Management of Spinal Hemangioblastoma in Von Hippel-Lindau Disease: A Case Report

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Abstract- Vertebral body location of hemangioblastomas (HB) is extremely rare. The authors report a case of spinal mass involving lower thoracic region with cord compression, approved to be spinal HB. A 57-year-old man presented to our center with eight months history of progressive intractable back pain and paraparesis. Admission computed tomography and magnetic resonance imaging (MRI) of the thoracolumbar spine demonstrated a lytic and expansile spinal mass with pedicle expansion and vivid contrast enhancement involving T11 and T12 vertebral bodies on the right side. He was a known case of von Hippel-Lindau (VHL) and he had history of 4th ventricular asymptomatic hemangioblastoma near Obex, multiple pancreatic cystic adenomas, multiple liver cysts, and right non-chromaffin adrenal mass. The patient underwent a T11-T12 partial transpedicular corpectomy with T5 to L3 posterior spinal fixation to bridge the invaded segment. The pathological and immunohistochemical findings were consistent with vertebral HB. Spinal HB although extremely rare, may be managed with subtotal tumor resection and fixation of normal adjacent vertebrae by cemented screws.

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Keywords: Vertebrae; Hemangioblastoma; Von hippel-lindau disease; Clear cell carcinoma

Introduction

Hemangioblastomas (HBs) arise generally within central nervous system parenchyma and are rare tumors with slow growth rate. They are most commonly found in the cerebellum, brainstem and spinal cord. (1). Six cases of spinal HB have been described previously, none of them having any evidence of VHL disease (1-6). We report the first case of von Hippel-Lindau (VHL) disease, presenting with spinal mass involving T11 and T12 vertebral bodies and pedicles with spinal cord compression.

Case Report

The patient was a 57-year-old man with eight months history of progressive intractable back pain and weakness, expressing visual analogue scale (VAS) of 80. On physical examination there were signs of spinal cord involvement, including impaired pin-prick and light touch below the right knee with associated leg weakness, hyperreflexia and extensor plantar response. Spinal computed tomography (CT)-angiography with intra-venous contrast was performed which showed no major feeding arteries (Figure 1A). On magnetic resonance imaging (MRI), abnormal bone marrow signals were seen at T11 and T12, with right pedicular expansion. Also, significant paravertebral extension of the mass with foraminal narrowing at right T11 and T12 levels was observed (Figures 1B, 1C). Past medical records of our case revealed nonfunctional pituitary adenoma, T9 compressive traumatic fracture leading to incidental discovery of T11 spinal cord HB, and clear cell renal cell carcinoma (RCC). Also in our patient's family history, total of seven brothers (including our case), mother, two aunts, two uncles, and grandfather had VHL disease.

Pre-operative imaging revealed 4th ventricular HB (Figure 1D), multiple pancreatic cystic adenomas,

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multiple liver cysts, and right non-chromaffin adrenal mass with negative biopsy.

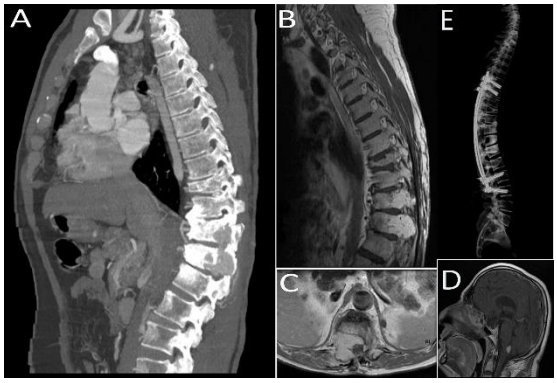

Figure 1. Imaging modalities of patient. A: Pre-operative sagittal CT-angiography with intra-venous contrast showed no major feeding arteries. B: On sagittal T1 weighted MRI with Gad, significant paravertebral extension of the mass with foraminal narrowing at right T11 and T12 levels was observed. C: On axial T1 weighted MRI with Gad, abnormal bone marrow signal changes were seen at T11 and T12, with right pedicular expansion. D: On sagittal T1 weighted brain MRI with Gad, revealed 4th ventricular hemangioblastoma near obex. E: On post-operative lateral spine X-ray, posterior spinal fixation with pedicular screws was performed from T5 to L3 levels

Clinical, laboratory and hormonal evidence were negative for pheochromocytoma and only urinary vanillylmandelic acid (VMA) was slightly elevated.

Through a posterior approach after sub-periosteal dissection of the affected spinal laminas, a purple vascularized mass was seen between T11 and T12 lateral masses. A subtotal resection of the extradural neoplasm was performed to expose dura matter, and then partial transpedicular corpectomy was performed for tumor resection. Because major feeding arteries were not identified in the preoperative vascular evaluation, selective preoperative embolization was not performed. Intraoperative bleeding necessitated five units of packed cell transfusion. Posterior spinal fixation with cemented pedicular screws was performed from T5 to L3 levels (sparing the involved osteoporotic levels) (Figure 1E). There were no major complications during or after surgery. His back pain resolved completely and there was no neurological deficit postoperatively.

Microscopic analysis of the specimens demonstrated that the lesion was intra-osseous with neovascularization, having dual cell populations; stromal foamy cells and endothelial cells (Figure 2A, B). Staining with CD31, and CD34 was positive in endothelial cells, staining with CD10, PAX8, SOX10, synaptophysin, chromogranin, D2-40, cytokeratin7, 20, and AE1/AE3 was negative. Immunohistochemical profile confirmed the vascular nature of the lesion and did not support metastatic carcinoma, especially renal clear cell carcinoma. Also, immunohistochemistry for inhibin (Figure 2C), and S100 was positive, further supporting the diagnosis of HB, World health organization (WHO) Grade I.

At follow-up visit one year later, the patient was without back pain (VAS=0) and his right leg paresthesia resolved completely.

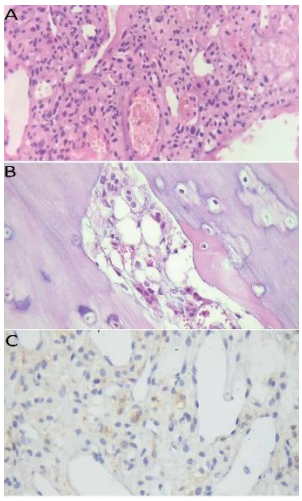

Figure 2. Photomicrographs of the resected tumor. A: Low-magnification (×100) H & E section showing a vasoformative lesion within bone matrix. B: High-magnification (×400) H & E sections showing endothelial cells and stromal cells with foamy cytoplasm. C: Cells stained positive with inhibin

Discussion

Hemangioblastoma is rare, histologically benign, highly vascular, slow-growing primary tumor of the central nervous system occurring either sporadically or as a component of VHL disease (7). The VHL related HB has more tendency to occur in eFIGxtra cerebellar regions (6). In the spinal cord, HBs occur as isolated tumors; most are intramedullary, extramedullary, on the surface of the cord or on nerve roots. Very rarely they occur extradurally within vertebrae, again usually in relation to spinal roots (1). Extradurally, pressure erosion of adjacent bony structures is not uncommon, but intraosseous involvement of a vertebral body as our case, has been documented only six times before (1-6). We have summarized previous reported cases of intraosseous HB in table 1. In none of 6 previous reported cases, the spinal HB was a component of VHL disease. In one example, Cho and colleagues reported a case of sporadic spinal HB in a thoracic vertebra, which was initially suspected to be a metastasis on imaging in a patient with a history of

RCC. In their study a complete workup of VHL, had not revealed any evidence related to this syndrome (1). Our patient had family history of VHL disease, as well as obex HB and left eye blindness due to retinal HB, proven spinal cord and vertebral HB, bilateral RCC, multiple pancreatic and liver cysts, all diagnostic of VHL.

Table 1. A summary of the previously reported spinal hemangioblastoma.

Author, Year	Age at diagnosis	Sex	VHL	Involved vertebral level(s)
Stevens *et al*., 1983 (2)	36	Male	No	T9
Higgins *et al*., 1995 (3)	30	Male	No	T9
Steinmetz *et al*., 2005 (4)	50	Female	No	T7-T9
Panelos *et al*., 2010 (5)	72	Female	No	S1
Cho *et al*., 2011(1)	55	Male	No	T11
Li *et al*., 2017 (6)	69	Female	No	C4
Current study, 2021	41	Male	Yes	T10-T11

Abbreviations: VHL, von Hippel-Lindau

Histologically, HB may closely resemble metastatic RCC, a problem confounding diagnosis in some cases of VHL syndrome (3). In our case, however, despite a known history of primary kidney tumor (RCC), the histological findings and the lack of immunoreactivity for CD10, PAX8, AE1/AE3, and positive inhibin, metastasis from kidney origin was excluded.

Due to technical difficulties for surgical resection in spinal HBs, pre-operative diagnosis and anatomical localization, employing pedicle screws in adjacent vertebra have a crucial role for stabilization after subtotal resections. Small or asymptomatic tumors may be followed with imaging to ensure stability, until they become enlarged or symptomatic (8,9). For solitary tumors in surgically accessible locations, surgical removal often offers definitive treatment (8,10). On the other hand, radiation therapy is considered a feasible alternative for surgically inaccessible lesions or VHL-related, multifocal lesions (11,1).

The management of spinal HB associated with VHL disease requires careful neurological observation and timely selective removal of symptomatic lesions (10). This patient became symptomatic after pathologic fracture secondary to spinal HB. Recent tumor growth and cord compression necessitated immediate surgical resection and stabilization of the adjacent vertebrae.

Spinal HB should be considered in the differential diagnosis of isolated vertebral lesions in patients with VHL syndrome. These challenging cases may be managed with subtotal tumor resection and fixation of normal adjacent vertebrae.

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