Acta Neurochir (Wien) (1990) 106: 68–72

Acta Neurochirurgica

Choroid Plexus Tumours in Children. Review of 24 Cases

G. Lena, L. Genitori, J. Molina, J. R. S. Legatte*, and M. Choux

Service de Neurochirurgie Pédiatrique, Hôpital des Enfants de La Timone, Marseille, France

Summary

24 cases of choroid plexus tumours (16 papillomas and 8 carcinomas) were observed in the Department of Paediatric Neurosurgery, Hôpital des Enfants de la Timone, Marseille France between 1975 and 1989. The case records were reviewed and the clinical, surgical, and pathological results are presented here. 14 (58%) were infants and 10 (42%) of these were aged less than 1 year at presentation. In 11 (46%) the tumour arose in the lateral ventricle, in 7 (29%) it was located in the IIId ventricle, and in 6 (25%) the tumour was found in the IVth ventricle. 5 of the 8 carcinomas had their origin in the IVth ventricle. 17 cases (75%) required ventriculoperitoneal shunt procedures before surgery of the tumour and in all these it was left in place post-operatively. One child required shunting de novo after removal of the tumour. A total excision was achieved in 20 children (83%) out of 23 operated upon. The operative mortality (mortality within one month of surgery) was 8%, the overall mortality was 25%. One child with a IIId ventricle papilloma died before surgery from a large gastro-intestinal haemorrhage. 5 children under the age of 3 years with carcinomas on the IVth ventricle died. The 3 children with supratentorial carcinomas are alive respectively 2 months, 8 years, and 13 years after treatment. Among 15 children with papillomas alive, 10 (67%) are neurologically intact and 5 (33%) have sequelae.

Keywords: Choroid plexus tumours; tumour removal; children hydrocephalus; shunt; results.

Introduction

Choroid plexus papillomas (CPP) are rare tumours. They represent 0.4 to 0.6% of intracranial tumours in all ages[6, 27, 43]. However, in children, they account for between 1 to 4%[12, 19, 21, 30, 31] and this incidence rises further in children less then 2 years old. Jooma[18] found 12 cases (12%) among 100 tumours of infants less than one year old.

There are several questions concerning these tumours which are the subject of some debate.

1. What is the pathogenesis of the hydrocephalus?
2. Is it necessary to shunt the patient?
3. Is it possible to remove these tumours totally, and
4. What is the prognosis?

24 personal cases of CPP in children have been reviewed and the clinical, surgical, and pathological findings form the basis of this report.

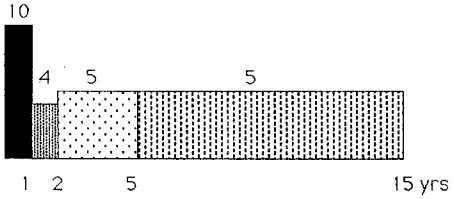
Material and Methods

This series comprises 24 children under 15 years of age seen in the Department of Paediatric Neurosurgery, Hôpital des Enfants de La Timone in Marseille between May 1975 and December 1989.

14 children (58%) were infants and 10 (42%) were less than 1 year old; 5 patients were aged between 3 and 5 years, and 5 between 6 and 15 years (Table 1). 13 children (54%) were male and 11 (46%) were female.

At presentation, 18 children (75%) had clinical evidence of intracranial hypertension, associated in 10 (42%) with increased head circumference. Developmental delay was present in 5 children (21%); 3 children, all with tumours located in the third ventricle, demonstrated Parinaud syndrome and one child was admitted for seizures. All the children underwent CT scan and in all the choroid plexus tumours were clearly demonstrated. Hydrocephalus was discovered in 18 cases (75%). Only 9 children of our series underwent cerebral angiography. Each time the feeding arteries (anterior or posterior choroidal arteries) were visible. One child with IVth ventricular tumour had an angiogram which showed a tumoural blush. 2 children (8%) demonstrated calcifications on plain skull X rays. Transcranial ultrasound examinations were performed in 4 infants and in all hydrocephalus was demonstrated. Furthermore in 3 cases the tumour was identified in the lateral ventricle but in one case the tumour

Table 1. *Age Distribution*



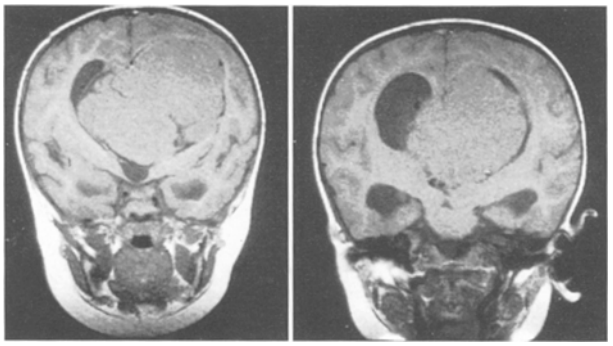


Fig. 1. MRI appearance of a huge papilloma in the third and lateral ventricles in a 8 months old infant

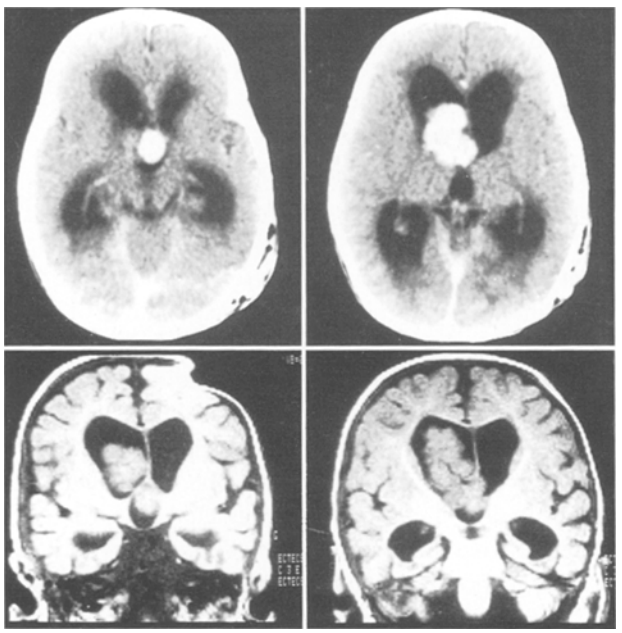


Fig. 2. CT Scan and MRI aspect of third ventricle papilloma involving the left ventricle

located in the IIId ventricle was not visible. The last four cases underwent MRI investigations, two were in the posterior fossa and two in the supratentorial compartment (Fig. 1).

In 11 children (46%) the tumour was located in the lateral ventricle, 6 in the left and 5 in the right. Seven tumours (29%) were found in the IIId ventricle (Fig. 2) and 6 (25%) arose in the IVth ventricle (Table 2).

In this series, 18 children (75%) required ventricular shunting, 16 with a ventriculo-peritoneal (VP) shunt before removal of the tumour and 1 with temporary external drainage. In one case the shunting procedure was necessary after tumour resection. 23 cases were operated on. A 10 year old boy, with a IIId ventricle choroid plexus papilloma (CPP), died from a large gastric haemorrhage prior to surgery. Total excision was performed in 20 cases (83%). Subtotal removal of the tumour was carried out in 3 children (13%) with choroid plexus carcinomas (CPC). The operative mortality within one month of surgery was 8%, namely 2 infants with carcinomas in the IVth ventricle died.

Table 2. *Localisation (24 cases)*

	Papilloma (16)	Carcinoma (8)
Lateral ventr. (L)	5	1
Lateral ventr. (R)	3	2
III Ventr.	7	
IV Ventr.	1	5

Pathological examination revealed benign papillomas in 16 children (67%), 8 (50%) were in the lateral ventricles, 7 (44%) in the IIId ventricle, and 1 in the fourth ventricle. In 8 children (33%) histology demonstrated CPC, 5 of these were found in the IVth ventricle and 3 in the lateral ventricle.

Radiation therapy was given in 3 children with CPC (two were located in the supratentorial space and one in the posterior fossa), in one of these cases treatment was combined with chemotherapy.

Results

3 children with supratentorial CPC are alive and one is neurologically intact 9 years after treatment. One case treated 14 years ago demonstrated psychomotor retardation with epilepsy. The last one is a recent case,

Table 3. *Actuarial Survival Rate Papilloma vs Carcinoma*

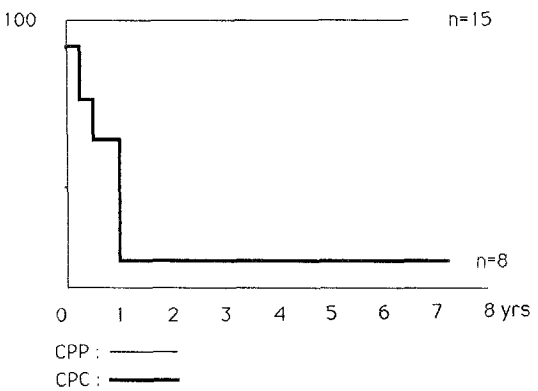
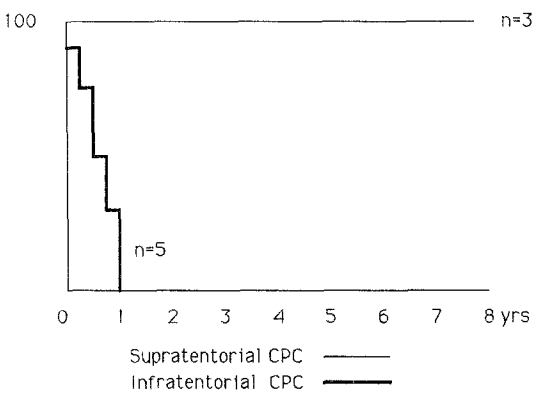


Table 4. *Actuarial Survival Rate of Carcinomas (8 Cases)*



a 6 year old girl, in good condition 2 month after surgery, actually treated by chemotherapy (8/1 protocol). Five children with carcinomas of the IVth ventricle aged respectively 5, 10, 11, 20, and 27 month died between 1 and 12 months after surgery. Thus the overall mortality was 25%.

The 15 patients with benign papillomas who underwent surgery alone are alive. Among these 10 (67%) are neurologically intact and 5 (33%) have motor sequelae associated in 4 cases with psychomotor retardation. One case has severe epilepsy as well (Tables 3 and 4).

Discussion

Although they occur in children more frequently than in adults, choroid plexus tumours are rare with their incidence varying form 1 to 4%[12, 19, 30]. In our experience, they represent 3,5% of the 685 cerebral tumours observed in children during the same period. They are much more frequent in infants. In paediatric series, infants represent 60 to 82% of the cases[7, 11, 30, 36, 37]. In our series, 60% of the cases were under 2 years old. In children, they are generally localized in the lateral ventricles. In the literature, this localization is found in 60 to 78% of the cases[7, 20, 21, 30, 35]. We have observed 43% only in this localization. Some cases of bilateral CPP have been published[7, 9, 20, 21, 31, 35]. They represent 7% of the cases according to Matson and Crofton[21]; in our experience we never observed bilateral tumours. IIIrd ventricle papillomas occur in 7 to 29% of the cases[7, 17, 20, 21, 30, 35, 37]. In our series 7 cases (29%) were at this site and 5 (71%) were in infants. IVth ventricular CPP represent 10 to 20% of the cases in paediatric series[7, 21, 28, 30, 41]; this location is usually observed in adults[25, 30, 41]. A similar incidence was found in our experience. Few cases of cerebello-pontine angle CPP have been published in children[7, 10, 29].

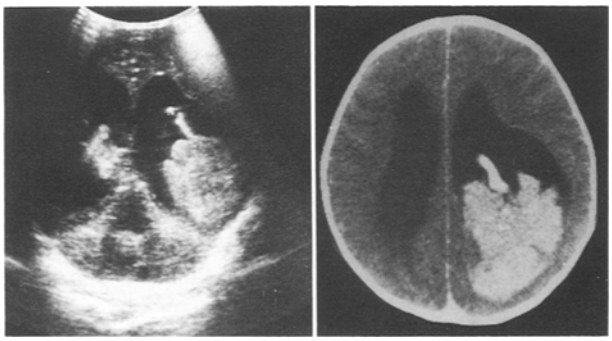


Fig. 3. Transfontanellar ultrasound and CT Scan of left lateral ventricle papilloma in a 4 months old baby. Note the similar aspect of the tumour and the feeding artery

At time of presentation, signs of increased ICP with or without macrocrania were present in 70% of the cases in the series of Velasco-Siles and Raimondi[40]. This was observed in 75% of our cases. Matson and Crofton[21] noted that the most common symptom was vomiting occurring in 62% of their patient. Developmental delay is never mentioned in the literature as the initial symptom in younger children, but 5 children in our series (21%) presented with this sign.

CT Scan and MR imaging have significantly simplified the diagnosis of these tumours. Transfontanellar ultrasonography may be considered in neonates and infants to detect these tumours[5, 14, 37]. In our series, this method has been used in 4 cases and in 3 the diagnosis was correctly established (Fig. 3). Cerebral angiography remains an important part of preoperative evaluation for these highly vascular tumours for several authors[15, 22, 28]. The angiographic appearance of these lesions has been described previously[30, 40] and Tomita and Naidich[38] showed the abnormal vascular supply from branches of the middle cerebral artery. We had performed angiography in only 9 cases, in 8 of these tumoural blush was demonstrated. In one, a 10 year old boy with IIId ventricular CPP, angiogram showed only signs of hydrocephalus without any tumoural blush. Actually, in our opinion, angiography will not change the surgical approach and operative technique, and especially in young baby, this method is not easy to perform and not without risks. Although some cases of CPP without hydrocephalus have been published[33, 34], they remain exceptional. 2 factors may explain this phenomenon: the first factor is the obstruction of CSF pathways either by the tumour in the ventricles, the haemorrhage into the CSF or by obstruction at the level of the arachnoid granulations of the basal cisterns from the high protein content[8, 19, 20, 24, 33]. The second one is the over-production of CSF[1, 8, 9, 20, 21, 23]. Evidence for this is the resolution of hydrocephalus following removal of the tumour[8, 9, 21, 25, 39]. Nevertheless, hydrocephalus persists in many cases[14, 17, 25] and a CSF shunt procedure is often necessary after surgical removal of the tumour[30, 40]. In our series, 18 (78%) out of 23 children operated on have a permanent CSF device. According to Koos *et al.*[19], we think that a preoperative shunting procedure allows resolution of the hydrocephalus and decreases the risk of post-operative subdural fluid collection in the majority of cases. Also the cortical mantle thickens and lessens the collapse of the cerebral hemispheres after removal of the

tumour. Suture of the pia mater has been proposed by Boyd and Steinbok[2] after the tumour removal to avoid this problem, but without real proof of its effectiveness. It is felt by the authors that the shunt is necessary particularly in infants to prevent the low pressure hydrocephalus responsible for some secondary worsening.

Total and en bloc tumour excision is currently the recommanded treatment[20, 21, 30]. It is the opinion of the authors that total excision is always possible in benign tumours. En bloc excision cannot be done in large tumours and this is especially true in neonates and infants[28, 37, 38]. In these cases morcellation is mandatory.

Preoperative radiation therapy has been proposed by some authors[4, 11] to decrease the risk of haemorrhage and the tumoral bulk, however cerebral irradiation is absolutely contra-indicated in neonates, infants and young children[37, 38, 41]. Post-operative irradiation is only indicated in malignant papillomas in childhood and adults[2, 3, 7, 24]. In this series, 2 children with lateral ventricle carcinomas were irradiated after subtotal resection and are alive with a follow up period of 8 and 13 years and one is neurologically intact.

In the literature operative mortality is between 0 and 19%[21, 22, 24, 26]. In this series the operative mortality within one month of surgery was 8%. The 2 infants who died had choroid plexus carcinomas located in the IVth ventricle. No operative mortality exists in cases of CPP and in supratentorial choroid plexus tumours. Major sequelae are rarely mentioned in the literature. Despite the fact that the majority of these tumours are benign the incidence of neurological sequelae varies between 29 to 63%[24, 26, 30]. Motor deficits, epilepsy, and psychomotor delays are the most frequent problems and these account for 36% in our series. Among 23 children operated on, 12 (52%) are neurologically intact.

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Correspondence and Reprints: Professeur Maurice Choux, Service de Neurochirurgie, Pédiatrique Hôpital des Enfants de la Timone, Bd. Jean Moulin, F-13005 Marseille, France.