

Case Report

Sporadic hemangioblastoma of the film terminale with peritumoral cyst

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Abstract

Background: Spinal hemangioblastoma originating from the film terminale are rare tumors. Here, we present a film terminale hemangioblastoma and review the appropriate literature.

Case Description: A 37-year-old female presented with bilateral lower extremity pain without a focal neurological deficit. The magnetic resonance (MR) image demonstrated an intradural spinal tumor at the L1 level, which was accompanied by peritumoral cysts. In addition, there were multiple serpentine flow voids (e.g., consistent with torturous and convoluted vessels), which is typical for hemangioblastoma. At surgery, a spinal hemangioblastoma originating from the film terminale with peritumoral cysts at the L1 level was fully excised without producing a focal postoperative neurological deficit. Histological examination revealed stromal cells with vacuolated cytoplasm and small nuclei in a rich capillary network accompanied by several enlarged vessels. These findings were compatible with a hemangioblastoma.

Conclusions: We reported a rare case of a hemangioblastoma originating from the conus presenting at the L1 level. Complete surgical resection was accomplished without any motor deficit.

Key Words: Film terminale, hemangioblastoma, indocyanine green videoangiography, peritumoral cyst, surgical resection

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INTRODUCTION

Spinal hemangioblastomas are rare tumors that account for approximately 3% of all intramedullary spinal tumors. They are often associated with von Hippel Lindau (vHL) disease. Spinal hemangioblastomas predominantly occur in the cervical and thoracic cord^[10] and are rarely found at the level of the conus.^[1-4,6,7,9] Here, we report a very rare sporadic hemangioblastoma originating from the film terminale at the L1 level, which readily diagnosed on magnetic resonance (MR), and fully excised without any postoperative neurological deficit and sequelae.

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CASE REPORT

A 37-year-old female presented with bilateral lower extremity night pain for 2 months without any focal neurological deficits, including normal sphincter function. She had no family history of central nervous system (CNS) tumors, and screening for vHL disease was negative. The MR imaging demonstrated an intradural spinal tumor at the level of the conus (L1). The tumor was 8 × 6 mm and isointense on both T1-weighted (T1) and T2-weighted (T2) MR images [Figure 1]. The T2 MR study showed a clear margin of the tumor and demonstrated both cranial and caudal peritumoral cysts without syrinx formation [Figure 2]. Serpentine flow voids of torturous and convoluted vessels were seen along the ventral aspect of the spinal cord. The edge of the conus medullaris showed deformation. Gadolinium-enhanced T1-weighted MR revealed homogeneously enhanced and lobulated tumor [Figure 3]. Preoperatively, a selective spinal angiogram confirmed the high vascularity of the tumor with a predominant supply from anterior spinal artery (e.g., arising from left T10 artery) [Figure 4].

Surgical excision of hemangioblastoma of the conus.

The patient underwent an L1 to L3 laminectomy. Under the operative microscope, the dura and arachnoid were opened, revealing a reddish tumor originating from the conus accompanied by cranial and caudal peritumoral cysts [Figure 5a and b]. The tumor measured 8 mm in diameter, and was soft, and well-circumscribed. Intraoperative indocyanine green (ICG) videoangiography showed the tortuous feeding arteries from the both poles and the draining vein along the film terminale [Figure 6]. The feeding arteries were coagulated and resected. An en bloc resection of the tumor including the part of the film terminale and enlarged veins was performed under the operating microscope without any significant changes on intraoperative monitoring (e.g., motor evoked potential and sensory evoked potentials were stable during surgery).

Histology

The histological examination of the tumor was consistent with a hemangioblastoma; it revealed stromal cells with vacuolated cytoplasm and small nuclei in a rich capillary network with several enlarged vessels; there were no mitotic elements [Figure 7]. The patient was discharged 1 week after the surgery, walking unassisted. She did have a sensory deficit around the anus which resolved within 1 month after surgery. She returned to her work at a nursery 2 months later.

DISCUSSION

There are only 11 case reports of conus hemangioblastomas in the literature.^[1-4,6,7,9] They develop as a subpial tumor of the spinal cord and are almost always associated with



Figure 1: T1-weighted (a) and T2-weighted (b) sagittal magnetic resonance images. An intradural spinal tumor existed at the level of L1 vertebral body. The tumor appeared isointense on both T1-weighted and T2-weighted MR images. Peritumoral cyst was seen at the cranial and caudal sides

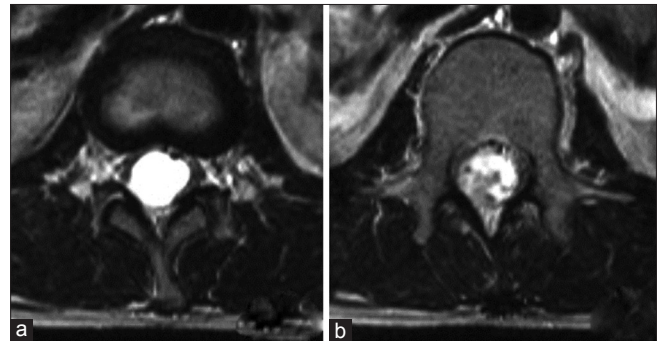


Figure 2: T2-weighted axial magnetic resonance images, T12-L1 intervertebral disc level (a) and L1 vertebral body level (b). Peritumoral cyst was evident

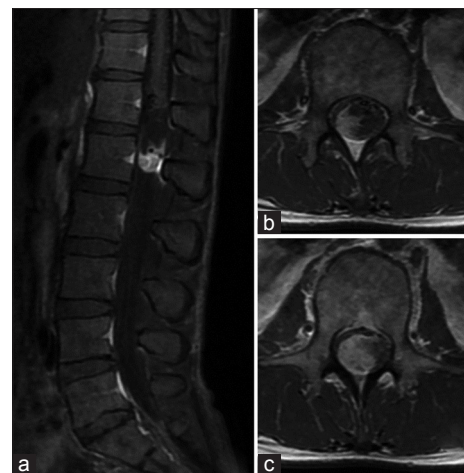


Figure 3: T1 image with gadolinium MR image, sagittal view (a) and axial views T12-L1 intervertebral disc level (b) and L1 vertebral body level (c). Homogeneously enhanced and lobulated tumor was clearly seen

a syrinx. Peritumoral cysts, as noted in this case cranial and caudal spread, are rare. More likely these tumors

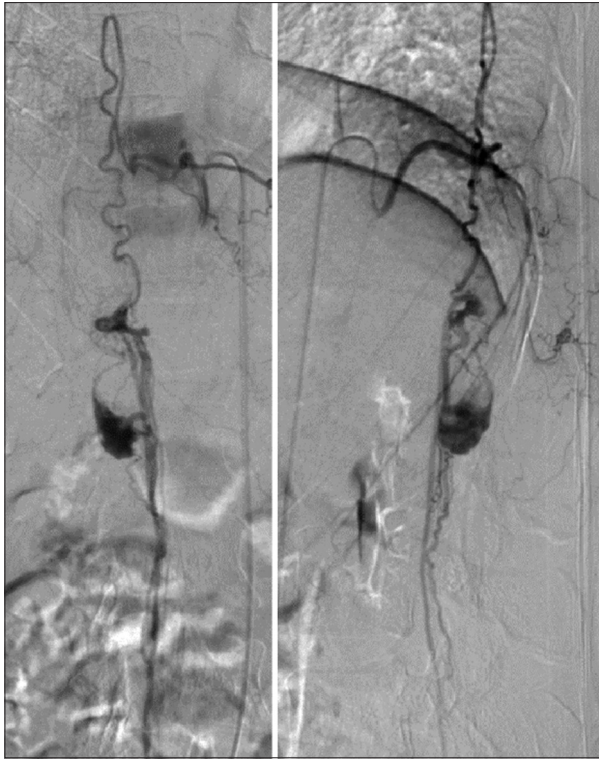


Figure 4: Preoperative selective spinal angiogram from left T10 artery. Abnormal vessels with tumor stain was observed

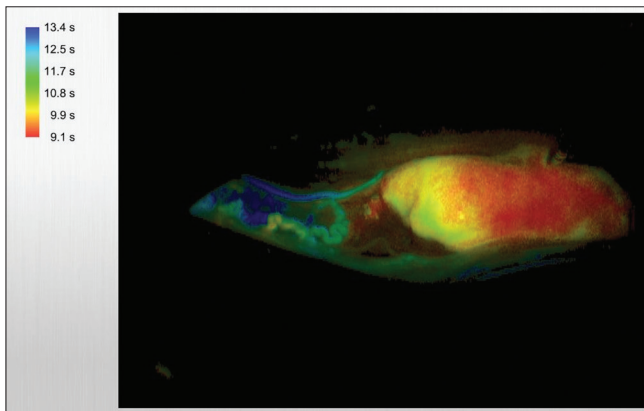


Figure 6: Intraoperative indocyanine green (ICG) videoangiography showing the tortuous feeding arteries from the both poles and the draining vein along the film terminale

are schwannomas (neurinoma) or ependymomas. In this case, we suspected a hemangioblastoma preoperatively as the MR clearly showed serpentine flow voids of tortuous and convoluted vessels cranial to the tumor.

Spinal angiography/embolization for hemangioblastomas

Spinal angiography is useful for detecting the location and vascularity of spinal hemangioblastomas tumor. In this case, it documented the hypervascularity of the lesion. It was also helpful in planning the surgical resection. Biondi *et al.* performed preoperative embolization in 4 cases

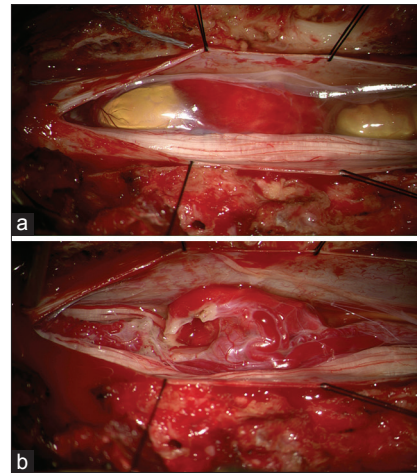


Figure 5: (a) Intraoperative view when opening the dura matter. A reddish tumor associated with peritumoral cysts at both cranial and caudal sides was found. (b) The tumor arose from the film terminale

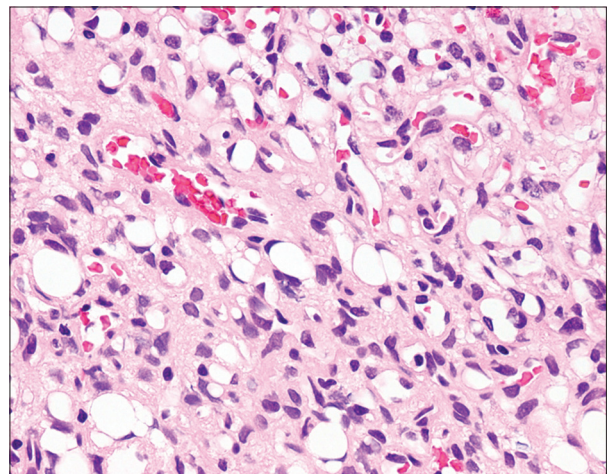


Figure 7: Histopathological findings of the tumor (HE stain). The tumor was composed of vacuolated stromal cells and small nucleus in a rich capillary network with several enlarged vessels

with hemangioblastoma of the lower spinal region and determined it to be useful.^[1] Alternatively, Saliou *et al.* reported 1 of 7 cases with hemangioblastoma associated with vHL disease who had a significant complication attributed to preoperative embolization with the patient ending up with a cerebellar syndrome/gait disturbance.^[8] They determined that preoperative embolization was recommended only in selected cases of hemangioblastomas. In the present case, the patient did not undergo preoperative embolization due to the high risk for the migration of embolic material into the anterior spinal artery.

Pathology/surgery for hemangioblastomas

Hemangioblastomas are benign tumors that should undergo partial/complete resection, as dictated by their location. The use of ICG videoangiography effectively identifies the feeding and draining vessels, facilitating tumor resection.^[5] Previous reports of resection of hemangioblastomas were favorable resulting in no motor

deficits. In this case, intraoperative neuromonitoring was stable and the patient's only deficit was a slight transient sensory loss around the anus.

CONCLUSION

We presented a rare hemangioblastoma arising from the film terminale associated with cepahald/caudad peritumoral cysts. The preoperative routine/enhanced MR readily documented serpentine flow voids of torturous/convoluted vessels characteristic for hemangioblastoma. Surgical resection was accomplished without preoperative embolization, and the patient exhibited no permanent neurological deficit.

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Conflicts of interest

There are no conflicts of interest.

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