Pediatr Radiol (2001) 31: 624–630

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Magnetic resonance imaging in the diagnosis and management of choroid plexus carcinoma in children

Received: 6 October 2000
Revised: 6 February 2001
Accepted: 5 March 2001

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Abstract *Background.* Choroid plexus carcinoma (CPC) is a rare tumour occurring in childhood. The MRI features are not well described.
Objective. To characterise the MRI features of CPC in children and to describe the role of MRI in post-surgical management.
Materials and methods. The pre- and post-operative MRI examinations of five children with histologically proven CPC treated at this hospital were reviewed.
Results. The tumour was supratentorial in two patients and infratentorial in three patients. Pre-operative MRI showed heterogeneous enhancing tumours with irregular margins, parenchymal invasion and surrounding oedema. Cystic change within the tumour was present in all patients. Curvilinear signal voids were seen in two patients and punctate signal voids in one. All patients had hydrocephalus. In one patient a fourth-ventricular metastasis was present at diagnosis. In all cases, surgical resection was incomplete. Follow-up found long-term survival in one patient who had been treated with chemotherapy and further surgery.
Conclusion. The MRI characteristics of CPC are nonspecific, but intraventricular location with tumour heterogeneity, parenchymal invasion and oedema may be useful features in diagnosis. MRI is valuable in the initial diagnosis, surgical planning and follow-up of children with these rare tumours.

Introduction

Choroid plexus carcinoma (CPC) is a rare tumour occurring predominantly in children. Choroid plexus tumours account for approximately 2–3 % of paediatric brain tumours. The majority of these are papillomas with carcinomas accounting for only 20–30 % [1–4]. The peak incidence is in the under 2 years age group [5, 6]. Because of the rarity of the tumour, most reports are limited to single cases or small series concentrating on the clinical course of the disease. We review five cases of choroid plexus carcinoma with particular reference to the pre-operative appearances of the tumours on MRI and describe the use of post-operative MRI in the further management of these patients.

Materials and methods

Between 1995 and 1998, five children with histologically proven CPC were treated at this institution. In each case the clinical diagnosis of brain tumour had been confirmed initially with cranial CT. MRI was performed 1–3 days pre-operatively using a 0.5-T machine (Gyroscan TS-NT, Phillips Medical Systems, The Netherlands). The imaging protocol included axial proton density and T2-weighted (T2W) sequences (turbo spin-echo; TR 3,101 ms, TE_1 23 ms, TE_2 120 ms) and coronal T1-weighted (T1W) sequences (spin-echo; TR 500 ms, TE 25 ms) prior to IV contrast medium administration. T1W images were then acquired in the axial, sagittal and coronal planes (spin-echo; TR 500 ms, TE 25 ms) following IV injection of dimeglumine gadopentate 0.1 mmol/kg (Magnevist, Schering Healthcare). Sagittal T1W contrast-enhanced images of the whole spine (turbo spin-echo; TR 754 ms, TE 12 ms) were also acquired to identify metastases. In one case the pre-operative MRI was performed at another institution and only contrast-en-

Table 1 Summary of patient presentation, treatment and outcome (*V-P shunt* ventriculo-peritoneal shunt)

	Age at presentation (months)	Sex	Signs, symptoms and duration	Treatment[a]	Outcome (time from diagnosis)
Patient 1	11	Male	6 weeks of vomiting, irritability and drowsiness; craniomegaly and bulging fontanelle	Chemotherapy; residual mass excised at 12 months	Alive; no evidence of tumour at 32 months; ild hemiparesis
Patient 2	4	Male	2 weeks squint, vomiting, irritability and lethargy	Chemotherapy; second surgical resection at 6 weeks	Died 2.5 months
Patient 3	18	Male	8 weeks of vomiting, squint and unsteadiness	Chemotherapy; palliative V-P shunt	Died 3 months
Patient 4	20	Male	4 weeks of vomiting, irritability, lethargy and unsteadiness	Chemotherapy; radiotherapy	Died 8 months
Patient 5	14	Male	2 weeks of vomiting, irritability; bulging fontanelle	Chemotherapy; radiotherapy	Died 16 months

[a]initial surgical debulking in all patients

hanced T1W images were available for review (patient 1). In all cases, post-operative MRI was performed at 0.5 T with the same protocol as pre-operatively. In four cases this was within 1 week of surgery.

All the MR images were reviewed by a consultant neuroradiologist and a consultant paediatric radiologist with experience in neuroimaging. A proforma was used to record the following: tumour site, size, margins and signal characteristics on each imaging sequence, presence of signal void and cystic areas, parenchymal extension, adjacent oedema, mass effect, extent of contrast enhancement, presence of hydrocephalus and presence and site of metastases.

Results

The patients were all male with a median age of 13.4 months at the time of presentation (range 4–20 months). The details of their clinical presentations and clinical courses are given in Table 1. Pre-operative MRI tumour characteristics are summarised in Table 2, and the pre-operative illustrations are cited in this table. In patient 2, correlation with the earlier CT showed areas of high attenuation thought to represent microcalcification corresponding to punctate signal voids on MRI (Fig. 1). In patient 5, enlarged enhancing vessels were seen on CT corresponding to curvilinear signal voids on MRI (Fig. 2).

All five patients initially underwent surgery. Surgical resection was incomplete in every case due to intra-operative bleeding or local invasion of vital structures. All five children received post-operative chemotherapy. All were followed up clinically and with MRI. Two patients underwent further attempts at resection (patients 1 and 2), and two received cranial radiotherapy (patients 4 and 5) on the basis of clinical and imaging findings. The post-operative MRI findings and subsequent management of each patient are summarised below.

Patient 1

Initial follow-up MRI was performed 7 weeks post-operatively. This showed a residual, 6 × 5 × 6-cm enhancing mass in the right hemisphere, adjacent to the posterior horn of the lateral ventricle. The 1-cm fourth ventricle metastasis was unchanged (Fig. 4). Chemotherapy was commenced at this time. Further MRI was performed at 2, 4, 6 and 8 months after commencing chemotherapy. At 2 months there was reduction in the degree of contrast enhancement and on the subsequent scans a slight reduction in the size of residual tumour was seen. At this stage further surgical resection of the hemispheric tumour was performed and was successful. The fourth ventricle metastasis was removed at a subsequent operation 3 weeks later. The patient has now been followed up for 3 years after first presentation and is well, with only a mild left hemiparesis. MRI to this time shows a marked reduction in hydrocephalus and no evidence of recurrent tumour.

Patient 2

MRI 1 day post-operatively revealed an irregular cavity measuring 4 × 1 × 1 cm in the posterior fossa with residual nodular enhancing tumour extending into the midbrain and left cerebral peduncle. Intraventricular haemorrhage and small subdural collections were present. Chemotherapy was commenced 4 weeks post-operatively. Two weeks later, repeat MRI showed enlargement of the mass, extending further into the brain stem and left thalamus (Fig. 5 a). This was not fully appreciated at the time and a further resection was attempted. This was unsuccessful due to involvement of vital structures. Further MRI, 1 week later, showed intraventricular haemorrhage and confirmed residual tha-

Table 2 Pre-operative MRI findings

	Tumour site/size (cm)	MRI characteristics of tumour	Hydrocephalus	Contrast enhancement	Metastases
Patient 1	Right lateral ventricle 10.0 × 8.0 × 7.5	Heterogeneous mass, irregular margins invading adjacent parenchyma; contains punctate and curvilinear signal voids and small cystic areas; mass effect, but minimal adjacent oedema	Yes	–	1 cm nodule in 4th ventricle
Patient 2 (Fig. 1)	4th ventricle, extending to midbrain and left cerebellar peduncle 4.5 × 4.2 × 6.0	Heterogeneous mass, isointense to grey matter on all sequences; irregular margin with some parenchymal extension; cystic areas but no signal voids; mild/moderate adjacent oedema and mass effect	Severe	Marked, uniform	None
Patient 3	4th ventricle inferior recess 3.8 × 3.4 × 3.2	Heterogeneous, predominantly isointense to grey matter on all sequences; irregular margins with parenchymal extension; cystic areas but no signal voids; mass effect, but only mild adjacent oedema	Yes	Minimal, heterogeneous	None
Patient 4 (Fig. 3)	4th ventricle, extending into right cerebellar-pontine angle 4.2 × 5.0 × 3.8	Heterogeneous, predominantly isointense to grey matter on all sequences; irregular margins with parenchymal extension; contains cystic areas and areas of high signal on T1W and PD thought to correspond to haemorrhage, no signal voids	Yes	Moderate, uniform	None
Patient 5 (Fig. 2)	Left lateral ventricle 8.0 × 6.5 × 7.0	Heterogeneous, predominantly isointense to grey matter on all sequences; lobulated margins, irregular in areas with parenchymal extension; mass effect and adjacent oedema; large cystic areas and areas of high signal on T1W and PD thought to correspond to haemorrhage; curvilinear signal voids	Yes	Moderate, uniform	None

lamic tumour (Fig. 5b). No further treatment was given and the patient died 4 weeks later.

Patient 3

Three days post-operatively, MRI showed a residual, irregular, minimally enhancing mass in the fourth ventricle measuring 3.3 × 2.2 × 2.2 cm, which was extending into the right cerebellar hemisphere and brain stem. Nodular enhancement was also seen around the superior sagittal sinus and there was a subdural collection. Chemotherapy was commenced 4 weeks post-operatively. Four weeks later, MRI showed the residual mass was unchanged and the nodular enhancement had resolved. Over the next 3 weeks, however, the patient's clinical condition deteriorated. A further MRI showed increasing hydrocephalus and tumour extension. The patient died 1 week later.

Patient 4

One day post-operatively, MRI revealed residual tumour in the right cerebellar hemisphere measuring 3.0 × 1.5 × 3.0 cm. There was surrounding oedema, a small local haematoma and a locule of air within the mass. There was a small right-sided subdural haematoma and intraventricular haemorrhage. Chemotherapy was commenced 4 weeks postoperatively. Eight weeks later, repeat MRI showed the tumour had reduced in size, measuring 1.6 × 1.1 × 1.4 cm, but was involving the brainstem. The cerebellar oedema had resolved. Cranial radiotherapy was administered. However, 10 weeks later, MRI showed tumour enlargement with increased surrounding oedema and mass effect. The tumour was displacing and directly invading the brainstem. The patient died 9 weeks later.

Patient 5

MRI was performed 6 days following subtotal excision. The tumour had reduced in size, particularly the cystic component, but there was considerable residual solid tumour measuring 6 × 8 × 5 cm. Chemotherapy was commenced 1 week post-operatively. MRI 6 weeks later showed a reduction in the size of the residual tumour but a new, 1-cm enhancing nodule was present in the basal cisterns and was thought to represent a metasta-

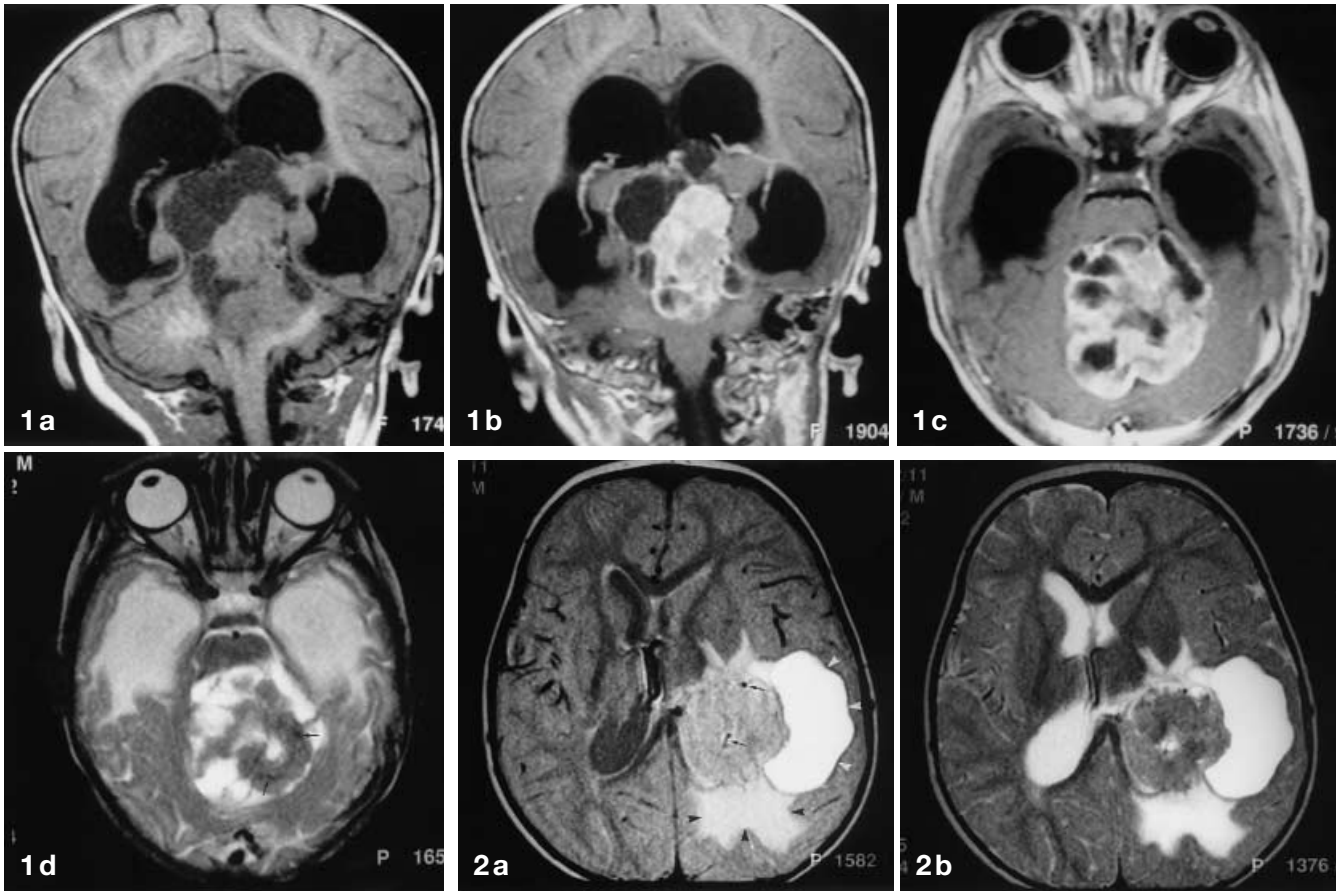

Fig. 1a–d Patient 2. Pre-operative appearances of a large fourth ventricular CPC. Coronal T1W images **a** before and **b** following contrast enhancement, showing a large complex enhancing mass involving the fourth ventricle, extending through the tentorial hiatus and causing marked hydrocephalus. **c** Axial T1W contrast-enhanced and **d** T2W images. The solid component of the tumour shows marked contrast enhancement. Punctate signal voids are seen on the T2W image (*arrows*), which corresponded to microcalcification on CT

Fig. 2a, b Patient 5. Left lateral ventricular CPC, pre-operative appearances. **a** Proton density and **b** T2W axial images, showing a complex mass with solid components isointense to grey matter and containing small curvilinear signal voids representing vessels (*arrows*). An associated tumour cyst (*white arrowheads*) is distinct from peritumoural oedema (*black arrowheads*) on the proton density image

sis. There was poor clinical response to chemotherapy and cranial radiotherapy was administered with good palliation. Nine months later, with the onset of clinical deterioration, further MRI showed marked enlargement of the cystic tumour in the left cerebral hemisphere with multiple enhancing metastases throughout the cerebrospinal fluid spaces. The patient died 2 months later.

Discussion

Choroid plexus carcinomas are rare tumours of neuroectodermal origin occurring predominantly in children. The tumour most commonly arises supratentorially, within the lateral ventricle, but around 15–20 % occur in the fourth ventricle [4, 7]. There is a slight male predominance with a male-to-female ratio of 1.3:1 [2, 4, 5]. Presenting features are nonspecific and generally reflect raised intracranial pressure, with focal neurology being variable [5, 7–10]. Surgery is the treatment of choice. The aim of surgery is to obtain gross total resection, the achievement of which is the sole most important prognostic factor [9, 11]. However, surgery is often limited at the first attempt because of peroperative blood loss, which can prove fatal, and because of the invasive nature of the tumour. If only a limited procedure is possible, but a biopsy is sufficient to establish the tissue diagnosis, then chemotherapy may reduce tumour vascularity and increase the likelihood of total resection at a second operation [7, 9]. In addition, a number of centres have adopted the use of post-operative chemotherapy in an attempt to control disease progression, but results have generally been disappointing. Radiotherapy is generally delayed or reserved for palliation

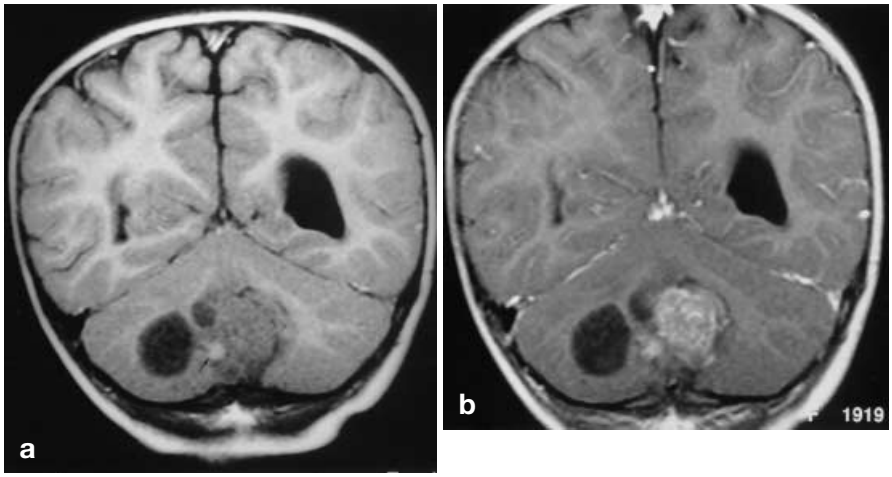

Fig. 3 a, b Patient 4. Pre-operative appearances of a fourth ventricular tumour. Coronal T1W images **a** before and **b** following contrast enhancement, showing tumour containing clear cystic areas and solid components, predominantly isointense to grey matter. A small area of high signal (*arrow*) is likely to represent haemorrhage. There is moderate uniform contrast enhancement of the tumour

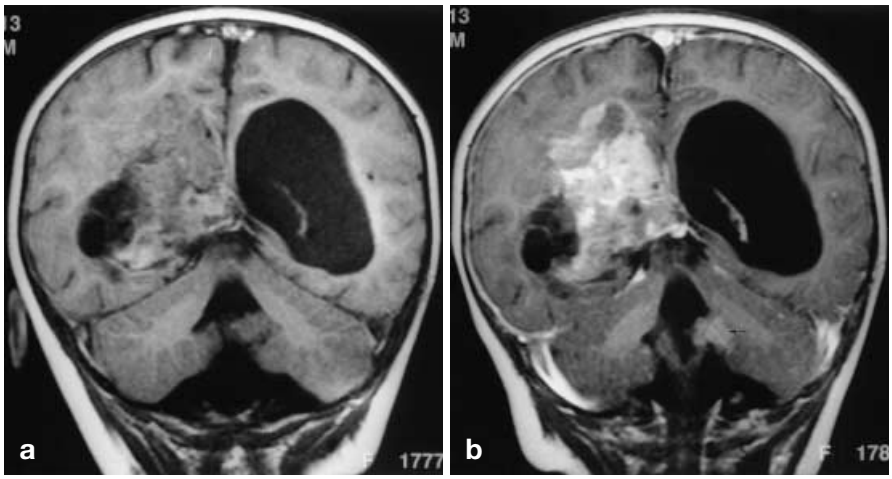

Fig. 4 a, b Patient 1. Large residual tumour involving the right lateral ventricle following initial subtotal resection. Coronal T1W images **a** before and **b** following contrast enhancement showing a predominantly solid residual mass, which is isointense to grey matter and shows avid enhancement. Cystic areas are seen laterally. There is marked hydrocephalus. An enhancing metastasis is seen in the wall of the fourth ventricle (*arrow*)

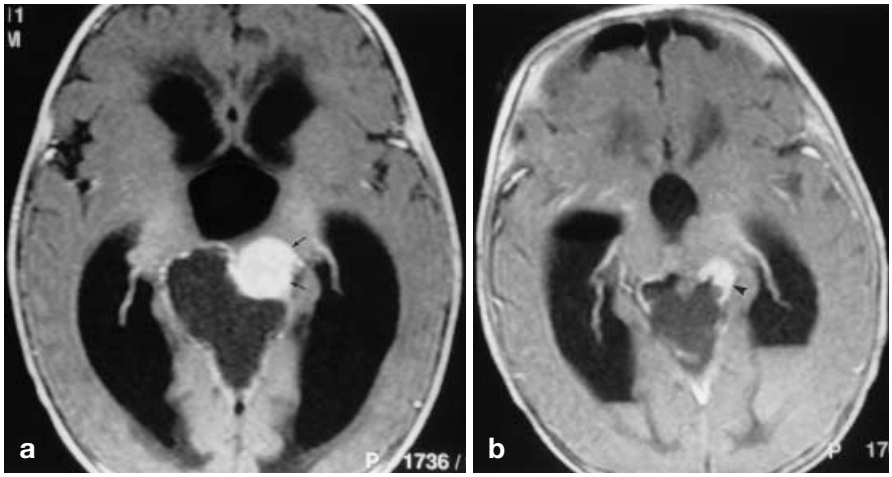

Fig. 5 a, b Patient 2. Axial T1W contrast-enhanced images **a** before and **b** following second attempt at surgical resection. Before second attempted resection, enhancing tumour is seen extending into the thalamus on the left (*arrows*). Postoperatively, an unresectable tumour residuum is demonstrated involving the thalamus (*black arrowhead*). Haemorrhage is noted within both lateral ventricles

because of its neurotoxic effects in this susceptible age group [2, 3, 5, 7–9, 12].

The distinction between choroid plexus papilloma and carcinoma can be difficult both radiologically and histologically. Macroscopically, papillomas can show parenchymal invasion and can also seed along CSF pathways [2, 7]. Radiologically, papillomas are typically homogeneous with well-demarcated margins on CT and MRI. They can, however, have irregular margins and show surrounding oedema, closely resembling CPC [13]. CPC is commonly large at presentation with extensive parenchymal invasion. It is therefore not always obvious on imaging studies that the tumour is arising from the choroid plexus. The differential diagnosis includes primitive neuroectodermal tumours (PNET), ependymomas and astrocytomas, as well as choroid plexus papillomas [13–17]. Reports of the imaging characteristics of CPC have inevitably been limited to case reports and small series. At angiography, which is now infrequently performed, the tumours have been shown to be hypervascular with supply predominantly from the choroidal arteries [7, 10].

On CT, the tumour is typically heterogeneous, but predominantly isodense or slightly hyperdense to grey matter. Calcification is frequent and tumour necrosis and cyst formation are also features. Contrast enhancement is usually marked [18, 19]. Hydrocephalus is nearly universal; the degree of hydrocephalus, however, has been noted to be less than that caused by choroid plexus papillomas [20].

The few previous reports that have detailed the MRI appearances state that the tumours have heterogeneous signal intensity on both T1W and T2W imaging, which is, in part, due to necrosis, haemorrhage and calcification. Generally the solid portions are intermediate signal intensity on T1W images (isointense to grey matter) and intermediate-to-high signal on T2W images. Areas of high signal intensity on T1W images may correspond to haemorrhage. Parenchymal invasion and vasogenic oedema are commonly seen [3, 6, 7, 14, 21–23]. In our patients, the tumours were heterogeneous, but predominantly intermediate signal on T1W, T2W and proton-density sequences. Irregular tumour margins, parenchymal invasion and surrounding oedema were seen in all our patients and these features help to distinguish CPC from choroid plexus papilloma [14].

Contrast enhancement following IV gadolinium chelates, in reported cases, is usually marked [3, 11, 23]. In our experience the degree of contrast enhancement was variable (Fig. 3).

In some previously reported cases, curvilinear signal voids representing dilated vessels were seen at MRI and these have been suggested as being characteristic [14–16], but these signal voids were evident in only two of our series. These findings are also seen in the other tumours listed in the differential diagnosis given earlier. The differential diagnosis may be narrowed down if the tumour can be located as intraventricular. Features suggesting this would include ventricular dilatation, particularly local to the tumour, tumour conforming to the ventricular shape and at least part of the tumour outlined by CSF within a ventricle. The diagnosis can then be narrowed to choroid plexus tumour or possibly meningioma, a most unusual tumour in this age group. Choroid plexus carcinomas are usually large at presentation with invasion of the adjacent parenchyma and oedema. This can make it difficult to distinguish from an aggressive parenchymal tumour growing exophytically into the ventricle.

Metastatic spread occurs by CSF dissemination and may occur early or late. The incidence of this is difficult to determine from the literature. In one series, leptomeningeal dissemination was found in 2 of 11 patients at presentation, but not all patients had been staged with MRI or myelography [3]. One of our patients had a metastasis within the fourth ventricle at presentation and another patient had multiple metastases within the CSF spaces soon before death. Pre-operative, post-gadolinium, T1W sagittal images of the entire spine were performed in all our patients, but no metastases were found below the foramen magnum.

Accurate assessment of residual disease is necessary and we have used MRI as the only imaging in the follow-up of these tumours. In four of the five cases, the first follow-up MRI was performed within 1 week of the operation; in the fifth case this was at 48 days. Further MRI was performed at intervals during chemotherapy to assess response. In four cases the residual tumour appeared to respond to chemotherapy, at least initially, as judged by a reduction in the size of residual mass. The decision to stop chemotherapy and give palliative radiotherapy was made in two cases when the residual disease subsequently enlarged. In one case 'second-look' surgery was attempted because of poor response to chemotherapy and because the full extent of the tumour was not appreciated on MRI. Surgery was, however, unsuccessful and retrospective review of the images showed that tumour was invading the thalamus and brainstem.

In summary, the MRI appearances of CPC are non-specific with a differential diagnosis including PNET, ependymoma and astrocytoma as well as choroid plexus papilloma [13–17]. The relationship of the tumour to the ventricle, particularly if it can be located as intraventricular, can help to distinguish it from other tumours occurring in childhood. CPC may be impossible to distinguish from choroid plexus papilloma, but tumour heterogeneity and signs of parenchymal invasion with vasogenic oedema within the adjacent brain are suggestive of malignancy.

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