Case Report

Sporadic hemangioblastoma of the film terminale with peritumoral cyst

Miho Zaimoku, Yoshiharu Kawaguchi, Shoji Seki, Yasuhito Yahara, Naoki Akioka¹, Johji Imura², Tomoatsu Kimura, Satoshi Kuroda¹

Department of Orthopaedic Surgery, ¹Neurosurgery and ²Diagnostic Pathology, University of Toyama, Toyama, Japan

E-mail: Miho Zaimoku - m.zaiki0309@gmail.com; *Yoshiharu Kawaguchi - zenji@u-toyama.ac.jp; Shoji Seki - seki@med.u-toyama.ac.jp; Yasuhito Yahara - yasuhito.yahara@duke.edu; Naoki Akioka - akioka@med.u-toyama.ac.jp; Johji Imura - imura@med.u-toyama.ac.jp; Tomoatsu Kimura - tkimura@med.u-toyama.ac.jp; Satoshi Kuroda - skuroda@med.u-toyama.ac.jp

*Corresponding author

Received: 20 June 17  Accepted: 06 July 17  Published: 10 January 18

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 finding 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

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 cord¹⁰ and are rarely found at the level of the conus.¹¹ 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.
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 tortuous 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.\(^ {1,2,4,6,7,9}\) They develop as a subpial tumor of the spinal cord and are almost always associated with a syrinx. Peritumoral cysts, as noted in this case cranial and caudal spread, are rare. More likely these tumors...
are schwannomas (neurinoma) or edenidymomas. 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 with hemangioblastoma of the lower spinal region and determined it to be useful. 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. 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. 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 surpentine flow voids of torturous/convoluted vessels characteristic for hemangioblastoma. Surgical resection was accomplished without preoperative embolization, and the patient exhibited no permanent neurological deficit.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Biondi A, Ricciardi GK, Faillot T, Capelle L, Van Effenterre R, Chiras J. Hemangioblastomas of the lower spinal region: Report of four cases with preoperative embolization and review of the literature. AJNR Am J Neuroradiol 2005;26:936-45.
2. Ciappetta P, Occhiogrosso G, Domeniciucci M, D’Andrea G, Bastianello S, Frati A. Hemangioblastoma of the film terminale. Case report and review of the literature. J Exp Clin Cancer Res 2007;26:281-5.
3. da Costa LB Jr, de Andrade A, Braga BP, Ribeiro CA. Cauda equina hemangioblastoma: Case report. Arq Neuropsiquiatr 2003;61:456-8.
4. Escott Ej, Kleinschmidt-DeMasters BK, Brega K, Lillehei KO. Proximal nerve root spinal hemangioblastomas: Presentation of three cases, MR appearance, and literature review. Surg Neurol 2004;61:262-73.
5. Hao S, Li D, Ma G, Yang J, Wang G. Application of intraoperative indocyanine green videoangiography for resection of spinal cord hemangioblastoma: Advantages and limitations. J Clin Neurosci 2013;20:1269-75.
6. Kunihiro N, Takami T, Yamagata T, Tsuyuguchi N, Ohata K. Spinal hemangioblastoma of cauda equina origin not associated with von Hippel-Lindau syndrome-case report. Neurol Med Chir (Tokyo) 2011;51:732-5.
7. Nadkarni TD, Menon RK, Desai KL, Goel A. Hemangioblastoma of the film terminale. J Clin Neurosci 2006;13:285-8.
8. Saliou G, Giammattei L, Ozanne A, Messerer M. Role of preoperative embolization of intramedullary hemangioblastoma. Neurochirurgie 2016 [Epub ahead of print].
9. Sergides IG, Wainwright KL, Biggs M. Incidental hemangioblastoma of the film terminale. Acta Neurol Belg 2009;109:55-6.
10. Siller S, Szelenyi A, Herlitzi L, Tonn JC, Zausinger S. Spinal cord hemangioblastomas: Significance of intraoperative neurophysiological monitoring for resection and long-term outcome. J Neurosurg Spine 2017;26:483-93.