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Posterior cranial fossa tumours in childhood

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Abstract. We reviewed clinical and CT findings in 133 posterior cranial fossa tumours in children. All had histological diagnosis, apart from 20 cases of brain stem glioma. The majority were intra-axial tumours, including 53 medulloblastomas (40%), 31 cerebellar astrocytomas (23%), 28 brain stem gliomas (21%), 14 ependymomas (11%), and single cases of ganglioglioma, haemangioblastoma and teratoma. Extra-axial tumours formed only 3%, including 2 chordomas and 2 schwannomas. The clinical data and CT findings are reviewed. Cerebellar astrocytoma involved the sexes equally, while medulloblastoma, brain stem glioma, and ependymoma were more common in males. Most cerebellar astrocytomas were in the midline, and presumably arose from the vermis. The frequency of calcification was similar to that in previous reports, being highest in ependymoma (69%), followed my medulloblastoma (29%), cerebellar astrocytoma (17%), and brain stem glioma (8%).

Key words: Brain tumour – Child's brain – Computed tomography

There have been several reviews of brain tumours in the Chinese [1–4], but none has focussed on posterior cranial fossa tumours in childhood. This study reviews such cases in a referral centre and compares the findings with those of previous reports.

Methods

We reviewed the clinical data and CT appearance of all cases of posterior cranial fossa tumours in patients less than 16 years of age seen in our hospital since 1979.

On CT, we reviewed the site of the primary lesion, the extent of involvement, its density of the mass and the degree of enhancement.

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Results

All the 133 cases of posterior cranial fossa tumour diagnosed in children in our hospital since 1979 had histological diagnosis, except for 20 cases of presumed brain stem glioma. The majority were intra-axial tumours: 53 medulloblastomas (40%), 31 cerebellar astrocytomas (23%), 28 brain stem gliomas (21%), 14 ependymomas (11%), and single cases of ganglioglioma, haemangioblastoma and teratoma. Extra-axial tumours were infrequent: we had 2 chordomas and 2 schwannomas. The numbers and sex distribution of the various tumours are shown in Table 1. Clinical features of the more common types are listed in Table 2.

The CT appearances are summarised in Table 3. The following findings are noteworthy.

Medulloblastoma

Most were in the midline of the posterior cranial fossa with only 8% located off the midline to one side. Before contrast medium the tumour was generally isodense with (33%), or denser than the surrounding brain tissue (67%) (Fig. 1).

Cerebellar astrocytoma

Histological examination showed grade I or II astrocytoma in 21 cases and glioblastoma multiforme or malignant

Table 1. Tumour type and sex distribution

	Male	Female	Total
Medulloblastoma	32 (60%)	21 (40%)	53
Cerebellar astrocytoma	14 (45%)	17 (55%)	31
Brain stem glioma	21 (75%)	7 (25%)	28
Ependymoma	11 (78%)	3 (22%)	14
Other intra-axial tumours	2 (66%)	1 (33%)	3
Extra-axial tumours	2 (50%)	2 (50%)	4
Total	82 (62%)	51 (38%)	133

Table 2. Clinical features of the four most common posterior cranial fossa tumours

	Medulloblastoma	Cerebellar astrocytoma	Brain stem glioma	Ependymoma
Vomiting	32 (60%)	14 (45%)	5 (18%)	7 (50%)
Headache	26 (49%)	14 (45%)	5 (18%)	4 (28%)
Gait disturbance	21 (40%)	10 (32%)	6 (21%)	7 (50%)
Focal neurological deficit	5 (9%)	6 (19%)	12 (43%)	1 (7%)
Dizziness	4 (8%)	2 (6%)	1 (4%)	2 (14%)
Visual disturbance	3 (6%)	3 (10%)	4 (14%)	0
Enlarged head	3 (6%)	2 (6%)	0	0
Ataxia	3 (6%)	1 (3%)	1 (4%)	0
Psychological problems	3 (6%)	0	0	0
Seizures	1 (2%)	1 (3%)	0	1 (7%)
Mental retardation	1 (2%)	0	0	0
Altered consciousness	0	1 (3%)	5 (18%)	0

Table 3. CT findings in common tumours

CT finding	Medulloblastoma (53)	Cerebellar astrocytoma (31)	Brain stem glioma (28)	Ependymoma (14)
Location				
Midline				
Anterior	0	0	25 (89%)	0
Anterior, middle	0	0	0	1 (7%)
Middle	14 (26%)	5 (16%)	0	5 (36%)
Middle, posterior	31 (58%)	8 (26%)	0	5 (36%)
Posterior	4 (8%)	4 (12%)	0	0
Anterior, middle, posterior	0	6 (20%)	3 (11%)	0
Off to one side	4 (8%)	8 (26%)	0	3 (21%)
Displacement of fourth ventricle				
Forward	42 (79%)	16 (52%)	0	4 (29%)
Forward, laterally to one side	0	1 (3%)	0	3 (21%)
Laterally	3 (6%)	5 (16%)	0	3 (21%)
Backwards, laterally	0	0	2 (7%)	0
Backwards	0	0	23 (82%)	0
Not recognizable	8 (15%)	9 (29%)	5 (18%)	4 (29%)
Hydrocephalus	34 (64%)	24 (77%)	8 (29%)	8 (57%)
Halo				
None	11 (21%)	15 (48%)	25 (89%)	4 (29%)
Complete	16 (30%)	4 (13%)	0	3 (21%)
Incomplete	26 (49%)	12 (39%)	3 (11%)	7 (50%)
Density				
(Numbers available	48	30	25	13
Low	0	17 (57%)	12 (48%)	0
Isodense	16 (33%)	7 (23%)	7 (28%)	5 (38%)
High	32 (67%)	2 (7%)	5 (20%)	4 (31%)
Mixed	0	4 (13%)	1 (4%)	4 (31%)
Calcification	14 (29%)	5 (17%)	2 (8%)	9 (69%)
Enhanced CT				
Numbers available	50	28	24	13
No enhancement	0	0	0	0
Slight	15 (30%)	11 (39%)	13 (54%)	1 (8%)
Moderate	35 (70%)	10 (36%)	8 (32%)	5 (38%)
Marked	0	7 (25%)	3 (13%)	7 (54%)
Focal lucency within tumour	34 (68%)	24 (86%)	8 (33%)	10 (77%)

astrocytoma in 6. Most occupied the midline (74%), but 26% were off the midline in one or other cerebellar hemisphere. Before contrast medium the tumour was less dense than the surrounding brain parenchyma in 57%, isodense (23%) or denser (7%); density was mixed in 13%. Foci of calcification were present in 17%. After intravenous contrast medium, enhancement was slight in 39%, moderate in 36%, and intense in 25% (Fig. 2).

Brain stem glioma

Five were low-grade astrocytomas, 1 was a malignant astrocytoma and the other 21 cases had no histological confirmation. The lesion occupied the anterior aspect of the midline in 89%, and the anterior, middle and posterior aspects in 11%. The tumour was less dense than the surrounding brain parenchyma in 48%, isodense in 28%, denser in 20%, and of mixed density in 6%. Calcification

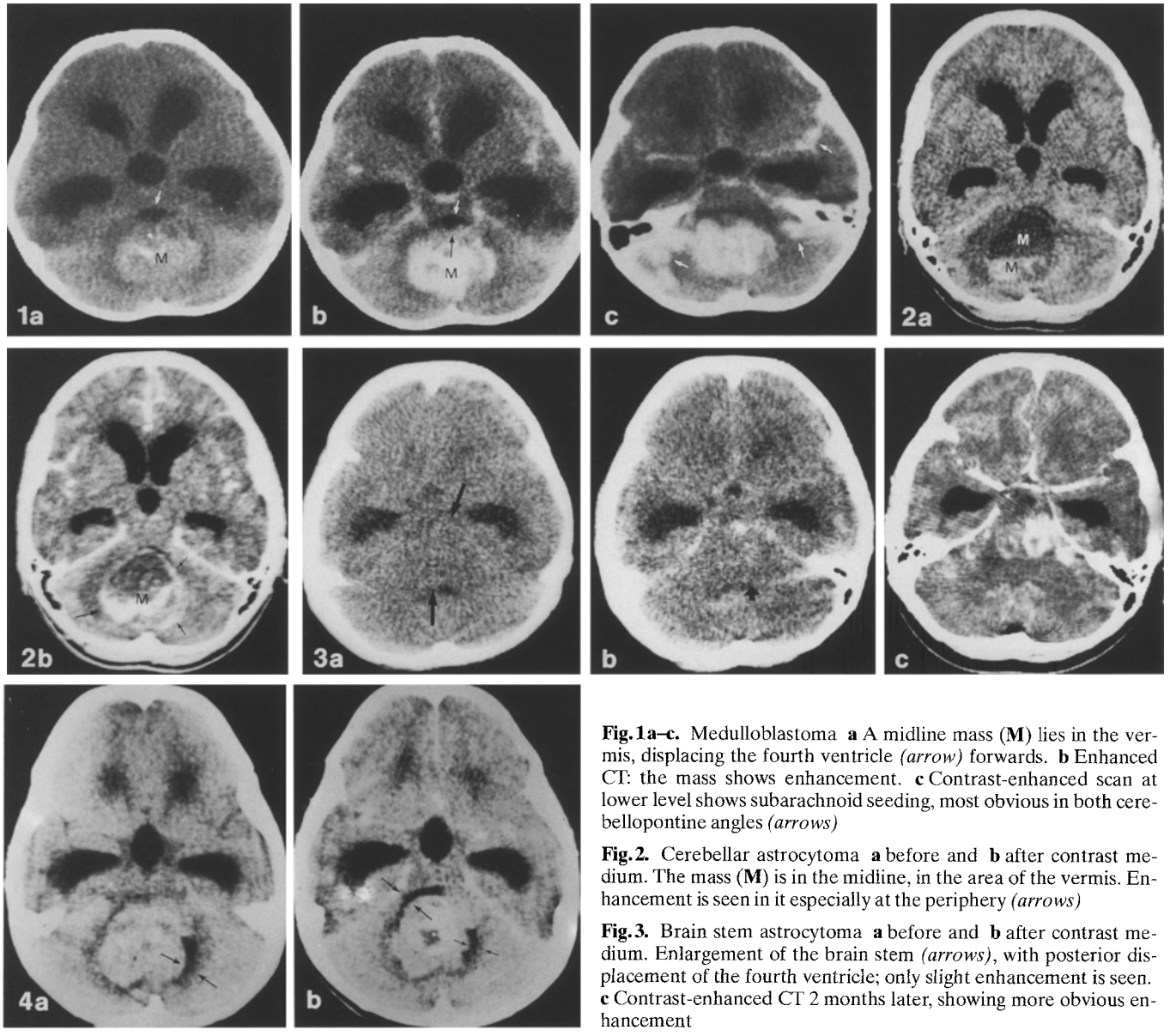

Fig. 1a–c. Medulloblastoma **a** A midline mass (**M**) lies in the vermis, displacing the fourth ventricle *(arrow)* forwards. **b** Enhanced CT: the mass shows enhancement. **c** Contrast-enhanced scan at lower level shows subarachnoid seeding, most obvious in both cerebellopontine angles *(arrows)*

Fig. 2. Cerebellar astrocytoma **a** before and **b** after contrast medium. The mass (**M**) is in the midline, in the area of the vermis. Enhancement is seen in it especially at the periphery *(arrows)*

Fig. 3. Brain stem astrocytoma **a** before and **b** after contrast medium. Enlargement of the brain stem *(arrows)*, with posterior displacement of the fourth ventricle; only slight enhancement is seen. **c** Contrast-enhanced CT 2 months later, showing more obvious enhancement

Fig. 4. Ependymoma **a** before and **b** after contrast medium. A mass of slightly increased density lies in the fourth ventricle, causing obstructive hydrocephalus. It shows inhomogeneous enhancement, with a lucency within the mass. A low density halo *(arrows)* surrounding the mass probably represents the enlarged fourth ventricle

was seen in 1 case. Enhancement was slight in 54%, moderate in 32%, and intense in 13% (Fig. 3).

Ependymoma

Most ependymomas were in the fourth ventricle and located in the midline. One fifth were off the midline, extending through the foramen of Luschka to the cerebellopontine angle cistern. They were isodense with brain parenchyma in 38%, denser in 31%, and of mixed density in 31%. Calcification was present in 69%. Slight enhancement was seen in 8%, moderate enhancement in 38%, and intense enhancement in 54% (Fig. 4).

Discussion

Previous reports on posterior cranial fossa tumours in children confirm that medulloblastoma, cerebellar astrocytoma, brain stem glioma, and ependymoma are the common lesions. They indicate that medulloblastomas comprise 28.9–40.3% [1, 3–7], cerebellar astrocytoma 13–27.7% [3, 4, 6, 7], brain stem gliomas 13.9–30% [3, 6–8]; in one report, brain stem gliomas were more common than cerebellar astrocytomas [6]. Ependymomas constitute 6.5–12.4% [3, 4, 6, 7], and are usually the least common type, but in one report, ependymomas were almost twice as common as cerebellar astrocytomas [1]. Our figures were medulloblastoma 40%, cerebellar as-

trocytoma 23%, brain stem glioma 21% and ependymoma 11%.

In children less than 2 years of age, cerebellar astrocytoma (7 cases) and medulloblastoma (6 cases) were most common, followed by brain stem glioma (3 cases) and ependymoma (1 case). These proportions are similar to those previously reported [9].

According to previous reports, medulloblastoma is more frequent in boys, who account for 61.5–88% [5, 7], cerebellar astrocytoma and brain stem glioma involve both sexes equally [7], but ependymoma shows a female preponderance of 80% [7]. Our results were somewhat different: boys predominated in the medulloblastomas (60%), brain stem gliomas (75%) and ependymomas (78%), while cerebellar astrocytoma involved both sexes equally with a slight – 55% – female predominance.

The density of the lesion relative to brain parenchyma was mostly higher in medulloblastomas, lower in cerebellar astrocytomas and brain stem gliomas and similar in ependymomas. According to previous reports, calcification is found most often in ependymoma (44–50%) [7, 8], and less often in the other tumours: 7.1–13.3% of medulloblastomas [5, 7, 8], 9% of cerebellar astrocytomas [7], and 12% of brain stem gliomas [7]. In our study, the frequency of calcification (ependymomas 70%, medulloblastomas 29%, cerebellar astrocytomas 17%) was higher than in other reports. Low density within the mass, suggesting cyst formation or necrosis, was seen in 86% of cerebellar astrocytomas, 77% of ependymomas, 68% of medulloblastomas, and 33% of brain stem gliomas.

A low-density halo surrounding the tumour completely or incompletely was seen with decreasing frequency in medulloblastomas, ependymomas, cerebellar astrocytomas, and brain stem gliomas. The halo might represent perifocal brain oedema or a dilated fourth ventricle when the mass lay completely within the ventricle.

As noted in previous reports, the maximum incidence of medulloblastoma was at 5–9 years of age [5]. Medulloblastomas are believed to arise from persistent primitive cell rests in relation to the inferior medullary velum at the base of the cerebellar vermis. The primitive cell rests normally migrate upwards and laterally from the neuroepithelial roof of the fourth ventricle to form the external granular layer of the cerebellum. These tumours, therefore, can arise anywhere along the pathway of migration, and tend to occur in the midline in younger individuals while there is a higher incidence of laterally located tumours in adults [5]. The majority in our series were in the midline and centrally located; forward displacement of the fourth ventricle was seen in 79%. A typical medulloblastoma is of uniformly high density homogeneously enhancing and sharply marginated after contrast medium [5, 8, 10, 11]. In our study, medulloblastoma was the most likely of the common tumours to be of high density: medulloblastoma (67%), cerebellar astrocytoma (7%), rain stem glioma (20%), ependymoma (31%).

Subarachnoid spread can be seen in 30% of medulloblastomas on the initial CT and in 39% on post operative follow-up [5, 11]. In our study the figures were 27% on initial CT and 52% later in the disease process. Subarachnoid spread was supratentorial (38%), infratentorial (33%), and to the spinal subarachnoid space (48%).

About two fifths of cerebellar astrocytomas are reported to be cystic or predominantly cystic [8, 11]. On CT, a typical cystic astrocytoma has a large, sharply marginated cyst containing low density fluid, and an enhancing tumour nodule is frequently observed at the margin of the cyst [8]. Very often, the mass is of low density, but could be solid or cystic mass. Cysts with proteinaceous fluid or previous haemorrhage may be denser than or isodense with brain. In our study, astrocytoma was the tumour most commonly of low density (57%). It was also most likely to show persistent focal lucency within the mass on enhanced scans (86%).

Cerebellar astrocytoma is typically a tumour of the cerebellar hemispheres [7]. Naidich et al. [12] found more than 78% to be predominantly hemispheric lesions with variable extension into the vermis. In our series, the majority arose in the midline: only 26% of the cerebellar astrocytoma were in the hemisphere, 19% displacing the fourth ventricle away from the midline. All our cases showed contrast enhancement of a solid portion or in the margin of a cyst.

Brain stem gliomas may be difficult to diagnose in the early stages because of their frequently slow development [13] and because of artifacts on CT; their demonstration is easier with MRI. Preoperative histological diagnosis of brain stem tumours was often impossible because of their inaccessibility.

Early CT diagnosis depends on displacement of the fourth ventricle and compression and distortion of the cisterns [13]. Mass effect and cisternal compression or obliteration can be appreciated in nearly every case of brain stem glioma. As in previous series [8] the fourth ventricle was compressed and displaced backwards (82%), backwards and to one side (7%), or obliterated (18%). Brain stem gliomas may show almost any density pattern. About half of them are of low density. Contrast enhancement of brain stem glioma is usually absent or minimal [13], but all our cases showed some enhancement slight in about half (54%).

Ependymomas may occur at any age [11], and were evenly distributed in this study. Most were in the fourth ventricle, therefore being midline. In previous reports, about three fifths of ependymomas have extended through the lateral recess of the fourth ventricle and the adjacent tissue into the cerebellopontine angle cistern on one or both sides [11]; in our study, only one fifth showed exophytic growth.

Postoperative myelography has been recommended in cases of ependymoma, before initiation of radiation therapy: a 36–43% incidence of positive findings has been reported [14]. In our study, 2 patients had seeding to the lumbar and 2 to the infratentorial subarachnoid space.

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