

Choroid plexus tumours: a surgically treated series

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Summary

Choroid plexus tumours -carcinomas and papillomas- are rare, especially in adults, and they pose some problems in their diagnosis and management.

We have reviewed a series of nine cases from our institution surgically treated during the last 18 years. Their clinical charts, neuroradiological examinations, surgical technique, neuropathology and follow-up were analysed.

In only one case total removal proved to be impossible, but even in cases of total removal recurrence appeared in two cases (one carcinoma and one papilloma). Morbidity is especially associated with posterior fossa tumours.

These rare tumours are managed surgically. They are usually associated with hydrocephalus, and it is difficult to forecast whether or not permanent CSF drainage will be required. A long-term follow-up is needed in patients with this type of tumour.

KEY WORDS: Choroid plexus; Carcinoma; Papilloma; Surgical removal.

Tumores de los plexos coroideos. Análisis de una serie de casos tratados quirúrgicamente.

Resumen

Los tumores de los plexos coroideos -carcinomas y papilomas- son raros especialmente en el adulto y plantean diversos problemas en su diagnóstico y tratamiento.

Hemos revisado una serie de nueve casos tratados quirúrgicamente en nuestro servicio durante los últimos 18 años, analizando los datos de la historia clínica, los exámenes neuroradiológicos, la técnica quirúrgica y el estudio neuropatológico.

Neurocirugía 2001; 12: 7-16

Solo en un caso se estimó imposible la resección tumoral y en otros dos ocurrió recurrencia (un carcinoma y un papiloma) a pesar de haberse llevado a cabo una resección aparentemente total. Los tumores localizados en la fosa posterior se asociaron a una mayor morbilidad.

Los tumores de los plexos coroideos deben ser tratados quirúrgicamente. Se asocian generalmente a hidrocefalia y es difícil de predecir cuando los pacientes van a necesitar derivación permanente de líquido cefalorraquídeo. Los pacientes con este tipo de tumor deben ser seguidos por períodos largos de tiempo.

PALABRAS CLAVE: Plexo coroideo. Papiloma. Carcinoma

Introduction

Choroid plexus tumours -papillomas and carcinomas- are relatively rare and occur more frequently in children than in adults, where they represent 3% and 0,5% of all intracranial tumours, respectively^{1,6,23,27,33,40,43,44}. More than 50% of choroid plexus neoplasms appear in the first 10 years of life. Incident peaks in the first 2 years^{7,40,41,43}, 8-17% of them may show features of malignancy^{17,27,30,33,34,39}. While the great majority are intraventricular they may occur outside the ventricles^{10,20,21,22,25,30}.

In children papillomas are usually located in the lateral ventricles, while in adults they show a predilection for the fourth ventricle^{6,26,27,32,40,41,44}.

Carcinomas appear more frequently in the lateral ventricles, but were also described in the third and fourth ventricle^{6,17,32,39,42,44}.

Their presence is most frequently heralded by raised intracranial pressure symptoms and signs, due to hydrocephalus, if they are located in the lateral or third ventricles, or by cerebellar and brainstem signs if in the posterior fossa^{23,29,41,43}.

We have reviewed the records of 9 patients with choroid plexus tumours surgically treated at our institution during the last 18 years. Their clinical features, diagnosis, treat-

Case	Sex	Age	Table 1		Imaging
			Signs & symptoms	Location	
1	M	30	Headaches, vomiting, for 3 months Papilledema	Left lateral ventricle	Ventriculography CT scan
2	M	47	Headaches, dizziness, visual disturbances, for 7 months Papilledema, Rt II, IX, X nerves palsies, gait ataxia, Rt motor incoordination	IV ventricle	CT scan Angiography MRI
3	M	25	Headaches, dizziness, for 2 months Papilledema. Nistagmus	III ventricle	X-ray CT scan
4	F	47	Headaches for 5 months. Papilledema	IV ventricle	CT scan
5	F	11	Headaches for 2 months Papilledema	IV ventricle	CT scan MRI
6	F	17	Headaches, decreased vision, menses irregularities, for 6 months Papilledema	III ventricle	CT scan MRI
7	M	31	Headaches, vomiting, unsteadiness for 3 months Papilledema, decreased hearing on the right, nystagmus, gait ataxia	IV ventricle + Right cerebello pontine angle	CT scan MRI
8	M	19	Headaches for 4 years Papilledema with left optic atrophy, decreased hearing on the left, decreased sensation on the left side (including face), gait ataxia	Right lateral ventricle	X-Ray CT scan MRI
9	M	65	Headaches dizziness, visual disturbances, unsteadiness, for 6 months Papilledema, nystagmus, VI bilateral palsy, decreased hearing on the right, bilateral motor incoordination, gait ataxia	Right cerebello pontine angle	CT scan MRI

ment, problems and outcome are presented. Total removal of these tumors should be the goal, but adjuvant therapy may be necessary.

Material and Methods

In the present study a series of 9 patients (6 males and 3 females) operated on for choroid plexus tumors during a period of 18 years was studied.

Their clinical symptoms and signs were reviewed. Neuroradiological examinations included ventriculography (one patient), CT scan (8 patients), MRI (6 patients), and angiography (1 patient).

All patients received medical treatment and were opera-

ted on. Different surgical approaches were used according to the tumor location, and ventricular shunting (external or internal) was inserted if necessary. A cystic-peritoneal shunt was also inserted in patients who developed a porencephalic cavitation after tumor removal.

Histopathological evaluation included the review of the formaline-fixed, hematoxylin-eosin stained slices embedded in paraffin, and immunocytochemical characterization with the peroxidase-antiperoxidase technique of 4 µm paraffin slices with a panel of 8 different antibodies, which included antibodies to cytokeratin (CYT), vimentin (VIM), transthyretin (TTR), S 100 protein (S 100), epithelial membrane antigen (EMA), carcino-embryonic antigen (CEA), glial fibrillary acidic protein (GFAP) and α -I antitripsyn.

Radiotherapy after surgery was offered to carcinoma patients.

Follow-up included clinical and radiological evaluation. Clinical evaluation included the neurological examination and Glasgow Outcome Scale (GOS) determination, and neuroradiological evaluation done by means of CT scan or MRI in order to evaluate local recurrence, growth of the remaining tumor and the presence of metastasis.

Results

From the 9 patients harboring choroid plexus tumours 6 had papillomas and 3 had carcinomas. Papillomas appeared in 3 females and 3 males, while the carcinoma patients were all males.

The patient's ages ranged from 11 to 65 (mean 35) years in papillomas, and from 19 to 31 (mean 27) in carcinomas.

Papillomas were located in the fourth ventricles (three cases), in the third ventricle (two cases) and in the cerebellopontine angle (one case).

Carcinomas arised from the lateral ventricles (two cases) and from the IV ventricle, extending to the CP angle (one case).

The clinical presentation in the majority of the cases was associated with intracranial hypertension, or with brainstem and cerebellar signs and symptoms if the lesion was situated in the posterior fossa. Headaches were invariably present in all patients. Vomiting, dizziness, visual disturbances, decreased hearing and unsteadiness were present in some of them. On clinical examination, papilledema was present in all patients, and cranial nerve abnormalities, ataxia and pyramidal signs were present in some of them. One of the patients (third ventricle) had diencephalic symptoms.

The duration of symptoms was usually short (2-7 months) except in one case where chronic headaches were present for four years, but in this case the diagnosis was made after a skull X-ray done after a mild head injury (case 8).

Diagnosis was made by ventriculography in one patient, CT scan in nine (the first one only had CT scans after tumour recurrence), and MR in six (in one of them -case 2- only after recurrence). Skull X-Ray was positive in two patients, showing a huge midline calcification in one patient and signs of intracranial hypertension in another one. Angiography was done in just one case (Table 1).

On CT scan tumours appeared as round or lobulated, iso or hiperdense mass lesions, with frequent calcifications, more rarely with cysts and edema, and without haemorrhage. After contrast injection there was a dramatic tumour enhancement in almost all the cases.

On MRI the lesions were usually heterogeneous. In T1 weighted images they were predominantly isointense, and in T2 they were even more heterogeneous with predominant iso and/or hyperintensity. Cysts were seen in four cases,

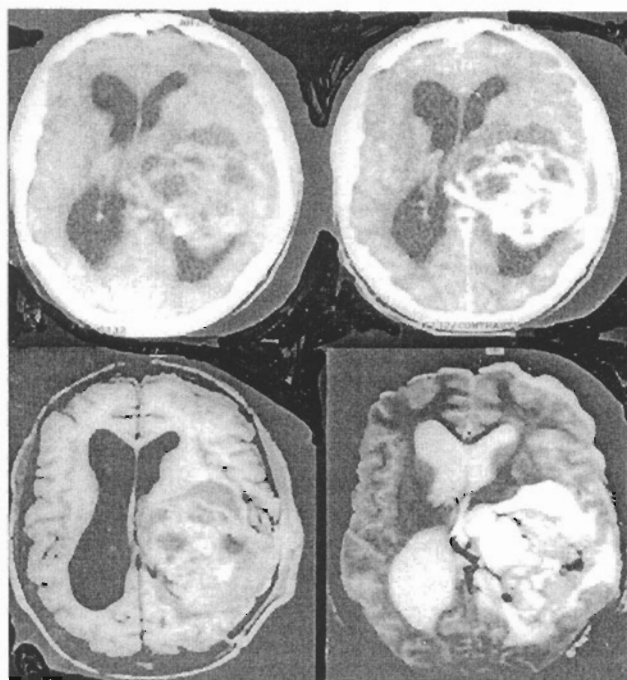


Fig. 1 . Upper part. CT scan of a plexus carcinoma of the lateral ventricle, before and after contrast administration, showing an heterogeneous solid lesion at the level of the right atrium, with an anterior cystic component, and with marked enhancement after contrast injection. Hydrocephalus is present.

Lower part. MRI of the same case, showing in both T1 and T2 the heterogeneity of this tumour, with predominant isointensity, as well as a marked heterogeneous enhancement after Gadolinium administration and the presence of edema.

and edema in two. Contrast enhancement after Gadolinium administration was heavy and usually heterogeneous (figures 1,2,3).

Hydrocephalus of some degree was present in all but one of the cases.

CT and MRI were not useful in differentiating papillomas from carcinomas, except in one case (case 8) where parenchymal invasion could suggest a carcinoma.

Surgical total removal was possible in all but one case (case 8), due to parenchymal invasion. In another case (case 2) the tumour was completely removed in the first operation, but proved to be impossible to remove completely at the second operation after recurrence, due to tumour invasiveness of the adjacent parenchyma. Tumours in the lateral ventricles were approached through temporal and parietal transcortical routes. The third ventricle tumours have been removed through a right frontal transcortical incision. For the posterior fossa tumours, a midline suboccipital craniectomy was used in three patients, while a lateral one was chosen in the patients with the tumour in, or extending to,

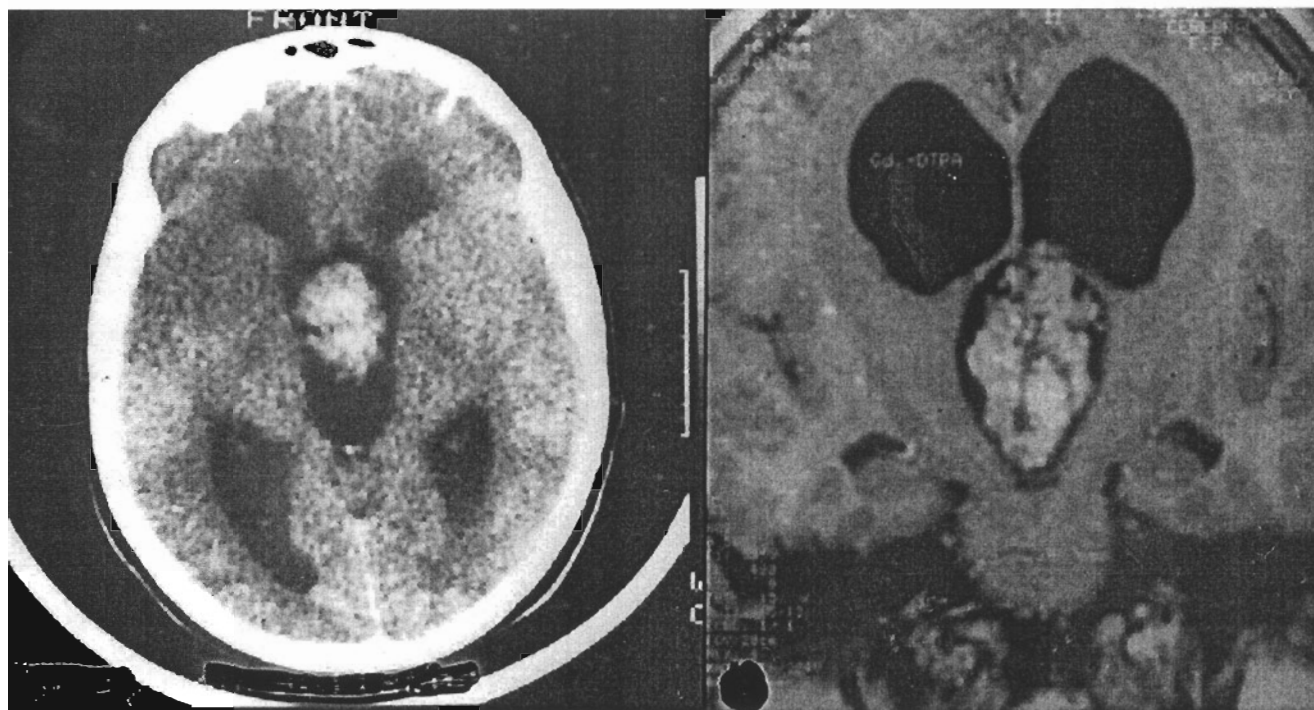


Fig. 2 Left. CT scan of a third ventricle papilloma, showing an hyperdense homogeneous tumour, with associated hydrocephalus. Right. T1 MRI after Gadolinium administration, showing the intense contrast uptake of this cauliflower-like tumour.

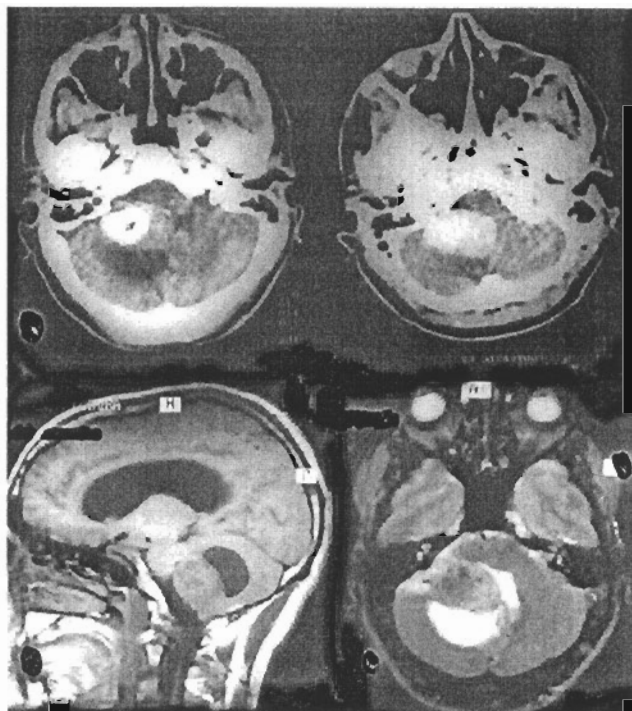


Fig. 3. Upper part. CT scan of a carcinoma of the fourth ventricle showing an isodense lesion with a huge calcification and enhancement after contrast administration. Lower part. T1 and T2 weighed MRI showing an isointense tumour extending into the right cerebellopontine angle

the cerebellopontine angle.

Five patients had an external ventricular drainage, inserted during tumour surgery, for only a few days in three cases, being converted to a ventriculoperitoneal shunt latter on in two cases. Another patient received a ventriculoperitoneal shunt previous to tumour surgical removal.

Porencephaly was seen after surgery in two patients, both of them harboring carcinomas in the lateral ventricles. A cystic-peritoneal shunt was inserted in both of them, sometime after tumour removal (Table 2).

The nature of the tumour was confirmed in all cases by histopathological examination. Six were classified as papillomas, with two of them showing some atypical features, and the remaining three were classified as carcinomas. While well differentiated papillomas mimicked the normal choroid plexus architecture and cytology, with a single layer of columnar or cuboidal epithelial cells on a connective stroma with occasional microcalcifications, the other two papillomas (cases 2 and 9) showed focal breakdown of papillary architecture with cytological atypia, rare mitosis and necrosis. Carcinomas showed loss of papillary differentiation, with patternless cellular sheets or tubular pattern, cellular pleomorphism, mitosis and necrosis. In one case invasion of the adjacent neuronal tissue could be demonstrated (Figures 4,5,6).

The panel of antibodies used for immunocytochemical characterization – cytokeratin (CYT), vimentin (VIM),

Table 2

Case	Location	Surgery	Pathology	Results
1	Left lateral ventricle	Transcortical temporal Total removal Transcortical occipital Total removal Transcortical occipital Total removal Cystic-peritoneal shunt	Metastasis Carcinoma	Right hemianopsia Two recurrences Porencephaly Radiation therapy Died six years after the first operation
2	IV ventricle	Suboccipital midline Total removal Suboccipital midline Subtotal removal	Papilloma	Meningitis Right hemiparesis Nystagmus Recurrence Right 12th palsy, dysarthria Focal epilepsy Died eight years after the first operation
3	III ventricle	Ventriculoperitoneal shunt Transcortical frontal Total removal	Papilloma (atypical)	Epilepsy GOS = 5 fifteen years after the first operation
4	IV ventricle	Suboccipital midline Total removal External ventricular shunt Ventriculoperitoneal shunt Shunt revision	Papilloma	Bilateral 6th, right 7th, 9th and 10th paresis Nystagmus, gait ataxia GOS = 4 ten years after the first operation
5	IV ventricle	Suboccipital midline Total removal External ventricular shunt	Papilloma	GOS = 5 eight years after the operation
6	III ventricle	Transcortical frontal Total removal	Papilloma	GOS = 5 eight years after the operation
7	IV ventricle	Suboccipital lateral Total removal External ventricular shunt	Carcinoma	Cerebellar hematoma 7th, 9th, 10th and 11th paresis Right superior limb incoordination Swallowing difficulties Erosive gastritis with GI haemorrhage
8	Right lateral ventricle	Transcortical parietal Partial removal External ventricular shunt Transcortical parietal Partial removal Ventriculoperitoneal shunt Transcortical parietal Partial removal Cystic-peritoneal shunt	Carcinoma	GOS = 4 four years after the operation Left hemianopsia Left superior limb mild (4+) paresis Porencephaly Radiation therapy GOS = 5 four years after the first surgery
9	Right cerebello pontine angle	Suboccipital lateral Total removal External ventricular shunt Suboccipital lateral Total removal	Papilloma (atypical)	Recurrence GOS = 5 three years after the first operation

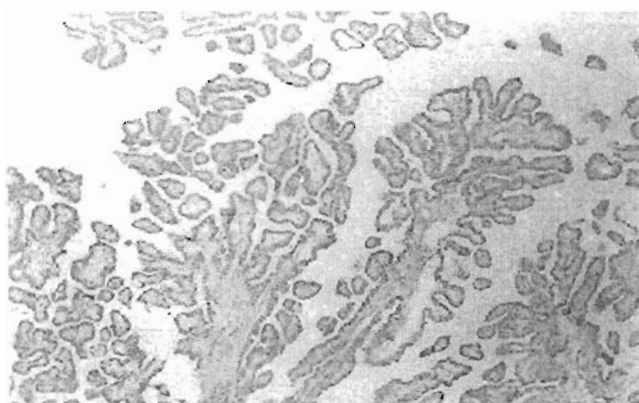


Fig. 4. Photomicrograph of a Hematoxylin/Eosin stained slide at 100 x magnification from a plexus papilloma, showing a papillary growth pattern, regular epithelial lining and fibrous stroma.

transthyretin (TTR), S 100 protein (S 100), epithelial membrane antigen (EMA), carcino-embryonic antigen (CEA), glial fibrillary acidic protein (GFAP) and α -1 antitrypsin - depicted a quite variable, non specific expression (Table 3).

No metastasis were seen in our patients. Two patients with carcinoma received radiotherapy.

Recurrences happened after total removal in both papillomas and carcinomas.

The period of follow-up ranges from 18 months to 15 years. Two of the patients died during this period, due to the tumour itself, one 6 years, and the other 8 years after the first operation. Both of them had recurrences of their tumours, and were reoperated (one had two recurrences). The first one had a papilloma (atypical), and the other one a carcinoma.

Four patients remained with permanent new neurological problems (epilepsy, cranial nerves abnormalities, and pyramidal tract signs) after surgery. From these patients, two have a GOS of 5, and the other two have a GOS of 4.

The other three patients are symptom-free, and have a normal life (Table 2).

Three detailed cases are described, because they presented special difficulties of treatment.

Case 1

A 30 year-old man presented with a 2 month history of headache and vomiting. On clinical examination he was found to have bilateral papilloedema, without any other neurological signs. A ventriculogram demonstrated symmetrical dilatation of the lateral ventricles and the left temporal and occipital horns did not fill completely. A transcortical left temporal craniotomy was performed, and a plexus choroid tumour was totally removed. Postoperatively he was left with a right hemianopsia. The histopathological examination was suggestive of a metastasis from a carcinoma.

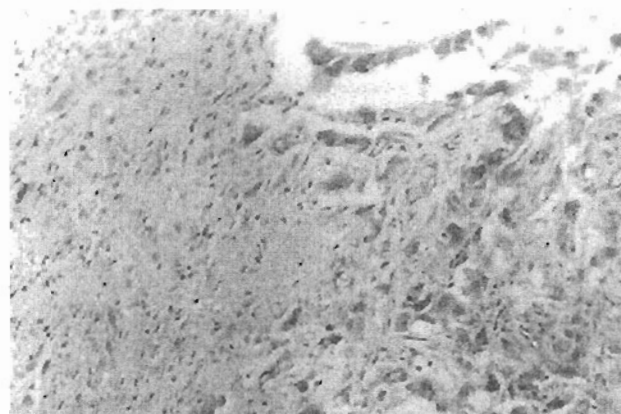


Fig. 5. Atypical papilloma with papillary structure, pseudostratification of endothelium and nuclear pleomorphism (Hematoxylin / Eosin x 200)

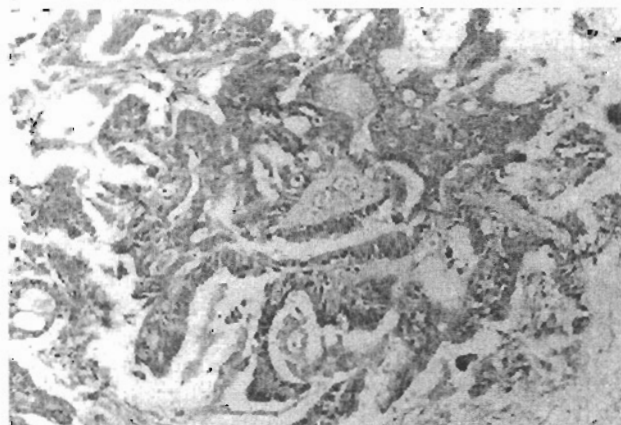


Fig. 6. Photomicrograph of a plexus carcinoma, showing cells with severe cytological atypia invading the parenchyma, where gliosis occurs (Hematoxylin / Eosin x 250).

He received radiotherapy. Three years later a CT scan revealed tumour recurrence at the occipital horn, and a new surgical procedure was carried out, again with total removal of the tumour, now through a small occipital lobectomy. At this time the neuropathological examination revealed a choroid plexus carcinoma. A new recurrence was shown by another CT scan, with an interval of ten months, and this was once more completely extirpated. A last CT scan, two years later, showed a CSF filled cystic cavitation adjacent to the left temporal horn, and a shunt from the cystic cavity to the peritoneum was inserted. No additional complications resulted from these last operations, but the patient died six years after the first operation, at home, and no autopsy was done.

Case 2

This 47 years-old male patient presented with a 7 months history of headaches, dizziness, and visual disturbances in

Table 3

CASE	DIAGNOSIS	CYT	VIM	TTR	S100	EMA	CEA	GFAP	α 1AT
1	carcinoma	-	+	+	++	++	-	-	-
2	papilloma	++	+/-	++	++	?	-	++	?
3	papilloma	++	+	-	++	?	-	++	?
4	papilloma	++	+	-	++	++	-	++	?
5	papilloma	++	-	-	++	+	-	++	-
6	papilloma	++	-	-	++	-	-	-	-
7	carcinoma	++	+	-	++	+	-	+	-
8	carcinoma	++	-	+/-	++	?	-	+/-	?
9	papilloma	++	++	?	++	-		+	+

CYT - cytokeratin. VIM - vimentin. TTR - transthyretin. S100 - protein S 100. EMA - epithelial membrane antigen. CEA - carcino-embryonic antigen. GFAP - glial fibrillary acidic protein. α 1AT - α 1 antitrypsin.

his left eye, which he described as seeing "brigh circles" crossing his eye. He was found to have bilateral papilledema, right seventh, ninth and tenth cranial nerve palsies, gait ataxia and right motor incoordination. CT scan depicted a fourth ventricular mass, with associated hydrocephalus. Total macroscopic removal of a choroid plexus papilloma was achieved through a median suboccipital craniectomy. The patient was left with a mild right hemiparesis, swallowing difficulties, dysarthria, and developed meningitis in the postoperative period, which was successfully treated with antibiotics, and he could live an independent life. A recurrence was diagnosed by CT scan eight years latter, and he was re-operated with a sub-total removal of the tumour at this time, due to tumour invasiveness. In the postoperative period he was left with a right 12th nerve paresis, and developed focal epilepsy, gastro-esophagic haemorrhage and ARDS, ending on a tracheostomy. He died seven months latter.

Case 8

A 19 years old boy has suffered a vehicle accident, and his skull X-ray demonstrated signs of high intracranial pressure. After inquiring, chronic headaches were known to exist, and an urgent CT scan was requested. It showed a huge right ventricular tumour occupying the temporal and occipital horns, as well as the atrium, associated with hydrocephalus. He was admitted for further investigation (MRI). His clinical examination revealed left optic atrophy, left fifth and eighth cranial nerves disturbances, left side decreased touch and pain sensation, and gait ataxia. He was operated on 12 days latter. A transcortical right parietal craniotomy was chosen, and through a sulcus the lateral ventricle was approached. Only partial removal was achieved. An external ventricular drainage was left. Histopathological examination showed a plexus carcinoma. In a second operation 3 weeks latter, the tumour proved to be impossible to remove completely, due to parenchymal invasion.

A ventriculoperitoneal shunt was inserted in the left lateral ventricle. The patient was left with a quite mild (4+) left superior limb paresis and a left homonymous hemianopsy. An MRI done three months latter revealed the residual tumour, with an anterior cyst and an associated CSF filled cavitation. It was tried again to remove the tumour, but just a partial excision was once more possible. The anterior cyst was aspirated and a cystic-peritoneal shunt was inserted, from the CSF cavitation. No added deficits resulted from this last operation, and he was sent for radiotherapy. Four years after the diagnosis he is doing very well.

Discussion

Choroid plexus tumours are neuroepithelial tumours^{7,25,27,33,37,40,41} and the first tumour of this kind was described, according to Davis and Cushing (9), by Guérard in 1833.

They are rare tumours, especially in adults, and are usually located inside the ventricles. The few cases described outside the ventricles are thought to arise from an embryonic rest of the choroid plexus, from a small tuft of choroid plexus extending from the foramen of Luschka into the cerebellopontine angle, or from dissemination through CSF^{5,10,20,21,22,30,41}.

Malignant papillomas or carcinomas are even rarer and some of the reported cases may not be in fact true primary choroid plexus carcinomas^{2,3,6,8,11,15,17,19,26,27,31,33,34,39,42,43,45}.

Very unusually choroid plexus tumours may be associated with other tumours³⁸, or being part of Li-Fraumeni syndrome⁴⁵ or von Hippel-Lindau disease⁴.

A few cases of multiple tumours have been described, accounting for less than 4% of all choroid plexus tumours^{8,41}. Familial occurrence is another rare possibility, raising the hypothesis of genetic predisposition in this kind of tumours⁴⁶.

One of the most constant features of this kind of tumours is the concomitant existence of hydrocephalus. The reasons for its development include tumour obstruction of the for-

men of Monro, third and fourth ventricles lumens, CSF overproduction, and blockade of its absorption due to high concentration of proteins and blood breakdown products from small intratumoural haemorrhages^{5,6,7,12,14,23,26,41,44}.

Other possible complications of choroid plexus tumours include subarachnoid haemorrhage^{1,13,35} and "seeding"^{5,16,29,32,34}.

On CT scans the usual appearance is of a well-circumscribed, round or lobulated, iso or hyperdense mass lesion, with marked enhancement after IV contrast. Intratumoural calcification is frequent, and small cystic areas may also appear^{17,20,22,25,26,44}.

MRI is the best diagnostic tool, because it shows the tumour in the three planes and may depict parenchymal invasion. On T1 weighted images these tumours appear as isointense heterogeneous well defined masses. Regions of low signal corresponds to the presence of calcification and/or vascular areas, and hyperintense zones are compatible with haemorrhagic component. There is usually marked heterogeneous enhancement with Gadolinium injection. On proton density and T2 weighted images tumours may be iso or hyperintense, and usually heterogeneous^{20,22,25,27,42,44}.

There are no special features to distinguish papillomas from carcinomas with CT or MRI scanning.

Lesions to be taken in account for the differential diagnosis include metastasis, ependymomas, medulloblastomas and other gliomas^{6,18,26,31,36,41}.

Microscopic examination in papillomas shows the normal choroid plexus cellular pattern, with a single layer of columnar or cuboidal epithelial cells devoid of cilia and blepharoblasts, lining a collagen stroma where microcalcifications may appear.

In this group some atypical features may be present - loss of the papillary structure, pseudostratification of the epithelium, cysts, necrosis and some mitotic activity and nuclear pleomorphism - and these tumours may be classified as atypical papillomas. Carcinomas usually have frank invasion of the adjacent neural parenchyma and unequivocal cytological and histological malignancy. High cellularity, cells forming large solid sheets with occasional differentiation, and anaplastic features including increased mitotic activity, cellular pleomorphism, nuclear atypia and necrosis are characteristic. They may spread along the neuro-axis^{3,11,26,27,39,40,42}.

Immunohistochemical studies may be useful for the differential diagnosis of intraventricular tumours and also in distinguishing between both choroid plexus papillomas and carcinomas. Various antibodies have been used, but are contradictory in many studies. These tumours usually coexpress both epithelial and glial types of reactivity. TTR (transthyretin) has been claimed to be a specific marker because it would be produced in the central nervous system at the site of the choroid plexus. Nevertheless, it is now well-known that reactivity may occur in myxopapillary

ependymomas and in metastatic carcinomas. Although it may be useful for the differential diagnosis of choroid plexus tumours, it is not very helpful in distinguishing papillomas from carcinomas, contrary to what has been previously stated by some authors^{15,18,22,27,28,31,33,40,41,44}.

Ultrastructural studies have been reported but there is no complete agreement upon typical choroid tumours characteristics^{7,22,27}.

Total removal of the lesion is the treatment of choice for choroid plexus tumours, being the surgical approach dependent on various factors such as tumour location, presence of hydrocephalus, neurological deficit, hemispheric dominance, and the surgeon's preference. Lateral ventricle tumours may be approached through a transcortical incision, or through the transcallosal route. Third ventricle tumours may also be approached by those two techniques combined or not, the last one, with a transylvian approach. Lesions in the fourth ventricle are approached via an inferior median suboccipital route, while tumours in the cerebellopontine angle are accessed via a lateral suboccipital craniectomy. Precocious interruption of arterial supply is of great help in facilitating tumour removal, but is quite difficult in lateral ventricle tumours^{17,23,26,36,40,41,42,44}.

There is still some controversy about the need and timing for shunting these patients. Some authors suggest that a shunt should be placed as soon as possible in third and fourth ventricular tumours, and removed a few days after surgery, definitive surgery being postponed to one or two weeks after shunting³⁶. Others suggest that a shunt must be inserted only after tumour removal, if hydrocephalus persists, using or not an external drainage temporarily, inserted at admission or during tumour surgery^{24,35,41,42}.

Radiation therapy is usually reserved for malignant tumours and for recurrent benign ones^{15,21,27,29,34,35,41,42,43}. Nevertheless, due to the possibility of seeding, entire neuro-axis radiotherapy has been proposed for all cases³.

Chemotherapy has been used with some good results in pediatric patients^{2,15,34,39,42,45}.

While some authors state that prognosis is usually good for papillomas and poor for carcinomas, despite surgical removal and adjuvant therapy^{5,6,27,40}, there is evidence that histologically malignant tumours do not always behave as malignant tumours from a clinical point of view^{2,15,39,41,42,44}.

The findings in our cases differ from those reported in the literature in three main points, which are the patient's age, the percentage of third ventricle and cerebellopontine angle location for papillomas, and the percentage of malignant tumours.

The first aspect is due to the fact that we do not see children under 11 years of age, at our institution. Papillomas in rare locations, such as the third ventricle and the cerebellopontine angle represent 50% in our series, which is much higher than in the literature^{16,29,44}. Carcinomas repre-

sent 33% of our plexus tumours, which is also a higher percentage than usual^{17,27,29}.

In conclusion, we believe that these rare tumours once diagnosed must be surgically treated, and total removal should be the goal because complete surgical resection is the major prognostic factor for long-term survival. If residual tumor is found a second surgery is indicated. Necessity for shunting must be evaluated individually.

Radiation therapy must be reserved for incomplete papilloma removals or recurrences, and for carcinomas whatever the surgical removal has been. Chemotherapy is another adjuvant therapeutic modality which may be used alone or in combination with radiotherapy.

Due to the possibility of recurrence, seeding, late hydrocephalus or CSF cystic cavitation development, patients with choroid plexus tumours must have a long-term follow-up.

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