

CASE STUDY

Hemangioblastoma of the Fourth Ventricle Mimicking a Posterior Fossa Medulloblastoma in a Child

Karim Baayoud*, Ismael Abdou Mahazou, Salma Lahlou, Chaimaa Amry, Abdelmajid Chellaoui, Abdelkoudous Laaidi, Abdessamad Naja, and Abdelhakim Lakhdar

ABSTRACT

Hemangioblastomas are benign vascular tumors of the central nervous system, rarely affecting children with an incidence of less than 1 per 1,000,000. The development of an intraventricular hemangioblastoma is exceptional, just a few cases have been reported in adults. We report a case of hemangioblastoma of the fourth ventricle in a 9-year-old girl without a pathological history. Revealed by rapidly progressive signs of intracranial hypertension and a static cerebellar syndrome. Magnetic resonance imaging (MRI) of the brain showed a homogeneous mass of the fourth ventricle measuring $44 \times 36 \times 32$ mm. We first performed a ventriculoperitoneal shunt, and after a few days, we proceeded to a partial resection of this lesion. A histopathological study confirmed the diagnosis of hemangioblastoma. Pediatric hemangioblastoma (PHB) of the fourth ventricle is the only case reported in the literature. This diagnosis can be evoked in front of a solid lesion of the fourth ventricle, especially in the context of von Hippel-Lindau's disease.

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Department of Neurosurgery, Ibn Rochd University Hospital Center, Morocco.

*Corresponding Author:
e-mail: kbd9684@gmail.com

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1. INTRODUCTION

Hemangioblastomas are benign vascular tumors of the central nervous system, rarely affecting children with an incidence of less than 1 per 1,000,000 [1], [2]. This tumor is preferentially located in the cerebellum, spinal cord and brainstem [3]. The intraventricular topography is extremely rare, only 3 case reports of hemangioblastoma of the fourth ventricle in adults have been described in the literature [4]. However, to our knowledge, hemangioblastoma of the fourth ventricle in children has never been reported. Here, we describe the first case of "hemangioblastoma of the fourth ventricle mimicking a posterior fossa medulloblastoma in children".

2. CASE REPORT

A 9-year-old girl with no medical history, presented for 1 month a rapidly progressive onset of headaches, associated with vomiting. Ten days before her admission to our department, she presented a visual impairment and diplopia. On examination, she had a static cerebellar syndrome and no swallowing disorder. The ophthalmological

examination revealed a drop in her visual acuity, more pronounced on the right side (5/10 on the right and 9/10 on the left) associated with a bilateral papilledema.

The brain computed tomography (CT) showed a slightly hyperdense rounded mass, measuring 4.4 cm in large diameter, well limited within the fourth ventricle associated with perilesional edema. The lesion enhanced intensely after injection of the contrast. This mass was responsible for a triventricular passive hydrocephalus (Fig. 1).

The magnetic resonance imaging of the head revealed a mass in the fourth ventricle measuring $44 \times 36 \times 32$ mm. This mass had an intermediate signal on T1-weighted and was iso-intense on T2-weighted images. The T1-weighted imaging following gadolinium injection demonstrated a homogeneous enhancement of the lesion. The intraventricular mass is responsible for a triventricular hydrocephalus (Fig. 2).

Spinal MRI and thoracoabdominal CT were carried out in search of other associated lesions, everything was normal.

We first performed a ventriculoperitoneal shunt (Fig. 1D) to relieve not only the intracranial tension but also to ease later tumor removal. After 2 weeks, the patient



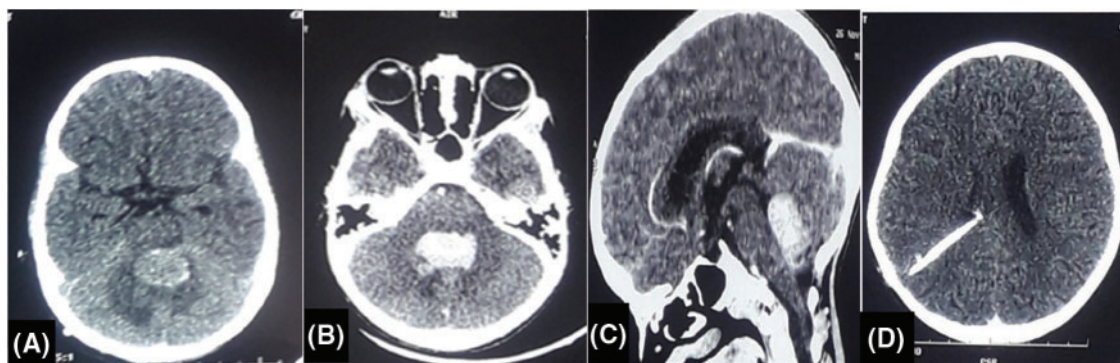


Fig. 1. Brain CT scan (A) shows a spontaneously hyperdense mass; contrast-enhanced CT scans (B) and (C) demonstrate intense homogeneous enhancement of this lesion into the fourth ventricle; (D) postoperative brain CT scan shows the ventricular catheter in the occipital horn of the right lateral ventricle.

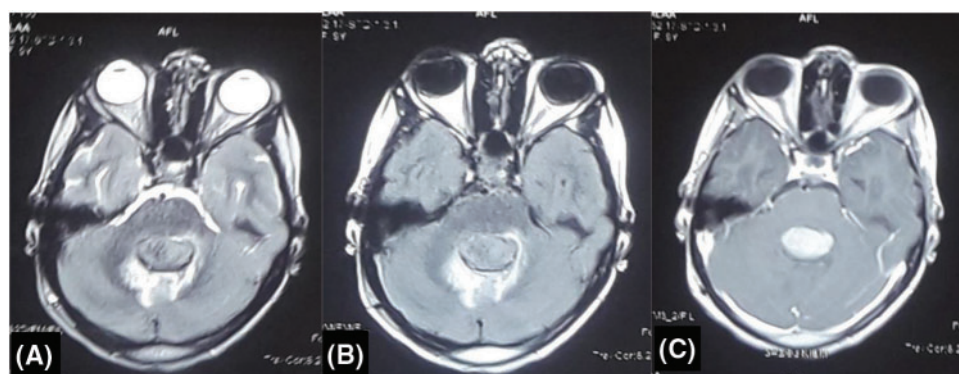


Fig. 2. Preoperative MR images of axial T2-weighted (A) axial FLAIR-weighted (B) and axial (C) postcontrast T1-weighted showed a homogeneous mass located into the fourth ventricle that enhances after gadolinium.

proceeded to the resection of the tumor. We have opted for a telovelar approach. The skin incision was performed from the inion down to the level of C2/C3. We carried out a midline suboccipital craniectomy. The dura was opened in a Y-shaped manner. Under microscope visualization, we dissected the telovelar-tonsillar fissures bilaterally. After the dissection, we discovered a highly vascularised red-orange lesion with tortuous vessels. The resection of this lesion was not complete given its adhesion to the floor of the fourth ventricle, but also due to its massive bleeding during excision. We achieved a large resection with satisfactory intraventricular hemostasis. A duraplasty was done, and then we completed the closure.

The post-operative care was uneventful. A brain CT scan was performed 48 hours post-operatively showing a

haemorrhagic remodelling at the surgical site associated with a residual tumor (Fig. 3).

Histopathological examination showed a highly vascularised tumor proliferation, composed of small congestive vessels, between which stromal cells with dense, vacuolated and moderately eosinophilic cytoplasm. In addition, there were patches of haemorrhagic suffusion and numerous deposits of haemosiderin. The diagnosis of a hemangioblastoma was confirmed and classified as World Health Organization (WHO) grade I.

At three months follow-up, there was a marked clinical improvement, no motor or sensitive deficit. No adjuvant treatment was proposed.

Six months later, the patient was admitted to the emergency room with a disturbed consciousness and

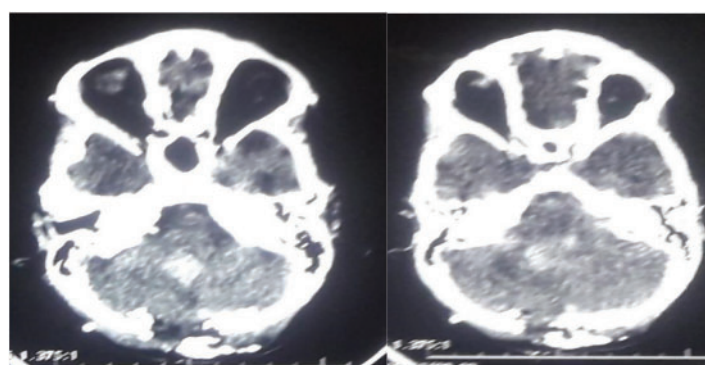


Fig. 3. Immediate postoperative CT scan showing a haemorrhagic remodelling at the surgical site and residual tumor.

cardio-respiratory instability, she died 3 hours after her admission to the intensive care unit.

3. DISCUSSION AND CONCLUSION

Paediatric hemangioblastomas (PHB) represent less than 1 per 1,000,000 [1], [2]. Although hemangioblastomas can arise along the central nervous system, they have a predilection for the posterior cerebral fossa and the spinal cord. In a series of 25 cases, they were mainly located in the cerebellum (40%) and the spinal cord (32%), followed by the brainstem (28%) [5]. Hemangioblastomas present in 70%–80% in sporadic form, and 10%–20% are associated with von Hippel-Lindau disease [6]. PHB is diagnosed at a median age of 12.6 ± 4.7 years, range of 1–16 years with a female predominance [5], [7]. Hemangioblastomas are grouped into four types of lesions: simple cystic (12.5%), simple solid (28.8%), solid with cystic component (3.8%), and cyst with mural nodule (54.8%) [8]. So, solid hemangioblastomas are quite common.

On examination, the most frequent symptoms were headache, nausea or vomiting, ataxia, vertigo, sensory changes and motor deficit [5]. One of the characteristics of “solid” hemangioblastoma, MRI showed an intermediate signal on T1 and rather hyperintense on T2, very strongly enhanced after injection of contrast product. It can be highlighted abnormal vessels dilated in contact with this nodule [9]. In our case, the lesion was an intermediate signal on T1-weighted and iso-intense on T2-weighted images. The T1-weighted imaging following gadolinium injection demonstrated a homogeneous enhancement of the lesion. In adults, a few cases of intra-ventricular localization have been described, specifically in the lateral ventricle, third ventricle and fourth ventricle [4], [10]–[14]. One of the characteristics of paediatric hemangioblastomas is the high prevalence of hydrocephalus [5]. In children, the surgical removal of paediatric hemangioblastoma represents a great challenge for the neurosurgeon. Massive bleeding during tumor removal can be life-threatening due to their low circulatory volume, pre-operative embolization may reduce this risk [15]. Patients with sporadic CNS hemangioblastomas were followed up for an average of 39 months, and patients with VHL syndrome were followed up for an average of 128 months [16].

Fourth ventricle tumors in children are dominated by the medulloblastoma and ependymoma, pediatric hemangioblastoma (PHB) of the fourth ventricle has never been reported in the literature.

CONFLICT OF INTEREST

Authors declare that they do not have any conflict of interest.

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