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Case Report

Cerebellar Hemangioblastoma: The Reality of Management in Madagascar

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Abstract

Introduction: Hemangioblastomas are rare tumors of the central nervous system located preferentially in the cerebellum. The objective of this work is to describe the reality of the management of a cerebellar hemangioblastoma according to the local means available in Tamatave through this first operated case.

Observation: It was a 53 years old woman who had an emergency entry for intracranial hypertension syndrome associated with cerebellar syndrome. This patient had left cerebellar hemangioblastoma confirmed by pathological examination. She underwent emergency ventriculo-peritoneal shunt and secondary tumor resection.

Conclusion: Management of cerebellum hemangioblastoma is possible; but it is difficult in a center with a limited technical platform like ours.

Keywords: *Hemangioblastoma; Madagascar; Management*

Introduction

Hemangioblastomas are vascular tumors of the nervous system. Rare tumors represent only 2.34% of all tumors of the central nervous system and only 1.35% of intracranial tumors. Hemangioblastomas are hyper vascularized, benign histological tumors and preferentially at the level of the posterior fossa (92.6%) [1]. They affect the young adults with a mean age of 34.9 years of age for cerebellar locations [2]. The objective of this work is to describe the reality of the management of a cerebellar hemangioblastoma according to the local means available in Tamatave through this first case.

Case Report

We report the case of a 53 years old woman with dizziness developed over one month associated with jet vomiting and diffuse headache rebellious to usual analgesics, all in connection with signs of intracranial hypertension. She had no particular personal or family antecedents.

On physical examination, the patient was confused with a Glasgow score 14/15, the pupils were symmetrical, equal and reactive. She had a cerebellar syndrome with an ebbing and widening of the levitation polygon, dysmetry, and dysarthria. Romberg's maneuver was negative. No signs of neurological deficit or signs of involvement of the cranial nerves were found.

Biologically, we did not find polycythemia. In the absence of magnetic resonance imaging (MRI), we performed a CT scan that found in the posterior fossa, an expansive process of cystic nature with wall nodule enhanced intensively and early after injection of the product. Contrast in left lateral cerebellar evoking in the first place a hemangioblastoma. He was responsible for compression of the fourth ventricle with upstream ventricular tricephalic hydrocephalus (Figure 1). Thus, we retained the diagnosis of a hemangioblastoma in front of its clusters of clinical and paraclinical arguments.

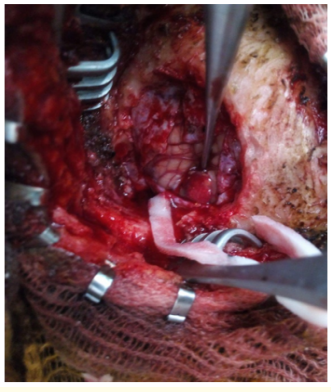

Figure 1: *Intraoperative view of a left cerebellar hemangioblastoma after cyst collaps.*

Compared to the treatment, initially a ventriculo-peritoneal by a non-adjustable medium pressure valve (the only available on site) was practiced the same day. The postoperative course was marked by an improvement of the vigilance of the patient, against the cerebellar syndrome persisted. Subsequently, we performed a tumor resection nine days after ventriculo-peritonial shunt. This surgery was performed under general anesthesia in right lateral decubitus, under an operating microscope. After opening the dura mater, the arteries and draining veins were identified and the feeding artery was well defined and then coagulated. Bulk resection could be performed (Figure 2); this was facilitated by the drainage of the peri-tumoral cyst. The postoperative course was simple. Anatomopathological examination of the operative specimen confirmed the vascular nature and the benign nature of the tumor. The histological aspect was that of a hemangioblastoma (Figure 3).

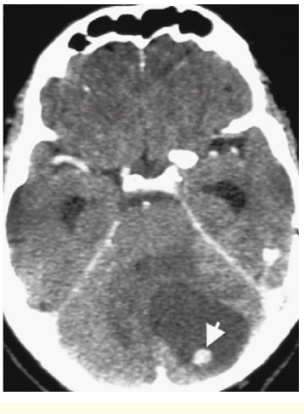

Figure 2: *Expansive process of spontaneously hypodense tumoral appearance of cystic pace measuring 38 X 28 X 27 mm, associated with a strongly raised wall nodule after contrast product: hemangioblastoma.*

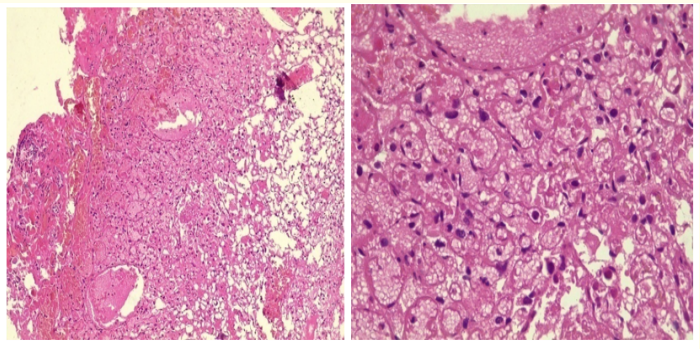

Figure 3: *Tumor proliferation, composed by polyhedral cells, sometimes spindle-shaped, with networks of capillaries anastomosed. The presence of balloon and microvascular stomal cells is noted. The histological aspect is that of a hemangioblastoma.*

After two months follow-up, a complete disappearance of the cerebellar syndrome and dysarthria was reported. Eighteen months after the surgery, the patient has no longer any symptoms related to hemangioblastoma and the systematic assessments carried out of point to a sporadic localization of the latter.

Discussion

Hemangioblastomas can occur in Von Hippel Lindau disease (VHL) where they are seen in 40% of patients; or sporadically [3-5]. Preferential localizations are the cerebellum and the spinal cord [6]. Supra-tentorial localization is rare [7]. In the context of VHL disease, which a hereditary inherited autosomal dominant disease is predisposing to the development of hemangioblastomas, they can be observed in 40% of patients. These can reach the central nervous system (60 to 80% of patients) and the retina (50%), the endolymphatic sac (5 - 11%). One can also observe renal cell carcinomas clear (50 - 75%), pheochromocytomas (25%) and cysts (75%) and pancreatic endocrine tumors (12%) [4,8].

The positive diagnosis of hemagioblastoma is radiological and is made by magnetic resonance imaging (MRI) which is the gold standard. It is characterized by the presence of a nodular image strongly enhance after injection of contrast medium. In case of doubt about the vascular architecture surrounding the fleshy portion of the tumor, arteriography is also an examination of choice because it allows doing at the same time pre-operative embolization when the tumor is highly vascularized, thus increasing the safety surgical excision [9]. In our case, in the absence of MRI and cerebral arteriography, the cerebral scanner allowed us to suspect it in front of a wall nodule image intensely and early enhancement after injection of contrast medium but the analysis of the arterial inferences and venous drainage remains difficult.

As for the management, in front of a communicating triventricular hydrocephalus (on an obstacle at the level of the passage of the cerebrospinal fluid in the fourth ventricle), an endoscopic ventriculocysternostomy remains the treatment of first intention: it has a good long-term efficiency especially for adults and low morbidity [10]. This makes it possible to avoid setting up a ventriculoperitoneal shunt system which is a foreign body that is not devoid of any mechanical and/or infectious complications. In addition, after surgical excision of the fleshy potion and collapse of the cystic portion of the cerebellar hemangioblastoma, the circulation of the cerebrospinal fluid could be reestablished so ventriculoperitoneal shunt system would no longer be useful. In our patient, given the absence of a neuroendoscope, we have implemented the only type of system available on site with a non-adjustable medium pressure valve. But normally, for adults, an adjustable valve system would change its opening pressure based on its clinical and radiological effectiveness. Surprisingly we have a good efficiency of the system and we did not observe any signs of intolerance or complications until 18 months of retreat.

The genetic confirmation test for VHL disease is not available on the spot, this prompted us to include in our semi-annual surveillance report: an ophthalmological examination for retinal angioma, an otoneurological and audiological examination at the search for

hypoacusis secondary to tumors of the endolymphatic sac; measuring blood pressure and heart rate for pheochromocytoma; an ultrasound of the abdomen looking for renal, pancreatic, or adrenal localization. An MRI of the nervous system or of the abdomen as well as the determination of catecholamine metabolites in the urine should be performed in the capital (Antananarivo) if there is suspicion [11].

Total block excision of the fleshy tumor potion remains the optimal treatment for cerebellar hemangioblastoma in sporadic hemangioblastomas [12]; which has been the case for our patient despite local means available. This explains the good neurological evolution and the absence of recurrence at 18 months of retreat.

Conclusion

Management of hemangioblastoma of the cerebellum is possible; but difficult in a center with a limited technical platform like ours. The brain CT scan with contrast medium still has a place in the positive and topographic diagnosis of this tumor. Magnetic resonance imaging, cerebral arteriography and neuroendoscopy should be introduced to facilitate and optimize this management.

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