Review Article

Purely extradural spinal nerve root hemangioblastomas

ABSTRACT

Spinal nerve root hemangioblastomas present mostly as intradural-extradurally. Purely extradural spinal nerve root hemangioblastoma is a very rare entity. In this study, we aimed to analyze epidemiological perspectives of purely extradural spinal nerve root hemangioblastomas presented in English medical literature in addition to our own exemplary case. PubMed/MEDLINE was searched using the terms "hemangioblastoma," "extradural," "spinal," and "nerve root." Demographical variables of age, gender, concomitant presence of von Hippel–Lindau (VHL) disease; spinal imaging and/or intraoperative findings for tumor location were surveyed from retrieved articles. There are 38 patients with purely extradural spinal nerve root hemangioblastoma. The median age is 45 years (range = 24–72 years). Female:male ratio is 0.6. Spinal levels for purely extradural spinal nerve root hemangioblastomas, in order of decreasing frequency, are thoracic (48.6%), cervical (13.5%), lumbar (13.5%), lumbosacral (10.8%), sacral (8.1%), and thoracolumbar (5.4%). Concomitant presence of VHL disease is 45%. Purely extradural spinal nerve root hemangioblastomas are very rare and can be confused with other more common extradural spinal cord tumors. Concomitant presence of VHL disease is observed in less than half of the patients with purely extradural spinal nerve root hemangioblastomas. Surgery is the first-line treatment in these tumors.

Key words: Extradural; hemangioblastoma; spine; surgery; von Hippel–Lindau.

Introduction

Hemangioblastoma is a benign hypervascular tumor seen concomitant with von Hippel–Lindau (VHL) disease in ¼ of the patients and mostly presents in the posterior cranial fossa and the spinal cord (¾ intramedullary, ¼ extramedullary).[1,2] Spinal nerve root hemangioblastomas present mostly as intradural-extradurally. Purely extradural spinal nerve root hemangioblastoma is a very rare entity. In this study, we aimed to analyze epidemiological perspectives of purely extradural spinal nerve root hemangioblastomas presented in English medical literature in addition to our own exemplary case.

Materials and Methods

PubMed/MEDLINE was searched using the terms "hemangioblastoma," "extradural," "spinal," and "nerve root." Patient demographical variables of age, gender, concomitant presence of VHL disease; spinal imaging and/or intraoperative findings for tumor location were surveyed. Spinal nerve root hemangioblastoma cases with intradural origins were excluded from the final analysis.

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Results

Thirty-eight patients with purely extradural spinal nerve root hemangioblastoma were retrieved from literature survey and our own case.[1,3-21] The median age is 45 years (range = 24–72 years). The female:male ratio is 0.6. Most common spinal location for purely extradural hemangioblastoma is thoracic level (48.6%). Other spinal locations are cervical (13.5%), lumbar (13.5%), lumbosacral (10.8%), sacral (8.1%), and thoracolumbar (5.4%). Concomitant presence of VHL disease is seen in 45% of the patients [Table 1].

Exemplary case

A 43-year-old man was admitted with dorsal and low back pain with progressive bilateral leg weakness. On neurological examination, he had 1/5 paraparesia and right foot clonus (2–3 beats). Whole spinal magnetic resonance imaging (MRI) was obtained, which depicted a mass lesion at thoracic 7–8 levels. The mass lesion enhanced intensely following intravenous (IV) contrast administration and it was located in the right posterolateral side of the spinal canal with some invasion into the same side intervertebral foramen [Figure 1]. The patient underwent surgery. Total laminectomy of T7–T8 and *en bloc* removal of mass lesion was performed by posterior approach. The mass lesion was adherent to the right T7 nerve root with slight infiltration to the dura mater [Figure 2]. No intraoperative

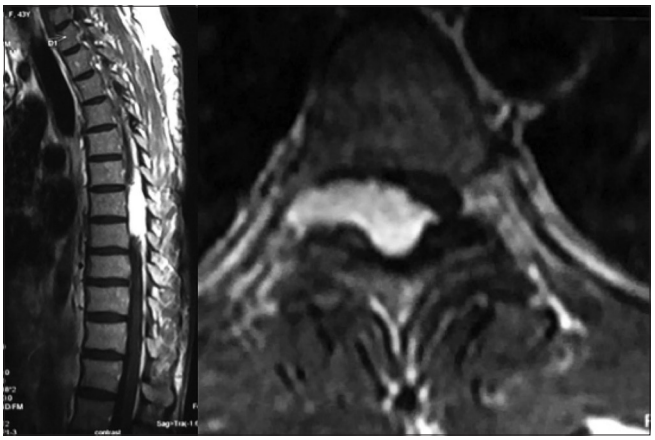

Figure 1: Homogenous enhancement is observed at right T7 nerve root location displacing the spinal cord to the left side of the spinal canal

Table 1: Purely extradural spinal nerve root hemangioblastoma cases presented in the literature

Authors/year	Age/gender	Spinal level	VHL presence
Tarlov/1947	42/male	T11–T12	N/A
Smith and Estridge/1963	N/A	N/A	N/A
Murota and Symon/1989	33/male	T10	None
	56/female	C5	Present
Higgins *et al.*/1996	30/male	T9	N/A
Chazono *et al.*/1999	48/female	L5	None
Chu *et al.*/2001	24/female	S1	Present
Hermier *et al.*/2002	58/male	S1	None
Lee *et al.*/2003	29/male	T12–L1, L5–S1	Present
Escott *et al.*/2004	62/male	C8	None
Glasker *et al.*/2005	35/male	T10	Present
	52/male	C7	None
	40/female	T6	Present
	48/female	L5	Present
	44/male	T6	None
	46/female	S1	Present
Kern *et al.*/2006	34/female	T10	Present
Purandare *et al.*/2012	32/male	T2	N/A*
Choudhury *et al.*/2012	N/A/female	Thoracolumbar**	Present
Mitchell *et al.*/2013	72/male	L3	None
Sun *et al.*/2014	63/female	T4	None
Law *et al.*/2014	59/male	L4	None
Roman-de Aragon *et al.*/2014	48/male	L4	N/A
Zakaria *et al.*/2014	40s/male	T3	N/A
Doyle and Fletcher/2014	N/A (12 patients)	C (2 patients), T (6 patients), LS (4 patients)	N/A
Laviv and Rappaport/2015	N/A	T	None
Present case	43/male	T7	None

*Had concomitant cerebellar hemangioblastoma, **Exact location of the tumor was not mentioned. VHL - Von Hippel–Lindau, C - Cervical, T - Thoracic, L – Lumbar, S - Sacral, N/A - Not available

complication occurred. Postoperative neurological status of the patient was same as the preoperative condition. The histopathology of the specimen was compatible with grade-I hemangioblastoma (WHO 2007 classification) [Figure 3]. Neither he nor his family had a history of VHL disease. At the fourth postoperative day, he was discharged with ongoing physical rehabilitation.

Discussion

Lindau defined the hemangioblastoma in 1926.[12,22] Origin of hemangioblastomas could not have been defined, yet. Hemangioblastomas present not only in the nervous system but also in other organs systems, so glial cells might not be the source of the tumor cells. Both mesodermal and neuroectodermal tissues have been proposed to be possible sources of hemangioblastoma, recently.[12]

Hemangioblastomas are sometimes seen concomitant with VHL disease, which is an autosomal dominant disorder of tumor suppressor gene, VHL.[13] More than half of the patients with spinal nerve root hemangioblastoma were found to have concomitant VHL disease.[11] However, in this study, concomitant VHL disease rate has been found as only 45%.

Spinal hemangioblastomas compose 1.6–5.8% of all spinal cord tumors and mostly present intramedullary in the dorsal aspect of cervical and thoracic spinal cord segments.[1,11] Sometimes, spinal hemangioblastomas present in both intramedullary and extramedullary (6–8% of spinal hemangioblastomas). Remaining rare locations are cauda equina, filum terminale, and proximal spinal nerve roots.[1,11] The most common site for purely extradural spinal nerve root hemangioblastomas is thoracic level (48.6%).

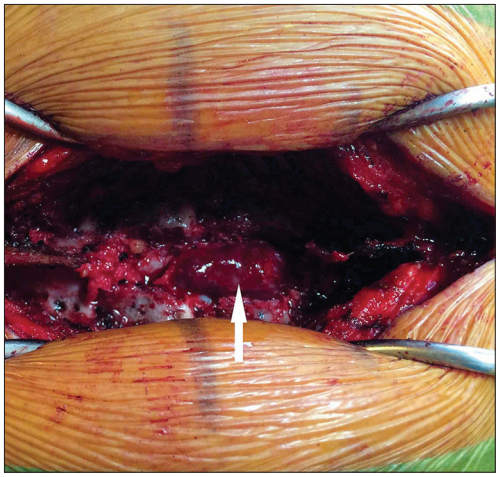

Figure 2: A vascular lesion is observed just over the dura. The lesion is adherent to the dura, yet not invading it

On MRI, spinal hemangioblastomas appear as hypo- to isointense on both T1- and T2-weighted scans with near homogeneous enhancement following IV contrast material administration. In the presence of intratumoral hemorrhage, intensities may differ on T1- and T2-weighted MRI.[11] Presence of prominent vessels in the subarachnoid space is diagnostic for hemangioblastomas.[8,23] Opposite to cystic components found in intracranial and intramedullary hemangioblastomas; extramedullary hemangioblastomas are solid in nature.[24] Angiography could be useful to depict vascularity of the tumor, and even preoperative embolization can be conveyed where needed.[11]

In differential diagnosis; meningioma, schwannoma, ependymoma, paraganglioglioma, and metastasis (especially renal cell carcinoma, due to its high vascularity) should be considered.[12]

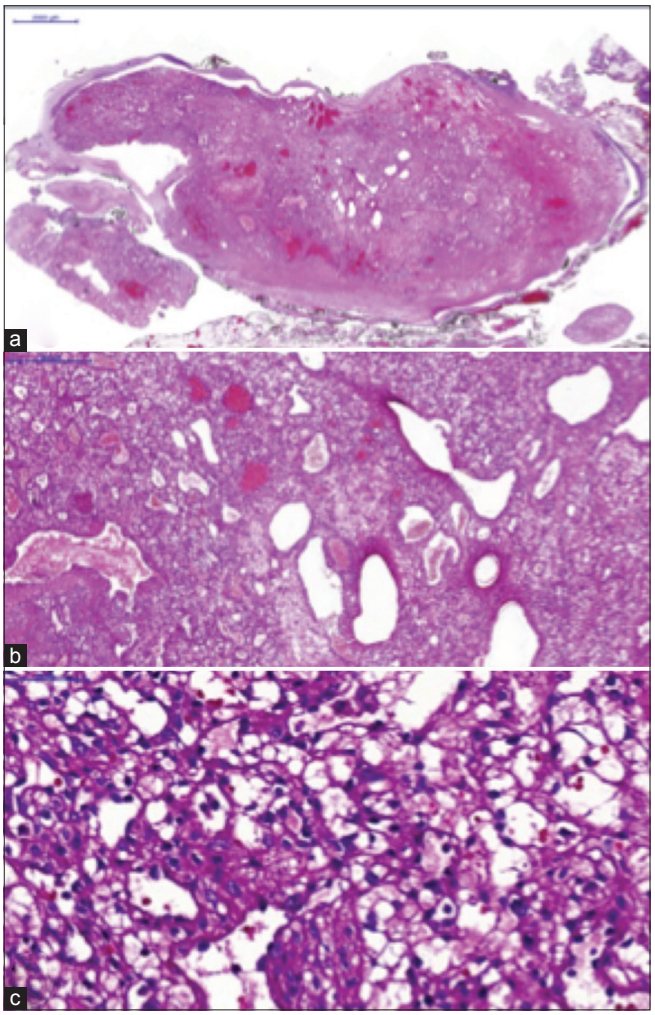

Figure 3: Encapsulated hemangioblastoma nodule attached to fibrous tissue (a - H and E, ×0.8). Tumor tissue showing thin-walled blood vessels of various diameters (b - H and E, ×4.2). High magnification reveals cells with cytoplasmic vacuolation typical for a hemangioblastoma (c - H and E, ×44.3)

Management of spinal nerve root hemangioblastomas starts with appropriate preoperative imaging to prevent massive intra-operative bleeding. It should be kept in mind that hemangioblastomas may show invasion into surrounding soft and bony tissue. Surgical experience is the mainstay of success in excision of these tumors.[13] There is a tendency to recur especially in cases with an inadequate amount of resection.[12]

Conclusion

Purely extradural spinal nerve root hemangioblastomas are very rare, and they might be confused with other more common extradural spinal tumors. Concomitant presence of VHL disease is observed in less than half of the patients with purely extradural spinal nerve root hemangioblastoma. Surgery, with careful preoperative planning, is the first line treatment in these tumors.

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Conflicts of interest
There are no conflicts of interest.

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