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Case Report

Sporadic hemangioblastoma of cauda equina: An atypical case report

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ABSTRACT

Background: Hemangioblastomas account for about 1%–3% of all central nervous system tumors. They are usually associated with the Von Hippel–Lindau syndrome and typically occur in the posterior fossa, or throughout the spinal neuraxis. Here, we report the unusual case of a sporadic cauda equina hemangioblastoma.

Case Description: A 28-year-old Caucasian female patient presented with progressive low back pain of 2 months duration. The magnetic resonance (MR) revealed a heterogeneous intradural and extramedullary lesion at the L2 level; with intravenous contrast, there were vascular flow voids and surrounding vasogenic edema (i.e., measuring 4.1 cm × 3.5 cm). The patient underwent an L2 right hemilaminectomy under intraoperative neurophysiological monitoring. She was discharged the 4th postoperative day, neurologically intact. Literature describes 21 previous reports of sporadic isolated spinal hemangioblastomas.

Conclusion: Although rare, sporadic, and isolated hemangioblastomas of the cauda equina should be included among the differential diagnoses of intradural spinal lesions, particularly when enhanced MR studies document serpentine flow voids.

Keywords: Cauda equina, pathology, spinal cord neoplasm, treatment

INTRODUCTION

Hemangioblastomas account for about 1%–3% of all central nervous system tumors.[1] They are usually associated with the Von Hippel–Lindau syndrome and are mainly found in the posterior fossa or throughout the spinal neuraxis.[1-5] They are typically seen in an intradural/intramedullary rather than intradural/extramedullary location.[4] Only 21 such cases of extramedullary hemangioblastomas of the cauda equina without Von Hippel–Lindau syndrome had been previously reported.[1] Here, we report yet another such case with literature review.

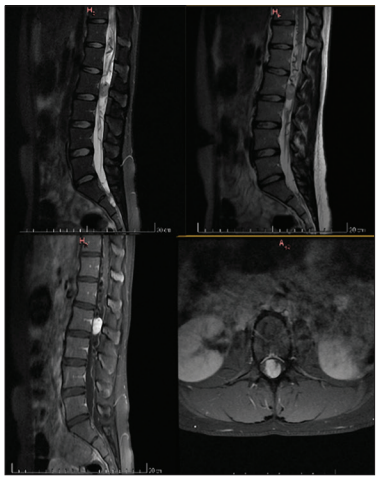

Figure 1: Magnetic resonance images. Above, sagittal T2 images and below, T1 images revealing gadolinium enhancement of L2 lesion.

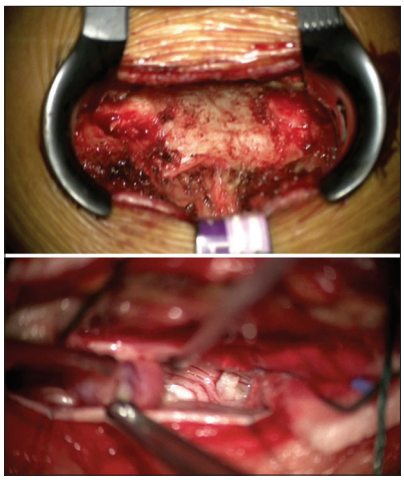

Figure 2: Above, L2 hemilaminectomy. Below, after dural sac opening and visualization of cauda equina roots and tumor fragments.

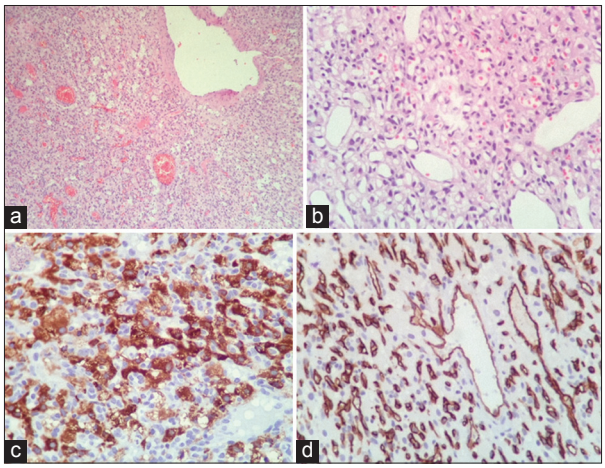

Figure 3: Histology image. In (a) and (b), hematoxylin-eosin images of hemangioblastoma. In (c), immunohistochemistry signaling with inhibin. In (d), immunohistochemistry signaling with CD34.

CASE DESCRIPTION

A 28-year-old Caucasian female presented with progressive low back pain of 2 months duration. She had no significant medical history and exhibited no focal neurological deficits. The magnetic resonance imaging (MRI) of the lumbar spine revealed a heterogeneous intradural/extramedullary lesion at the L2 level (measuring 4.1 cm × 3.5 cm) that enhanced with contrast. Flow voids were also demonstrated along with surrounding vasogenic edema [Figure 1]. Differential diagnoses included arteriovenous malformations, cavernous malformations, and/or other highly vascular lesions (e.g., hemangioblastomas).

Surgery

An L2 right hemilaminectomy was performed utilizing intraoperative neuromonitoring [Figure 2]. A richly vascularized tumor was identified on opening the dura. There was a clear cleavage plane between the tumor and the filum terminale, facilitating full resection. The patient was discharged home on the 4th postoperative day without a neurological deficit.

Pathology

Hematoxylin and eosin stains revealed a highly vascular and cellular tumor [Figure 3]. The immunohistochemical examination showed the presence of inhibin. The tumor was CD34 positive and showed a rich vasculature. Together these results were diagnostic for a hemangioblastoma.

DISCUSSION

Isolated tumors of the cauda equina represent 10% of all spinal canal lesions and typically include schwannomas, meningiomas, or ependymomas. Hemangioblastomas appear isointense on T1 and hyperintense on T2 MRI scans and/or heterogeneous hyperintensity on T2 studies secondary to flow voids attributed to their high vascularity (e.g., serpentine/tortuous/convoluted vessels).[1-5]

For the total of 22 cases of filum terminal hemangioblastomas (e.g., including this case), patients averaged 51 years of age [Table 1]. Notably, half of these tumors involved the cauda equina, and the other half, the filum terminale. In the majority of cases, patients exhibited sensory symptoms and neurological deficits postoperatively, these complaints typically improved.[1]

Surgical resection of these lesions is considered safe and effective with low accompanying complication/morbidity rates (e.g., few significant postoperative neurological deficits).

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal.

Table 1: Data of patients with spinal hemangioblastomas.

Case	Reference	Age (years)/sex	Spinal level	Origin of tumor	Preoperative symptoms	Tumor origin resected	Postoperative outcome
1	Sasaki *et al.*, 1978	40/M	L3	Cauda equina	Sensory and sphincter	No	Temporary sensory disturbance
2	Wolbers *et al.*, 1985	36/M	L1	Filum terminale	Motor and sensory	No	Improved
3	Rohde *et al.*, 1995	38/F	L3–4	Cauda equina	Sensory only	Yes	Continued sensory changes
4	Chazono *et al.*, 1999	48/F	L5	Cauda equina	Motor and sensory	Yes	Continued sensory changes
5	Tibbs *et al.*, 1999	35/M	L2–3	Filum terminale	Sensory only	No	Improved
6	Baker *et al.*, 2000	48/F	L5–S1	Unknown	Unknown	Unknown	Unknown
7	Farneti *et al.*, 2001	57/M	L4	Filum terminale	Sensory only	Yes	Improved
8	Hermier *et al.*, 2002	58/M	S1	Cauda equina	Motor, sensory, and sphincter	Yes	Improved
9	Costa *et al.*, 2003	40/F	L2-3	Cauda equina	Motor and sensory	No	Temporary motor changes
10	Escott *et al.*, 2004	70/M	L1	Unknown	Motor only	Yes	Improved
11	Escott *et al.*, 2004	45/M	L4–5	Cauda equina	Sensory only	Yes	Improved
12	Biondi *et al.*, 2005	61/M	L3	Filum terminale	Motor and sensory	Unknown	Unknown
13	Biondi *et al.*, 2005	Unknown/M	Unknown	Filum terminale	Sensory only	Unknown	Unknown
14	Nadkarni *et al.*, 2006	52/M	L2–3	Filum terminale	Sensory and sphincter	No	Improved
15	Ortega-Martinez *et al.*, 2007	41/F	L3 and S1	Filum terminale	Sensory only	Yes	Continued sensory changes
16	Ciappetta *et al.*, 2007	62/F	L2–3	Filum terminale	Sensory only	No	Improved
17	Wong *et al.*, 2007	64/M	L4	Filum terminale	Sensory only	Yes	Temporary urinary incontinence
18	Sergides *et al.*, 2009	75/M	L3	Filum terminale	None	Yes	None
19	Kunihiro *et al.*, 2011	50/M	L3-4	Cauda equina	Sensory only	Yes	Continued sensory changes
20	Wu *et al.*, 2013	53/M	L1–2	Cauda equina	Sensory only	Yes	Unknown
21	Blaty, 2016	82/M	L4	Cauda equina	Sensory only	No	Improved sensory changes; temporary urinary and bowel retention
22	Brock *et al.*, 2018	28/F	L2	Cauda equine	Pain	Yes	Improved

M: Male and F: Female. Adapted from Blaty *et al.*[1]

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Conflicts of interest

The authors declare no conflicts of interest.

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