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Third Ventricle Craniopharyngioma Intraventricular Tumor: A Case Report and a Brief Literature Review

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Abstract

Craniopharyngioma is a rare embryonic malformation, usually benign, of the sellar or parasellar regions. In this study, an uncommon case of third ventricle craniopharyngioma and a succinct review of its prevalence are presented. Even with low mortality rates, these injuries involve a high degree of endocrinological, visual, and neuropsychological morbidities, which have a huge impact on the patient's quality of life. Thus, surgical resection is the preferred therapy for tumors that compromise the flow of cerebrospinal fluid. However, due to the location of the craniopharyngioma, surgical management is accompanied by the risk of injury to important adjacent areas with postoperative repercussions. Therefore, the neurosurgeon's experience and the choice of the best surgical approach, are crucial for increasing prognosis.

Keywords
- craniopharyngioma
- third ventricle
- cerebral ventricle neoplasms
- neurosurgery

Introduction

Craniopharyngioma (CP) is a rare embryonic malformation of the sellar/parasellar region with a low histological grade, with up to half the 0.5 to 2.0 new cases per million population per year occur in children and adolescents and is a usually benign central nervous system tumor accounting for 2 to 3% of intracranial neoplasms.[1–3] These commonly arise from the epithelial nests of Rathke's pouch, located on the anterior pituitary gland, and surround the hypothalamic stalk, purely

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intraventricular CPs are rare.[2] These tumors show an intact third ventricle floor, a patent suprasellar cistern, a normal pituitary stalk, and an absence of sellar abnormalities. Surgeries for these lesions pose significant technical challenges because of important surrounding structures.[4]

Today, the literature shows two main surgical approaches for these CP types, the translamina terminalis and the transventricular approaches. The translamina terminalis approach using subfrontal, pterional, or basal interhemispheric openness approaches has been used to treat suprasellar or intraventricular CPs. Although this is an easier option for accessing tumors in the inferior part of the third ventricle, it is difficult to remove large lesions. On the other hand, transventricular approach through a transcortical or transcallosal openness approach allows access to the third ventricle with transforaminal, subchoroidal, or transfornician approaches. The usefulness of these approaches has been reported for an intraventricular tumor removal, but there is a risk of injuries to the surrounding neural or vascular structures, such as the body of the fornix.[4]

This article focuses on a rare case of a third ventricle CP and a brief review with its prevalence.

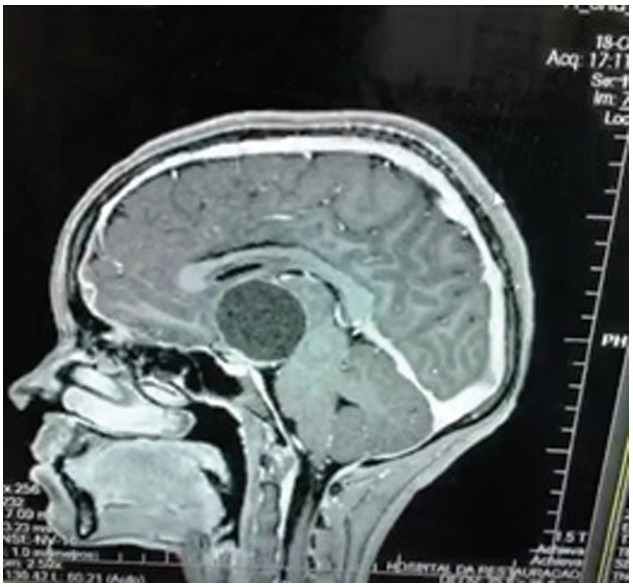

Fig. 1 Magnetic resonance imaging (MRI), and contrast-enhanced T1-weighted on the sagittal section shows ring-like uptake, intraventricular oval cystic lesion with compressive effect.

Report Case

T.M.A., female, 15-year-old, was admitted at a neurosurgery care unit reporting several occurrences of a holocranial headache, in tightness, strong intensity, that makes her wake up during the morning with vomiting and no association of picture or phonophobia and she reports worsening at the end of the day, without analgesics improvement associated with dizziness in 3 days of admission and weight loss of approximately 26.4 lb in the last 6 months. She described a delayed pubertal development without Menstruation and no family history related to this patient's condition.

On examination with 15-lead electroencephalogram, we found isofoton reagent pupils and preserved motricity. Fundoscopy with signs of papilledema and presence of "setting sun" were also found. Reflexes, campimetry, coordination, sensitivity, and motricity are unchanged. Was solicited a computerized tomography showing an intraventricular dilatation, a cyst in the third ventricle, and signs of ependymal transudation with Evans 0.38. Was solicited a magnetic resonance imaging, and contrast-enhanced T1-weighted on the sagittal section shows ring-like uptake, intraventricular oval cystic lesion with compressive effect (►**Fig. 1**). Axial T2-weighted fluid-attenuated inversion recovery image showed a hyperintense lesion in the third ventricle (►**Fig. 2**) and contrast-enhanced coronal T2-weighted image highlighted a cystic lesion of the third ventricle and a subdural collection (►**Fig. 3**).

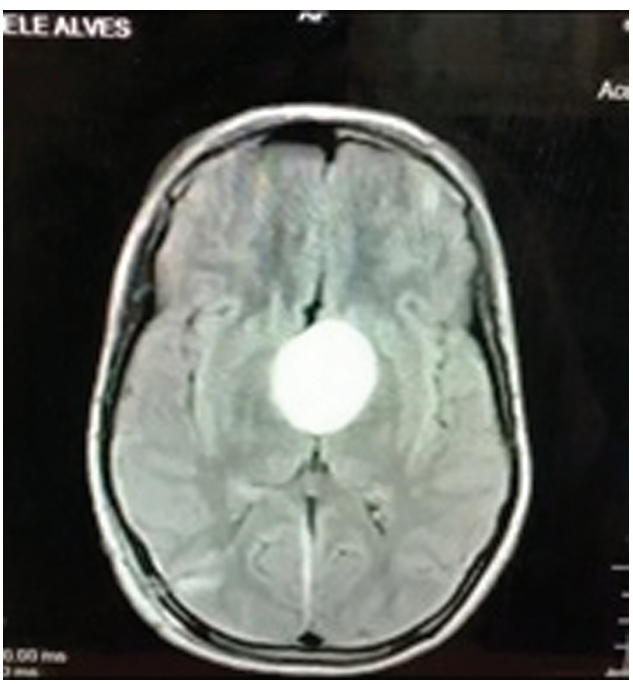

Fig. 2 Axial T2-weighted fluid-attenuated inversion recovery (FLAIR) image showed a hyperintense lesion in the third ventricle.

The patient was submitted to a ventriculoperitoneal shunt and a microneurosurgical with a transcallosal approach to complete resection of the tumor, without intercurrences. Beginning with patient in lateral Recumbency, in cephalic lateralization to the right, with sagittal suture in parallel position to the ground (►**Fig. 4**). The craniotomy is placed two-thirds in front and one-third behind the coronal suture and the durotomy with dissection of the interhemispheric plane to expose the corpus callosum (►**Figs. 5** and **6**). Callosotomy was performed and identification of the foramen of Monro, by the choroid plexus (►**Fig. 7**). Enlargement of the callosotomy was performed to identify the cystic component of the lesion. Aspiration of the content and subsequent resection of the capsule (►**Fig. 8**), and finally had a External Ventricular Drain catheter insertion (►**Fig. 9**).

The histopathology verified an adamantinomatous CP subtype. In the last medical follow-up after surgery, she

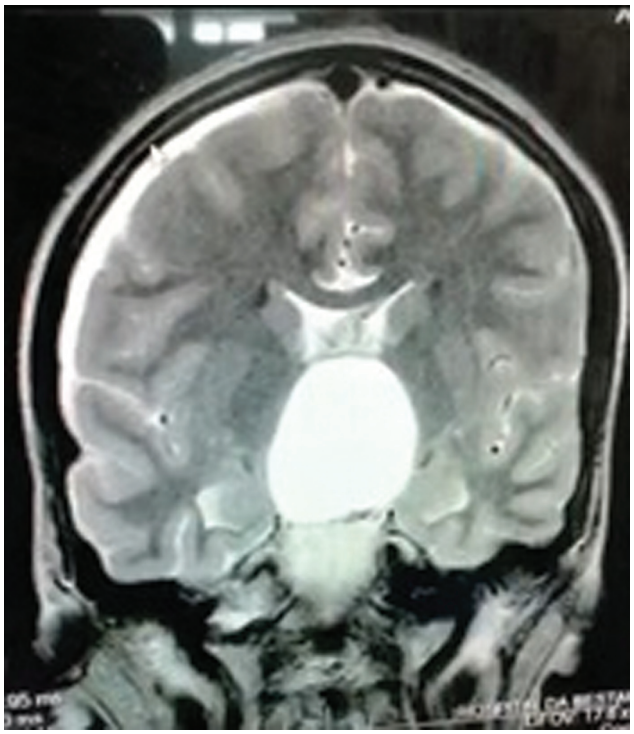

Fig. 3 Contrast-enhanced coronal T2-weighted image highlighted a cystic lesion of the third ventricle and a subdural collection.

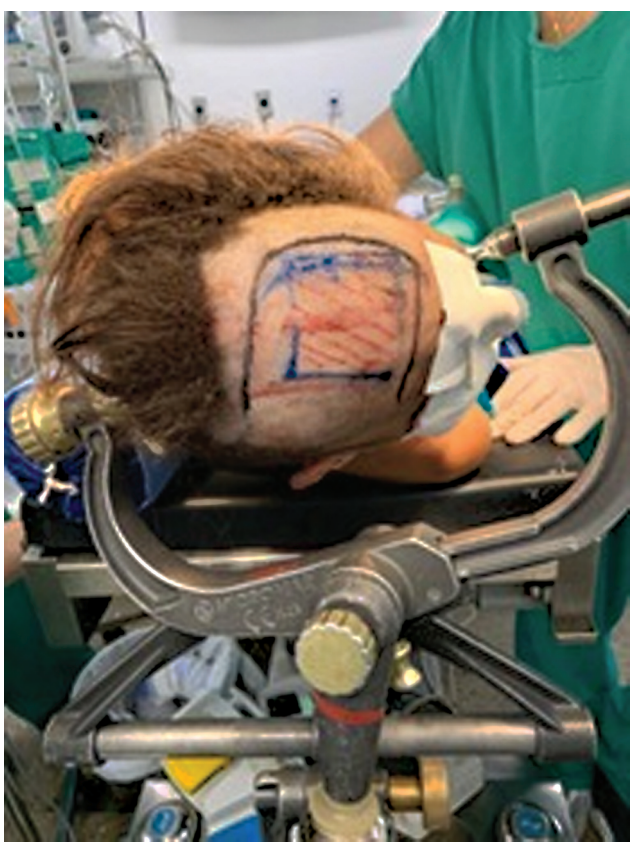

Fig. 4 Patient in lateral decubitus, in cephalic lateralization to the right, with sagittal suture in parallel position to the ground.

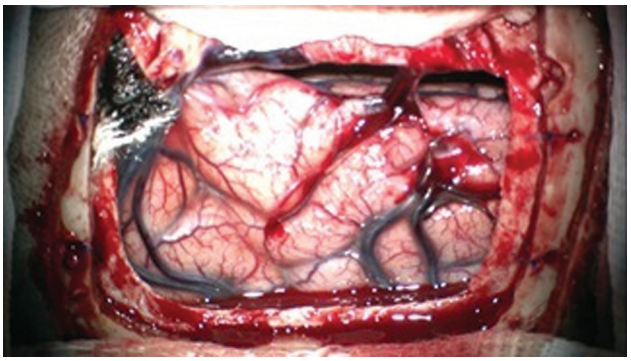

Fig. 5 Craniotomy placed two-thirds in front and one-third behind the coronal suture.

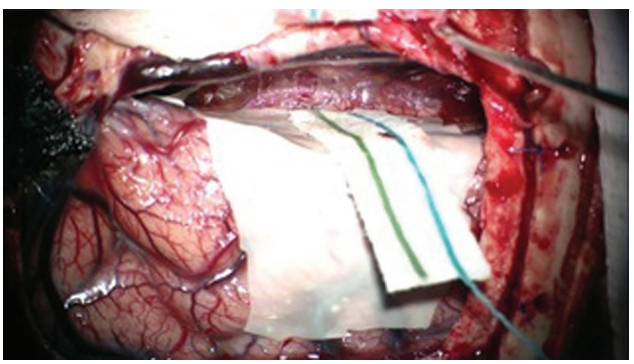

Fig. 6 Durotomy with dissection of the interhemispheric plane to expose the corpus callosum.

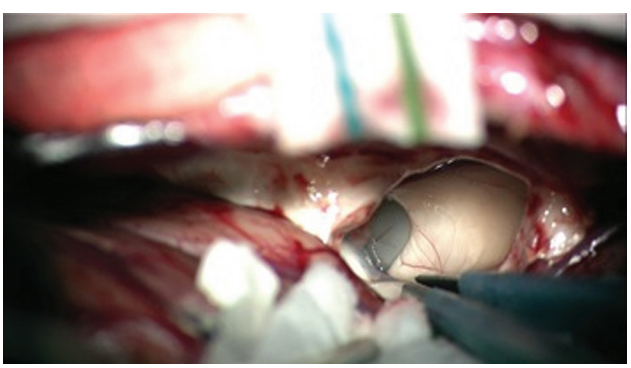

Fig. 7 Callosotomy was performed and identification of the foramen of Monro, by the choroid plexus.

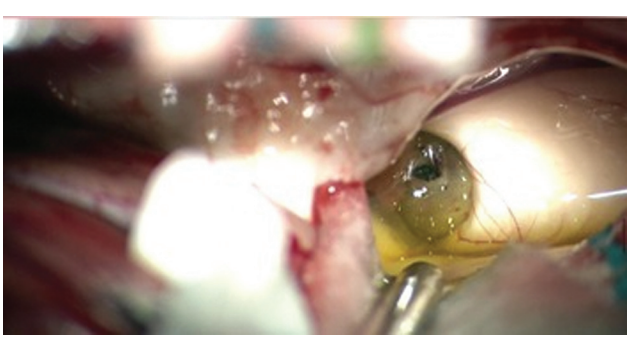

Fig. 8 Aspiration of the content and subsequent resection of the capsule.

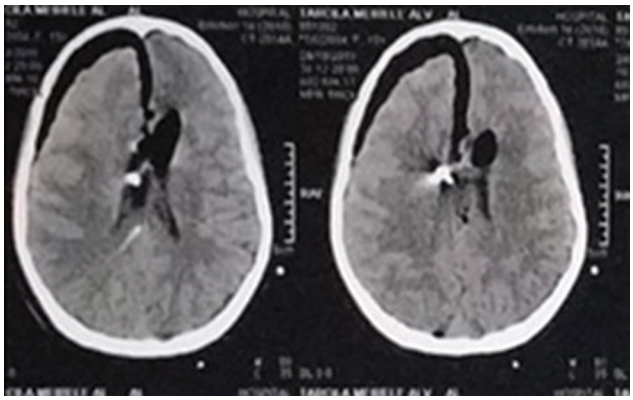

Fig. 9 PIC-DVE catheter insertion.

was doing well, although diagnosed with diabetes insipidus, which was treated with desmopressin (DDAVP).

Discussion

The incidence of purely intraventricular CPs accounts for 0.5 to 11% of all CPs. Third ventricle tumors are surgical challenges because of the complex surrounding structures, including the hypothalamus, infundibulum, optic pathways, limbic system, and nearby vasculature. Tumor dissection within the third ventricle can cause various injuries, such as hemiparesis, memory loss, increased endocrinopathy, or visual loss. In general, intraventricular CP occurs at a greater age, and its most common presenting symptoms are headache and raised intracranial pressure, rather than primary visual deterioration. The visual field defects in such cases could be due to chiasmatic compression, due in turn to the bulging of the tumor.[2,4]

The most appropriate therapeutic option in the treatment of patients with CP remains a controversial subject in the literature. Even after surgical treatment, recurrences are frequent, resulting in a great impact on morbidity and mortality and quality of life of these patients. In neoplasms of the anterior portion of the third ventricle, as well as the suprasellar region, occlusion of the interventricular orifices may occur. This technique consists of placing two siliconized rubber catheters in the lateral ventricles or subdural spaces, by means of paramedian trepanizations in the posterior parietal region; these catheters are connected to a T-shaped probe (Kehr probe) which, in turn, is connected to a Holter valve.

Basically, there are three major approaches to the third ventricle: transcallosal, transcortical, and endoscopic approaches. For primary tumors, the transcallosal technique is the most suitable, because it allows direct access to the origin of the tumor through the exposition of the third ventricle in addition to the frontal horn and body of the lateral ventricle. It begins with a craniotomy, which is placed two-thirds in front and one-third behind the coronal suture, succeeded by the opening of the dura mater and its reflection over the superior sagittal sinus. Once the first steps are completed, the interhemispheric dissection begins, with the subsequent identification of the pericallosal arteries and the corpus callosum's surface. Thereby, after making a little incision of 2.5 cm behind the genu of the corpus callosum and entering the lateral ventricle, there are three options for accessing the third ventricle: transforaminal, transcoroideo transvelum interpositum, and interforniceal approach. The transchoroidal approach accesses the third ventricle through an incision on the choroidal fissure, which ends up allowing the visualization of the roof of the third ventricle with the allocation of the fornix to the contralateral side. Transchoroidal approach could be associated with hemiparesis, mutism, and infarction of the basal ganglia.[5] In contrast, after the visualization of the septum pellucidum, in the interforniceal approach, an incision in the midline of this structure is performed, to dissect until reaching the fornix fibers. The dissection must be performed carefully, so as not to damage the anterior commissure or the region posterior to the foramen of Monro. This approach can be associated with hemiparesis and transient memory loss.[5]

The transcortical approach of the third ventricle CP could be made by pterional, frontolateral, or orbitozygomatic craniotomies, and each of these different techniques has its own specific benefits. The pterional craniotomy provides a shorter path toward the parasellar region. Alternatively, the orbitozygomatic craniotomy is useful in tumors growing in the vertical axis of the third ventricle. Lastly, in the frontolateral or supraorbital technique, the patient is placed with neck extension and rotation at 30 degrees contralaterally to the side of the craniotomy. After the patient's placement, the incision of the skin is started and performed with the lower margin inserted in the axial plane of the orbital root and the craniotomy is fulfilled in a semicircular pattern. Thus, the free bone flap is positioned over the central region of the middle frontal gyrus. Thereby, the medial border of the flap is placed 2 cm off the midline of the coronal suture, whereas the posterior border of the flap is positioned 2 cm after this suture. Therefore, the route to be taken by the transcortical approach must be carefully evaluated to avoid involvement of important cortical areas.[5]

The endoscopic approach is a recommended minimal access technique for resection of a small suprasellar CP, which may extend to the third ventricle.[6] This approach is performed by the posterior planum, athwart the tuberculum sellae and the anterior wall of the sella. Thus, after a sphenoidotomy is accomplished and the dura mater is exposed and pierced, the CP becomes visible and resection is possible. However, endoscopic management is frequently associated with a higher recurrence rate.

CP is a rare embryonic malformation, usually benign, of sellar and suprasellar origin. This tumor has a slow growth rate and rarely extends beyond the third ventricle. However, due to its location, adherence, and infiltration of eloquent structures belonging to the central nervous system, CP starts to present various endocrinological, visual, and neuropsychological manifestations, in addition to repercussions due to the mass effect on intracranial pressure, such as headache, nausea, vomiting, and papilledema. Thus, CP with hypothalamic invasion and penetration of the third ventricle is a tumor that has a high degree of progression and morbidity, which requires an individual management based on its size,

predominant composition, location, and anatomical characteristics to define surgical access.

Conclusion

The third ventricle CP is a rare tumor, which requires experience and ability to treat. It is a challenge controlling border lesions and avoiding damage to healthy structures while accessing the third ventricle. The choice of the approach is very important to surgical outcome and prognosis. So, the transcallosal approach and ventricular peritoneal shunt were with less complications. Furthermore, interdisciplinary neurointensive and oncology cares were fundamental to patient recovery.

Conflict of Interest
None declared.

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