CASE REPORT

Hemangioblastoma of the Cauda Equina : A Case Report and Review of the Literature

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Abstract : Introduction : Hemangioblastoma in the spine mainly occurs at the cervical and thoracic levels and is often associated with von Hippel-Lindau (VHL) syndrome. Here, we reported a quite rare case of spinal sporadic hemangioblastoma arising from the cauda equina. Case presentation : A 66-year-old woman presented with a 5-year history of low back and leg pain. Imaging revealed a hypervascular intradural extramedullary tumor in the lumbar region. Preoperative angiography helped to identify the feeding arteries and draining vein, and so facilitated subsequent tumor resection. The pain was dramatically improved but weakness of the left tibialis anterior and left extensor hallucis longus muscles persisted. Discussion : We reported a rare case of spinal hemangioblastoma arising from the cauda equina. Preoperative angiography may be useful for diagnosis and understanding of the anatomy of feeding veins. J. Med. Invest. 69 : 312-315, August, 2022

Keywords **:** *hemangioblastoma, cauda equina, lumbar spine*

INTRODUCTION

Hemangioblastoma is a slow-growing benign tumor that develops in adults, typically occurring in the brainstem, cerebellum, or spinal cord (1). Approximately 70% of all hemangioblastomas are sporadic and the remaining 30% occur in association with the inherited disorder von Hippel-Lindau (VHL) syndrome (1).

In the spinal cord, hemangioblastomas in the lumbosacral region without VHL syndrome are relatively rare. Here, we report on a case of intradural extramedullary hemangioblastoma in the lumbar spinal canal not associated with VHL syndrome. Control of intraoperative bleeding during surgery control is often a major challenge because hemangioblastomas are highly vascular capillary-rich tumors and differentiation of vessels and nerves is sometimes difficult. In this case, we used preoperative angiography to identify feeding arteries, draining veins, and nerve supply and successfully resect the tumor.

CASE PRESENTATION

A 66-year-old woman presented to our institution with a 5-year history of low back pain and pain in the left lower limb. Symptoms deteriorated after she carried out agricultural work 1 month prior to presentation. Conservative treatment including non-steroidal anti-inflammatory drugs was not effective in alleviating the pain. Magnetic resonance imaging (MRI) showed a tumor mass in the cauda equina, so she was referred to our department.

Neurological examination revealed no motor weakness. The straight leg raising test was positive and hypalgesia was observed over L5 and S1 sensory dermatomes on the left side. She had neither intermittent claudication nor bladder and bowel dysfunction. MRI revealed an intradural extramedullary spinal mass at L3 (Figure 1). On T1-weighted image (T1WI), the mass appeared as an iso signal intensity area compared to spinal cord. On T2-wightd image (T2WI), the mass appeared as higher signal intensity compared with spinal cord and lower signal intensity compared with cerebral spinal fluid (CSF). Inside and adjacent to the mass, there were flow voids suggesting feeding artery or draining veins. On fat suppressed T1WI after administration of the gadolinium contrast medium, the mass showed strong and homogeneous enhancement. These MR imaging findings

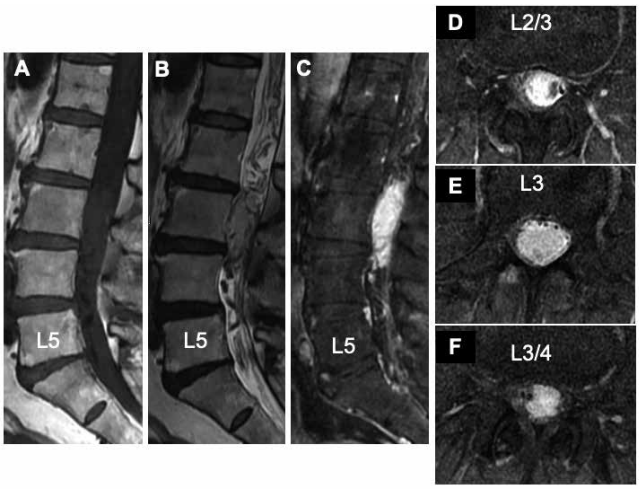

Figure 1. Magnetic resonance imaging (MRI) showing an intradural tumor at L3. (A) The lesion appears as an iso-intense area on sagittal T1-weighted imaging and (B) hyperintense with a low intensity area on sagittal T2-weighted imaging (T2WI). Serpentine vascular structures (area of flow voids on T2WI) distinguished from the cauda equina appear at both the cranial and caudal parts of the tumor. (C) Contrast-enhanced fat-suppressed (CE-FS) T1WI. The tumor shows strong gadolinium enhancement (D, E, F). Axial view of CE-FS T1WI. The tumor occupies the intradural space and flow voids are seen in and around the tumor.

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indicated that the spinal mass was hypervascular tumor. Spinal nerves of cauda equina were involved in the tumor. Myelography revealed complete block of the enhancement because of the occupied mass effect of this tumor in the subarachnoid space.

The patient underwent preoperative angiography 1 day before scheduled surgery. Four feeder arteries were seen arising from the left 9th and 10th intercostal arteries and the bilateral iliac arteries (Figure 2 A-D), with a draining vein in the caudal part of the tumor, followed by left lateral sacral vein and left internal iliac vein. CT angiography showed tumor feeder arteries and a draining vein (Figure 2 E, F). Because the tumor feeders arose from the left 9th and 10th intercostal arteries, followed by anterior spinal artery and left posterior spinal artery respectively, embolization was not performed to avoid iatrogenic neurological deficit.

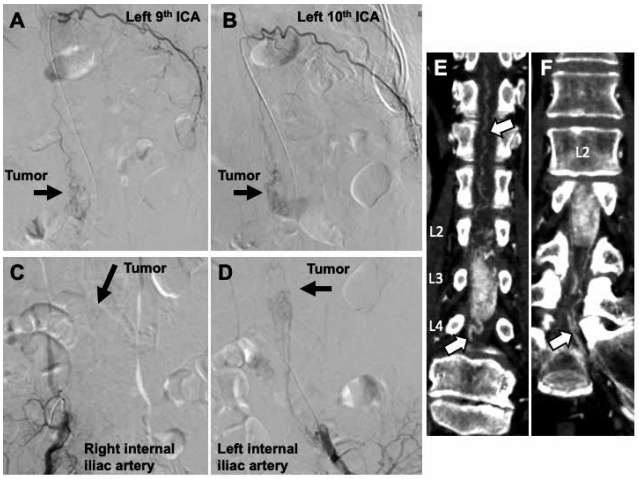

Figure 2. Preoperative angiography (A-D) and post-contrast CT images (E, F). Tumor was indicated by black arrows (A-D). Tumor feeders arose from the left 9th and 10th intercostal arteries (ICAs) (A, B) and the internal iliac artery on both sides (C, D). Spinal arteries were followed by the left 9th and 10th ICAs. Strong enhancement of the tumor by contrast media were shown from left 10th ICA and left internal iliac artery. CT angiography showed feeder arteries (E, F) and a draining vain (F), which were indicated by white arrows.

After the informed consent, we performed marginal resection of the tumor (Figure 3). After laminectomy of L2 through L4, the dura was incised and intraoperative Doppler ultrasound identified 4 feeder arteries and 1 draining vein of the encapsulated tumor, which were successfully coagulated and dissected. Some spinal nerves had to be sacrificed since they were tightly intertwined and penetrated into the tumor. Macroscopically, the tumor was a well-circumscribed yellow-red mass. Intraoperative findings were consistent with intradural extramedullary hemangioblastoma.

Histological examination demonstrated short spindle-shaped tumor cells and numerous vessels on hematoxylin and eosin staining (Figure 4). The tumor cells were enriched with lipid-containing vacuoles and were positive for S100 protein and partially positive for inhibin alpha staining. The final diagnosis was consistent with hemangioblastoma, WHO grade 1.

After surgery, low back and left leg pain immediately improved. However, she developed complications of left leg muscle weakness and bladder and bowel dysfunction. Muscle strength was grade 1/5 for both the tibialis anterior and extensor hallucis longus on the left side. Bladder and bowel dysfunction improved after 6 months but the muscle weakness remained at grade 1/5 for both tibialis anterior and extensor hallucis longus. She has, however, been able to walk with a brace. MRI showed no tumor recurrence after 2 years.

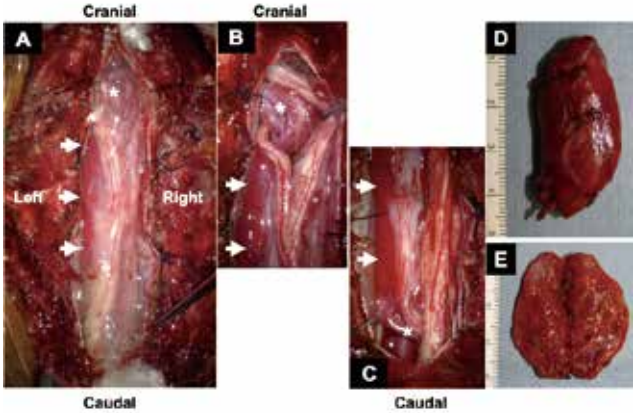

Figure 3. Intraoperative and postoperative findings. (A) After dural incision, the mass is observed under the arachnoid membrane. (B) Vessels are seen in the cranial part of the tumor. (C) Vessels are also seen in the caudal part of the tumor. (D) The 4.0 × 2.0 cm mass is well-circumscribed and slightly elastic, and (E) the cut surface is reddish brown (E). * Feeder vessels around the tumor ; arrows indicate the tumor.

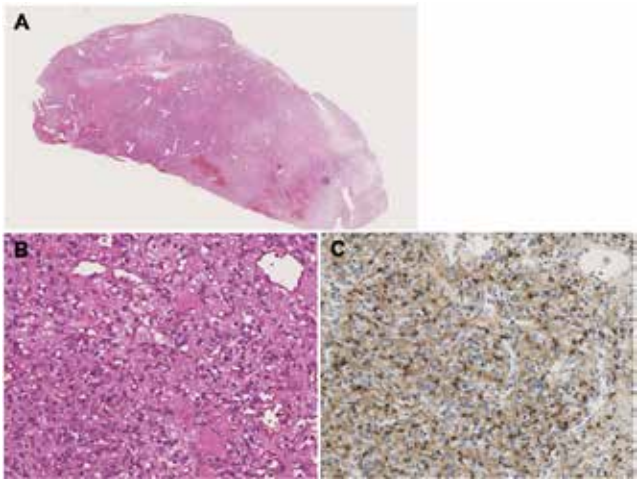

Figure 4. Histological findings of the resected tumor. (A) Low-power photomicrograph showing abundant vessels (hematoxylin and eosin staining). (B) The tumor consists of short spindle-shaped tumor cells with lipid-containing vacuoles and numerous vessels (hematoxylin and eosin staining, magnification ×200). (C) The tumor cells are positive to S100 protein in both the nucleus and cytoplasm.

DISCUSSION

Here, we report on a rare case of intradural extramedullary hemangioblastoma in the lumbar spinal canal without VHL syndrome. According to a study on 678 surgical spinal tumors by Hirano *et al*., the incidence of hemangioblastoma was 3.4%, followed by schwannoma (57.2%), meningioma (11.7%), ependymoma (8.0%), and hemangioma (4.0%) (2). Of these hemangioblastomas, only 4.2% were found in the lumbar region, whereas

33.3% were cervical, 50.0% were thoracic, and 8.3% were thoracolumbar (2). Focusing on the lumbar lesion, there could be only 1 case of hemangioblastoma compared with 173 of schwannoma, 12 of ependymoma, 4 of hemangioma, 4 of neurofibroma, and 1 of meningioma by calculating from their data of 678 cases. They also reported that 91.3 % of all spinal hemangioblastomas were intramedullary and the remaining 8.7% were intradural extramedullary (2). Furthermore, 39.1% of the cases were diagnosed in association with VHL syndrome (2). In a case report on hemangioblastoma of the cauda equina without VHL and review of the literature, Martins *et al*. reported only 22 cases including their own case (10 cases arising from the cauda equina, 10 from the filum terminale, and 2 of unknown origin) (3). Therefore, sporadic intradural extramedullary hemangioblastoma of the cauda equina, such as our case, is quite rare.

The first step to appropriate treatment is accurate diagnosis using MRI. In past case reports, hemangioblastoma typically shows high iso-intense signals on T2WI and strong gadolinium enhancement with a small area of low intensity as well as flow voids on T2WI (3-11). High vascularity is one of the characteristics of hemangioblastoma. In lumbar or sacral hemangioblastoma, schwannoma and meningioma often occur but rarely show marked enhancement or enlarged vessels (12). Malignant tumors such as metastasis or malignant nerve sheath tumor may infiltrate and erode bony structures, whereas hemangioblastoma only scallop the vertebral bodies (13). Vascular malformation is one of the differential diagnoses of a hypervascular mass, but the mass lesion is rarely well defined or and is typically heterogeneous on unenhanced MRI (12).

Angiography is the second step to diagnosis and is also important for surgical planning. In practice, MRI alone may be sufficient for diagnosing hemangioblastoma. However, detecting vessels before surgery is useful during it (5, 14). In this report, we successfully found feeder arteries and the draining vein using preoperative angiography, which was helpful to coagulate and divide these vessels during the operation. Embolization can reduce intraoperative bleeding but there are few reports on embolization for hemangioblastoma, especially in the spine (5). Only 3 cases of preoperative embolization have been reported in the lumbar region, including dumbbell-shaped tumor (5, 7, 15). Complications associated with embolization are rare, but 1 of the 3 cases of lumbar hemangioma showed transient moderate worsening. Spinal tumor embolization is safe, has few complications, and could improve surgical outcomes (5, 16), but surgeons should consider and evaluate the risk of devascularization. In cases of lumbar hemangioblastoma, embolization may be performed but only in difficult cases such as dumbbell-shaped tumor.

There are various postoperative outcomes after sporadic lumbar hemangioblastoma. In a review of 22 cases (3), symptoms improved in 10 cases, transiently worsened in 4, persisted in 4, and the status was unknown in 4. In the present case, low back and leg pain improved but the patient developed transient urinary dysfunction with persisting muscle weakness. Surgical resection of lumbar hemangioblastoma is safe and effective but there remains some possibility of postoperative neurological deficit.

Few studies have reported on the recurrence of sporadic lumbar hemangioblastoma, so the recurrence rate remains unclear (4). Spinal hemangioblastoma recurred in 6.25%-20% of cases with sporadic disease and one-third of patients with VHL syndrome. The risk factor for recurrence was subtotal resection and therefore marginal but complete resection may be mandatory. If complete resection is not possible, radiotherapy is an option, although there is insufficient evidence to support this method (4, 17).

CONCLUSION

We reported a rare case of spinal hemangioblastoma arising from the cauda equina. Angiography may be useful for preoperative diagnosis and detection of feeding veins for surgical planning.

DATA AVAILABILITY

The datasets generated and analyzed during this study are available from the corresponding author upon request.

COMPETING INTERESTS

None.

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None.

ETHICAL APPROVAL

This study was approved by the Ethics Committee of Tokushima University (number 1942-4, April 28, 2014).

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