Ependymomas of the posterior cranial fossa: CT and MRI findings

Abstract We studied nine children with posterior cranial fossa ependymomas to identify specific neuroradiological features. Patients were studied preoperatively with CT and MRI; T1-, T2- and proton-density (PD)-weighted images were obtained. All children underwent surgery and a definite histopathological diagnosis was made. All the tumours grew into the fourth ventricle and caused dilatation of its upper part, which resembled a cap. All but one were separated from the vermis by a cleavage plane. In eight cases there was desmoplastic development through the foramina of the fourth ventricle, and five were heterogeneous due to necrosis and cyst change; one had a haemorrhagic area. In most cases the solid portion was isointense with grey matter on T1-weighted images, hyperintense on PD weighting, and isointense on T2-weighted images. On CT the tumour was isodense in six cases and calcification was detected in four. The presence of both desmoplastic development and a tumour/vermis cleavage plane in a posterior cranial fossa tumour isodense on CT is highly suggestive of ependymoma.

Key words Ependymoma · Posterior cranial fossa · Children · Magnetic resonance imaging · Computed tomography · Brain tumours

Introduction

Ependymomas predominate in childhood and adolescence, with a peak incidence from 1 to 15 years according to various reports [1-7]. About half are found in children aged less than 3 years [8]. They are more frequent in males, with a male/female ratio of about 3:2 [7]. They are rare, accounting for 1.9-7.8 % of central nervous system (CNS) tumours; however, in children this percentage increases to 13 % [9].

In 60-70 % of cases they are infratentorial and they account for 15 % of all posterior fossa tumors, following medulloblastoma, cerebellar pilocytic astrocytoma and brain stem tumours [2, 10].

They may arise anywhere in the ventricular system, but the fourth ventricle, especially the caudal part of its floor [7, 11], is the commonest site, followed, in order of frequency, by the lateral ventricles, aqueduct, third ventricle, spinal cord, filum terminale and cerebral hemispheres. Ectopic remnants of ependymal cells in the lateral recesses of the fourth ventricle and in the parenchyma may be responsible for extraventricular origin of these tumours [2, 4, 5, 12].

Ependymomas are neuroepithelial neoplasms forming a limited, but well-defined group of gliomas, whose classification, prognostic criteria, and treatment are still controversial.

Of the 310 CNS neoplasms we have seen since 1976, 15 (4.8 %) had a histological diagnosis of ependymoma; 9, all studied preoperatively by MRI, were infratentorial, accounting for 9.2 % of the 162 posterior cranial fossa tumours treated in that period.
Table 1 Presenting signs in nine patients with ependymoma of the fourth ventricle

<table>
<thead>
<tr>
<th>Clinical disturbance</th>
<th>No. of cases (%)</th>
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<tbody>
<tr>
<td>Increased intracranial pressure</td>
<td>9 (100)</td>
</tr>
<tr>
<td>Truncal/limb ataxia</td>
<td>6 (67)</td>
</tr>
<tr>
<td>Cranial nerve palsies</td>
<td>5 (56)</td>
</tr>
<tr>
<td>Nuchal rigidity</td>
<td>4 (44)</td>
</tr>
<tr>
<td>Torticollis</td>
<td>3 (33)</td>
</tr>
</tbody>
</table>

Methods

We analysed the preoperative MRI studies of 9 young patients with ependymoma of the fourth ventricle. All patients underwent surgery, and a histological diagnosis was made; two tumours were anaplastic. The patients were aged 18 months – 17 years (mean 5.5 years), and there were 6 males and 3 females.

MRI was performed at 0.5 Tesla (T) in eight cases and at 1.5 T in one. Spin-echo images acquired at 0.5 T were 5 mm thick and usually had a 256 x 256 matrix. All the children underwent T1-weighted (500/20/4, TR/TE/NEX) and T2-weighted images (2000/120/2) in the axial plane and in axial, sagittal and coronal planes after administration of 0.1 mM/kg gadolinium-DTPA (Gd-DTPA). In four cases PD-weighted images (2000/30/2) were also performed in the axial plane. Images acquired at 1.5 T were 5 mm thick. T1-weighted (500/20/2), T2-weighted (3000/100/1) and PD-weighted images (2300/20/1) were performed. The patient was examined in three planes before and after Gd-DTPA administration.

We performed 4 CT examinations both before and after intravenous contrast medium; 3 other patients had previously undergone CT.

Results

The clinical presentations and neuroradiological findings are summarised in Tables 1–3. Removal of the mass was radical in six cases, whereas residual tumour was detected on postoperative MRI in three. Adjuvant treatment included irradiation in four cases, while chemotherapy was added in four others. One patient, aged less than 3 years at the time of diagnosis, was given only postsurgical chemotherapy. Follow-up ranges from 1 to 4.3 years (mean 2.3 years); currently, eight of the nine patients show no evidence of recurrence, but the patient treated with surgery and chemotherapy relapsed 1 year after the operation and subsequently died.

We observed that the neoplasm, always of considerable size, tended to fill the fourth ventricle and caused dilatation of its upper portion and the lower part of the aqueduct (“capping fourth ventricle”: Fig. 1a). This is due to combined obstruction of the cerebrospinal fluid (CSF) pathways, and mechanical dilatation of the ventricle by the tumour. Hydrocephalus was present in all cases.

We found either a plane of cleavage (Figs. 3–6) or a CSF cleft (Fig. 2a) between tumour and vermis in all cases but one, in which the mass was very large (Fig. 1a).

In one case the origin of the tumour from the floor of the fourth ventricle was visible (Fig. 3).

In eight cases typical “desmoplastic development” was demonstrated (Figs. 1, 3, 4, 6). This term is used to describe the way in which ependymomas extend through the outlet foramina of the fourth ventricle into the subarachnoid space of the posterior cranial fossa [14, 15]. In four cases, the tumour extended only through the foramen of Magendie, and in one through one lateral foramen of Luschka; in three cases, it spread through both the foramen of Magendie and one lateral foramen.

In one case the neoplasm appeared to have developed in the vallecula and cisterna magna, lifting the vermis without occupying the fourth ventricle, which was only deformed (Fig. 2a).

In two cases the cerebellopontine angle cistern was occupied to the level of the inner auditory meatus (Fig. 4). In three, the tumour spread through the foramen of Magendie down to C2–3 (Figs. 1, 2). Encasement of the vertebrobasilar vessels was seen in three cases (Figs. 1, 2).

Calcification was detected in four of seven tumours investigated by CT (Fig. 4). The solid portion was isodense in six cases (Fig. 4) and iso- or hypodense in three.

On MRI, the cerebellar cortex was used as comparison: the mass was most often isointense on T1-weighted images, giving high signal on PD-weighted images and isointense on T2-weighted images, but three patients showed markedly high signal with this latter sequence (Fig. 5b). The structure was heterogeneous, with areas consistent with necrosis and cystic change, in five cases, showing markedly increased signal on T2-weighted images (Fig. 5); they were better defined on contrast-enhanced images.

Contrast enhancement of the solid portions of the tumour was totally absent in two cases. In one, only the borders of a necrotic portion enhanced (Fig. 5).

No difference in any neuroradiological feature was found between ependymoma and anaplastic ependymomas.

Discussion

Ependymomas are nodular, lobulated, reddish neoplasms. Although generally solid, about 20% are soft [2] and adapt their shape to that of surrounding structures [7]; they often resemble a placenta or a cauliflower. Their growth is slow [7, 14].

A typical but not specific feature of ependymomas [7] is plasticity: their tendency to spread as ribbon-like extensions through the outlet foramina of the fourth ventricle into the subarachnoid space of the posterior cranial fossa and the cervical spinal canal. In about 15% of cases, these tumours extend through the foramina of