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Original Research Article

Conus Medullaris Hemangioblastoma a Case Report

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Abstract: Hemangioblastomas of the spinal cord are rare lesions. Most commonly these tumors are present in patients with von Hippel-Lindau (VHL) syndrome. We describe here the case of a 35 years old woman with a pure radicular hemangioblastoma, not associated with VHL, presenting with radicular pain, diagnosed with magnetic ressonance imaging (MRI) and submitted to total resection with a very good outcome. The histological examination proved hemangioblastoma of the conus medullaris in a patient without clinical criteria for VHL.

Keywords: Conus Medullaris, Hemangioblastoma, Von Hippel-Lindau Syndrome.

INTRODUCTION

Hemangioblastomas are rare, benign, highly vascularized tumors classified as grade I according to World Health Organization classification systems. About 3% of all intramedullary tumors are hemangioblastomas. Spinal cord hemangioblastomas are either sporadic or manifestations of von Hippel-Lindau (VHL) disease in 20% to 45% of patients [1-3].

MATERIALS AND METHODS

A 35 years-old woman presented with right sciatic pain non responsive to the usual analgesics for twenty days prior to hospital admission. Her physical examination showed paraparesia, No sphincter disorder, Pyramidal syndrom in both lower limbs on the clinical examination.

Clinical Evaluation

Detailed neurological examinations were performed before surgery and in the postoperative period. Patient was followed in 6- to 12-month intervals after surgery. To determine neurological function after surgery and distinguish this outcome from the progression of additional CNS tumors in VHL disease, functional outcome was recorded immediately (0-30 days) after surgery, in the short-term (6 months) after surgery, and over the long-term (most recent follow-up) after surgery.

Imaging and Explorations:

Preoperative evaluation of the precise tumor location is important for total resection of tumor and improving the surgical outcome. Cerebro-Spinal CT scan and MRI were performed. Metanephrines in urines and genetique tests were explored.

Tumor location and size were determined by preoperative contrast-enhanced T1-weighted magnetic resonance imaging (MRI). Tumor volume was determined by the product of greatest anteroposterior diameter × greatest craniocaudal diameter × greatest mediolateral diameter × 0.5.13.

Surgical Technique

Surgical resection was performed as described as follows: a laminectomy was performed at the level of the tumor based on preoperative fluoroscopy. Ultrasound was then used to confirm the adequacy of the dural exposure. Following dural opening, microdissection was performed to isolate the tumor from uninvolved nerve roots. The involved nerve rootlet(s) was then tested for motor function usingelectromyography (including anal sphincter, external urinary sphincter, and lower extremities), as well as testing for intravesicular pressure

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(detrusor contraction). If involved roots did not show evidence of motor function with stimulation, they were cauterized, sectioned, and removed with tumor.

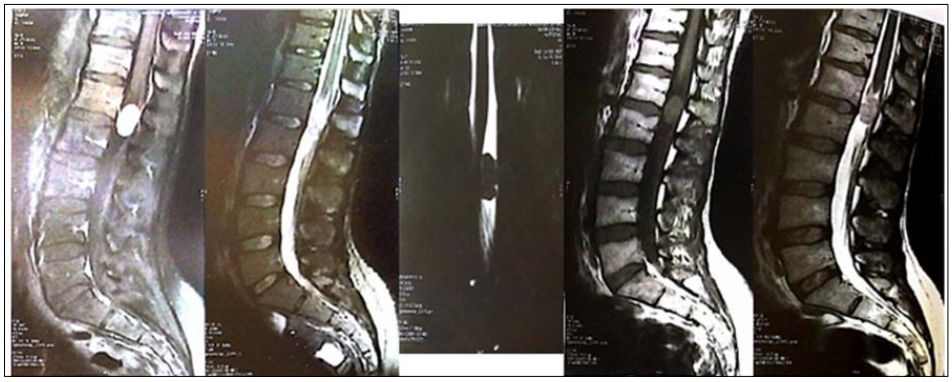

Figure 1: Pre operative MRI findings for Intradural-extramedullary spinal hemangioblastoma.

Sagittal MR images showed that tumor gave intramedullary T2 high intensity areas (HIAs) spreading toward the craniocaudal sides of the tumor,

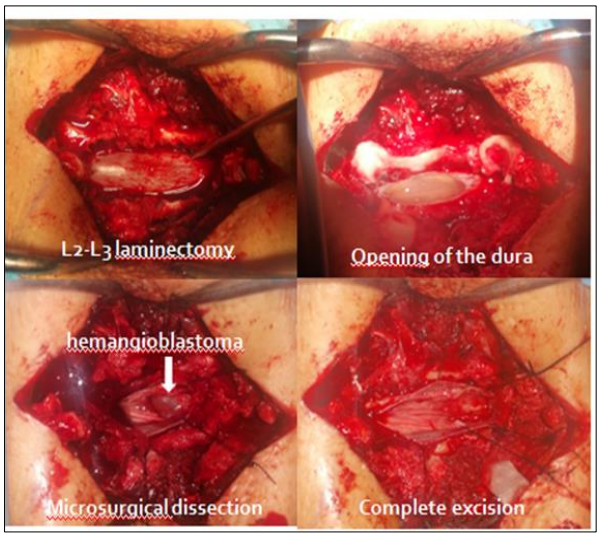

Figure 2: Microsurgical resection stages of spinal cord hemangioblastoma

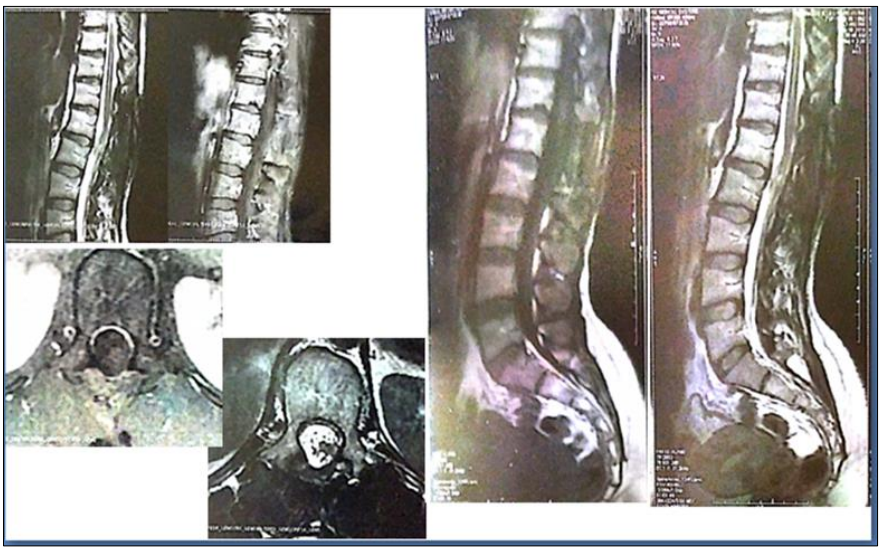

Figure 3: Post operative MRI findings for Intradural-extramedullary spinal hemangioblastoma

RESULTS

A 35-years-old woman presented with right sciatic pain non responsive to the usual analgesics, paraparesia without sphincter disorder. Screening for VHL disease was negative. Magnetic resonance imaging showed contrast enhancing intramedullary tumor with adjacent cyst in L2. We performed one single Stage L2-L3 Laminectomy. We noticed that after the opening of the dura, the tumor appeared in well circumscribed postero-lateral position without capsule. (Figure 2). The surgical resection followed rules that apply to resection of arteriovascular malformations: coagulation of arterial feeders precedes the coagulation of the draining vein, which is preserved until the end of surgery Complete macroscopique excision was performed.

The patient presented with transient post operatory sensory deficit (sciatic pain until 6 months post op) with an improvement of the paraparesia.

At 1 year follow up functional results were satisfactory and no recurrence (Figure 3).

DISCUSSION

From 1.6% to 2.1% of all spinal cord tumors are hemangioblastomas. Although hemangioblastomas have been reported in all spinal levels and compartments, with cervical and thoracic lesions predominating, the majority of these lesions are intramedullary. Extramedullary disease makes for around one fifth of all spinal hemangioblastomas, which is remarkably uncommon [1-7].

Hemangioblastomas are benign lesions that are partially cystic and histologically distinguished by stromal cells with foamy cytoplasm in endothelium-lined vascular channels. Lesions of the central nervous system primarily affect the retina and cerebellum; less frequently, they affect the medulla, spinal cord, and very infrequently, the supratentorium. In patients with VHL illness, medullary and spinal cord lesions are more frequent, while spontaneous lesions are usually cerebellar.

Von Hippel-Lindau disease is an heredofamilial autosomal dominant disease with incomplete penetrance, characterized by intracranial and intraspinal hemangioblastoma, often multiple, retinal hemangioblastoma, cystic lesions in the kidneys, liver, pancreas and epididymis, benign and malignant renal cell tumors. The vascular tumor may be the sole manifestation of this syndrome [8]. Althought histologically benign, in the central nervous system these lesions may be devastating, especially if presenting in the posterior fossa or intramedullary.

In our review, we found only three cases of intradural hemangioblastomas of the nerve root in patients without the stigmata of VHL syndrome, and only one was documented with magnetic ressonance imaging, as ours own [1-9]. Some authors recommend the super seletive spinal angiography to distinguish between intradural hemangioblastoma and spinal arteriovenous fistulae [1], but in this case, the MRI was quite sufficient for surgical planning.

The treatment of choice for these tumors is total surgical resection, which, if accomplished, is usually curative. At surgery, intralesional debulking should not be performed. These tumors should be dissected and removed en bloc, once the intralesional resection, even with the smaller lesions, will be associated with profuse bleeding [1-10].

CONCLUSION

Hemangioblastoma of the conus medullaris is very rare, It constitutes a real challenge because of the risque of significant post operative neurological worsening. The Complete excision of the tumor is possible in more than 90% of cases and it prevents the recurrence of the tumor.

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