Clinical Case Reports

ISSN: 2675-3626

Supratentorial extra ventricular intraparenchymal ependymoma in a child: clinical case report

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Background: Ependymal tumors are uncommon in intracranial tumors. Ependymal origin usually involves an intraventricular localization. Exceptional intraparenchymal extra ventricular locations have been described. We report a case of extra ventricular ependymoma managed in our department.
Case Description. An 8-year-old girl was admitted in January 2019 for left hemiparesis that had been progressing for two weeks associated with chronic headaches. Neurological examination noted a muscle strength rated at 4/5 in left hemi body. Medical imaging made it possible to highlight a voluminous mixed, intraparenchymal, right parietal temporal mass first evoking diffuse astrocytoma (WHO grade II). After gross total excision, histology and immunohistochemistry concluded that it was WHO grade II ependymoma. At five months postoperatively, the patient was asymptomatic and control CT scan was unremarkable. After a 4-year follow-up, the patient remained asymptomatic.
Conclusion. Extra ventricular ependymoma diagnosis was a histological surprise. Surgical removal of this lesion was not a problem. Course was satisfactory in the absence of adjuvant therapy (chemo and/or radiotherapy).

Keywords: ependymoma, extra ventricular, surgery, immunohistochemistry

INTRODUCTION/BACKGROUND

Ependymal tumors belong to the group of glial tumors. Ependymal tumors are uncommon among intracranial tumors. They are found more in children than in adults and are preferentially located in the posterior cerebral fossa in the fourth ventricle. Ependymal origin usually involves intraventricular localization. Extra ventricular, intraparenchymal, supratentorial ependymal tumors are relatively rare [2,9].

There is no specific clinical expression to intracranial ependymal tumors. However, in all topographical forms, one telltale syndrome dominates intracranial hypertension syndrome. The scanner can direct towards the diagnosis. MRI makes it possible to better analyze the lesion.

In addition to excision problems posed by some of their locations, intracranial ependymal tumors are poorly identified therapeutically because of recurrence risk or distant neurological transplantation from the initial localization. Radiotherapy effectiveness is also discussed by some authors [3,6,9].

CASE REPORT

An 8-year-old girl, student, was admitted in January 2019 for left hemiparesis evolving for two weeks associated with chronic, persistent headaches resistant to usual analgesics. She reports no pathological history. Neurological examination noted a muscle strength rated at 4/5 in the left hemi body.

CT scan followed by cerebral MRI (figures 1 and 2) revealed a large mixed (cystic and fleshy), extra ventricular, well limited mass of, 64 millimeters in anteroposterior diameter, 50 millimeters in thickness and 60 millimeters in transverse diameter. This mass was intraparenchymal on right parietotemporal with homogeneous contrast uptake at the level of fleshy portion. They were perilesional edema, mass effect on lateral ventricles and medial line deviation. This lesion first suggested a diffuse astrocytoma (WHO grade II).

A macroscopically total excision of this lesion was performed approximately 2 weeks after the patient's admission. The CT scan performed on the second

Received: 09 January 2023

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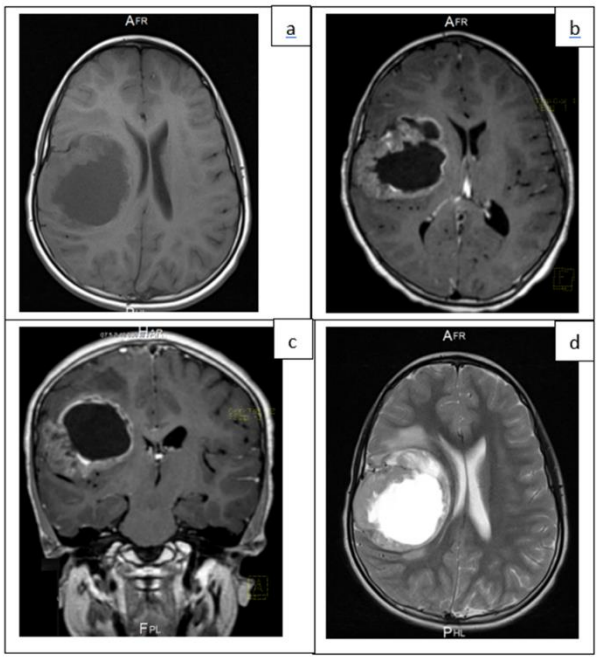

Figure 1- Preoperative MRI. (a) T1-weighted axial section showing a voluminous mixed (cystic and fleshy) extra ventricular intraparenchymal parietotemporal mass, well-limited, perilesional edema, mass effect on lateral ventricles and medial line deviation. On T1 sequences injected in axial (b) and coronal section (c) there is a homogeneous contrast taking at the level of the fleshy portion. T2 -weighted sequence (d) shows a cystic appearance of the lesion part.

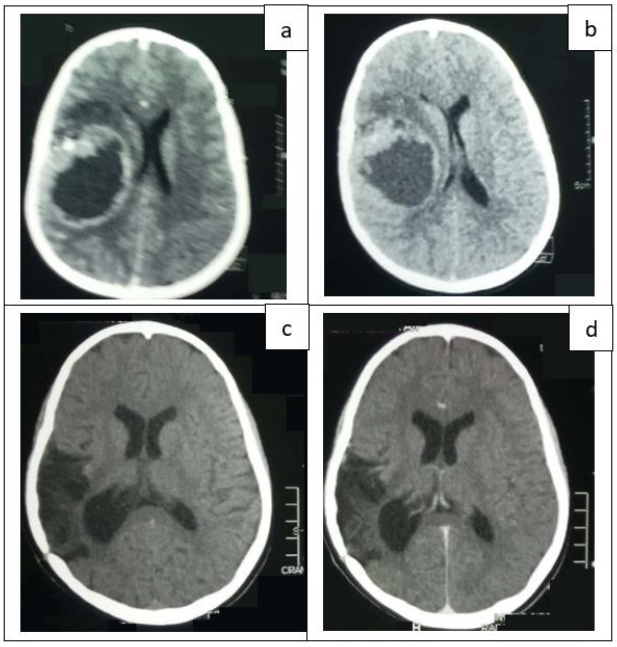

Figure 2- Pre- and post-operative CT scan. Preoperative parenchymal window CT without (a) and with contrast injection (b) showing a voluminous right parietotemporal mixed (cystic and fleshy) extra ventricular intraparenchymal, taking contrast at the level of the fleshy portion, well limited mass with perilesional edema, a mass effect on lateral ventricles and medial line deviation. CT images at 5 months postoperative parenchymal window without (c) and with contrast injection (d) showing right sequellar parietotemporal porencephalic cavity without tumor recurrence signs.

postoperative day had objectified a small hemorrhagic edematous remodeling of the operative site without tumor residue.

Histologic examination of the surgical specimen had permitted to observe under microscopy a tumoral proliferation made up of perivascular pseudo-fruit bats and sometimes organized into a tube and into ependymal fruit bats. The tumor cells were cubo-cylindrical. They were had monomorphic nuclei, ovoid in shape, with fine chromatin producing the so-called "pepper and salt" appearance. These nuclei sometimes had a small nucleolus. The vessels had a hyalinized wall. There was no capillary endothelium proliferation, necrosis, or abnormal mitotic activity. It was associated with a clear cell component (Figure 3). This led to the conclusion of an ependymoma (WHO 2016 grade II) subject to immunohistochemical confirmation. The block of the anatomy pathological specimen was sent to Europe where immunohistochemistry confirmed this diagnosis in view of the positivity of the GFAP, the PS100 and the EMA.

At five months postoperatively, the patient had fully recovered from her left hemi body motor deficit and had returned to school with excellent academic results as before

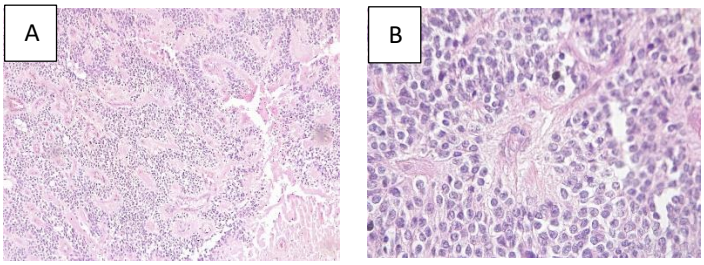

Figure 3- Histological aspect of the surgical specimen. (a) low magnification (GX40): Proliferation of monomorphic cells around vessels with hyalinized walls. (b) high magnification (GX100): Aspects of perivascular pseudo-fruit bats

the operation. Control CT scan done five months postoperatively (figure 2) had objectified a sequela porencephalic cavity at the operation site without a tumor-like lesion. After a 4-year follow-up, the patient still remains asymptomatic.

DISCUSSION

Extra ventricular ependymal tumors are relatively rare. Studies note an average of 1 to 3 cases per year. Thus, there were 15 cases in 15 years [2], 11 cases in 11 years [5], 6 cases in 12 years [7], 56 cases in 17 years [9], 66 cases in 20 years

[10], 48 cases in 20 years [8]. Other authors have published single cases in male adults (23, 42, 61 years) [3,4,6] or in a 16-year-old patient [1]. Patients consisted of 30 men and 25 women with an average age of 30.0 ± 23.6 years (range 1 to 74 years) [9]. All three patients were males aged 7, 15 and 17 years [7]. The 15 patients included 6 men (40%) and 9 women (60%), with a median age of 15 years [2].

Clinically, our case was revealed by persistent headaches associated with hemi body deficiency. Persistent headache was the most consistent symptom [4,6]. Sometimes a neurological deficit was associated [6]. In medical imaging, the main differential diagnosis was astrocytoma [5,6,11]. CT scans suggested the diagnosis of extra ventricular ependymoma in 3 out of 11 cases [5]. Although MRI characteristics of extra ventricular ependymomas varied and were not specific, multiparametric MRI techniques contribute better than CT to diagnosis [5,10,11]. Frontal localizations [1,6,9] and parietal localizations [7,9] were the most common. However, all other lobes may be the localization of an extra ventricular ependymal tumor: temporal lobe [4]; occipital [3]. An infratentorial location has also been described [5]. Intra tumoral hemorrhage was sometimes noted [3,4]. Out of 11 MRI cases [5], images displayed quasi-circular (4 cases), irregularly lobulated (7 cases) variable-intensity masses. The masses presented with cysts or necrosis (8 cases), hemorrhage (7 cases), marked (9 cases) or mild (2 cases) enhancement, and moderate (4 cases), mild (3 cases) or absent (4 cases) peritumoral edema. The tumors were also frequently closely associated with the lateral ventricle (6 cases). Tumors appeared isointense to hypointense on T1-weighted imaging (T1WI) and heterogeneously hyperintense or hypointense on T2WI, demonstrating wreath-like and ring-like characteristics, with intra tumoral nodules (3 cases) or marked flake-like inhomogeneous (6 cases) enhancement on post-contrast MRI. Only 2 solid lesions showed mild enhancement (2 cases).

Surgical resection was most often macroscopically total [1,2,4,6,9]. Resection was macroscopically total in 76% and subtotal 24% [9]. Nine patients (60%) underwent gross total resection, 5 patients (33.3%) underwent subtotal resection, and 1 patient (6.7%) underwent biopsy [2]. A contralateral transfalcine interhemispheric approach was chosen, which resulted in total tumor resection [1].

In our case, histology and immunohistochemistry had concluded to a grade II class ependymoma of the 2016 WHO classification. Our case, which was managed in 2019, that is to say before the latest WHO classification which has been in force since 2021, was not classified according to this latest classification. Similarly, all of the articles that we found in the literature had not used the WHO classification of 2021 but that of 2016. Thus, the pathological anatomy had concluded to an anaplastic ependymoma (grade III of WHO 2016) in most series [3,4,6,9]. Ependymoma (WHO 2016 grade II) was sometimes noted [1]. According to WHO classification, there were 69% anaplastic ependymomas and 31% grade II (ependymoma) [9].

Depending on pathological anatomy results, adjuvant therapy (chemotherapy or radiotherapy) may be combined with surgery. However, this adjuvant treatment does not seem to really influence occurrence or no recurrence. Thus, seven years after adjuvant radiotherapy, a regular follow-up MRI showed a partially cystic mass of 3 cm in the same area as the initial tumor requiring revision surgery, as decribed by Seo et al. [6]. Despite additional radiotherapy and adjuvant chemotherapy, ten months later tumor recurred with hemorrhage into the spinal canal, as described by Iwamoto at al. [4]. Four years after radiotherapy, anaplastic ependymoma recurred at the cervicomedullary junction and T8-T9 levels, as describe by Han at al. [3].

According to our revision, patients who underwent macroscopically total resection had better progression-free survival than those who had subtotal resection [8]. Grade II ependymal tumors had a better progression-free survival (PFS) and overall survival (OS) compared to grade III tumors with statistically significant differences ($P < 0.01$ and 0.02, respectively) [2]. Anaplastic histology (WHO grade III) was a poor prognostic factor for PFS ($P = 0.04$) [2]. Tumor resection quality and tumor grade were identified as prognostic factors for tumors-free survival (PFS) and overall survival (OS) while age, sex, tumors side and postoperative radiotherapy were not prognostic factors for PFS and OS [9].

CONCLUSION

Supratentorial extra ventricular, intraparenchymal ependymoma diagnosis, was a histological surprise that required immunohistochemistry confirmation in Europe. Therapeutically, his surgical removal did not pose a problem. In the absence of adjuvant therapy (chemo and/or radiotherapy), the course was satisfactory clinically and CT scans. At five months postoperatively, no image of recurrence or tumor progressive recovery been objectified on control imaging.

DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Work exempted from ethics committee authorization

Consent to participate

The patients gave consent to use their information and images for research purposes. *Consent for publication*

The patient gave consent to use his information and images for publication.

Conflict of interest

The authors declare no conflicts of interest with respect to the content, authorship, and/or publication of this article.

Funding

This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors

CONTRIBUTIONS

-Denlewende Sylvain Zabsonre: Conceptualization, Data curation, Writing – original draft, Writing – review & editing

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REFERENCES

1. Baldoncini M, Ferini G, Umana GE, Chaurasia B, Demichelis ME, Villalonga JF and al. Contralateral Interhemispheric Transfalcine Approach for Supratentorial Extraventricular Ependymoma Resection. Anticancer Res. 2022 Jun;42(6):3203-3207.8
2. Byun J, Kim JH, Kim YH, Cho YH, Hong SH, Kim CJ.Supratentorial Extraventricular Ependymoma: Retrospective Analysis of 15 Patients at a Single Institution. World Neurosurg. 2018 Oct;118:e1-e9.5
3. Han MH, Park KS, Park SH, Hwang JH. Supratentorial extraventricular anaplastic ependymoma presenting with repeated intratumoral hemorrhage. . Brain Tumor Res Treat. 2014 Oct;2(2):81-6.6
4. Iwamoto N, Murai Y, Yamamoto Y, Adachi K, Teramoto A. Supratentorial extraventricular anaplastic ependymoma in an adult with repeated intratumoral hemorrhage. Brain Tumor Pathol. 2014 Apr;31(2):138-43.4
5. Leng X, Tan X, Zhang C, Lin H, Qiu S. Magnetic resonance imaging findings of extraventricular anaplastic ependymoma: A report of 11 cases. Oncol Lett. 2016 Sep;12(3):2048-2054. 10
6. Seo SW, Kang HJ, Lee MS, Suh SJ, Lee YS, Lee JH, Kang DG. A Case of Recurrent Supratentorial Extraventricular Anaplastic Ependymoma in Adult. Brain Tumor Res Treat. 2019 Apr;7(1):44-47.9
7. Takaki Y, Tsutsumi S, Teramoto S, Nonaka S, Okura H, Suzuki T and al. Supratentorial extraventricular ependymoma presenting calvarial erosion: A report of 3 cases. Radiol Case Rep. 2021 Aug 7;16(10):3003-3006.2
8. Wang L, Han S, Yan C, Yang Y, Li Z, Yang Z. The role of clinical factors and immunocheckpoint molecules in the prognosis of patients with supratentorial extraventricular ependymoma: a single-center retrospective study.J Cancer Res Clin Oncol. 2021 Apr;147(4):1259-1270.7
9. Wang M, Zhang R, Liu X, Li D, Qiu C, Zhao P and al. Supratentorial extraventricular ependymomas: A retrospective study focused on long-term outcomes and prognostic factors. Clin Neurol Neurosurg. 2018 Feb;165:1-6. 1
10. Xing Z, Zhou X, Xiao Z, She D, Wang X, Cao D. Comparison of Conventional, Diffusion, and Perfusion MRI Between Low-Grade and Anaplastic Extraventricular Ependymoma. AJR Am J Roentgenol. 2020 Oct;215(4):978-984.3
11. 11. Zhou X, Su Y, Huang W, Lin X, Xing Z, Cao D. Differentiation between supratentorial pilocytic astrocytoma and extraventricular ependymoma using multiparametric MRI. Acta Radiol. 2022 Dec;63(12):1661-1668. 11