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Choroid plexus papillomas of the III ventricle in infants

Report of three cases

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Abstract The III ventricle is an uncommon location for choroid plexus papilloma at any age. We describe three new cases of choroid plexus papillomas of the III ventricle (CPPs). All children were boys under 4 months of age and all presented with increased intracranial pressure, hydrocephalus and macrocephaly. The three were examined by preoperative computed tomography (CT) and ultrasonography. Two of them were investigated with magnetic resonance imaging (MRI). The first case was treated with a right corticofrontal transventricular approach and subtotal resection, so that he required a second operation through a transcallosal approach. In the other two cases a transcallosal approach was used. Two children needed permanent ventriculo-peritoneal shunts. The average follow-up of 4.3 years has revealed no neurological deficits in any case. The timing of and the need for shunting are major considerations. Clinical and imaging follow-up (CT and/or ultrasonography) are very helpful in controlling postoperative hydrocephalus and subdural effusion, avoiding unnecessary shunting in many cases. The operative approaches, transcortical and transcallosal, are discussed.

Key words Brain neoplasm · Childhood · Choroid plexus papilloma · Hydrocephalus · III Ventricular tumors

Introduction

Choroid plexus tumors (CPTs) are uncommon neoplasms of the central nervous system; they arise in children within the ventricles or cerebellopontine angle [10, 12].

CPTs are reported to be more common in children than in adults [1, 11], accounting for 2.3–3% of intracranial tumors in children under the age of 17 years [9, 14], as against 0.4–1% in all-age brain tumor series [5, 7].

Several large series have found that 11–16% of childhood brain tumors occur in children less than 2 years of age, but 70% of patients with choroid plexus papillomas (CPPs) are less than 2 years old [1, 15]. Matson and Crofton [9] and Humphreys et al. [5] have described the first year as the most common time of life (40–50%) for CPPs. Only 29–39% of CPTs are choroid plexus carcinomas [6, 13].

The III ventricle is an uncommon location for CPPs at any age [1, 5, 11, 16]. This report will focus on three cases of III ventricle CPPs from our series of eight CPTs in infants and children diagnosed and treated since 1974 in our institution.

Summary of cases

Three cases of CPPs of the III ventricle have been managed at the Hospital Sant Joan de Deu of Barcelona during the last 20 years.

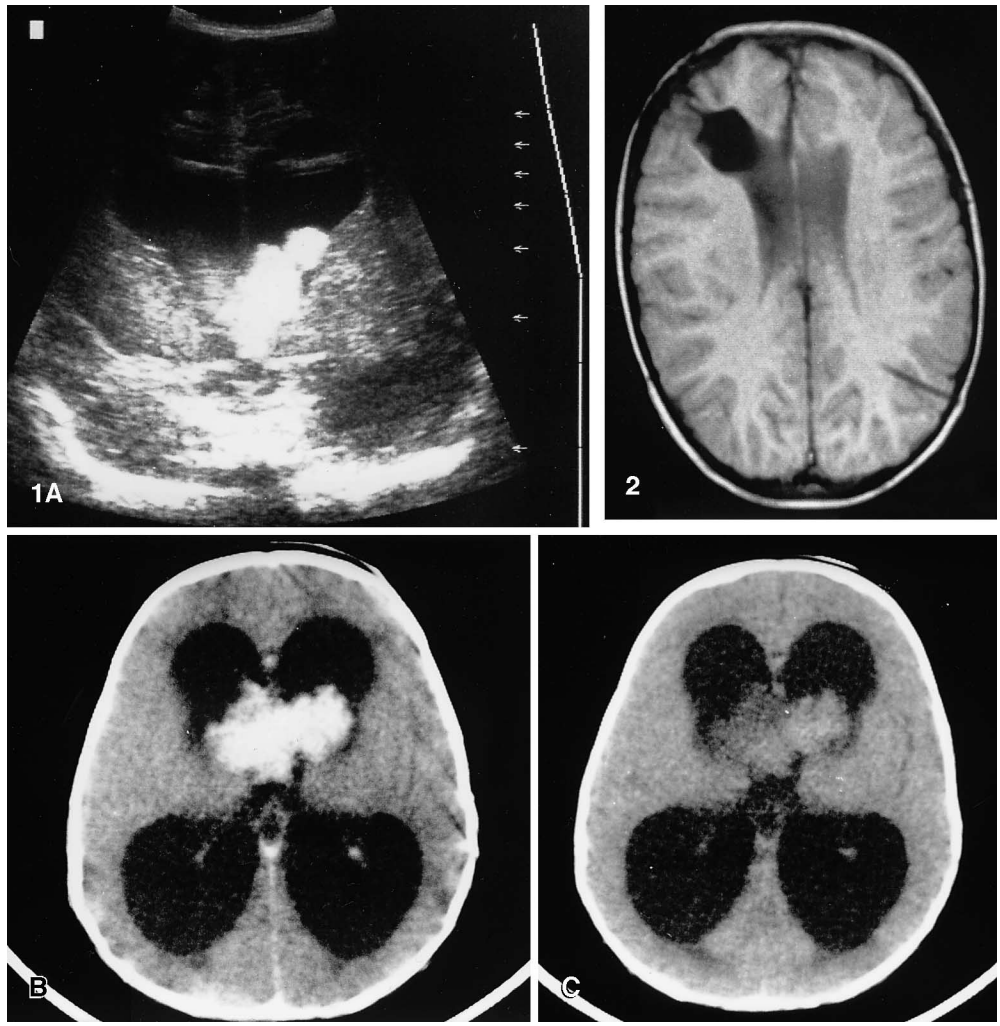

Fig. 1A–C Case 1: **A** Intracranial ultrasonography shows an hyperechogenic mass in the III ventricle, extending toward the lateral ventricle through the foramen of Monro. **B** CT scan post EV infusion of contrast medium shows important ventricle enlargement and an enhanced cauliflower-like mass arising from the III ventricle. **C** Without contrast enhancement, the lesion is isodense with the brain parenchyma

Fig. 2 Case 1: MRI 20 months after surgery: no subdural effusions are seen. The parenchymal lesion caused by the transcortical frontal approach is evident

Case 1

A 2-month-old white baby boy was admitted to our institution because of a 1-month history of progressively enlarged head. Physical examination on admission revealed a head circumference of 41 cm (+3δ), sunset eyes and a normal neurological examination. A cranial ultrasound study and subsequent CT scan (Fig. 1) showed massive obstructive hydrocephalus secondary to a III ventricle tumor.

A right frontal transcortical approach was used and partial removal of the tumor was carried out as there were difficulties in identifying the pedicle through the foramen of Monro. Fifteen days later the rest of the tumor was totally removed by an interhemispheric transcallosal approach. Owing to persistent hydrocephalus following removal of the papilloma a ventriculo-peritoneal (VP) shunt was inserted.

Postoperatively, an important asymptomatic subdural effusion was detected, which disappeared 4 months later without specific treatment (Fig. 2).

The pathological diagnosis was CPP.

Nine years later the patient is asymptomatic with a good neuropsychological development, but is shunt dependent.

Case 2

This 4-month-old boy had a 3-month history of irritability and increasing head size. The clinical examination showed an enlarged head with bulging fontanels and splayed sutures. He had poor upward gaze. No other clinical findings were detected. A CT scan showed massive obstructive hydrocephalus secondary to a large III ventricle tumor. On MRI the mass was localized to the III ventricle and appeared to be hyperechogenic on both T1- and T2-weighted pulse sequences (Fig. 3). The T1-weighted image obtained after the perfusion of paramagnetic contrast medium (Gd-DTPA) demonstrates significant enhancement of the tumor.

The hydrocephalus was treated before the craniotomy with a VP shunt. The tumor was felt to be completely resected through a transcallosal approach, and the postoperative course was uneventful.

Histological investigation of the tumor showed fibrovascular fronds covered with numerous papillae, resembling normal choroid plexus.

Postoperatively asymptomatic subdural effusions were present, which disappeared 6 months later (Fig. 4).

His current condition, 2 years postoperatively, is good.

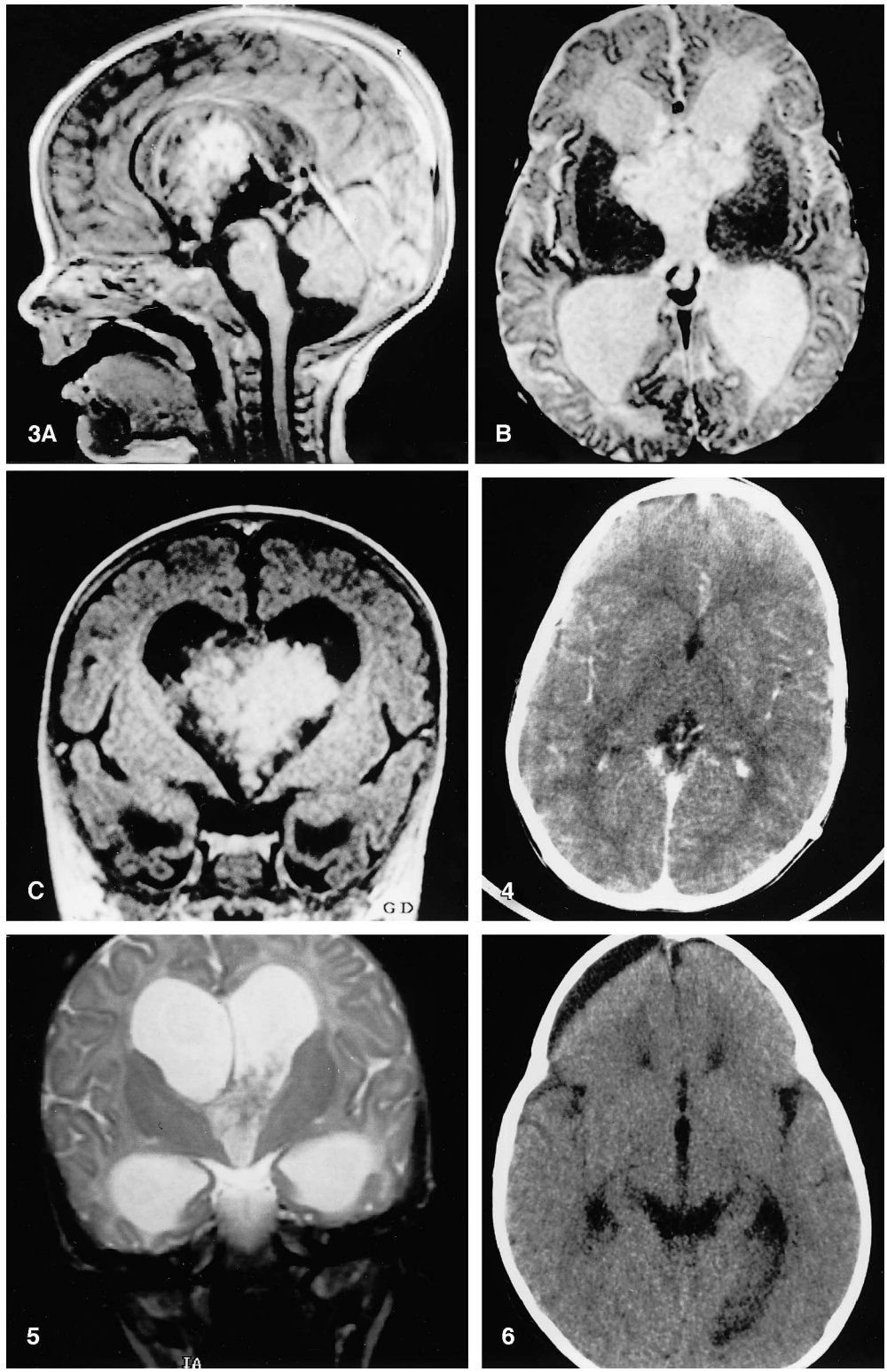

Fig. 3A–C Case 2: preoperative MRI, **A** sagittal, **B** axial, and **C** coronal views. A large mass is observed in the III ventricle, protruding into the lateral ventricles through the foramen of Monro. There is important dilatation of the ventricular system

Fig. 4 Case 2: Enhanced CT scan 28 months after surgery. The ventricular system is normal (the patient is shunted) and no lesion is observed in the III ventricle. No subdural effusions are seen

Fig. 5 Case 3: MRI in coronal section, demonstrating the origin of the tumor from the III ventricle

Fig. 6 Case 3: CT scan 24 months after surgery (the patient was not shunted). No hydrocephalus is seen. There is only a small asymptomatic subdural collection

Case 3

A 3-month-old white baby boy presented with a 1-month history of increased fronto-occipital circumference and bulging fontanels. On examination his general condition was excellent and no neurological deficits were found. MRI showed a large lesion in the III ventricle, which was compatible with CPP. Note the vascular nature of the tumor (Fig. 5). A transcallosal approach was used, and the tumor was totally resected by the piecemeal method. Postoperatively, the child had a important subdural fluid collection; it was clinically significant, requiring a subdural shunt for 3 weeks.

The typical pattern of CPPs was found on histological examination.

The postoperative course was benign. Two years later the child is asymptomatic, and the subdural effusions have almost disappeared (Fig. 6).

Discussion

Estimates of the incidence of CPP in large series of brain tumor cases include 0.5% among 200 intracranial tumors [3]. Davis and Cushing [4] reported 12 CPPs in their series of 2023 brain tumors, 2 of which were in children.

CPPs are reported to be more common in children than in adults [1, 9, 11], and in pediatric series a greater proportion of brain tumors are CPPs [2, 6, 11]. They appear predominantly within the first decade of life, but 70% of patients are less than 2 years old [1, 9, 15, 18]. The first year is the most common time for CPPs [5, 9, 16], as in our three cases.

CPPs of childhood usually occur in the lateral ventricle, as opposed to those of adults, for which the IV ventricle is a more common location [10, 16, 18]. CPPs are rarely found in the III ventricle [1, 6]. None appeared in the large series reported by Raimondi and Gutierrez [11], Matson and Crofton [9] and McGirr et al. [10]. Tomita et al. [16] found a relatively high incidence: 29%. A high proportion of CPPs of the III ventricle is found in patients under 1 year of age. All our cases are younger than 1 year [1, 5, 7, 11, 16].

Clinical findings

Children with CPTs usually present with signs and symptoms of increased intracranial pressure because of obstructive hydrocephalus [1, 6, 7, 10], and especially children less than 2 years of age, as our patients were. None of them had vomiting or developmental delay, owing to the cranial plasticity typical of this age-group. Signs such as vomiting, ataxia and any others are less frequent and usually appear in patients older than 2 years [6]. Except for macrocranium and sunset eyes, the neurological examination in our three cases was normal. The reason why the patients with CPPs located in the III ventricle are younger than those with CPPs in the lateral ventricles may lie in the mechanism of development of the hydrocephalus: while the tumors located in the lateral ventricles make for overproduction of CSF, those located in the III ventricle cause an obstruction.

Neuroimaging

CT and MRI are currently the investigative procedures of choice for the diagnosis of a CPP. Owing to the early age at presentation with these tumors, a very useful diagnostic procedure is ultrasonography, which has the advantages of easier examination, accuracy and lower cost (Fig. 1A).

CT allows accurate, non-invasive localization of the tumor and narrows the differential diagnosis. CPPs are isodense and enhance homogeneously [6] (Fig. 1B,C).

MRI provides anatomical resolution and vascular detail, but heavy sedation is required, which means that MRI must not be used as the first diagnostic test or for short-term control, but it is certainly the most accurate diagnostic procedure. The preliminary diagnosis can be made by ultrasonography and confirmed by MRI, and for short-term controls ultrasonography and/or CT can be used. The MRI of CPPs is generally one of intermediate signal intensity on T1-weighted images and of either intermediate or increased signal intensity with T2 weighting [17].

Hydrocephalus and subdural effusions

Hydrocephalus associated with CPPs has attracted researchers' attention. Hydrocephalus can be life threatening and warrant immediate attention. Clinical evidence confirming CSF overproduction by CPPs is provided by the observation that postoperatively the hydrocephalus has resolved in several cases. The CT appearance indicates that patients with CPPs in the lateral ventricle have bilateral ventriculomegaly without obstruction of the CSF pathways.

However, CPPs of the III or IV ventricle can cause hydrocephalus by a combination of CSF pathway obstruction and overproduction [16] (Fig. 3A); in fact, two of our cases required shunt placement, probably because of this combination. In cases shunted before craniotomy there is some doubt as to what would have happened had they been treated without precraniotomy shunting.

The problem of postoperative subdural fluid collections is not often discussed in the literature [6, 9, 16]. A subdural shunt can be necessary to control a progressively increasing head size. In a review, Koos et al. [8] recommended shunting of the subdural space if a subdural fluid collection became a problem. It appears that a postoperative subdural fluid collection results from persistence of a ventriculosubdural fistula.

It is questionable whether preoperative shunting will aid in prevention of this complication, and in fact the patient recorded here as case 2, whose hydrocephalus was drained

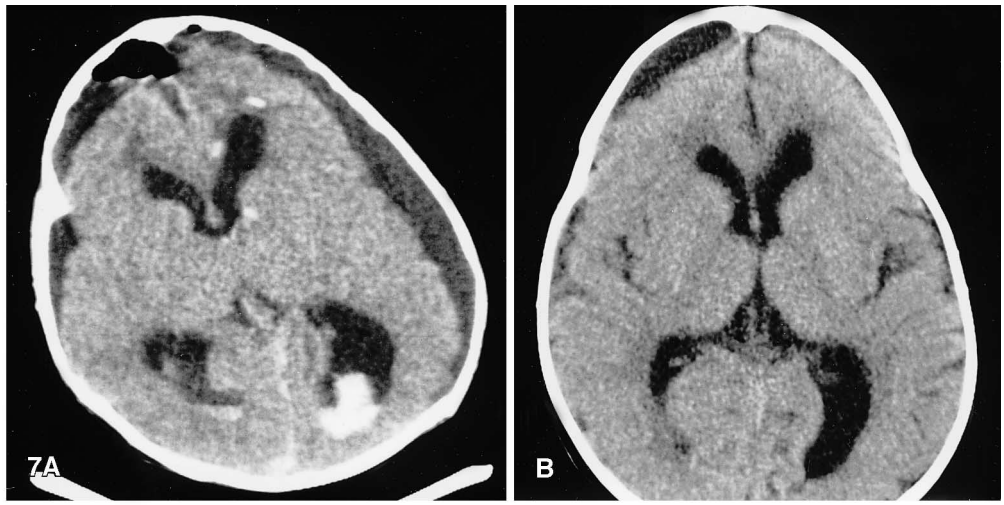

Fig. 7A, B Case 3: **A** Immediate postoperative CT scan showing the presence of moderate ventricular dilatation and bilateral subdural effusions. **B** CT scan done 24 months after surgery shows the absence of hydrocephalus and near-total resolution of the subdural collections

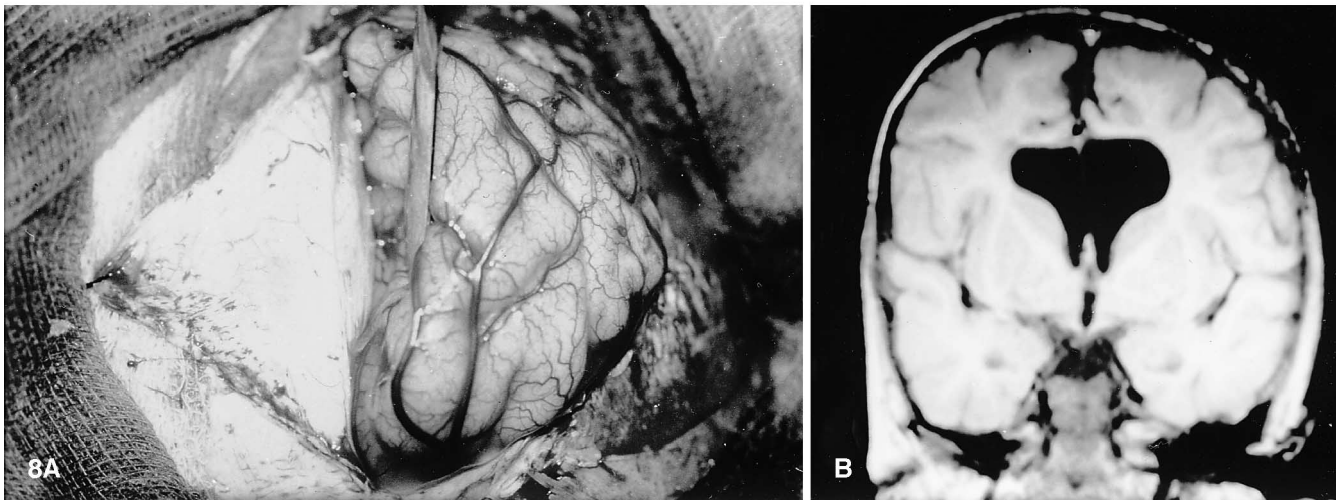

Fig. 8 **A** Intraoperative picture of a typical transcallosal approach (case 3). **B** Postoperative MRI in case 3: there is no evidence of tumor in the III ventricle, and the surgical lesion of the corpus callosum is evident

by a shunt preoperatively, developed a subdural effusion just as did those who were not previously shunted. In our opinion there is no evidence that preoperative shunting prevents subdural effusions.

Subdural effusion usually resolves spontaneously over 3–12 months postoperatively, as in our three cases (Figs. 2, 4, 6, 7).

Surgical approach

Surgical resection remains the treatment of choice for CPPs [6, 7, 10, 16]. Current microsurgical and neuroanesthetic techniques have significantly reduced the operative mortality and morbidity in patients with CPPs.

Two approaches have been advocated for resection of III ventricle papillomas: transcortical-transventricular, and interhemispheric transcallosal (Fig. 8) [1, 6, 9, 16, 18]. The transcortical-transventricular approach can cause postoperative seizures, and if the tumor extends into the contralateral ventricle it may be difficult to resect that portion by this route.

The transcallosal approach was utilized as first procedure in two of our cases. In our opinion this is the better approach for lesions of this kind, because it allows perfect bilateral intraoperative control of the lesion.

If are CPPs of the III ventricle properly recognized and treated the results should be excellent. These patients enjoy excellent neurological and psychomotor development.

In conclusion, CPPs are uncommon benign tumors that are more frequent in children than in adults. Clinically the development of hydrocephalus is typical, which in patients younger than 2 years of age leads to macrocranium and bulging fontanels. This hydrocephalus is thought sometimes to be caused by an overproduction of CSF in the tumoral plexus, but in those cases in which the tumor is sited in the III ventricle an obstructive factor can be added. Typically, subdural collections are present postoperatively, which need to be treated only if they are clinically significant, because in most cases they disappear spontaneously

over 3–12 months postoperatively. Our preferred surgical approach to CPPs sited in the III ventricle is the transcallosal approach. Even if complete tumor resection is achieved it is possible that shunting will be needed because of persistent hydrocephalus. The prognosis is excellent if the tumors are correctly treated.

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