ISSN 1220-8841 (Print)
ISSN 2344-4959 (Online)

ROMANIAN
NEUROSURGERY

Vol. XXXVI | No. 4 December 2022

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DOI: 10.33962/roneuro-2022-075

Romanian Neurosurgery (2022) XXXVI (4): pp. 411-417
DOI: 10.33962/roneuro-2022-075
www.journals.lapub.co.uk/index.php/roneurosurgery

Surgical management of spinal cord hemangioblastoma. Our clinic experience and case report

George Popescu[1], Marius C. Zaharia[1], Andrei Giovani[1], Ioana Miron[1], Cezar Vîjlănescu[1], Radu M. Gorgan[1,2]

[1] Bagdasar-Arseni Clinical Emergency Hospital, Department of Neurosurgery, Bucharest, ROMANIA
[2] Carol Davila University of Medicine and Pharmacy, Department of Neurosurgery, Bucharest, ROMANIA

Keywords
hemangioblastoma, von Hippel-Lindau, stereotactic radiosurgery

ABSTRACT

Background. Hemangioblastomas are considered rare tumours that are located, in most cases, in the posterior cranial fossa. In most of cases, spinal hemangioblastomas are identified on the thoracic and cervical levels and can be associated in one-third of cases with von Hippel Lindau Syndrome.

Material and methods. In this paper, we are presenting our clinic`s experience with spinal hemangioblastomas and the follow-up of the patients in the last 10 years.

Results. In our study, we included six men and three women with a mean age of 45 years, ranging between 36 to 61 years. The mean hospitalization days was 17 days. Among nine patients, there were identified 12 tumors. Six patients presented cervical spine hemangioblastomas, two of them had thoracic spine lesions and in one case, the tumour was identified at the lumbar level. Furthermore, two patients presented multiple lesions. Two patients had been associated with von Hippel Lindau Syndrome. In all cases, surgery was the therapeutic approach.

Discussions. Despite the rapid advance of technology and the new alternatives for the treatment of these lesions, surgical resection of hemangioblastomas remains the gold standard treatment.

Conclusions. Hemangiolbastomas are benign tumours that can be associated with von Hippel Lindau. A whole-body scan is required to confirm or exclude this syndrome. Even though the surgical treatment for asymptomatic patients remains debatable, surgical intervention is the only treatment that can lead to the total removal of the tumour.

Corresponding author:
G. Popescu

4th Neurosurgical Department, "Bagdasar Arseni" Clinical Emergency Hospital, Bucharest, Romania

george_popescu39@yahoo.com

ISSN online 2344-4959
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First published
December 2022 by
London Academic Publishing
www.lapub.co.uk

BACKGROUND

Hemangioblastomas are rare intra-axial tumors that usually occur in the posterior fossa [1]. Histologically wise, these are benign, highly-vascularized tumors that can present a muriform nodule. Moreover, spinal cord hemangioblastomas are rarely identified and represent one to six procents of all spinal cord tumors and just 1-2,5% of all central nervous system tumors [2]. In most cases, spinal hemangioblastomas

are identified on the cervical and thoracic segments and are usually described as intramedullary lesions situated at the posterior pole of the spinal cord, due to their development from dorsal root precursors [5], [7]. Spinal hemangioblastomas may be sporadic lesions but in up to 30% of the cases, they represent a feature of von Hippel Lindau Syndrome [3].

Von Hippel-Lindau (VHL) disease is a rare autosomal dominant tumor syndrome characterized by the development of benign and malignant tumors in the parenchyma of the various organs and systems, including the central nervous system [4]. It usually affects young adults, and in the VHL context, multiple hemangioblastomas are identified [12]. Over 50% of the people with VHL (63%) present central nervous system tumors, and usually the first manifestations of the syndrome are determined by hemangioblastomas [6], [11]. One of the most affected organs by VHL is the kidney. VHL determines renal cell cysts and clear cell carcinoma [8]. Other organs usually affected by the syndrome are the pancreas and retina [9], [10].

The most effective tool to identify these lesions is magnetic resonance. The rapid and constant evolution and accessibility of magnetic resonance imaging (MRI) raised the discovery rate of hemangioblastomas and more accurate descriptions were obtained [13], [14]. On T1-weighted MR imaging, the tumors have the aspect of bright enhancing lesions, and T2-weighted imaging can be used for a better characterization of the tumor, as well as emphasize any accompanying edema or syringomyelia [14]. However, spinal hemangioblastomas` features are not always well defined and difficult clinical diagnosis may occur.

The symptoms of patients with hemangioblastomas may range based on the segment where the lesion is located. Despite de rapid advance of technology and the new alternatives for the treatment of these lesions (such as embolization, stereotactic radiosurgery, and even some molecules such as bevacizumab and other inhibitors of angiogenetic signaling pathways), total removal of the hemangioblastomas remains the most appropriate therapy. However, even after the most laborious microsurgical excision, postoperative neurological degradation is not uncommon, being reported in up to 50% of the patients [15], [16]. Therefore, the management of asymptomatic lesions is still under debate. Furthermore, the long-term outcome of the patients with intramedullary hemangioblastomas is not well defined, due to the small number of cases.

Materials and methods

In this article, our target is to describe our clinic`s experience with spinal hemangioblastomas and the follow-up of patients with spinal hemangioblastomas in the last 10 years.

We performed a 10 years retrospective study that included 9 patients who benefited from surgical treatment for intramedullary spinal cord hemangioblastoma at the 4th Neurosurgery Department of "Bagdasar-Arseni" Clinical Emergency Hospital from Bucharest.

Results

In our study were included three women and six men with a mean age of 45 years, ranging between 36 to 61 years. The mean hospitalization days was 17 days, ranging from 7 to 36 days. Detailed hospitalization dates are available in Fig. 1.

Table 1. Patient's presentation state.

#	Age/ Sex	Sex	Signs/ Symptoms	Duration (months)	Location of tumor	No. of tum-ours
1	42	M	headache, gait impairment, right hemiparesis	6	cervical	1
2	43	F	headache, dysphagia, dysphonia	4	cervical	1
3	61	M	gait impairment, dysphagia, tetraparesis	2	cervical	1
4	41	F	paraparesis, sphincter dysfunction	4	lumbar	1
5	45	M	headache, ataxia, right hemiparesis	3	cervical	1
6	39	M	paraparesis, numbness of lower limbs	5	thoracic	2
7	36	M	paraparesis, sphincter dysfunction	6	thoracic	3
8	48	F	headache, paraparesis, sphincter dysfunction	3	cervical	1
9	51	M	paraparesis, numbness of upper/lower limbs, sphincter dysfunction	4	cervical	1

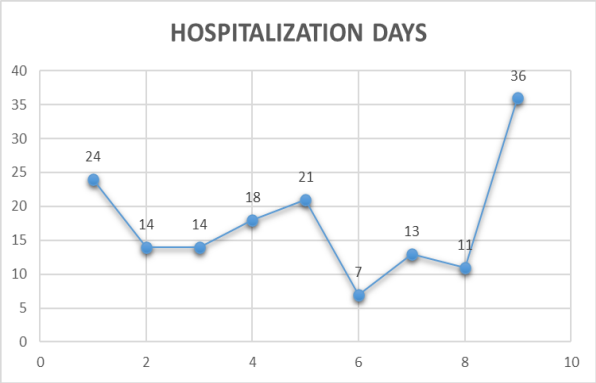

Figure 1. Hospitalization chart for each patient.

The signs and symptoms of the patients at admission vary depending on the level of the lesion and they will be detailed in Table 1. Karnofsky's performance score and McCormick's functional score for each patient as well as their discharge state and follow-up are presented in Fig. 2 and Fig. 3.

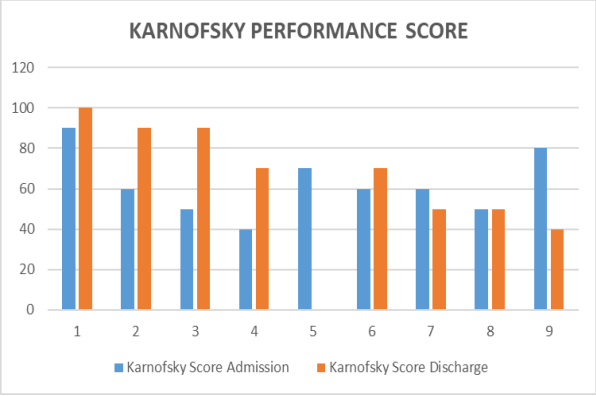

Figure 2. Karnofsky Performance Score at admission and discharge.

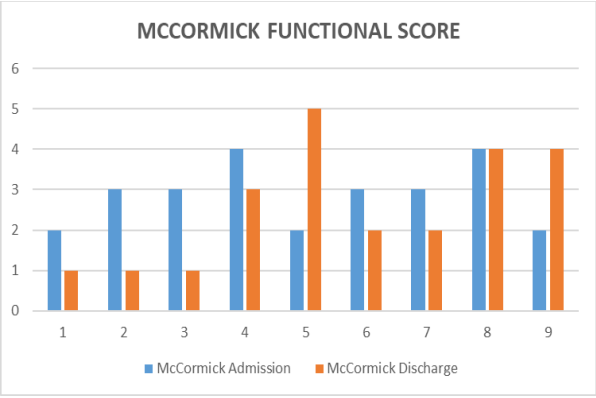

Figure 3. McCormick Functional Score at admission and discharge.

The presence of the tumours, localization, distribution and other possible characteristics such as the presence of syringomyelia were evaluated for each patient using MR imaging. Among nine patients, there were identified 12 tumors. The MRI described seven cases of hemangioblastomas associated with cystic component. Six patients presented cervical spine hemangioblastomas, two of them had thoracic spine lesions and in one case, the tumor was identified at the lumbar level. Furthermore, two patients presented multiple lesions. It is worth noting that both cases presented thoracic hemangioblastomas. One of them presented two lesions while the other had 3 thoracic spinal hemangioblastomas, located on the posterior edge of the spine. Because of the risk of multiple lesions for these patients, they were imagistically checked for vHL syndrome, such as brain tumors, retinal angiomas, pancreatic lesions renal masses and. All of them benefited from CT scans in order to emphasize the other possible tumors. Two patients were linked with vHL syndrome, one presented with pancreatic cyst and the second patient had multiple kidney cysts. None of them had a documented family history of the disease. In all cases, surgery was the therapeutic approach. The patients were placed in ventral position. After midline incision was performed, spinal laminectomy and posterior midline approach for the resection of spinal tumors were used. Laminectomies were performed at the level of the tumor as well as adjacent levels to ensure better visualization of the tumor and to avoid postoperative compressive edema. Complete resection was achieved in all cases, under microscope magnification. One patient had concomitent removal of both hemangioblastomas from the thoracic segment while the second patient with multiple thoracic lesions underwent two surgical interventions in order to completely remove his three tumoral masses. Due to preserved articular capsules, none of the patients required instrumentation, and no signs of instability were accused on the follow-up. The postoperative evolution of the patients was heterogeneous. Two patients developed hydrocephalus and 3 of them developed. Unfortunately, one patient died during hospitalization, due to multiple comorbidities. The follow-up was between 6 to 36 months. Four patients were lost after 12 months of follow-up. During this

time, two patients presented tumoral recurrence which required another surgical intervention.

Case presentation

A 51-year-old male presented to our clinic for headache, progressive numbness of the limbs, and paraparesis high-level Frankel C. The onset of the signs and symptoms was 4 months ago. He had no significant pathological history and no traumatism was reported. Head and cervical MRIs were performed. Head MRI had presented no tumoral masses Intra/extra-axial and no other lesions of the brain, dura mater, or bone were identified. On the other hand, the cervical spine MRI emphasized an intramedullary subdural tumor at the C2 level with medullary infiltration (Fig. 4).

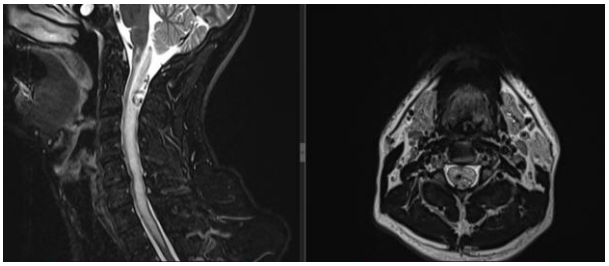

Figure 4. Cervical spine MRI - intramedullary subdural tumour at the C2 level with medullary infiltration.

Compelling the clinical and neurological state of the patient with the imagistic finding, the patient had surgical indication. After the written consent of him and his family, the patient underwent surgery. Under total anesthesia, he was placed in a ventral position. A C1-C3 midline incision was performed. The paravertebral muscles were detached bilaterally at these levels and laminectomies of C2 and C3 levels were performed in order to ensure better control of the tumor. Dura mater was carefully incised on the midline and the tumor was revealed, covered by the arachnoid. After the arachnoid layer was dissected as well, a large muriform highly vascularized tumor with a high caliber feeder vessel in the cranial side of the tumor was exposed. Using microscope magnification, circumferential dissection was performed with bipolar forceps and cotton pads. The accurate localization of the main nutrient artery is a crucial step in surgery. The main feeder was carefully dissected at cut using bipolar forceps and surgical scissors after tumor dissection was complete. The tumor was sent to the anatomopathological laboratory. After laborious hemostasis, the dura mater was sutured in a watertight proof fashion and the surgical wound was closed layer by layer, performed respecting the anatomical planes.

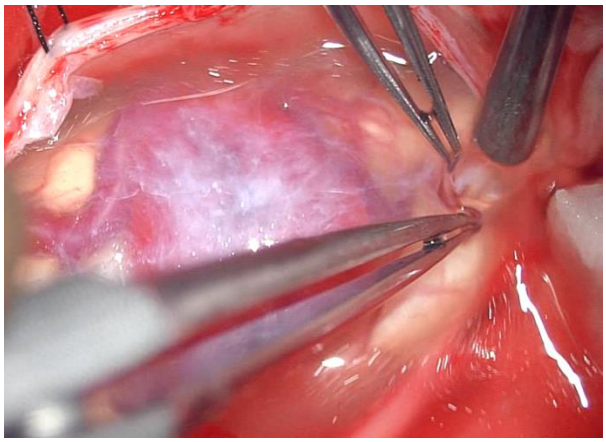

Figure 5. Dura mater was opened. The CSF exited with high pressure when the arachnoid layer was incised.

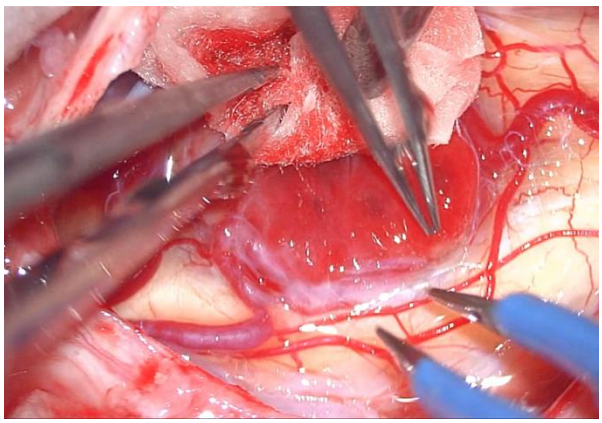

Figure 6. After complete exposure of the tumour, carefully dissection was performed using bipolar forceps.

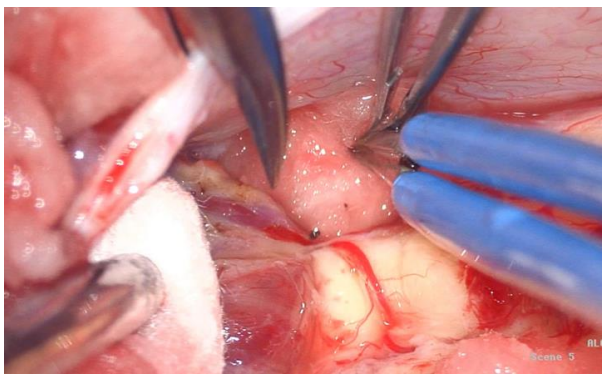

Figure 7. Complete resection of the lesion was achieved. The tumour is pushed cranially to obtain better exposure to the main feeder. Dissection of the main feeder.

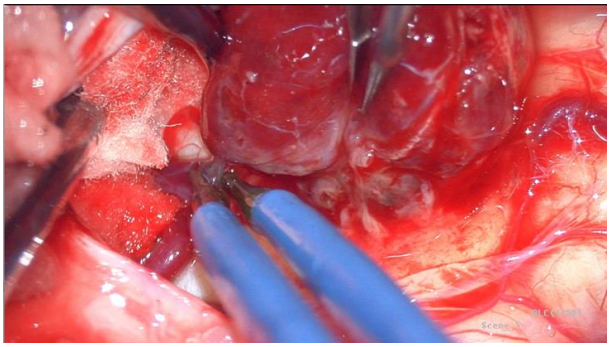

Figure 8. The main feeder was coagulated and sharply cut using a surgical scissor.

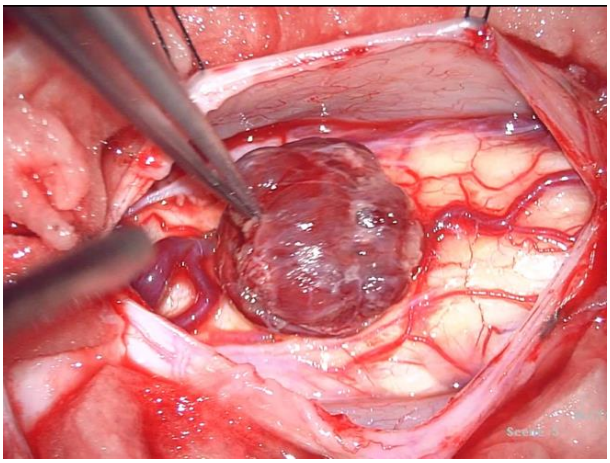

Figure 9. Complete dissection of the tumour was achieved. It was sent to histopathological analysis.

Postoperative, clinical and neurological state of the patient remained stationary. Postoperative MRI firmly established the total removal of the lesion and some blood that respected the surgical passage without any medullary compression

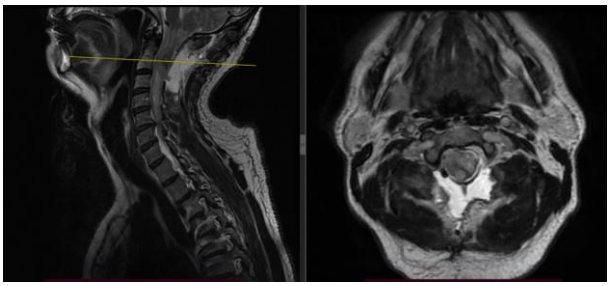

Figure 10. Postoperative MRI.

One week after the surgical intervention, the patient started to feel drowsy, he accused headache and nausea as well. A brain CT scan was performed (Fig. 11). CT scan showed slight enlargement of the ventricles and periventricular edema, with clear signs of hydrocephalus. Moreover, hemorrhage was identified in the right occipital corn.

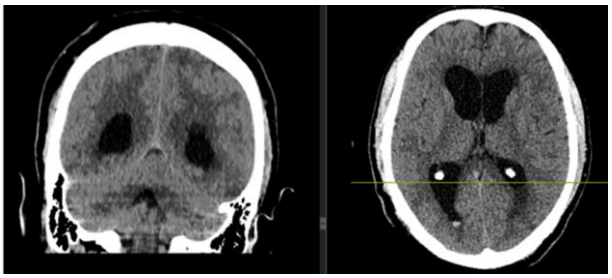

Figure 11. Discrete enlargement of the ventricles. Periventricular edema. Intraventricular hemorrhage in the right occipital corn.

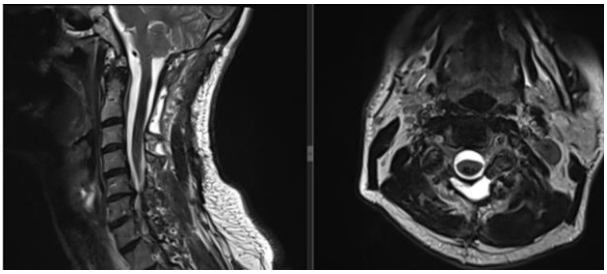

Figure 12. Postoperative MRI – 2 months after surgical intervention. The spinal cord presents a normal signal, without signs of compression, and no blood signal in the tumoral bed.

The patient underwent a new surgical intervention to evacuate the blood clots and to preserve CSF drainage in order to preserve the normal CSF flow. Postoperative, the newly installed symptoms were remitted. The patient was transferred to Neuromotory Rehabilitation Clinic, stationary compared to the admission state. After two months of kinesiotherapy, the patient is able to walk again and take care of himself by performing simple yet critical actions, such as changing his clothes or holding a glass of water.

Discussions

Hemangioblastomas are rare, vascularized tumors that usually develop in the posterior cranial fossa. Spinal cord hemangioblastomas constitute about 1-6% of all spinal cord tumoral pathology. Therefore, there is a lack of large cohort studies regarding the subject in the specialty literature. The accurate localization of the main nutrient artery is a crucial step in surgery. Especially in the cervical region, hemangioblastomas present a rich network of blood

vessels and the removal of one of them may prove to be difficult and with major consequences for spinal cord vascularization. Total resection may be very difficult due to intraoperative hemorrhage that can occur and blur the interface between the spinal cord and the lesion. To prevent intraoperative bleeding, preoperative embolization can be performed. However, preoperative embolization presented multiple complications such as neurological deterioration and intradural hemorrhage and its use remains controversial [17], [18].

Histologically wise, hemangioblastomas are benign tumors and the recurrence rate is low. However, it was noted a higher recurrence among the patients with von Hippel Lindau Syndrome [19]. Moreover, 20-30% of spinal hemangioblastomas are linked with vHL [20].

In order to avoid intraoperative and postoperative risks, alternative treatments were developed. In recent years, stereotactic radiosurgery was used as a therapeutic adjuvant, optimizing tumor dose while sparing the spinal cord [21], [22]. The biggest downside of stereotactic radiosurgery is the radiation-inducted myelopathy with potential permanent neurological impairment, which has been reported already in some cases [23], [24]. In addition, the overall recurrence rate after stereotactic radiosurgery is higher compared to classic surgery as few articles suggested [25], [26], [27], [28].

Conclusions

Despite the multiple treatment options available today for hemangioblastomas, the surgical approach remains the gold standard therapy. A whole-body scan is required to confirm or exclude von Hippel Lindau syndrome. Even though the surgical treatment for asymptomatic patients remains debatable, surgical intervention is the only treatment that can lead to the total resetion of the lesion. However, the clinical and neurological condition of the patient is not always improved. A multidisciplinary team is required in order to achieve the best results for the victim of this pathology and the best quality of life.

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