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Choroid plexus papillomas in infancy and childhood *

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Abstract. The present study deals with 15 cases of choroid plexus papilloma, out of approximately 500 cases of brain tumors observed in children up to 16 years old. Several features are considered, including clinical symptoms and signs – mostly related to increased intracranial pressure – radiological diagnosis, pathology and surgical treatment, and results. Surgery may be radical in most cases, with the exception of histologically malignant papillomas. The management results are usually satisfactory. An adequate choice of surgical approach is mandatory and surgical technique should be meticulous, also in order to avoid the risk of intraoperative tumoral seeding.

Key words: Choroid plexus papillomas – Pediatric age

Choroid plexus papilloma is a relatively rare tumor. Data in the literature report an incidence ranging between 0.3% and 0.7% of all intracranial tumors [1, 2, 8, 10]. This percentage increases if only brain tumors of infancy and childhood are considered. In fact, the incidence of choroid plexus papillomas in children under the age of 16 is relatively high, with a peak under the age of 2.

In this study, we report 15 cases of choroid plexus papilloma in children under age 16 out of approximately 500 cases of histologically verified brain tumors, observed during a 35-year interval. Several features such as clinical symptoms and signs, radiological evaluation, pathology, surgical treatment, and results are reviewed.

Clinical material and methods

Fifteen cases of histologically verified choroid plexus papillomas represent in our series 3% of the 500 surgically treated pediatric brain tumors. The location of the lesions of our series is summarized in Table 1. There were 6 males and 9 females, with an average age of 6.5 years.

Clinical findings

A constant feature of choroid plexus papillomas was, in our experience, the paucity of localizing signs and symptoms. The clinical history may be short. It ranged in this series from 20 days to 3 years, averaging 8.6 months. The clinical symptoms were related mostly to increased intracranial pressure (ICP). Actually, head enlargement or headache in older children and/or vomiting, usually represented the first complaint. Focal signs in the form of seizures and/or

Table 1. Topography of choroid plexus papillomas in present series of cases

No. of cases	Age/sex			
1	9 years/F	■		
2	12 years/F	■		
3	17 months/F	■		
4	13 years/M			■
5	3 years/F			■
6	5 months/M	■		
7	4 years/M	■		
8	11 months/M	■		
9	15 years/F			■
10	7 years/F			■
11	3 years/F		■	
12	15 months/M	■		
13	7 years/M	■		
14	11 years/F			■ [a]
15	5 years/F	■		
Total		9	1	5

[a] Cerebellopontine angle

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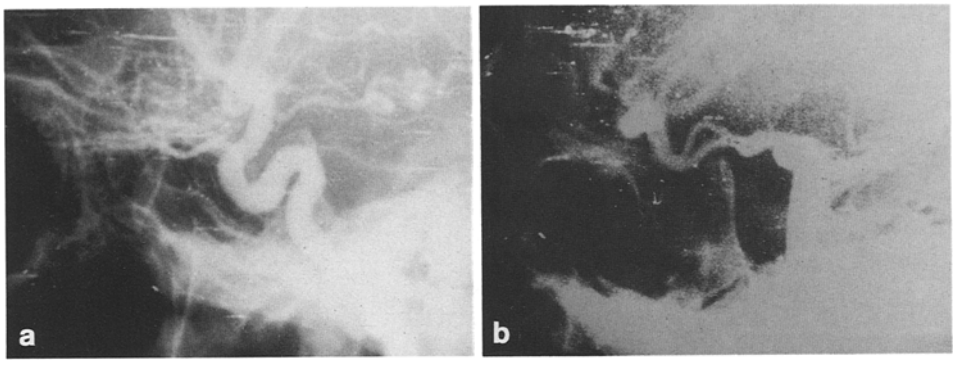

Fig. 1. **a** Left carotid angiography, lateral view: marked hypertrophy of AChA feeding a choroid plexus papilloma of the lateral ventricle. **b** Left vertebral angiography, lateral view, showing the posterior feeders of the choroid plexus papilloma

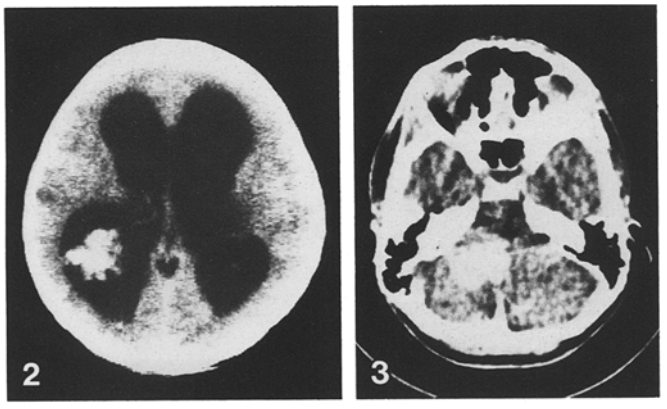

Fig. 2. CT scan demonstrating the typical hyperdense lobulated aspect of a choroid plexus papilloma of the lateral ventricle

Fig. 3. CT scan displaying a choroid plexus papilloma of the cerebellopontine angle

deficits were infrequent (two cases). Neurological signs were also related to raised ICP. Enlargement of the head (obviously in younger subjects) was found in three instances. Papilledema at fundus inspection was present in four cases. Bilateral paresis of the VI cranial nerve affected three children. Unilateral prevalence of deep tendon reflexes was present in only four cases.

Diagnostic aids

EEG usually was not specific. In those cases submitted to lumbar puncture early in this series, common features were an evident increse of ICP (up to 500 cm/H_2O) and the hyperalbuminosis of the CSF (up to 5 g/l).

Radiological diagnosis

Plain X-rays of the skull showed signs of raised ICP and/or hydrocephalus, such as spreading of sutures and, less frequently, increased digital impressions of the cranial vault. In some cases, there were tumoral calcifications, particularly evident in one case.

Gas or iodinated contrast ventriculography were of great diagnostic aid in the past, but now, after introduction of the CT scan, they are no longer indicated. Angiography demonstrated, in our cases, the typical picture of hydrocephalus that may be asymmetric in choroid plexus papillomas of the lateral ventricle. Angiographic signs directly related to the tumor were the presence of a "blush" at the tumor site, the hypertrophy of the arterial feeders of the mass, and evidence of early filling of veins draining from the tumor.

Figure 1 shows hypertrophy of the anterior choroidal artery (AchA) and, to some extent, of the posterior choroidal artery (PChA), in a supratentorial choroid plexus papilloma. On the other hand, infratentorial tumors show hypertrophy of vermian or medullary branches of the PICA and/or the precentral branches of SCA. Early drainage is evident in occipital horn and thalamostriatal veins in supratentorial tumors, while infratentorial cases show an early filling of incisural veins draining into the great vein of Galen or into the tentorial venous sinuses.

CT scan features were variable in the choroid plexus papillomas examined. More commonly, the lesions were hyperdense, greatly enhancing after contrast medium injection, lobulated, and with a suggestive location (Fig. 2). An exceptional case of choroid plexus papilloma of the cerebellopontine angle was documented by CT scan (Fig. 3).

Pathology

Macroscopically, the tumor appears as an irregularly lobulated, pink-gray mass resembling a cauliflower and is usually encapsulated. The ventricular trigone area represents a common locus of tumor implantation. The consistency is usually soft and friable if there are no calcifications. Microscopic aspects closely resemble normal choroid plexus with villi formed by a single layer of cuboidal or cylindrical epithelial cells with a vascular-connective tissue axis. Ultrastructurally, the homology with the normal choroid plexus is preserved (cellular polarity, microvilli on the free epithelial surface) [4].

No malignant choroid plexus papilloma was encountered in this series. Tumoral seeding, in our experience, does not appear to be related to histological malignancy. In one instance, a child bearing a benign choroid plexus papilloma of the III ventricle was submitted to control CT scan 4 years after an apparently radical operation. The tomography displayed a tumor with the features of a choroid plexus papilloma of the IV ventricle, most likely a result of tumoral seeding. However, due to refusal of re-operation, the second tumor could not be confirmed histologically.

Surgical therapy

A gross total removal of the lesions was performed in all of the present cases. The lesions were removed "en bloc", whenever possible, following careful obliteration of the vascular pedicle. In order to minimize the risk of seeding during surgical manipulation of the tumor. We carefully protected surrounding structures with cottonoids.

As for lateral ventricle choroid plexus papillomas, among different approaches suggested for these lesions [7, 8, 12, 13, 17] (Fig. 4), we used indifferently the parietal posterior or median temporal routes. For lesions of the dominant hemisphere, parietal access was preferred. An anatomosurgical study of the cadaver enabled us to verify that the distance between cortical surface at incision site and the ventricle do not differ either with posterior parietal or with the temporal approach.

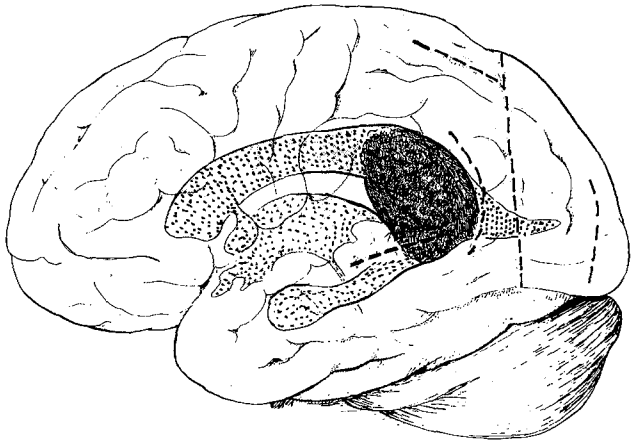

Fig. 4. Scheme of the different surgical approaches to the lateral ventrical choroid plexus papilloma

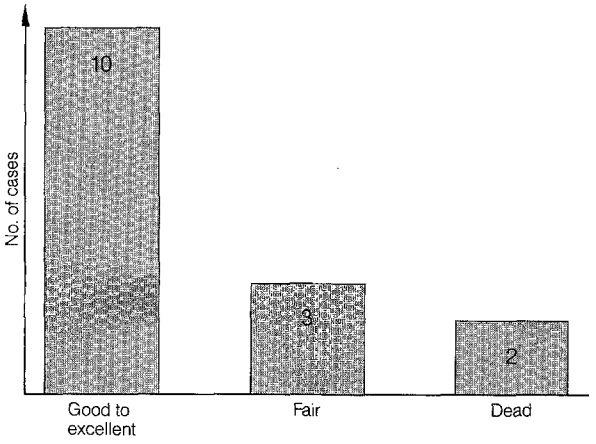

Fig. 5. Results of the present series

Choroid plexus papillomas of the IV ventricle were approached through a suboccipital craniectomy and splitting of the vermis. The only instance of III ventricle papilloma of our series was removed via the transcallosal route. The retromastoid approach was used for the choroid plexus papilloma of the cerebellopontine angle.

Patients of our series were never shunted preoperatively. However, postoperatively, an intraventricular catheter at the tumoral site was maintained for 3 days.

Results

The results of the present series are shown in Fig. 5. Follow-up ranged between 3 and 24 years (average, 14 years). Good results were obtained in ⅔ of patients. It is worth noting that results in children over 5 years of age were consistently good. Among seven cases operated on under 5 years of age, only two could have been classified as good results. Surgical mortality was two cases.

Three cases showed relevant sequelae such as pyramidal weakness and mental retardation, two of which suffered from meningitis and persisting hydrocephalus following removal of lateral ventricle papillomas. In one case hydrocephalus was detected 1 year after removal of the only III ventricle papilloma in this series. Since this case later showed a IV ventricle tumor, a role of this latter in the production of the hydrocephalus could not be ruled out. These three cases were the only ones requiring shunt in the present series.

We have no experience with radiotherapy.

Discussion

The incidence of these tumors in our series is comparable to that reported in the literature [1, 2, 10, 11, 13, 15, 16]. In the pediatric age, the site of predilection of papillomas is the lateral ventricle with an incidence varying between 67% and 75% [1, 8, 9]. Papillomas of the IV ventricle account for 15%, while data in the literature assign an incidence of 8% to III ventricle choroid plexus papillomas [1, 8, 10, 13]. Both sexes were comparably represented in our series, as is the case in the literature [5, 8, 10, 12, 17].

Symptoms and signs of these tumors appeared not to be specific either in our or other series [6, 8, 10, 12, 17], mainly suggesting an increased ICP due to the often coexisting communicating hydrocephalus. Therefore, the chances of diagnosis rely upon radiologic tools [14]. CT scan definitely replace such invasive procedures as ventriculography and pneumoencephalography. The variability of the choroid plexus papilloma image at scan depends upon different enhancing factors inside the tumor, such as calcifications and hemorrhages. However, the typical localizations of the lesions are usually suggestive. We have no experience with NMR diagnosis of these tumors.

The reported incidence of malignant transformation rates between 10% and 30% [9, 18]. No malignant tumor was encountered in our pediatric series. The likelihood or recurrence or tumoral seeding appears not to be strictly related to histological malignancy [8, 9]. Arseni et al. [2] mentioned a case of recurrence of a benign papilloma 19 years after a macroscopically total surgical removal.

Radical surgery represents the primary objective with these tumors, which may be hindered by their high vascularity, expecially when dealing with large lesions. Nevertheless, comparison among larger series of choroid plexus papillomas confirms that aggressive surgical policy is correlated with better follow-up results (Table 2). Operative mortality is usually high, but it is improving, owing to the spreading use of microsurgical techniques. Neurological sequelae such as postoperative hydrocephalus and epilepsy were present in our series, as in the literature [8, 10, 12, 17], mainly in large tumors with intraoperative complications.

As for surgical technique, care should be given to an early control of the vascular peduncle and to minimizing the cortical incision and parenchymal damage in supratentorial tumors.

There have been suggestions that therapeutic preoperative irradiation of choroid plexus papillomas may facilitate surgery by reducing the tumor vascularization. The risk of cerebral damage from irradation, especially in younger patients with benign lesions, in our opinion

Table 2. Data regarding topography and prognosis from major surgical series of choroid plexus papillomas

Authors	No. of operated cases	Lateral ventricle tumors	III Ventricle tumors	IV Ventricle and PCF tumors	Total removal	Surgical mortality	Mortality due to recurrence	Long-term good results
Matson and Crofton [10] (age below 12 years)	15	14	0	1	73%	20% (3)	7% (1)	40% (6)
Raimondi and Gutierrez [13] (up to puberty)	22	19	0	3	?	0	5% (1)	?
Hawkins [8] (age below 8½ years)	16	12	0	4	44%	25% (4)	12% (2)	37% (6)
Present series (age below 16 years)	15	9	1	5	100%	13.3% (2)	0	66.6% (10)

makes radiation therapy a treatment which must be limited to cases with verified histological malignancy.

Preoperative routine CSF shunting has been used [13], but the reduction of ventricular size, limiting the space for surgical manipulations, may be not advisable, particularly when dealing with supratentorial, intraventricular tumors.

In conclusion, choroid plexus papillomas represent a peculiar group of infantile tumors, since they are potentially curable. Therefore, an early diagnosis and meticulous technique are essential to achieve a radial surgical removal without invalidating sequelae for the child.

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