Radiology Case Reports 19 (2024) 4151–4157

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Case Report

Intraventricular neurocytoma: A diagnostic challenge with prognostic value ☆

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ARTICLE INFO

Article history:
Received 7 June 2024
Accepted 22 June 2024

Keywords:
Atypical neurocytoma
Imaging features
Complicated neurocytoma
Intracranial hypertension
Magnetic resonance
Perfusion MRI

ABSTRACT

Intraventricular neurocytoma is a low incidence central nervous system tumor. It predominantly affects young adults with no apparent gender predilection. The main symptoms include headache, nausea and vomiting. These result from hydrocephalus due to the obstruction of cerebrospinal fluid flow. On diagnostic imaging, neurocytoma can be suspected by some features, such as peripheral cysts, lobulated contours and septa that bridge the ventricular wall, giving a "scalloped" appearance. There are other characteristics, but they are less specific for the diagnosis. The atypical variant of neurocytoma is even rarer and leads to a worst prognosis. Atypical neurocytomas develop higher proliferative potential identified by the Ki-67 biomarker and higher recurrence rate. There are few studies about the imaging characteristics of atypical neurocytomas. At this point, there are no reliable distinctive features to differentiate atypical neurocytomas, especially due to their low incidence.

We present the case of a 20-year-old female patient with symptoms of intracraneal hypertension. CT and MRI of the brain revealed a mass occupying the body of the left lateral ventricle, adjacent to the foramen of Monro. The mass was primarily solid with discrete peripheral cyst and a few scalloped areas. It also showed signs of supratentorial obstructive hydrocephalus. The tumor was partially removed because of bleeding and compromise of vascular structures. Immunohistochemistry revealed positive synaptophysin, elevated Ki-67 (7%), increased number of blood vessels and moderate nuclear atypia. After surgery, the patient persisted with signs of intracranial hypertension, not improving with clinical management and requiring aggressive surgical procedures.

While rare, atypical neurocytoma requires a better characterization, especially through imaging, to optimize immediate management and explore new therapeutic options.

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☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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https://doi.org/10.1016/j.radcr.2024.06.055

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Introduction

Intraventricular neurocytoma has an incidence between 0.25% and 0.5% of all primary malignancies in adults [1–3]. It is categorized as grade 2 WHO, being part of neuronal and glioneuronal tumors [3,4]. It affects mainly young adults with ages between 20 and 40 years old [1,3]. It is frequently a slow growing tumor, with clear margins, lobulated contours, with broad attachment to the septum pellucidum and contact with the foramen of Monro [1,5]. Its intraventricular location causes obstruction of cerebrospinal fluid circulation with hydrocephalus and symptoms derived from these conditions [5]. The prognosis in this tumor is frequently benign [6]. However, nearly 25% of tumors in this group behave more aggressively and recurrence is usual. This situation is related to increased levels of proliferation marker Ki-67. The risk of relapse is increased with values more than 2% and encompasses worst prognosis [1,3,7]. Some imaging features could be related to atypical neurocytomas and should be considered for a differential management of this subgroup of patients

Case report

Female patient, 20 year old without relevant personal or familiar medical history. Three weeks before being admitted to the hospital, showed symptoms of nausea, vomiting on several occasions and headache of moderate intensity. Pain relieved partially with analgesics. Focal neurological deficit signs were absent. Computed tomography (CT) showed a soft tissue density mass, partially calcified, located in the left ventricle, occupied the foramen of Monro and had a broad attachment to the septum pellucidum. It caused an important ventricular dilatation, effacement of sulci and transependymal edema (Fig. 1). Magnetic resonance imaging (MRI) also revealed a voluminous intraventricular mass, isointense to gray matter on T1 weighted images and slightly hyperintense on T2 weighted images. It was possible to identify subtle cystic components on the periphery of the lesion, better delineated than on CT images. It also had septa reaching the wall of the lateral ventricle. These gave the contours of the lesion a scalloped appearance. MRI also revealed signs of supratentorial hydrocephalus with dilatation of lateral ventricles and periventricular hyperintensity on T2 weighted images (Fig. 2). Susceptibility images revealed prominent voids of signal inside the lesion related to vascular structures and calcifications. Diffusion-weighted imaging and apparent diffusion coefficient (ADC) manifested increased restriction. The mass exhibited heterogeneous enhancement with intravenous contrast and prominent vessels within (Fig. 3). It received arterial supply from many small branches of anterior and middle cerebral arteries, with venous drainage to the left internal cerebral vein. The vascular structure of the lesion precluded embolization prior surgery. Dynamic susceptibility contrast perfusion study showed a relative cerebral blood volume (rCVB) of 3.4 for the lesion compared to the contralateral white matter (Fig. 3). During surgery, a friable reddish mass was discovered within the left lateral ventricle, with broad adherence to the septum pellucidum and abutting to the contralateral ventricle. Frozen section reasserted diagnosis of neurocytoma. Surgeons discovered strong adherence of the mass to thalamostriate and anterior septal veins. Because of the risk of deep vein drainage system lesion, the mass was partially resected. Subsequent immunohistochemical analysis displayed: Glial Fibrillary Acidic Protein (GFAP) negative, Synaptophysin positive, Ki-67: 7%. Microscopic biopsy images showed increased vessel density and moderate nuclear atypia. Postoperative CT revealed enduring signs of cerebral edema although external

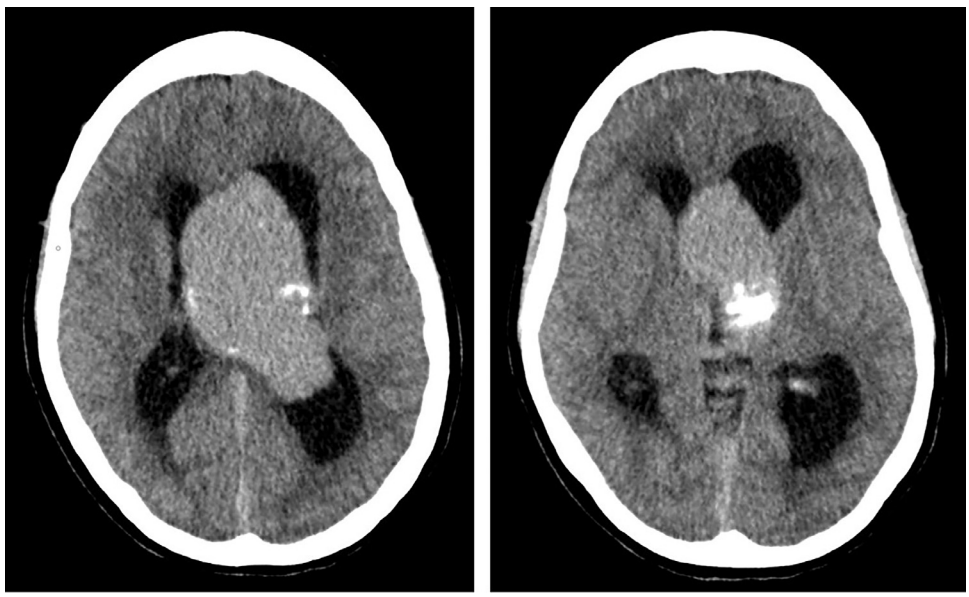

Fig. 1 – Brain CT : intraventricular mass with calcifications occupying the body of the left ventricle with broad contact with the septum pellucidum and the foramen of Monro. Lateral ventricular dilatation is present with transependymal edema and effacement of the sulci.

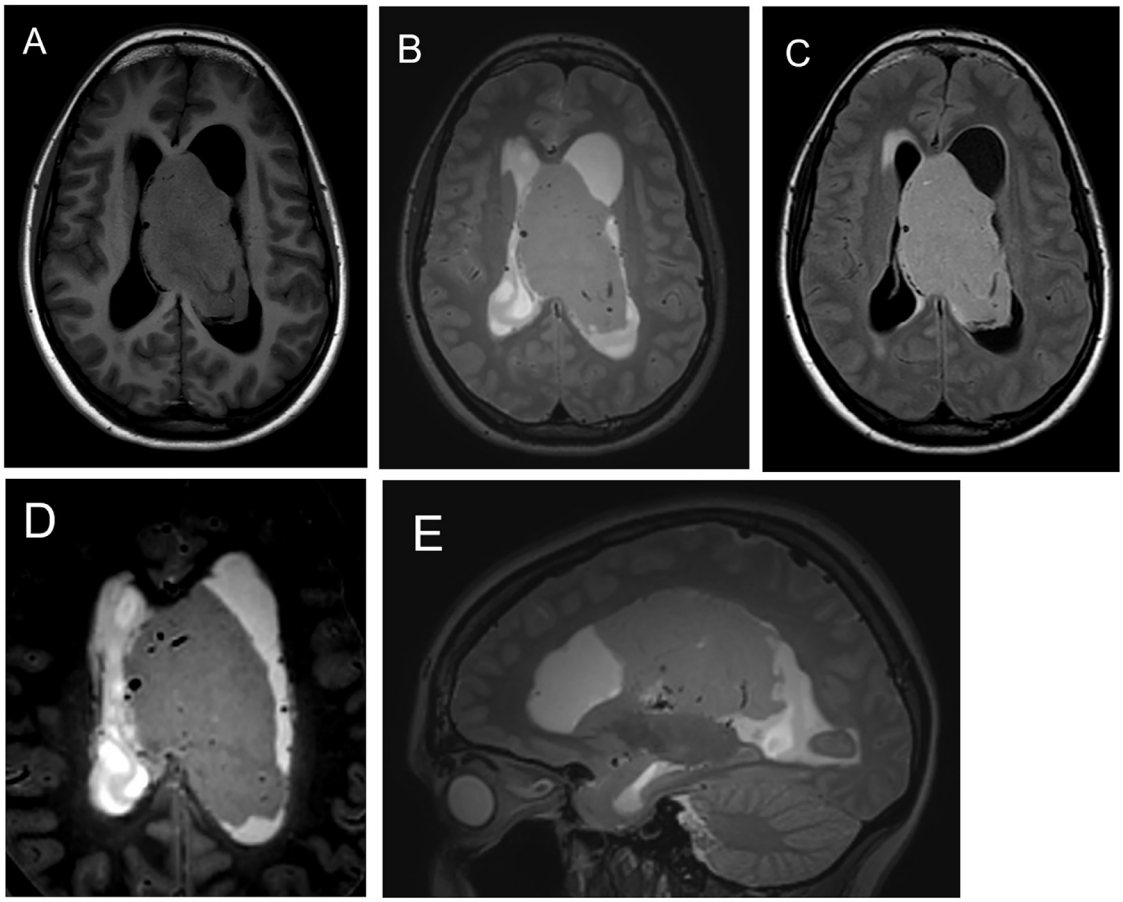

Fig. 2 – Brain MRI. (A-C) T1, T2 and FLAIR weighted images respectively: intraventricular lesion, mainly solid, isointense to gray matter on T1, slightly hyperintense on T2. Transependymal edema as hyperintensities adjacent to the frontal horns of the lateral ventricles. (D) Subtle cystic area on the periphery of the lesion (E) thin septa bridging the lesion and the wall of the ventricle.

ventricular drain was placed during surgery. MRI control study demonstrated blood products and remnants of the mass occupying the ventricle (Fig. 4). Perfusion and diffusion images helped to make the distinction between them (Fig. 5). Owing to persistent signs and symptoms of increased intracranial pressure, patient required bilateral decompressive frontal craniotomy. After two weeks the external ventricular drain was removed and replaced by a ventriculoperitoneal shunt to treat the hydrocephalus.

Discussion

Neuroepithelial intraventricular tumors are rare. They represent 2%-7% of brain tumors. Central neurocytomas are even rarer, with a prevalence of 0.25%-0.5% [8,9]. Most tumors in this group exhibit a low proliferative potential, with a benign behavior and good prognosis. The last is consistent with complete excision of the mass [10]. Unfortunately, complete excision is only achieved in 50% of cases, with successful surgery rates of 30%-70% varying according to institutions [6,11].

Central neurocytomas include a subtype called *atypical neurocytomas*. These tumors develop greater proliferative potential recognizable mainly with the monoclonal antibody Ki-67/MIB1. This marker detects proteins related to tumoral cell proliferation [2]. This is one of the reasons for these tumors to demonstrate lower rates of complete excisions, higher percentage of local recurrence and potential arachnoid dissemination [11,12]. Overall survival rate in 10 years for patients in this subgroup is 63%, in contrast with 90% of typical neurocytoma [12]. A meta-analysis of Rades and Schild revealed that patients with intraventricular neurocytomas excised completely have a survival rate even greater than 90% [13]. On the contrary, research conducted by Rades, Schild, and Fehlauer involving 129 patients revealed that neurocytomas with a Ki-67 value exceeding 3% exhibit a recurrence rate of 48%, whereas tumors with a Ki-67 value below 3% demonstrate a recurrence rate of 12% [14].

Cellular atypia has not shown to be an appropriate marker to consider a neurocytoma as atypical. A retrospective study of Mackenzie reported that a Ki-67 value greater than 2% is better related with local recurrence of neurocytoma compared with cellular atypia. The study also proposes the term *proliferating*

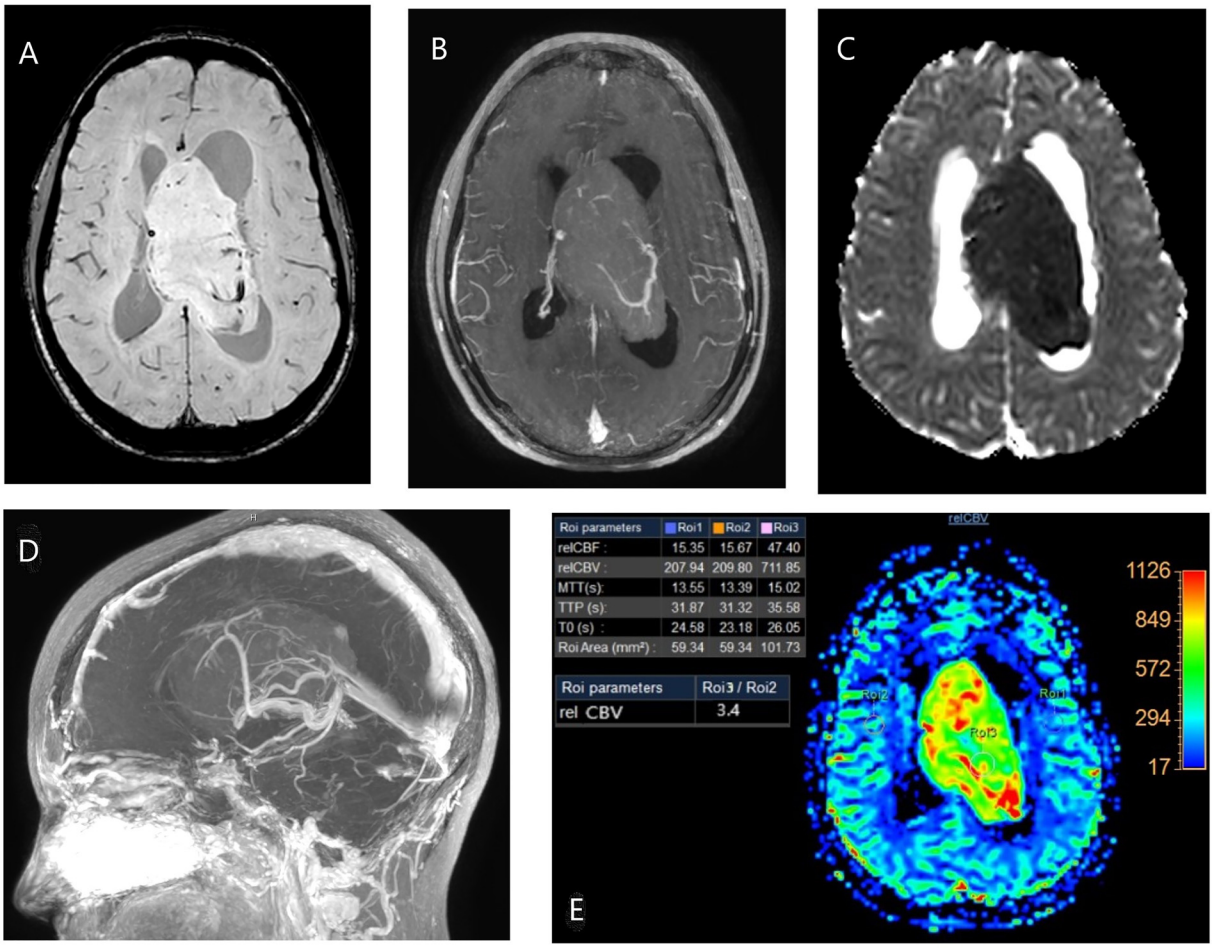

Fig. 3 – Brain MRI. (A) Susceptibility image with hypointensities inside the lesion corresponding to calcification and void of signal of the vessels. (B) contrast image with moderate enhancement (C) ADC image with significant restriction (D) 3D reconstruction demonstrates important drainage to the deep venous system. (E) Perfusion with increased vascularity inside the lesion.

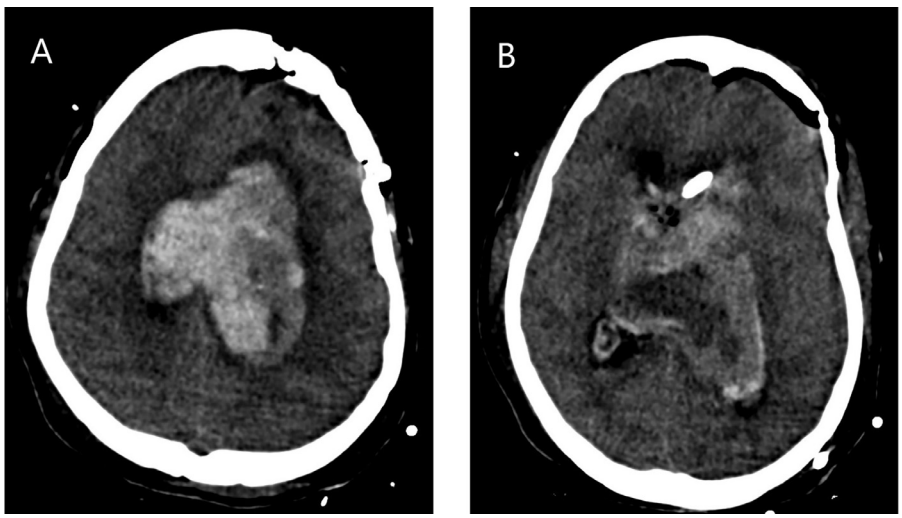

Fig. 4 – A and B one-day control brain CT: (A) periventricular hypodensity and effacement of sulci, consistent with increased intracranial pressure (A and B) Differentiating between blood products and remnants of the mass occupying the ventricle proves challenging.

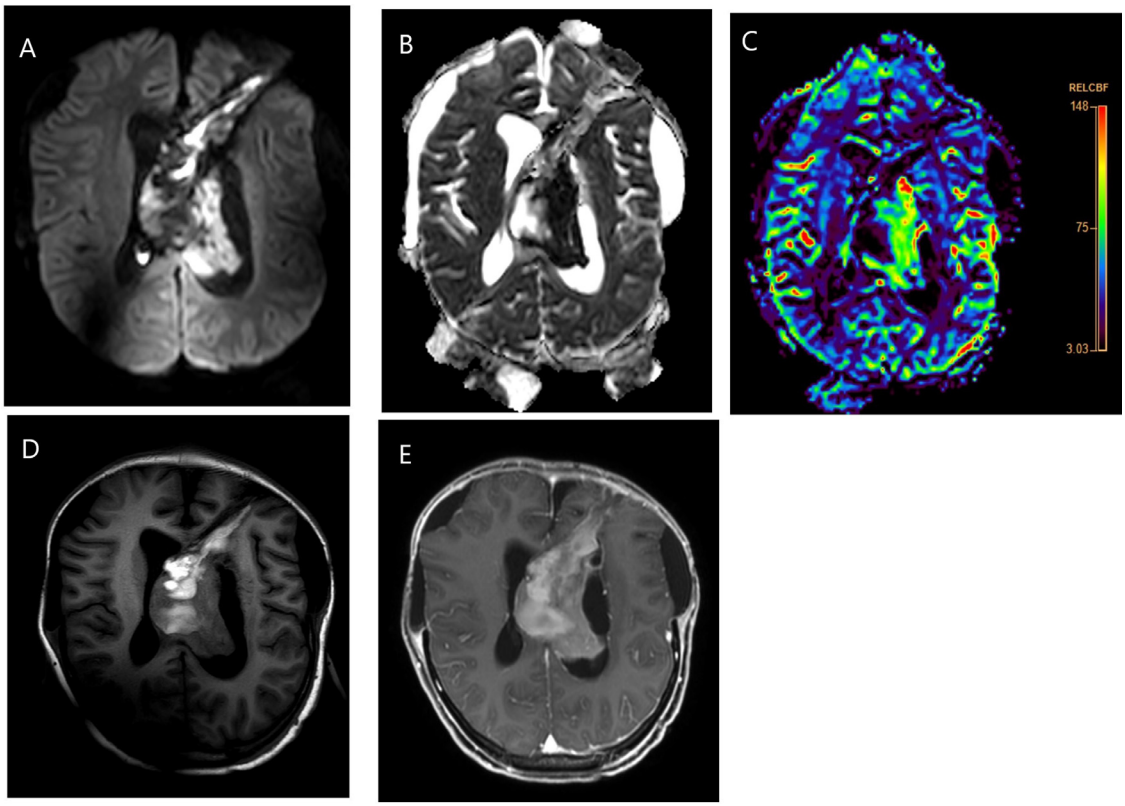

Fig. 5 – Two-months after the initial surgery. Patient required a second surgery with bilateral decompressive frontal craniotomy (A and B) Diffusion weighted image and ADC reveal central zone of restriction. (C) Perfusion image shows increased vascularity areas corresponding with tumor remnants. All images show the external ventricular drain placed during the first surgical procedure. (D) T1 weighted image and T1 with contrast (E) respectively. Blood products are spontaneously hyperintense on T1. The remnants of the mass demonstrates slight contrast enhancement.

neurocytoma instead of the expressions atypical or anaplastic [15].

There are some imaging features to take into consideration for the diagnosis of intraventricular neurocytoma. It is described as a soft tissue, well-defined margin mass. It shows broad contact with septum pellucidum or the ventricular walls because of its origin on the bipotential cells of the germinal matrix. Its location is mainly in the frontal horns of the lateral ventricles with some variability inside the ventricular system [9]. Neurocytoma demonstrates cystic components in the form of clusters, giving the mass a "soap bubble" appearance, specially in bigger tumors. This component tends to be located peripherally because it is not secondary to hemorrhage or necrosis unlike other tumors [3]. The presence of scalloped contours with septa that bridge the mass to the ventricular wall is one of the most specific signs of neurocytoma [3,16]. Calcification is present in 50% of neurocytomas but is also a feature of other tumors. On MRI intraventricular neurocytoma is iso to slightly hyperintense to gray matter on T1-T2 weighted images. Susceptibility weighted images demonstrate signal void for the vessels inside the lesion giving it a granular appearance [16]. With intravenous contrast neurocytoma shows moderate and heterogeneous enhancement [7]. The lesion presents restriction on diffusion weighted images. Sakamoto et al proposed that the minimal value for ADC in neurocytoma has an inverse relationship with the malignant potential of the tumor. Therefore, lower values of ADC relate with malignant potential and greater values of Ki-67 in patients with neurocytoma [17].

In the case presented, there were several features that heightened suspicion of neurocytoma. The location of the mass within the ventricular body and its broad attachment to the septum pellucidum were notable. The mass appeared predominantly solid, with a scant cystic component on the periphery, which made it difficult to recognize the classic "bubble soap" appearance typical of neurocytoma. However, it was possible to identify some septa bridging the lesion to the ventricular wall. This finding gave the mass a scalloped appearance, which is more characteristic of neurocytoma. Prominent vessels and calcifications within the lesion are features commonly observed in many intraventricular tumors. Additionally, the mass was well-defined, uniform, and did not exhibit signs of compromise of the adjacent brain parenchyma, findings that made diagnosis of neurocytoma more likely.

Among the main differential diagnoses for intraventricular masses on young adult patients there are: oligodendroglioma, ependymoma, subependymal giant cell astrocytoma and intraventricular meningioma [7]. Oligodendroglioma is rarely intraventricular. If it is the case, it can be found mainly in the body of the lateral ventricle. It has gross calcifications in 90% of

cases. This lesion is very heterogeneous with important areas of hemorrhage and necrosis. On T2 weighted images the irregularity of its content is more conspicuous. This tumor tends to be more aggressive and can infiltrate adjacent brain areas [3,18]. Ependymoma is generally located in the fourth ventricle. It has calcifications and hemorrhage [19]. Subependymal giant cell astrocytoma is mostly situated near the foramen of Monro. It avidly enhances with intravenous contrast and is strongly linked to tuberous sclerosis [3,16]. Intraventricular meningioma presents as a uniform mass that frequently calcifies. It has a tendency to manifest in a slightly older age group (30-60 years). It is situated frequently in the ventricular trigone. With intravenous contrast it enhances avidly. Perfusion values demonstrate higher rCBV (between 6 and 9) in comparison with neurocytoma [3].

This article presents the case of a female adult woman with headache, nausea, and vomiting. These symptoms are secondary to intracranial hypertension and are frequent, but not exclusive of neurocytoma [10]. Imaging studies revealed a mass in the body of the lateral ventricle, adjacent to the foramen of Monro. The lesion was mainly solid, with discrete peripheral cystic component and septa bridging to the wall of the ventricle. It also exhibited calcifications within. The mass was quite uniform, with no areas of hemorrhage or necrosis as in oligondendroglioma. Its location made the diagnoses of meningioma an ependymoma less likely. Perfusion study revealed intermediate value unlike meningiomas which show a higher rCBV.

Unfortunately, even with the imaging diagnosis of neurocytoma, it is difficult to predict the behavior and prognosis of the tumor. In this case, immunohistochemical analysis revealed Ki-67 value of 7%, consistent with at as a complication in atypical neurocytomas has not been described previously and should be considered for future evaluations. More investigations about this subtype of tumors are necessary. Its prompt recognition could potentially improve the management of these patients and offer a differential consideration in the therapeutic algorithm.

Conclusion

Atypical intraventricular neurocytoma is an unusual lesion with different behavior and prognosis in comparison with most central neurocytomas. The low incidence of these tumors makes it difficult to obtain differential information about them. It is important to increase the reports of cases of this subgroup of tumors. Obtaining more information about its imaging features, clinical evolution, therapeutic management and subsequent evolution will allow the establishment of differential algorithms for this group of patients.

Patient consent

As the author of the article, I confirm that I have obtained written and signed informed consent for publication of the article from the patient(s), their guardian(s), or legal representative(s).

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