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Cervicomedullary junction intramedullary hemangioblastoma. A 10 years report of cases and review of literature

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ABSTRACT

Hemangioblastoma is a rare, benign, highly vascularized tumour, that usually presents a muriform nodule. The most frequent localization is the posterior cranial fossa. The frequency of this spinal lesion is very low, representing less than 5% of spinal cord tumours. The presentation of hemangioblastoma can widely vary, from a solid tumour to a lesion with a cystic component. We present the case of a 43 years old patient, admitted to our clinic for left hemiparesis, swallowing disorders, and dysphonia. An MRI is performed and it showed a cervicomedullary junction tumour with a solid nodule and an anterior cystic cavity pushing towards the 4th ventricle. The patient underwent surgery for the total removal of the tumour. Post-operative CT confirmed the total ablation of the tumour. The patient is discharged with improved symptomatology. In the last 10 years, in our clinic were admitted 23 cases of hemangioblastoma, 14 males (61% of the patients) and 9 females (39%) with an age at presentation varied from 14 to 78 years (mean 48,2 years). Only 3 patients (13%) out of the 23 had associated von Hippel–Lindau syndrome, with hemangioblastomas also present in other locations. The most common location was the posterior fossa, in 13 cases (56,6%). Headache (69,5%), ataxia (56%) and balance disorders (52%) were the most common symptoms accused by patients. The follow-up varied from 6 to 84 months (mean 20 months). Only one patient died during hospitalization and one tumour recurrence was noted. Most patients improved or remained clinically stable postoperatively.

BACKGROUND

Hemangioblastoma is a rare, histologically benign, highly vascularized tumor, that usually presents an enhancing muriform nodule, having the most frequent localization in the posterior cranial fossa [1].

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Moreover, hemangioblastoma is the most common primary intra-axial posterior fossa tumor in adult population. It is usually identified in the cerebellar hemisphere (including vermis) and brainstem [2], [3], [4]. On the other hand, supratentorial localization is rare, in the literature being described less than 100 cases. Furthermore, only 3-13% are reported in the spinal cord [5].

The presentation of hemangioblastoma can widely vary, from a solid tumor to a lesion with a cystic component. Usually, sporadic hemangioblastoma is a solid tumor with a pseudocapsule. Despite the lack of a proper capsule, the lesion is well circumscribed. The solid nodule is red and very well vascularised, localized close to the pial layer. The cystic cavity usually occurs due to the thinness of the vessel wall that allows water to leak [5], [6]. The walls of the cyst are tapped with non-neoplastic compressed cerebellum cells and the cyst fluid is usually yellow with a high concentration of proteins [6], [7].

Both CT (computed tomography) scan and MRI (magnetic resonance imaging) can be useful for emphasizing the lesion. On CT scan, the mural nodule is isodense with fluid density surrounding the cyst [8], [20]. On contrast scan, cyst walls usually not enhance and the presence of calcification is rare. The MRI scan is superior to the CT scan and it shows vascular signal voids, especially in the periphery of the lesion, as well as, the hemosiderin deposits that can occur due to recurrent hemorrhages [6], [8], [9].

The most common symptoms determined by these tumors are: headaches, hydrocephalus, cerebellar dysfunction and altered mental state.

Surgical approach is the gold standard for sporadic cases of hemangioblastoma, but not for multiple lesions associated with von Hippel Lindau syndrome. The outcome of the surgery is good, given that hemangioblastomas are benign lesions [10]. Despite that tumoral cells may be spreaded through CSF, it still remains a benign tumor. Solid lesions are more difficult to excise compared to cystic ones [11].

Hemangioblastomas are usually sporadic lesions but around 20% of them are associated with Von Hippel Lindau syndrome [5]. Moreover, 6% of cerebellar hemangioblastomas are associated with retinal hemangioblastoma. The retinal hemangioblastoma is usually placed peripherally. It tends to rupture and cause hemorrhage that can lead to retinal detachment.

Von Hipple Lindau syndrome is a multysistem neoplastic condition with autosomal inheritance, described by the occurrence of multiple hemangioblastomas in different localisation such as brain, spinal cord and retina [12]. In these cases, individual resection of the central nervous lesions is not the optimal treatment until it became symptomatic, due to the tendency of reccurence and the inconsistent growth pattern of tumor [13], [14].

Case presentation

We present the case of a 43 years old patient, admitted to our clinic for left hemiparesis, swallowing disorders, and dysphonia. The first symptoms occurred 8 months ago with left intercostal neuralgia.

An MRI is performed and it shows a cervicomedullary junction tumor with a solid nodule and an anterior cystic cavity pushing towards the 4th ventricle, which lead to our presumptive diagnosis of hemangioblastoma. In order to rule out von Hipple Lindau syndrome, a whole-body CT scan was performed. No other lesions were identified. The decision was made to perform surgery in order to remove the tumor.

The patient was placed in the prone position, and a midline incision was made, with a standard occipital craniectomy and resection of the C-1 posterior arch. After the dura mater was opened, the tumor was identified on the midline, at cervicomedullary junction. Under the operating microscope, the hemangioblastoma was dissected, the integrity of the lesion was preserved. The tumor was gently retracted laterally, and the feeding vessels were identified coagulated with bipolar forceps and divided sharply. The main feeder is exposed, clipped, coagulated, and cut. The tumor was sent for pathologic examination and the diagnosis of hemangioblastoma was confirmed. Closure of the surgical wound was performed respecting the anatomical planes and leaving an epidural external drain in place. Finally, the skin suture was draped in a sterile fashion. After careful hemostasis, the surgical wound is closed layer by layer. No perioperative incidents were reported. The post-operative MRI confirmed the total resection of the tumor.

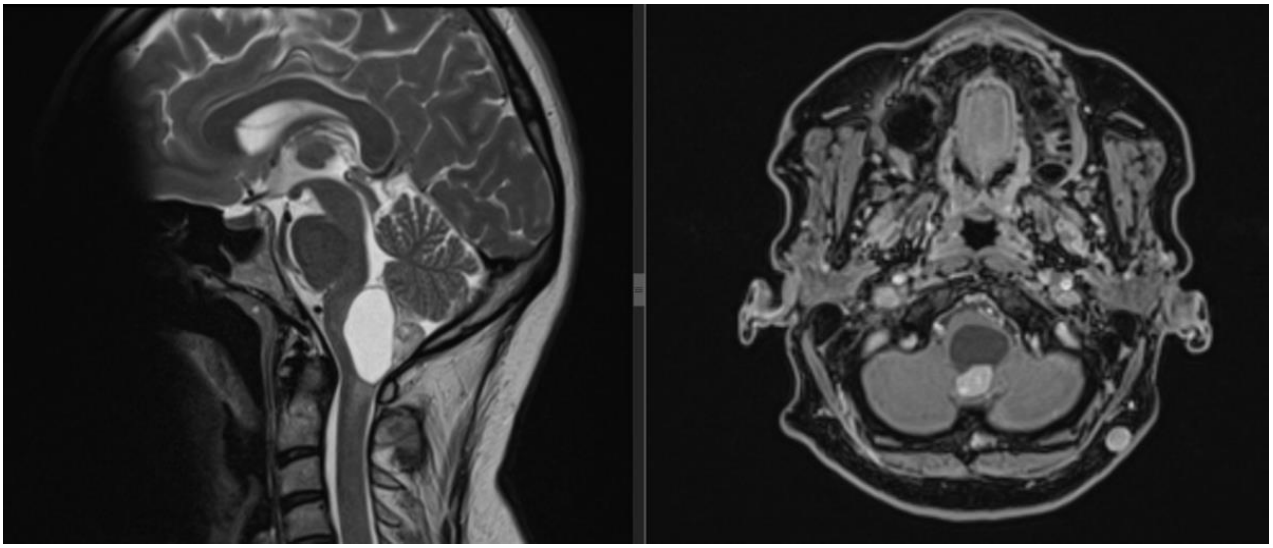

Figure 1. Cervicomedullary junction tumor with a solid nodule and an anterior cystic cavity pushing towards the 4th ventricle.

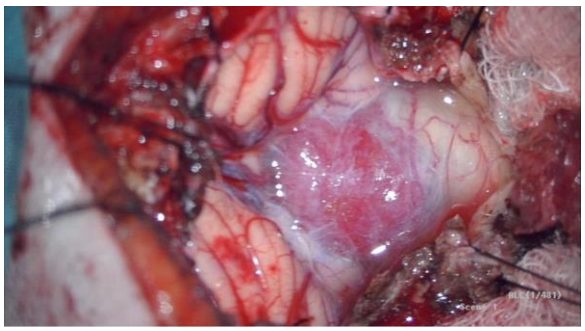

Figure 2. The cervicomedullary junction lesion, after dura mater was opened.

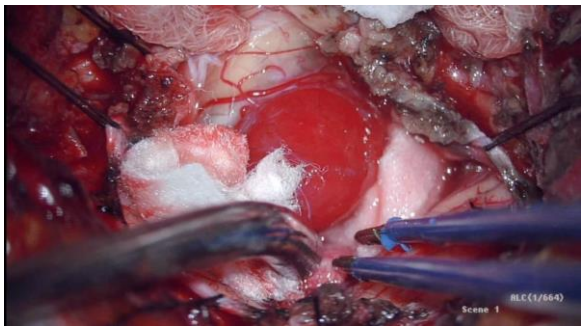

Figure 3. Gently dissection of the lesion. The feeders are coagulated and sharply cut.

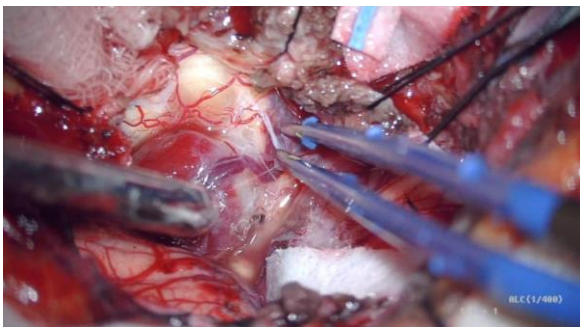

Figure 4. After circumferential dissection, the main feeder is exposed.

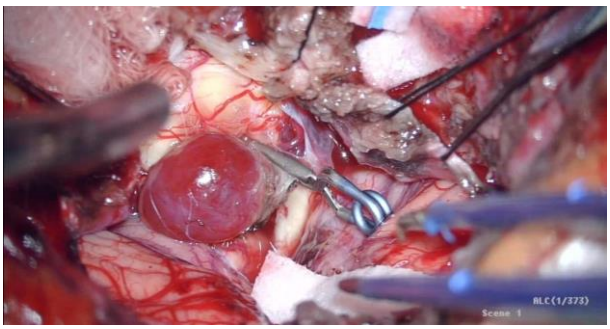

Figure 5. The main feeder is clipped and cut. The tumor is removed "en bloque".

The external drain was removed 24 hours later. Symptomatology was improved in the first week postoperative by the patient. Mobilization of the patient was allowed 24 hours after the surgery. The skin sutures were suppressed at seven days postop.

This is just one out of 23 cases we had in our clinic in the last 10 years. 14 men (representing more than 60% of the study group) and 9 women. Their median age was 48, ranging from 14 to 78 years. The average time of hospitalization was 15 days, between 7 to 20 days.

Headache and ataxia were the most common symptoms accused by patients, accused by more than 70% of the patients. Other symptoms were dysphonia, dysphagia, seizures, cecity and hearing loss.

Table 1. Table of the main symptoms accused by patients

Symptom	Cases	Percentage
Headache	18	78,2
Ataxia	17	73,9
Dysphonia	2	8,69
Dysphagia	2	8,69
Seizures	2	8,69
Loss of visual acuity	1	4,34
Hearing loss	1	4,34

MRI was performed in all cases. It is worth mentioning that in 17 cases hemangioblastoma presented a cystic component. Also, in 3 cases were identified multiple lesions. There were 16 cases of intracranial hemangioblastoma, 2 with supratentorial location (frontal and parietal lobe), and in 14 cases, the tumor was located in the posterior fossa, in the cerebellum. Just 4 craniospinal junction cases were identified. At the spine level, there were 3 tumors, 2 located in the thoracic spine and 1 in the cervical spine.

All of the patients underwent surgery. In 22 out of 23, complete ablation of the tumor was performed. In just one case, total excision was not possible, and a subtotal ablation was performed. The postoperative evolution was good, without complications in 20 cases. 3 patients developed hydrocephalus and in one case a recurrence occurred, a few months after the surgical intervention. Postoperative follow-up was between 6 months to 7 years, with a mean follow-up of 20 months. Neurological improvement was noted in 18 cases, whereas in 2 cases, the neurological state remained stationary and in 3 cases, the neurological status had worsened. One patient died.

As we mentioned before, hemangioblastoma can be associated with von Hipple Lindau syndrome, a rare genetic disorder with multisystem involvement. It is characterized by visceral cysts and benign tumors with the potential for malignant transformation. 3 patients had associated this syndrome with pancreatic and kidney cysts.

Discussions

One of the greatest complications of hemangioblastomas is the risk of hemorrhage. However, the risk of hemorrhage is lower for the lesions of the spinal cord, where catastrophic neurological impairment is the main risk [6], [15], [16]. In 2007, Dr. Cornelius et al [17] compared the outcome of perioperative embolization of hemangioblastomas for spinal and cerebellar lesions. The result of embolization was favorable in patients with spinal cord hemangioblastomas, but, for the hemangioblastomas located at the cerebellar level, there were reported acute tumoral bleeding that led to death of the patients. The outcomes following embolization are very different for these two locations possibly because of the different capillary sizes. Due to high mortality, the procedure is no longer used.

In some cases, due to the clinical state of the patient or the localization of the tumor, surgical intervention cannot be achieved. In 2020, Mak et al[18] published the case of an immunocompetent patient that presented with a one-year history of progressive nausea and vomiting. After an MRI scan, a homogeneous enhancing intra-axial mass located at cervicomedullary junction was revealed. The surgical removal attempt failed due to multiple bradycardia episodes. However, a biopsy was performed and it confirmed that the lesion was a hemangioblastoma. Conservative treatment with bevacizumab, a humanized monoclonal antibody that targets vascular endothelial growth factor (VEGF) was initiated and the neurological status of the patient had been improved and the size of the tumor remained stable. Moreover, the reduction of the surrounding edema was noticed. Hemangiobalstomas are highly vascularized tumors with a rich capillary network. The endothelial cells express the VEGF receptor. Based on the histopathological characteristics, new treatment approaches are proposed for patients that are not suitable for surgical intervention.

In the last couple of decades, stereotactic radiosurgical treatment represented an alternative for the surgically inaccessible locations or for multiple localisations that typically occur in von Hippel-Lindau disease. Moss et al [13] published their almost 20 years experience of radiosurgical treatment for hemangioblastomas. Their study comprehends 92 hemangioblastomas from 31

patients, 26 of them being diagnosed with von Hipple Lindau syndrome. The mean age of the patients was 41 years. The radiation dose used to the periphery of the tumor average 23,4 Gy with a mean tumor volume of 1,8 cm3. The tumoral response was measured with contrast-enhanced computed tomography scans. In 62% of the cases, the tumor volume remained stable, in 22% the tumor shrank, and 16% of the cases, hemangioblastoma contiuned to grow. These results emphasized that stereotactic radiosurgical treatment is a safe and effective alternative to open surgery.

Even though there are multiple approaches for the treatment of hemangioblastoma, surgical treatment remains a safe option for patients with a good outcome. In 2020, Xiangdong Yin et al [19] published a meta-analysis composed of 13 studies that included 473 cases. In this paper, they analyzed the surgical outcomes, including gross total resection, mortality, neurological morbidity, and functional outcome. Gross total resection was performed in 98% of the cases. In our study, total resection was achieved in 95,6% of the cases (22 out of 23). Neurological improvement was reported in 85% of the cases in their study, compared to our clinic, where 86% of the patients reported an improved neurological state. Mortality was also similar between the statistics of meta- analysis and our clinic. In the mentioned study, mortality was 4% (23 out 473) compared to the 4,4% (1 out 23) mortality rate in our clinic. Xiangdong Yin et al postulated that surgical treatment for hemangioblastomas is still feasible and effective, a statement supported by good statistical outcomes from both their study and from our clinic experience.

Conclusions

Hemangioblastoma is a benign, highly vascularized tumor that usually occurs in the posterior fossa but its localization may widely vary. In the most cases, total resection is possible with a significant improvement of the neurological state of the patient. The rate of recurrence is low, even though tumoral cells can be spread by CSF. Nowadays there are multiple treatment options and every case should be carefully investigated in order to choose the best approach for improving the quality of the patient`s life.

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