can be involved with these tumors[1,2]. About 30% of reported cases are associated with VHL syndrome. However, sporadic hemangioblastomas can be found with no association with VHL syndrome, and the exact incidence has not been clarified. Recent studies of VHL syndrome involving the central nervous system suggest that patients with hemangioblastomas associated with VHL syndrome are usually younger, have more multiple hemangioblastomas, and are more commonly affected in their cerebellum than patients with sporadic hemangioblastomas[3,6]. Undoubtedly, a sporadic hemangioblastoma at the filum terminale is a very rare condition. Ortega et al. documented from a literature review that only nine cases of hemangioblastomas arising in the filum terminale were reported in 2007[7]. None of the patients in the literature, including ours, with a hemangioblastoma at the filum terminale had any association with VHL syndrome. It seems that a solitary filum terminale hemangioblastoma is only a sufficient, but not a necessary, condition for VHL syndrome.

The origin of extramedullary hemangioblastomas has not been clarified. Hemangioblastomas commonly arise from the central nervous system's medullary tissues and develop into the marrow cells as intramedullary tumors. A recent biomechanical study suggests a clue for the origin of extramedullary hemangioblastomas[4]. Extramedullarily secreted vascular endothelial growth factor (VEGF) from stromal cells might play an important role in endothelial cell proliferation. Some kind of genetic mutation or alteration, like that exhibited in VHL syndrome, might be involved in this process; however, genetic studies need to clarify the origin of extramedullary hemangioblastomas.

Radiologic findings of filum terminale hemangioblastomas are often confusing. Patterns of tumoral enhancement are not unique or different from those of other neurogenic tumors, and tortuous vessels around the mass are noticeable in other tumors because of the venous engorgement that results from the space the mass occupies in the spinal canal.

Several authors have reported that the spinal angiography and preoperative embolization of tumors can help to diagnose them accurately and resect them effectively[5,7]. Preoperative embolization might help reducing intraoperative bleeding; however, routine conventional angiography is an invasive procedure and preoperative embolization seems to be unnecessary except for the large tumors in a critical location. The most reported cases prove that a hemangioblastoma can be diagnosed accurately by preoperative CT and MRI alone[9,12]. Moreover, an exact diagnosis of hemangioblastoma should be based on histopathological, rather than radiological, findings. Two major components of hemangioblastomas are stromal cells and capillary endothelial cells. The differences between these components are significantly identified with an antigen expression pattern. Stromal cells usually have patterns that lack antigen expression, including endothelium-associated factors such as CD31 and CD34. Most cases reported in the literature suggest a diagnosis of hemangioblastoma made only from common staining, such as hematoxylin and eosin (HE) and periodic acid Schiff (PAS) staining. Only a case that Taniguchi S et al[11]. recently reported presents the specified immunohistopathological features of a spinal hemangioblastoma.

Microsurgical en bloc resection is a standard treatment method with low morbidity and good functional outcomes[10]. Debulking surgery of tumoral masses is not indicated in the most cases because hemangioblastomas usually have rich vascularity and debulking surgery is associated with abundant bleeding[1,5,10,12,13]. Instead, surgeons should circumferentially dissect tumoral masses with adhesive rootlets and remove them en bloc. Incomplete resection is strongly associated with a high rate of recurrence[2,13]. Our patient's tumoral mass was small and located slightly on the right side, leaving enough space for en bloc removal with the ULBD technique to avoid wide laminectomy and eliminating the need for an additional instrumentation.

CONCLUSION

Isolated spinal hemangioblastomas at the filum terminale are rare. Physiological and neurological examinations of our patient revealed no association with VHL syndrome. These tumors' morphologic and radiologic features are confusing and additional immunohistopathological stains are mandatory for accurate diagnosis. Surgeons can achieve complete microsurgical en bloc resection with no morbidity. Preoperative angiography and tumor embolization are optional for diagnosing

and treating spinal hemangioblastomas.

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