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(CASE REPORT)

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When rarity complicates diagnosis: A rare intraventricular tumor in pediatrics

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Abstract

Introduction: Meningiomas are rare tumors in children, accounting for around 3% of all pediatric brain tumors. Among them, primary intraventricular meningiomas (PIM) are even more exceptional, with an estimated incidence of between 0.5% and 5% of all meningiomas.

This article presents a case of intraventricular meningioma in a 7-year-old child, with a review of the literature.

Case Report: We report the case of an intraventricular meningioma in a 7-year-old child. He presented with symptoms of intracranial hypertension, and a cerebral MRI revealed an enhanced tumor at the intraventricular level of the right lateral ventricle. After surgical excision, histology showed a WHO grade I transitional meningioma. The postoperative course was favorable, with satisfactory radiological and clinical control.

Conclusion: Intraventricular meningiomas are rare in the pediatric population. These tumors are often large and aggressive when they occur in children. The management of these tumors is a surgical challenge.

Keywords: Intraventricular Tumor; Pediatric; Meningioma; Surgery

1. Introduction

Intracranial meningiomas are rare tumors in children, accounting for less than 3% of pediatric brain tumors. Intraventricular localizations represent approximately 0.5% to 5% of these cases (1). The management of these tumors is challenging, as they are generally large at the time of diagnosis.

We present the case of a 7-year-old girl with a large intraventricular meningioma, who presented with signs of intracranial hypertension (HTIC). We also reviewed the literature on pediatric intraventricular meningiomas.

2. Case report

In this case, a 7-year-old girl from a non-consanguineous marriage consulted us with headaches that had been evolving for 20 days. Initially moderate and frontal, they progressively increased in intensity, becoming more intense on awakening and associated with vomiting, which temporarily relieved them. There was no history of loss of consciousness or convulsions. Clinical examination revealed a conscious, stable patient with no sensory-motor deficits.

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However, a progressive decline in visual acuity was noted. Ophthalmological examination revealed a bilateral absence of light perception, with areactive semi-mydriasis. Bilateral grade III papilledema was observed.

A cerebral CT scan with contrast medium injection (Figure 1) revealed a large lesion in the left lateral ventricle, with very intense contrast uptake. This mass caused ventricular dilatation with a mass effect. Brain MRI with contrast injection (Figure 2) revealed a large lesion in the left lateral ventricle measuring 5.2 cm x 4.8 cm x 6.5 cm, hypointense in T1, isosignal in T2, with intense enhancement. The mass caused ventricular dilatation with a mass effect. The radiological diagnosis was suggestive of choroid plexus papilloma, ependymoma, or intraventricular meningioma.

Due to the HTIC syndrome associated with a visual threat, the patient benefited from a ventriculoperitoneal shunt (DVP) as a first-line treatment to relieve the HTIC and preserve visual acuity. After obtaining informed consent, surgery was performed. An inter-hemispheric approach was performed with a callosotomy allowing access to the lesion. The tumor was entirely intraventricular in the left lateral ventricle, compressing the surrounding structures, notably Monro's foramen, especially on the left. Intraoperatively (Figure 3), the tumor was highly vascularized, with variable consistency (soft in places and firm to hard in other areas). Macroscopically complete excision was performed, followed by placement of an external ventricular drain (EVD) to ensure optimal management of intracranial pressure. The operation went smoothly and without incident. The patient was extubated without complication. The DVE was removed after 48 hours of monitoring, with a satisfactory follow-up CT scan (Figure 4). Histopathological analysis confirmed the diagnosis of grade I meningioma according to the WHO 2021 classification. The patient was seen in consultation after 6 months with a favorable clinical evolution, without recurrence of HTIC signs or complications. Post-operative contrast-enhanced brain MRI performed after 6 months (Figure 5) revealed no tumor residue, with satisfactory control.

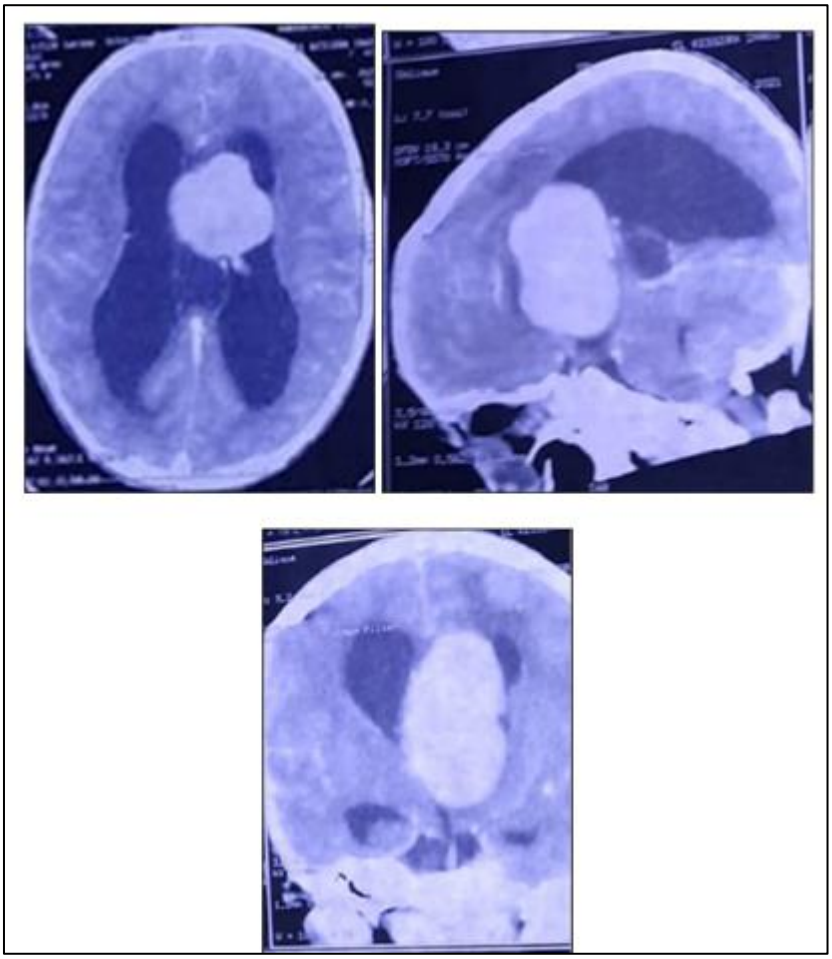

Figure 1 CT scan of the brain with contrast injection revealed a large lesion in the left lateral ventricle, intensely enhancing with contrast. This mass caused ventricular dilation with a mass effect

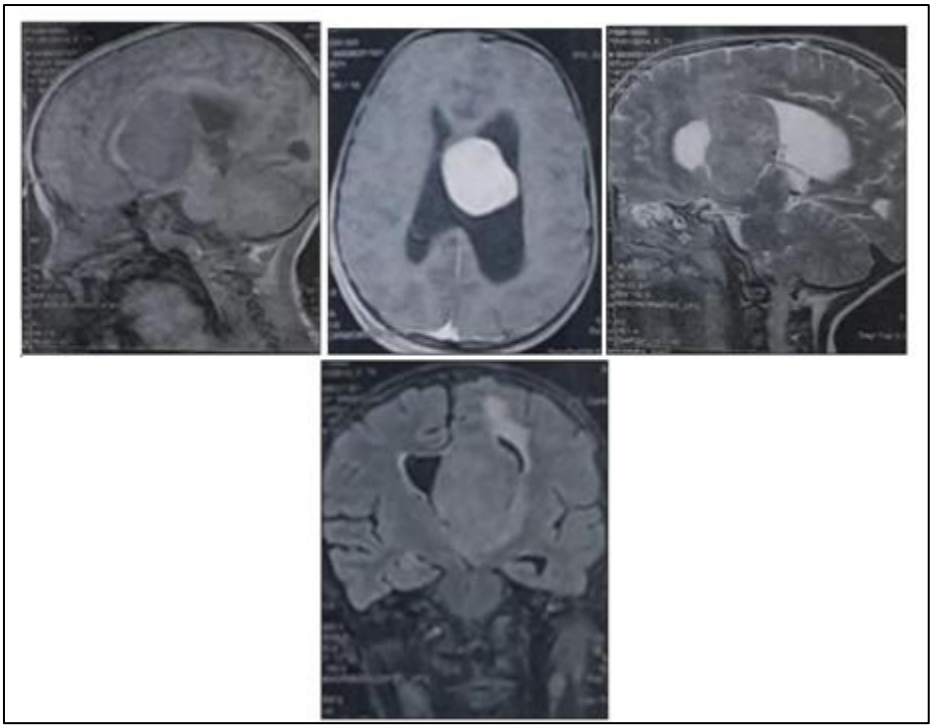

Figure 2 The brain MRI with contrast injection revealed a large lesion in the left lateral ventricle, hypointense on T1, isointense on T2, with intense enhancement

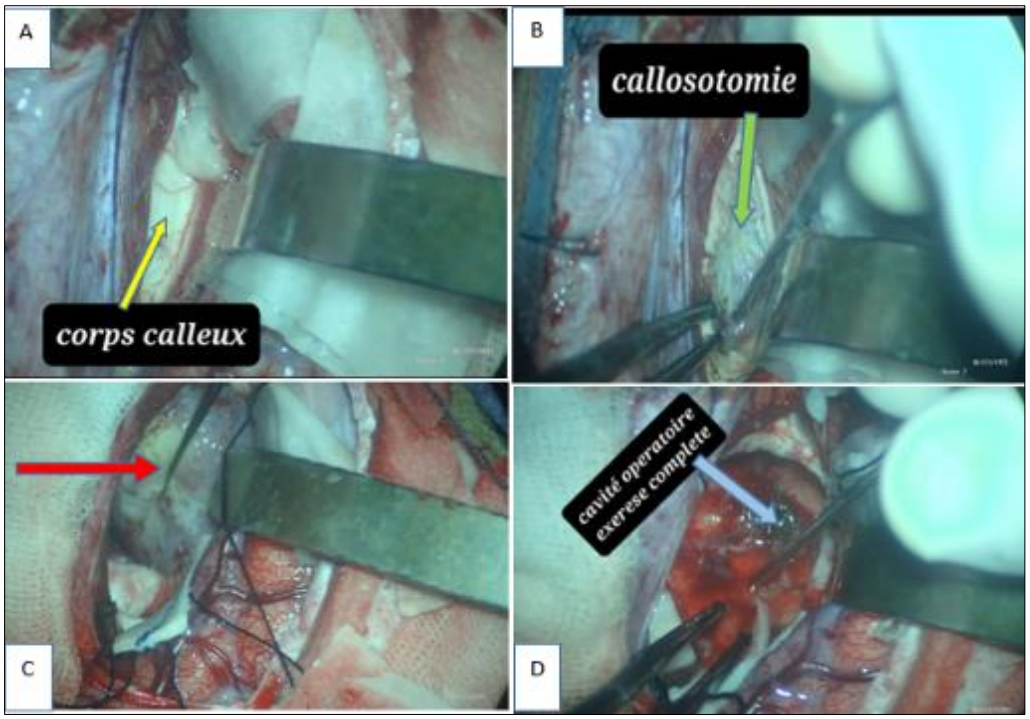

Figure 3 Intraoperative images: A) Corpus callosum (yellow arrow). b) Callosotomy (green arrow). c) Intraventricular meningioma (red arrow). d) Resection cavity (blue arrow)

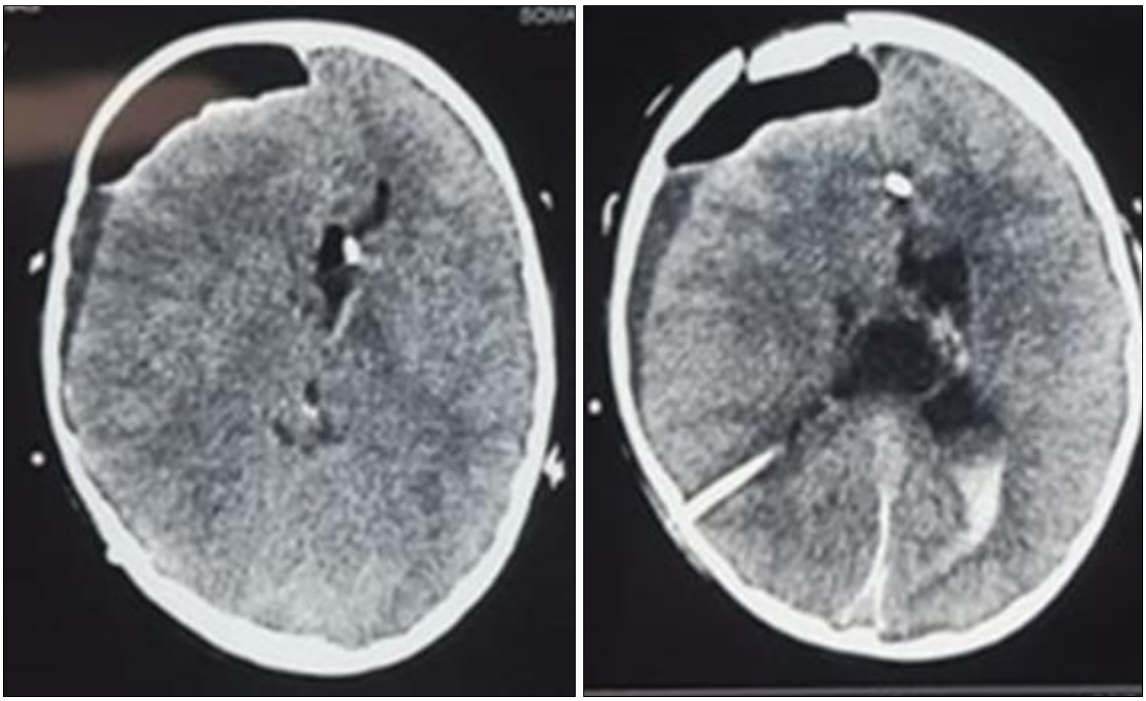

Figure 4 Postoperative CT scan showing complete excision of lesion

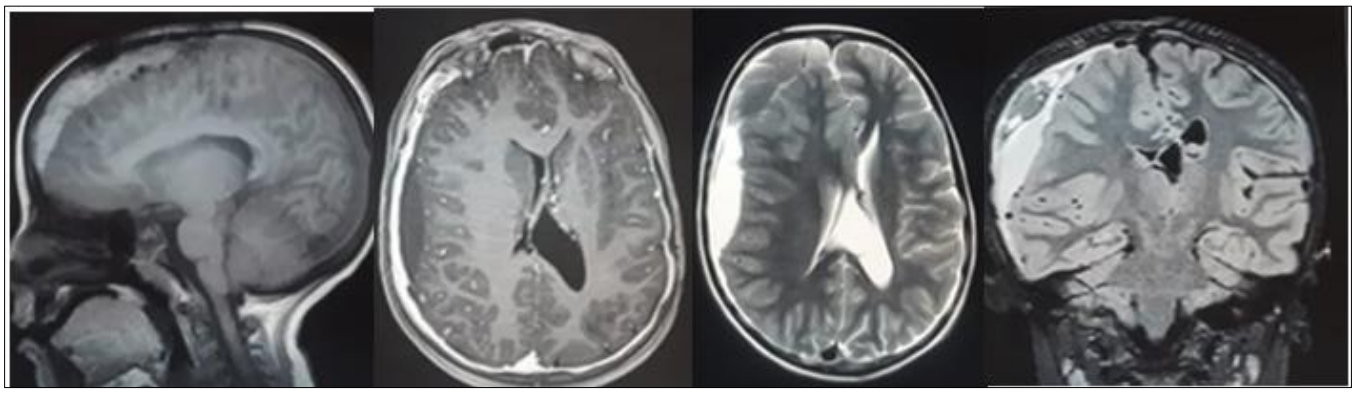

Figure 5 MRI control at 6 months post-operative showing complete excision of the lesion

3. Discussion

Intraventricular meningiomas are rare tumors, representing around 0.5% to 2% of all intracranial meningiomas (2,3). Their most frequent location is in the lateral ventricles, with a predilection for the left side. A review of 585 cases reported up to 2006 showed that 78.9% of intraventricular meningiomas were located in the lateral ventricles, 14.5% in the third ventricle, and 6.4% in the fourth ventricle, with a female predominance (4). However, this analysis mainly concerns adults and data on pediatric intraventricular meningiomas remain limited. Our study thus contributes to the literature on this rare entity in children. In children, intraventricular meningiomas are even more exceptional, representing less than 3% of pediatric brain tumors, with intraventricular localizations constituting around 0.5 to 5% of cases (2). The clinical presentation is often dominated by signs of intracranial hypertension (ICHT) (5,6), as observed in our case. Neurological deficits and localization signs are generally absent due to intraventricular localization. Some patients may present with seizures that are predominantly generalized in nature (7).

The origin of intraventricular meningiomas is uncertain, but they appear to arise from the stroma of the choroid plexus or from remnants of arachnoid tissue invaginated into the choroidal web, which would explain their frequency in the trigonal region and their pedicular attachment to the choroid plexus (8, 9). These tumors often develop slowly and reach enormous size before becoming symptomatic, unless the lesion is located in a region that blocks CSF circulation (9,10),

thus causing ventricular dilatation and HTIC. In our case, the tumor was located in the left lateral ventricle, compressing the surrounding structures, notably the foramen of Monro, thus explaining the signs of HTIC.

CT and MRI are currently safe and accurate means of diagnosing these tumors. The radiological appearance is similar to that of other meningiomas. On CT, these tumors are usually hyperdense, taking up contrast strongly, sometimes with calcifications (11-12). Hydrocephalus induced by lateral intraventricular meningiomas is usually localized to the ipsilateral side (11). On MRI, meningiomas are iso or hyposignal in T1-weighted sequences, and iso or hyposignal in T2-weighted sequences with high contrast uptake (13, 14, 15, 16). Differential diagnosis with other tumors located in the lateral ventricle should include choroid plexus papilloma, low-grade gliomas, and ependymomas in children (17, 18).

Various surgical approaches to lateral ventricular meningiomas have been advocated (15,16). A transfrontal approach was reported by Busch. Cushing proposed a temporoparietal approach. In 1960, Cramer described the merits of a posterior parieto-occipital incision, as did Fornari and others (19). Transcortical temporal, frontal, and occipital approaches have been proposed by several authors, offering easier access to the choroidal artery (20, 21). The posterior transcallous approach was described by Kempe and Blaylock (13) for the ablation of tumors in the ventricular trigone of the dominant hemisphere. Optimal resection of intraventricular meningiomas relies on careful planning, taking into account their size, location, and vascularization. Tumor reduction must be performed rapidly to avoid brain retraction, and thus minimize post-operative neurological deficits. Coagulation of the choroidal artery branches should be performed as soon as possible (22). In our case, we opted for an inter-hemispheric transcallous approach, enabling direct access and macroscopically complete excision while minimizing the risk of postoperative complications.

4. Conclusion

Intraventricular meningiomas are rare in children and often diagnosed late because of their slow growth. They must be taken into account in the differential diagnosis of intraventricular tumors. CT and/or MRI images enable a correct diagnostic approach in most cases. Microsurgical techniques enable these tumors to be completely removed, providing effective management.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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