Spinal Root Hemangioblastomas in von Hippel-Lindau Patients: Observations from Surgical and Neurophysiological Monitoring

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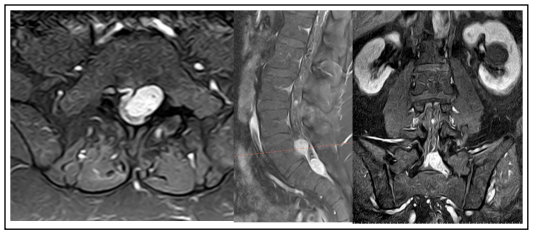
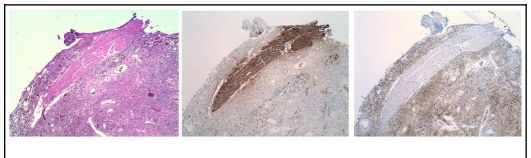
Introduction

Hemangioblastomas (HGB) are benign tumors, present at central nervous system in at least 75% of patients affected from von Hippel-Lindau (VHL) disease. Spinal hemangioblastomas are almost exclusively intramedullary lesions, and only on extremely rare occasions they have been reported outside the spinal cord, on spinal roots. The target of the present paper is to review the clinical and surgical treatment aspects iof spinal root HGBs in a VHL patient series.

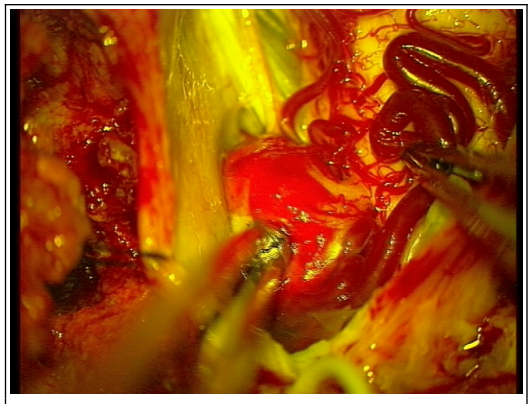


Methods

A retrospective review of clinical and surgical reports has been performed on patients treated for spinal root hemangioblastomas within a series of 81 VHL patients, followed and treated in a Familial NeuroOncology Unit. A review on published spinal root hemangioblastomas has been performed, in order to get a common pattern of behavior and management.



H&E, Neurophilament, and Inhibin Staining of a Root-HGB



HGB growing from a cervical root, with rootlets splaying through the tumor

Results

In our series, a total of five surgical procedures on 3 patients have been performed, to treat spinal root hemangioblastomas. Age of presentation was between 41 and 55. The most frequent symptoms were sensitive and motor deficit, both in two. The anatomical level was cervical in 1 case, lumbar in 2, and sacral in 2 more cases. Nerve root fibers were evidently splayed out troughout the tumor in each case. Cervical tumor was evidently growing from posterior root, and on neurophysiological intraoperative monitoring during tumor dissection in all cases, sensitive and no motor deficit was observed. In S1 and S2 tumors, motor root component was adherent but easily separated from the tumor. Therefore, in our cases tumor was related to sensitive root in all (5/5) of hemangioblastomas. Hystopathological evaluation showed neurofilament fibers encased within tumor tissue, in most of cases.

In postoperative follow-up, a clinical maintenance level was observed in pre -and postoperative evaluation.

Functional Pre-Op Level	Functional Post-Op Level				
	I	II	III	IV	Total
I No / mild focal deficit	2				2
II Mild/moderate gait difficulty, ...ambulates independently		1			1
III Severe neurological deficit requiring cane/brace for ambulation					-
IV Severe deficit requiring wheelchair, not independent					-
Total	2	1	-	-	3

Functional preop & postoperative level following McCormick´s Scale

Up to now, 46 other patients with 52 spinal root hemangioblastomas have been previously published. Of them, lumbosacral region was more frequently observed (27 tumors), followed by cervical (15) and thoracic levels (10), with age limits between 22 and 77. In 22 cases, tumor were described as related to sensitive roots, in three cases a relation with motor root was evident, and no information could be obtained for the rest of tumors. Also, no data about the eventual relationship with sporadic presentation or VHL disease were obtained, as diagnosis criteria were not adequately applied in most published cases.

Learning Objectives

Management of spinal root hemangioblastomas in von Hippel-Lindau patients, presented as spinal root tumors with flow voids in MRI in sporadic or VHL patients. Most of tumors are related to sensitive roots, and can be resected without motor deficits if carefull microsugical dissection is performed when resection with neurophysiological intraoperative monitoring.

Conclusions

Up to now, 52 spinal root hemangioblastomas have been previously published. We have described five additional cases. Tumors grow more frequently on lumbosacral region and from sensitive spinal root fibers, making possible a total resection of hemangioblastomas with minimal or no functional deficits in VHL patients. We suggest that surgical treatment on spinal root hemangioblastomas should be performed at clinical symptom beginning, or/and when tumor grows close to a diameter close to 1.5 cm at lumbosacral level although asymptomatic, always under neurophysiological monitoring, in order to get the best treatment results.

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