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Surgical treatment of sporadic and von Hippel–Lindau syndrome-associated intramedullary hemangioblastomas

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ABSTRACT

Object: Intramedullary hemangioblastomas are rare lesions. They can be related to von Hippel–Lindau syndrome or they may be sporadic. This study describes surgical treatment for this infrequent tumor.

Methods: Twelve consecutive patients received surgery to remove sporadic or von Hippel–Lindau syndrome-associated intramedullary hemangioblastomas. Patients were evaluated at four time points: before treatment, on postoperative day one, on the day of discharge, and at a follow-up examination.

Results: The patients showed good preoperative neurological status. The cohort had a slight female predominance. All tumors spanned at least one spinal segment. In all cases, total tumor removal was achieved, and a good outcome was obtained. None of the following factors had a significant effect on outcome: age, sex, tumor size, the presence of a syrinx, or the presence of von Hippel–Lindau syndrome.

Conclusions: The surgical removal of intramedullary hemangioblastomas resulted in satisfactory long-term functional outcomes. The best results were obtained before neurological symptoms occurred. Thus, we suggest that surgery should be considered for managing asymptomatic, surgically accessible, space-occupying lesions in sIH group, and isolated, space-occupying lesions in vHLS-IH group.

1. Introduction

Intramedullary hemangioblastomas are rare neoplasms. They account for about 2% of all spinal cord tumors [1]. Hemangioblastomas are highly vascularized tumors, and due to their vascular permeability, they produce peritumoral cysts, which occur in about 50% of cases [2]. About 70% of intramedullary hemangioblastomas are isolated (sporadic, sIH), and 30% are associated with von Hippel–Lindau

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Abbreviations: vHLS, von Hippel–Lindau syndrome; IH, intramedullary hemangioblastoma; sIH, sporadic intramedullary hemangioblastoma; mMS, modified McCormick scale.

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syndrome (vHLS-IH) [3]. The rarity of these tumors has precluded a precise definition of a detailed treatment scheme. The natural history of these tumors is difficult to define, because they have unpredictable growth rates [4]. The aim of this study was to evaluate the short- and long-term results of the surgical removal of sIH and vHLS-IH, with attention to outcome in patients with asymptomatic vHLS-associated tumors.

2. Materials and methods

This retrospective study reviewed medical charts for 12 consecutive patients that received surgery for sporadic-IH ($n = 6$) and vHLS-IH ($n = 6$).

2.1. Clinical evaluation

Patients were evaluated at four time points: before treatment, on postoperative day one, on the day of discharge (early outcome), and at a follow-up examination (late outcome). The early and late outcomes were compared separately for patients with sIH and vHLS-IH. Follow-up data were obtained from medical charts, from telephone interviews with the patients, and from interviews during individual visits. Patients were assessed with a modified McCormick scale (mMS, Table 1) [5]. The follow-up period ranged from 3 to 10 years (average 5 years). Complete follow-up data were obtained in all cases.

2.2. Imaging evaluation

All patients underwent contrast-enhanced MRI before and after operative treatment. Spinal angiography was performed in eight (67%) patients.

2.3. Surgery

All tumors were removed via a posterior approach. Patients were placed in a prone position for removing tumors located in the thoracic spine and at the level of the atlanto-occipital junction. Patients were placed in a sitting position for removing tumors located in the cervical spine and at the cervico-thoracic junction. Three patients received laminectomies with an extended suboccipital craniotomy. Surgery was initiated with a midline dural incision over the dorsal surface of the spinal cord, followed by lateral dural sutures. Then, an incision was performed through the arachnoid and pia mater, at a location that depended upon the location of the most superficial part of the tumor ($n = 6$ midline incisions and $n = 6$ non-midline incisions). When the tumor was identified, the arterial feeders were gently coagulated during dissection. The tumor was microsurgically detached from the surrounding tissue, shrunken with bipolar coagulation at low power, and removed en bloc in all cases. It was not necessary to employ an ultrasound aspirator or laser. Concomitant intramedullary syrinxes were not directly dissected. The dura was closed in a watertight fashion. Laminoplasty was not applied. The wound was closed in a standard fashion. Motor and somatosensory evoked potentials were monitored during all procedures. Preoperative embolization was not used.

Table 1 – Modified McCormick scale [5].

Grade	Modified McCormick scale
I	Intact neurologically, normal ambulation, minimal dysesthesia
II	Mild motor or sensory deficit, functional independence
III	Moderate deficit, limitation of function, independent w/external aid
IV	Severe motor or sensory deficit, limited function, dependent
V	Paraplegia or quadriplegia, sometimes w/flickering movement

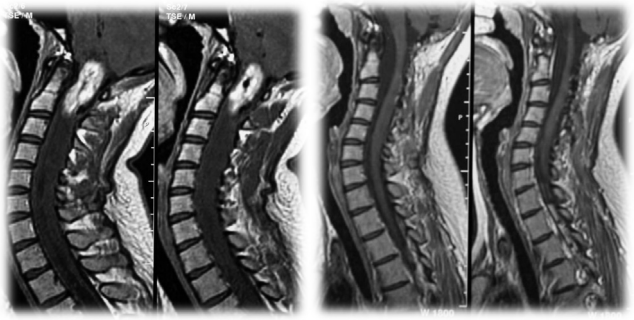

Fig. 1 – C1–C3 hemangioblastoma before and after treatment.

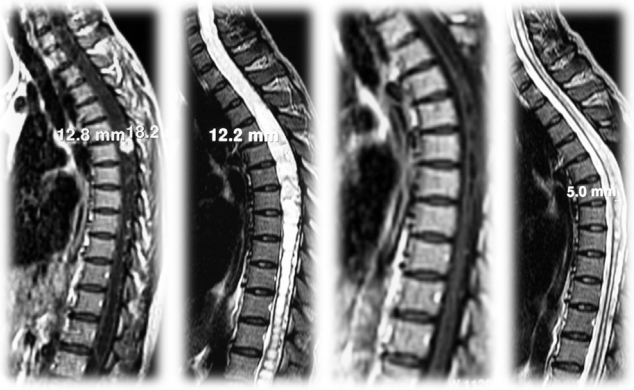

Fig. 2 – Th6 hemangioblastoma before and after treatment.

2.4. Statistical analysis

Fisher's exact test was used to evaluate significance. A p-value <0.05 was taken as statistically significant. Also, p-values between 0.06 and 0.07 were defined as a trend (Figs. 1 and 2).

3. Results

3.1. Patients and symptoms

We analyzed data for five men (42%) and seven women (58%), ages 18–59 years (mean 34 years). Six patients (50%) had sIH