Hemangioblastoma of the Filum Terminale Associated with Von Hippel-Lindau Disease: A Case Report

Von Hippel–Lindau hastalığına eşlik eden filum terminale hemangioblastomu: Bir olgu sunumu

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ABSTRACT

We report a 41-year-old man who presented with low back pain, lower extremity paresthesia, urinary retention and constipation. Magnetic resonance imaging showed a vascular intradural-extradural lesion at the second lumbar vertebral level. His medical history revealed that he had undergone surgery for a cerebellar hemangioblastoma 5 years ago. The patient underwent a spinal operation and a vascular tumor was removed from filum terminale. Pathologic examination of the tumor revealed a hemangioblastoma. Hemangioblastomas may occur sporadically or in association with von Hippel-Lindau disease. In the second case, they are often multiple and accompanied by cerebellar and brainstem lesions. The hemangioblastomas reported in the conus medullaris or in the extradural compartment adjacent to the conus medullaris are rare, tumors of the cauda equina are uncommon, and lesions of the filum terminale are extremely rare. We report a patient with von Hippel-Lindau disease having filum terminale hemangioblastoma and discuss the diagnosis, pathogenesis and treatment of hemangioblastoma.

KEYWORDS: Filum terminale, Hemangioblastoma, von Hippel–Lindau disease, Treatment

INTRODUCTION

Spinal cord hemangioblastomas (HBs) account for 1.6–2.1% of all spinal cord tumors, and may occur sporadically (67–75%) or as a manifestation of von Hippel-Lindau (VHL) disease (25–33%) (3,4,8,9,12,16). Seventy-five percent of spinal HBs are intramedullary, described in all spinal compartments and more commonly reported in the cervical or thoracic location (9, 19,27). Extradural-intradural HBs may arise from dorsal spinal cord pia or nerve roots (17,20). HBs located in the conus medullaris are rare (14,16,23,24,30), and tumors of the cauda equina (5,7,11,15) are uncommon. Hemangioblastoma of the filum terminale is extremely rare and very few cases were reported in the literature (1,10,18,26,25,28,29). We report a patient with VHL disease having filum terminale hemangioblastoma and discuss the diagnosis, pathogenesis and treatment of hemangioblastoma.

CASE REPORT

A 41-year-old male presented with a 3-month history of low back pain and lower extremity paresthesia. He had to strain to pass urine and had constipation. His pain had become progressively worse over the past three months. In his medical history he had undergone surgery for a cerebellar hemangioblastoma 5 years ago (Figure 1A–C). His postoperative period was uneventful after the cranial surgery but five year later he was examined for a neurogenic bladder complaint. His neurological examination showed...
bilateral lower extremity weakness, patellar hyporeflexia and hypoesthesia in the medial side of the legs. Magnetic resonance imaging (MRI) revealed a 1.5 cm diameter intradural tumor at L2 level, which was brilliantly enhancing after gadoliniunm infusion. The tumor was predominantly isointense on T1-weighted images and hypointense on T2-weighted images. Multiple dilated and tortuous vessels exited at the superior pole of the mass (Figure 2A, B). Afterwards, selective spinal angiography was performed, which confirmed the high vascularity of the tumor. The predominant arterial supply to the lesion was from the anterior spinal artery (Figure 2C). The patient underwent a spinal operation and L1-L3 laminectomies were performed. After opening of the dura mater, the feeding artery was identified by ultrasonography and coagulated

Figure 1: A) Axial CT section with contrast showing left cerebellar hypervascular lesion (arrow). B) Sagittal T1-weighted MRI of the cranium with contrast showing the lesion and arterial feeding (arrow). C) Postoperative axial CT section showing the lesion, which were totally removed.

Figure 2: A) Sagittal T1-weighted MRI of the lumbar spine with contrast showing a 1.5 cm diameter intradural tumor at L2 level, which was brilliantly enhancing after gadoliniunm infusion (small arrow is showing arterial feeding from superior pole of the mass, large arrow is showing the hypervascular lesion). B) Axial T1-weighted MRI of the lumbar spine with contrast showing an intradural lesion (arrow). C) Selective spinal angiography showing arterial supply (anterior spinal artery) and high vascularity of the tumor (arrow).
with bipolar cautery (Figure 3A). Dilated venous channels exiting from superior pole of the tumor were coagulated and divided. The tumor arising from filum terminale was circumferentially dissected, preserving all roots of the cauda equina and excised in an en bloc manner. Macroscopically, the tumor was a reddish-brown, highly vascular lesion (Figure 3B). Histopathology of the tumor showed a highly vascular tumor composed of vascular channels of varying sizes and thickness with intervening stromal polygonal cells with clear to eosinophilic cytoplasm and round to oval hyperchromatic nuclei. Moderate pleomorphism of the cells and clusters of foamy cells were seen. Some vessels were thrombosed and there were foci of hemorrhage. The histological diagnosis was hemangioblastoma. Von Hippel-Lindau clinical screening was positive. The patient’s low back pain and lower extremity paresthesia were immediately disappeared after surgery. The weakness in the lower extremities were recovered over 6 months. Bladder and bowel control of the patient were resolved slowly within four months period. Postoperative MRI confirmed complete excision of the mass.

DISCUSSION

HBs are low-grade, highly vascular tumors commonly associated with Von Hippel-Lindau Syndrome (VHL) and most commonly occurring in the cerebellum (21). VHL disease is caused by germline mutations of the VHL tumor suppressor gene located on the distal part of the short arm of chromosome 3 (3p25–26). This gene, encoding for a 213–amino acid protein, plays a major role in the regulation of vascular endothelial growth factor expression that plays an important role in endothelial cell proliferation (6,22). This finding explains the highly vascular nature of the HBs.

The majority of spinal HBs are intramedullary, and have been described at the cervical and thoracic levels (9,19,27). 21–28% of spinal HBs are intradural extramedullary and 9–13% of these found within the cauda equina (3). An association with von Hippel–Lindau disease is found in approximately 30% of all spinal hemangioblastomas. The origin of extramedullary hemangioblastoma is unclear. Extramedullary occurrence of HBs is more common in patients with Von Hippel Lindau disease; one autopsy report found multiple microscopic spinal root HBs (12). HB of filum terminale is very rarely reported (1,10,18, 25, 26,28,29). The reported cases were sporadic and not related with VHL. The case we present is part of VHL disease.

Some untreated filum terminale HBs may remain asymptomatic for several years. However as they grow, they may cause compress surrounding neural elements and cause symptoms and by time symptoms aggravate. Most common symptoms of filum terminale HB are progressive low back and lower extremity pain and weakness. If a filum terminale HB reaches a certain volume it may also cause cauda equina syndrome.

A spinal HB can be diagnosed accurately pre-operatively using CT and MRI scans alone. Spinal angiography provides information about feeding arteries (mostly anterior spinal artery) of the lesion and venous drainage systems for pre-operative strategy planning (2,13). Preoperative embolization is effective in the reduction of intraoperative bleeding and facilitates tumor resection, however it is generally not necessary. The risk of embolization should also be compared with the surgical risk in large and high vascular lesions treated surgically without preoperative devascularization.

Treatment of HB is surgical excision that is possible with low morbidity and results in excellent long-term functional outcome. Circumferential dissection and en bloc removal is the surgical technique for surgical resection. Intra-tumor debulking may cause profuse bleeding. Incomplete resection has been associated with a high rate of recurrence.
CONCLUSION

Hemangioblastomas of the filum terminale are extremely rare and reported cases are sporadic and not related with von Hippel-Lindau disease. We report a patient with von Hippel-Lindau disease having filum terminale hemangioblastoma and discuss diagnosis, pathogenesis and treatment of hemangioblastoma.

REFERENCES