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Hemangioblastoma of the lateral ventricle: case report and review of the literature

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Summary

We report a unique case of hemangioblastoma of the lateral ventricle in a 73year-old man with cognitive deficits and fluent dysphasia. He harboured an intraventricular tumor, placed at the trigone of the left lateral ventricle. The tumor was successfully excised, by means of a temporal craniotomy. The patient became mute in the immediate postoperative with restoration of speech within a few days. The literature has been reviewed and only three other similar cases have been reported. Discovery of lesions in such unusual location should raise a high degree of suspicion for von Hippel-Lindau disease.

KEY WORDS: Hemangioblastoma. Lateral ventricle. Von Hippel-Lindau disease. Mutism.

Hemangioblastoma del ventrículo lateral: caso clínico y revisión de la literatura

Resumen

En este trabajo describimos el caso de un paciente de 73 años con déficits cognitivos y disfasia sensitiva y que fue diagnosticado mediante tomografía computarizada y resonancia magnética cerebral de una lesión tumoral situada en la encrucijada ventricular izquierda. La lesión se abordó quirúrgicamente a través de una craneotomía temporal, realizándose una extirpación total. El estudio anatomopatológico confirmó el diagnóstico de un hemangioblastoma. Durante la evolución postquirúrgica el paciente presentó un cuadro de mutismo transitorio. Una revisión exhaustiva de la literatura ha evidenciado la existencia de tan sólo tres casos similares descritos previamente. El diagnóstico de este tipo de lesiones en una localización tan infrecuente debería hacernos sospechar la enfermedad de von Hippel-Lindau.

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PALABRAS CLAVE: Hemangioblastoma. Ventrículo lateral. Enfermedad de von Hippel-Lindau. Mutismo.

Introduction

Hemangioblastomas of the central nervous system (CNS) are the most frequent vascular tumors[19,22]. They are histologically benign tumors representing 1-2% of all intracranial neoplasms[5,14,24] but when posterior fossa tumors are considered alone, the percentage rises to 7.3[15]. Although they are usually isolated tumors, sometimes they can be associated with von Hippel-Lindau (VHL) disease in 3 to 38% as a major manifestation[3,14]. This inherited phakomatosis is clinically characterized by the formation of vascular tumors, including retinal and CNS hemangioblastomas. Other features include cyst of the kidneys, liver and pancreas, clear renal cell carcinomas, pheocromocytomas and endolymphatic sac tumors. It is thought to be a dysembryogenic defect of the mesenchyme occurring in about the third month of fetal life and affecting various developing organs in the body simultaneously. In 1993, the VHL gene was localized on the shorter arm of chromosome 3 (3p25-26)[10].

Although hemangiobastomas are predominantly found in the cerebellum, spinal cord and brainstem[3,24], some unusual locations have been reported[16,18]. Supratentorial location account for 4% to 13%[3]. However, hemangioblastomas of the lateral ventricle are very rare indeed. The first described case was an incidental finding during a postmorten examination[21]. Afterwards only three symptomatic cases have been published[6,9,12].

Case Report

This 73 year-old-man presented with a few days history of cognitive deficits and confusion without any other complain. Neurological examination disclosed a Glasgow Coma Scale of 14 (E, 4; V, 4 and M, 6) and fluent dysphasia. He had a history of a previous surgical procedure for

Abreviaturas. VHL: Hippel-Lindau disease.

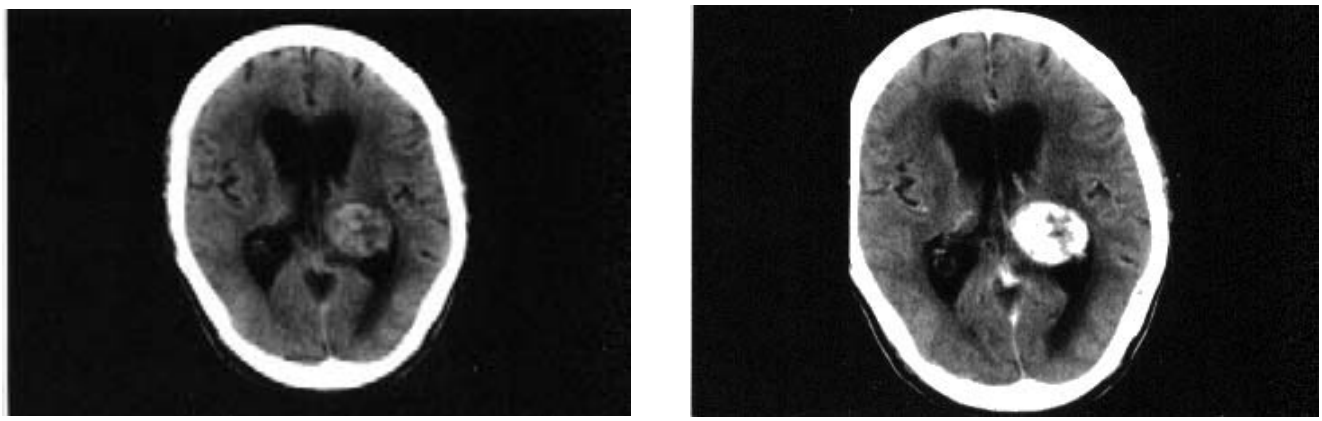

Fig. 1. *Preoperative computed tomography scan. Left: Plain scan showing and intraventricular tumor situated at the trigone of the left lateral ventricle with dilatation of the ipsilateral trigone and temporal horn; Right: Enhanced sean showing a homogeneous contrast enhancement.*

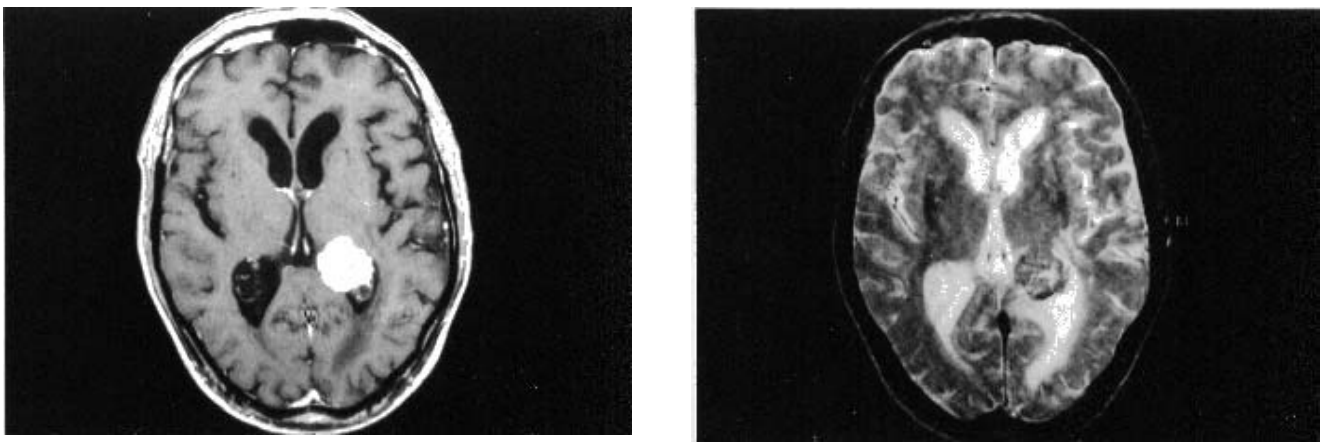

Fig. 2. *Preoperative magnetic resonance imaging. Left: T1-weighted sagittal section after gadolinium-DTPA infusion showing a solitary, solid, rounded and well-enhanced intraventricular mass 3 cm in diameter. Right: T2-weighted showing a hypo-isointense mass compared with the normal white matter with central hypointense areas.*

the removal of a renal cell carcinoma ten years ago. Laboratory investigations revealed a hemoglobin of 15.5 g/dL and a hematocrit of 46.2%. None of his relatives had had any relevant illness or any lesion of the VHL disease.

Computed Tomographic (CT) scan showed an intraventricular tumor placed at the trigone of the left lateral ventricle with dilatation of the ipsilateral trigone and temporal horn. It enhanced markedly with contrast medium (Fig. l). The posterior fossa was normal.

On Magnetic Resonance Imaging (MRI) the tumor had a heterogeneous signal. lt was hypo-isointense on T1-weighted images compared with the normal white matter and hypo-isointense on T2 weighted images with central hypointense areas. Long Repetition Time (TR) sequences showed periventricular edema. Gadolinium-enhanced T1-weighted MRI demonstrated a solitary, solid, rounded intraventricular tumor, 3 cm in diameter, placed at the trigone of the left lateral ventricle (Fig.2). The approximated volume of the lesion was 13.5 cm^3 which was calculated by a modified ellipsoid formula (length x width x height x 0.5).

Magnetic resonance angiography demonstrated a large vascular tumor fed by branches of the P2 segment of the posterior cerebral artery (Fig.3).

On June 2003, the patient underwent a left temporal craniotomy. Intraoperative ultrasonography was performed to confirm the precise location of the tumor. A 3 cm horizontal corticotomy was made over the middle temporal gyrus. This gave direct access to the left temporal horn. The choroids plexus was covering a reddish vascular tumor. The lesion was foamy and had an important vascularity. With careful dissection under the microscope, blood supply coming from the posterior choroidal vessels and the ependyma was interrupted. A large vein draining to the deep venous system was coagulated. It was relatively well demarcated except the anterior area, where the limits were somewhat imprecise. The whole tumor was then removed.

The patient became mute in the immediate postoperative period and control CT showed a large frontal pneumo-

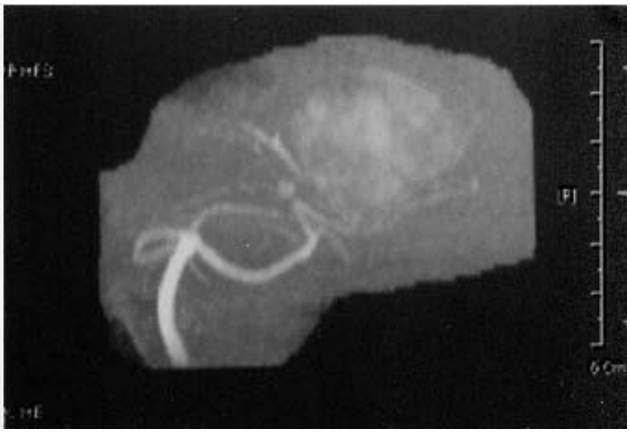

Fig. 3. *Magnetic resonance angiography demonstrating a vascular tumor fed by branches of P2 segment (posterolateral choroidal arteries).*

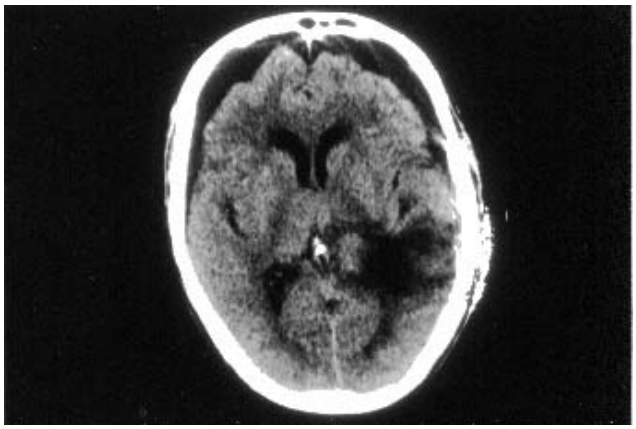

Fig.4. *Early postoperative computed tomography scan, demonstrating complete tumor removal and large pneumoencephalus.*

encephalus and the complete removal of the tumor (Fig.4). Twelve days after the operation, the speech returned but a mixed dysphasia was present. He improved in terms of cognitive disturbances but presented gait ataxia. Serial CTs demonstrated progressive disappearance of pneumoencephalus.

Histopathological examination showed a tissue composed of a fine network of blood spaces separated by numerous polygonal stromal cells, with lighty stained cytoplasm. Immunohistochemical staining showed a strong immunopositivity in several stromal cells for vimentin and EMA (Epithelial Membrane Antigen). The intercapillary component was immunopositive for endothelial cell markers CD31 and CD34. In addition, immunohistochemical expresion of a cell cycle related antigen (MIB-1) showed a low proliferative kinetic. It was typical of a bening hemangioblastoma.

Following the protocol of the National Institutes of Health Screening we performed an additional evaluation. It included ophthalmologic examination, CT scanning of the abdomen and 24-hour urinary analysis for vanillymandelic acid, metanephrines, and total catecholamines. The result of the entire screening workup was negative.

Discussion

Hemangioblastomas of the CNS are infrequent and they account for approximately 2% of primary CNS tumors[14,19,24]. The most frequently affected site is the posterior fossa with the cerebellar hemispheres as the most common location (83%), followed by the spinal cord and brainstem[14]. Supratentorial hemangioblastomas are rare, they account for 4% in patients with sporadic disease and 13% in VHL disease[3].The intraventricular location is very unusual and only three symptomatic lesions[6,9,12] and an incidental postmorten finding[21] have been described to date (Table l).

According to Melmon and Rosen's criteria[11], the patient's case can be classified as VHL disease, because he was previously operated of a renal cell carcinoma. lnterestingly, all lateral ventricle hemangioblastomas have been seen in patients with VHL. So a high degree of suspicion for VHL disease should be raised in patients with hemangioblastomas in that location. In addition spinal hemangioblastomas, multiple lesions[3] and leptomeningeal hemangioblastomatosis[16] are diagnostic of this condition. Within these non-invasive tumors, symptom production depends on lesion size and location. It has been reported that at the time of surgery cerebellar hemangioblastomas (mean tumor volume 3.4+6.8 cm^3) were much larger than those lesions in the brainstem (0.7±0.5 cm^3) or spinal cord (0.8+1.8 c m^3)[23]. All the lesions reported to be lying in the lateral ventricle had a greater volume, between 13.5 and 62.5 cm^3. lt is likely that lesions in the lateral ventricle need major volume to produce symptoms, because there is much space available to accommodate a mass effect in this anatomical compartment. Cysts have been frequently associated with tumor size either in cerebellum, brainstem or spinal cord, but it is noteworthy that the lateral ventricle lesions were solid, in spite of their large volume.

Mutism, developed by our patient in the postoperative period, is a state in which the subject is conscious but unable to speak[4]. Postoperative mutism is an infrequent and usually transient complication of surgery. It is most commonly seen following extensive callosotomy, ventrolateral thalamotomy (especially when performed bilaterally) and resection of supplementary motor area or posterior fossa lesion[4,7,17]. These areas are connected via dentatothalamocortical pathways and it appears that bilateral disruption of the elements of this pathway may be responsible for this

Table 1
Summary of clinical patient data in the reported cases of hemangioblastomas of the lateral ventricle

Author, Year & Reference	Age & Sex	Clinical presentation	Lesions associated with VHL disease	Radiological evaluation	Volume (cm3)	Number & Nature	Procedure & Resection	Postoperative recovery
B. Vecchi and G. Patrassi, 1935[21]	80, F	Incidental finding during postmorten examination.	?	None.	?	Multiple Solid	None	None
PR. Diehl arad L. Symon, 1981[6]	20, M	7-week history of progressive visual loss, headache and bilateral papilledema.	Retinal angiomatosis, epididymal cyst.	CT: right temporal tumor and dilated temporal horra. Heavy enhancement after contrast injection.	62.5	Solitary Solid	Through middle temporal gyrus. Total removal.	Transient partial left homonymous field defect
H. Murakami, S Toya et al 1985[12]	31, F	Headache and gait disturbance.	Positive family history.	CT: right cystic cerebellar lesion and a right intraventricular lesion.	?	Multiple ?	?	?
Y-S. Ho and C. Plets, 1990[9]	44, F	8-year history of depressive illness. In the last months, moderate bioccipital headache.	Positive family history.	CT and MRI: IVT at the right-TLV with dilatation of the ipsilateral temporal and occipital horras. Heavy enhancement.	13.5	Solitary Solid	Through posterior temporal lobe. Total removal	Left homonymous lower quadrantanopsia.
Present report, 2004	73, M	Few-days history of cognitive deficits and confusion.	Renal cell carcinoma.	CT and MRI: IVT situated at the left-TLV with dilatation of the ipsilateral ventricle arad showing heave. Heavy enhancement	13.5	Solitary Solid	Through the middle temporal gyrus. Total removal	Transient mutism.

VHL=von Hippel-Lindau; F=female; M=male CT=computed tomography; MRI=magnetic resonance image; IVT=intraventricular tumor; TVL=trigone of the lateral ventricle; ?=data not provided.

phenomenon[4]. It is also in relation to damage to the limbic system, caused by lesions in the septum pellucidum or both fornices[1,13]. Single photon emission tomography studies reported that patients with postoperative cerebellar mutism disclosed a marked reduction of cerebral perfusion in the fronto-parietal region[8,20]. We reviewed the literature looking for reports on patients with temporal transcortical surgery for tumors at the trigone of the lateral ventricle and postoperative mutism and did not find any report to elucidate our disappointing fact. The closeness between the tumor and the posterolateral region of the thalamus and the crura of fornix and/or the edema or ischemia in the fronto-temporo-parietal region, caused by surgical manoeuvres, could have been the main causes triggering this speech disturbance.

Conclusion

Hemangioblastomas of the lateral ventricle are exceptional tumors and their discovery should raise a high degree of suspicion for VHL disease.

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