Hemangioblastoma of the lateral ventricle: seventh reported case in the literature

Hemangioblastoma do ventrículo lateral: Relato do sétimo caso na literatura

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ABSTRACT

Introduction: Hemangioblastomas are capillary-rich benign tumors mainly found in the infratentorial compartment. When at the supratentorial space, they are rarely located at the lateral ventricle. We report on a case of a 25 years-old female patient treated at our institution. Case report: The patient came to our service with a three months history of progressive headache, bilateral visual loss, and right hemiparesis. Brain CT-scan and MRI showed a heterogeneous intraventricular contrast-enhancing lesion at the left lateral ventricle. Von Hippel-Lindau was investigated but she had neither family history of the disease nor any signs of it. She was submitted to microsurgical tumor resection with a parieto-occipital craniotomy via the intraparietal sulcus, with an uneventful recovery. Histological examination confirmed the diagnosis of hemangioblastoma. The patient is in outpatient clinic follow-up completely integrated to her familiar and professional life.

Discussion: According to a PubMed search, this is the seventh case of lateral ventricle hemangioblastoma reported on English literature. It has a cystic characteristic that differs from other cases, and a bigger volume compared to the previous ones.

Keywords: cerebral tumours, hemangioblastoma, lateral ventricle

INTRODUCTION

Hemangioblastomas are benign tumors of unknown origin characterized histologically by many capillaries, and are composed of three cell types: endothelial cells, pericytes, and stromal cells 1. It is the most frequent vascular tumor in the central nervous system 11, representing 1,5 to 2,5% of all intracranial primary tumors, and is associated to von Hippel-Lindau disease in 30% of cases 10. Hemangioblastomas usually are infratentorial lesions, occurring at the supratentorial compartment in only on 4 to 13% cases 2. Localization in the lateral ventricle is very rare with few reports on English literature. We present the clinical presentation, radiological features, surgical approach, and postoperative follow up of a 25 years-old female patient treated by our team.
CASE REPORT

A 25 years-old female patient presented to our service complaining of a 3 months-history of progressive headache, bilateral visual loss, somnolence, and right hemiparesis. Brain CT-scan and MRI showed a heterogeneous intraventricular contrast-enhancing lesion in the left lateral ventricle with cystic areas of 4 x 4 x 5 cm, compressing and dilating the left ventricle (Fig.1). Investigation with imaging and laboratory exams showed no sights of von Hippel-Lindau syndrome and also no family history. She was submitted to a parieto-occipital craniotomy followed by microsurgical tumor resection by a small corticotomy at the intraparietal sulcus, accessing the ventricular atrium and lateral ventricle with minimal brain retraction. The tumor was microscopically dissected around its periphery, progressively decompressed, and removed en bloc, but not enucleated, due to the risk of bleeding. She had an uneventful recovery without additional deficits (Fig. 2). Hystological examination confirmed the diagnosis of hemangioblastoma (Fig. 3). In her last outpatient follow-up, she still had a grade IV+ right hemiparesis, and completely reintegrated to her personal and professional life. The patient agreed with this case publication by informed consent.

Supratentorial hemangioblastomas are being more frequently reported on in the literature and its occurrence is not as rare as previously believed. The pituitary stalk is the most common site of supratentorial hemangioblastomas. A PubMed search revealed only five case reports of surgically treated symptomatic lateral ventricle hemangioblastomas and one case diagnosed by Vecchi on a post-mortem study. Considering this search, this is the seventh report of a lateral ventricle hemangioblastoma in English literature.

The previous articles presented big and solid lateral ventricle hemangioblastomas, all of them without cystic components. The case we present has cystic areas, and a bigger volume compared to the biggest tumor previously reported, 80 cm³ and 62.5 cm³, respectively. All previous cases were treated by transcortical approaches through temporal or parietal craniotomy. Two of these five operated cases presented postoperative mutism, justified by the author as caused by tumor proximity to the thalamic posterolateral area and to the crus of the fornix, or to postoperative fronto-temporo-parietal edema at Wernicke’s speech area. We have not noticed this complication in our case nor the visual deficits commonly associated to transtemporal approaches, most probably due to the use of a parieto-occipital craniotomy and microscopic approach through the intraparietal sulcus with small corticotomy.
Radiologically, hemangioblastomas may resemble brain metastases, gliomas, meningiomas, and other intraventricular lesions, so neurosurgeons who deal with intracerebral tumours must keep them in mind. Histopathologically, they may be confused with angiomatous angioblastic meningioma and renal cell carcinoma. Hemangioblastoma association to von Hippel-Lindau syndrome justifies the indication of a CT scan in any patient presenting with the referred syndrome. Considering all six previously reported cases of lateral ventricle hemangioblastoma, only one, as the one we are presenting, was not associated to von Hippel-Lindau disease.

REFERENCES