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Cerebral tumors: Specific features in children



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KEYWORDS

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Abstract Brain tumors are the second leading cause of cancer in children. Primary tumors predominate and are of very varied histological types. Their prognosis and treatment depend on the histological type and grade. The diagnostic approach to these includes analysis of the site of the lesion and appearances on computed tomography and MR, and taking account of the age and clinical features of the child. CT is used to diagnose the tumor in an emergency situation. Conventional MR provides a morphological approach and allows a staging assessment to be carried out before surgery. Advanced MR techniques (diffusion-weighted and perfusion imaging, MR spectroscopy) provide further information for the differential diagnosis, presumptive diagnosis of type and grade and to guide biopsy towards the most malignant areas in the lesion.

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Brain tumors are the commonest tumors (20–22%) in children after leukemias (30%), although they are still the leading cause of cancer deaths [1] and have a number of specific features compared to adults.

Clinical signs

The clinical signs are multiple and non-specific and depend on the site of the tumor and age of the child. These include signs of raised intracranial pressure (RICP) and various neurological signs (epilepsy, visual and endocrine disturbances, ataxia, and cranial nerve palsies). Signs are more misleading in young children (macrocephaly, bulging of the fontanelles, feeding difficulties, failure to acquire new skills, hypotonia and irritability).

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Type

Unlike adults, metastases are very rare in children. Brain tumors are almost invariably primary and extra-axial tumors are extremely rare and are predisposed to by genetic changes such as NF2.

Topography

Posterior fossa tumors are as common overall as supratentorial tumors, although this varies depending on age. Under the age of 3 years old, supratentorial tumors predominate, whereas posterior fossa tumors are more common between the ages of 3 and 11 years old. Subsequently, the incidence of supra- and infratentorial tumors is identical [2]. Tumors are located in the midline in 85% of cases in the posterior fossa in 50%, in the suprasellar region in 30% and in the pineal region in 5%. This is a risk factor for meningeal spread and requires a complementary spinal MR alongside the pre-treatment cerebral MR with a sagittal T1-weighted sequence after gadolinium injection. Hemispheric tumors constitute approximately 15% of tumors and often cause epilepsy. The site provides a guide as the type of tumor (Table 1). Posterior fossa tumors include pilocytic astrocytoma, brain stem gliomas, medulloblastomas and ependymomas. Malignant gliomas should be considered in the case of tumors in the basal ganglia and thalamus. Malignant gliomas should also be

considered in the deep hemisphere whereas, peripherally, tumors are usually benign, such as the DNET.

Histological varieties

Tumors are classified by their histological type and grade (level of malignancy) in the WHO 2007 classification [3]. Grades 1 and 2 are benign tumors and grades 3 and 4 are malignant. There is considerable heterogeneity in histological types in children, both benign and malignant. Primary neuroepithelial tumors are the most common (80%), followed by craniopharyngiomas and germ cell tumors (3–5%). The commonest of the neuroepithelial tumors are the gliomas (30–50%), particularly pilocytic astrocytoma (20% of primary neuroepithelial tumors), followed in descending order by neuronal and mixed glioneuronal tumors, which account for approximately 19% (gangliogliomas, DNET, gangliocytomas, dysplastic cerebellar gangliocytoma), embryonic tumors, which account for 17% (medulloblastoma, rhabdoid tumors or ATRT, primary neuroepidermal tumors or CNS PNET), and ependymomas which account for approximately 11%. The pilocytic astrocytomas, medulloblastomas, other gliomas and ependymomas account for over 80% of childhood tumors [4]. Glioblastoma is 100 times less common than in adults. The incidence of these different histological types depends on age. Under 3 months old, teratomas (30–50%), pilocytic astrocytomas

Table 1 Histology of childhood brain tumors by site.

Site	Site	Type
Posterior fossa	Cerebellum/vermis/V4	Pilocytic astrocytoma Medulloblastoma Ependymoma Rhabdoid tumor (ATRT)
	Brain stem	Infiltrating glioma Circumscribed glioma
Hemispheres	Superficial	Ganglioglioma DNET Pleomorphic xanthoastrocytoma Angiocentric glioma Oligodendroglioma
		Deep
	Deep gray nuclei	
Intraventricular		Craniopharyngioma Optic tract glioma Germinoma Hypothalamic hamartoma
Suprasellar		Germinoma Pineoblastoma Papillary pineal gland tumor
Pineal		

ATRT: atypical teratoid rhabdoid tumor; DNET: dysembryoplastic neuronal tumor; V4: fourth ventricle.

(18–47%), papillomas (5–20%) and embryonic tumors predominate [4,5]. Medulloblastoma is the predominant tumor between 0 and 4 years old, pilocytic astrocytoma between 5 and 9 years old, high grade glioma between 10 and 14 years old, and suprasellar tumors between 5 and 19 years old.

Genetic and molecular analysis

This is carried out as a complementary process to histological examination and provides a better assessment of the diagnosis/prognosis and response to treatment. Childhood gliomas have different molecular and genetic features to those in adults and the same histological type can have different histomolecular features depending on its site [6].

New aspects from the WHO (World Health Organization) 2007

These contain additions and new definitions or clarifications of existing diseases. Several tumor groups are concentrated in the pediatric population [7]. For the gliomas, these include two types of tumor, pilomyxoid astrocytoma and angiocentric glioma. Pilomyxoid astrocytoma (grade 2), which is a variant of pilocytic astrocytoma, is characterized by local recurrence and dissemination. Angiocentric glioma (grade 1, also known as angiocentric neuroepithelial tumor) is seen in children and young adults with drug resistant epilepsy. The diagnosis is generally made on histological examination. Of the intraventricular tumors, the atypical choroid plexus papilloma (grade 2) is characterized by increased mitotic activity, which is not differentiated or distinguished from a papilloma on imaging and the diagnosis is histological. The same applies within the hemispheric glioneuronal tumors to papillary glioneuronal tumors (PGNT), which are generally diagnosed as a ganglioglioma preoperatively. In the pineal region, the papillary tumor has no specific imaging features and the diagnosis is also histological. Amongst the embryonic tumors, medulloblastoma with extensive nodularity may appear similar on imaging to a classical medulloblastoma. A bunch-of-grapes appearance with multiple enhanced nodules, however, is characteristic. There are strong genetic predispositions to this tumor including the Fragile X syndrome and the Li-Fraumeni and the Gorlin syndromes. A new concept of the embryonic tumor with abundant neuropil and true rosettes (ETANTR, grade 4) is described amongst the other PNET. This is a hemisphere tumor with little or no peritumoral edema and weak or non-existent enhancement.

Specific childhood brain tumors

In infants, these are the desmoplastic infantile ganglioglioma (grade 1) and the atypical rhabdoid tumor (ATRT) (grade 4), which carry a poor prognosis, choroid plexus papillomas (grade 1) and teratomas. Regardless of age, the other tumors which are unique to the pediatric population are the pilocytic astrocytoma and secretory or non-secretory germ cell tumors.

Syndromic context

Occasionally, specific brain tumors may occur in predisposing syndromes for cancer: NF1 (optic tract glioma), NF2 (meningiomas, ependymomas), tuberous sclerosis (giant cell subependymal tumor) and Gorlin or Li-Fraumeni syndrome (medulloblastoma) [8].

Prognosis and treatment

These depend on the histological grade and type and on the staging assessment but also on the child's age. Tumors in children under 6 months old carry a poor prognosis because of the higher incidence of malignancy, larger tumor size and the contraindication to radiotherapy because of the adverse effects on the developing brain [9].

Imaging strategy

The prognosis and treatment of brain tumors depends on the histological grade, which is based on the WHO 2007 criteria, obtained after biopsy or surgical resection. Generally, treatment always starts with surgery, possibly supplemented by chemotherapy and/or radiotherapy depending on histological type and grade of the tumor. It is important, however, to obtain an accurate preoperative diagnosis of the type of assumed tumor for several reasons. It is firstly essential to distinguish between true tumors and pseudotumors (abscesses, demyelinating lesions, encephalitis, etc.), which may have the same clinical presentation and must not be treated surgically. This therefore requires multiparametric MR [10,11]. Knowledge of the type and grade of tumor enables surgery (the surgical approach) to be planned, or biopsies to be guided towards the most malignant parts of the lesion. In a presumed benign tumor, surgery may be less aggressive in some areas. This appears to be essential to preserve the child's future neurological outcome. In addition, some tumors are not treated surgically and are occasionally not biopsied because of their site (optic tract, brain stem) and the diagnosis and choice of the treatment is based on imaging.

The diagnostic approach includes analysis of the site of the tumor and its MR and CT appearances, incorporating factors relating to the patient's clinical findings and age.

Cerebral CT

Because of its ready availability and speed, CT is the first investigation to be performed for a suspected brain tumor. It provides a positive diagnosis of the tumor and demonstrates whether there are signs of brain herniation, which determine the urgency of surgery. This is performed in unenhanced mode in order to identify hemorrhage or calcifications, followed by contrast enhancement. These features are difficult to distinguish on MRI [12] even using T2*-weighted gradient-echo and susceptibility (SWI) weighted imaging. CT can also be used to assess tumor cellularity, a hyperdense tumor on CT reflecting hypercellularity and very often a high grade.

Cerebral MR

MR is the investigation of choice for the preoperative assessment of a childhood brain tumor. It includes conventional multiplanar imaging and advanced techniques, which should form part of the initial assessment of any brain tumor. T1- and T2-weighted images and T1 images with gadolinium enhancement provide a morphological and staging assessment of the lesion but appear however to be limited in their ability to provide a more detailed characterization, particularly in terms of type and histological grade. Advanced MR techniques such as diffusion-weighted imaging, MR spectroscopy and perfusion MR offer a functional approach which improves the characterization of cellularity, vascular hemodynamics and tissue metabolism, improving the specificity of the MR.

Conventional MR

The imaging protocol should be adapted to take account of the site of the tumor. This should include a minimum of T1-weighted images without gadolinium, T2-weighted and T2* and FLAIR images, 3D volume images, or at least 2× T1-weighted orthogonal planes after gadolinium enhancement. Interpretation should include an analysis of the boundaries of the lesion (infiltrating or expansive), recording its site, whether it is unifocal or multifocal, its size and intensity on T1, T2 and FLAIR imaging, whether it is homogeneous or heterogeneous in appearance and whether it contains necrosis or cysts within the tumor, macroscopically abnormal blood vessels, contrast enhancement, edema around the tumor, a space-occupying effect or meningeal sites of disease.

The overall appearance of the lesion may have characteristic features in a clearly defined site (a pilocytic astrocytoma in the posterior fossa, teratoma, craniopharyngioma, etc.) (Box 1).

Outside of these conditions, evidence is sought to support the benign or malignant nature of the lesion, which also depends on tumor site:

- a hemorrhagic tumor with peripheral edema containing multiple ectatic vessels is suggestive of a malignant tumor although, in a recent study, edema around the tumor and a mass effect did not correlate with high grade malignancy in children, as edema around the tumor was present more in benign (50%) than malignant (31%) tumors [13];
- on T2-weighted imaging (T2WI), hypercellular tumors (often high grade) are iso-/hypointense compared to the cortex, whereas paucicellular tumors (often low grade) are hyperintense [14]. Tumor microvasculature is also clearly seen on this sequence;
- the level of contrast enhancement does not correlate with grade of tumor in children. Pilocytic astrocytoma and choroid plexus papilloma enhance intensely after gadolinium injection, whereas these are low grade tumors. Conversely, high grade tumors such as the anaplastic astrocytoma, medulloblastoma or ETANTR, may or may not enhance;
- intra- and peritumoral cysts which are isointense with CSF (cerebrospinal fluid) are conventionally characteristic of pilocytic astrocytomas, gangliogliomas (regardless of grade) and DNET (in which generally the intratumoral cysts are less than 5 mm in size);

Box 1. Imaging features of the main childhood tumors.

- Posterior fossa tumor
 - Medulloblastoma: low ADC.
 - Pilocytic astrocytoma: cyst with wall contrast enhancement, high ADC.
 - Ependymoma: extension into the foramina of Luschka and Magendie.
- Suprasellar tumor
 - Craniopharyngioma: a hyperintense cyst on T1WI, calcified.
 - Germinoma: solid part hypointense on T2 imaging and ADC ≤ normal brain.
 - Pilocytic astrocytoma: high choline and ADC in the tissular part.
- Pineal tumors
 - Germinoma: hypointense solid part on T2WI and ADC ≤ normal brain.
 - Teratoma: cyst, calcifications, tissular components.
 - Pineoblastoma: very low ADC.
- Cortical tumors
 - Ganglioglioma: calcifications.
 - DNET: pseudocystic and cystic.
 - Pleomorphic xanthoastrocytoma: superficial and meningeal enhancement.
 - Angiocentric glioma: hyperintense ring on T1WI.
- Intraventricular tumors
 - Choroid plexus papilloma: intense enhancement in the ventricular trigone.
 - Subependymal giant cell astrocytoma: foramen of Monro, tuberous sclerosis.
- DGN tumors: gliomas, germinoma.
- Hemispheric tumors: gliomas, PNET (low ADC, necrosis and hemorrhage), ependymomas (intraparenchymal, close to the ventricular trigone).

DNET: dysembryoplastic neuronal tumor; PNET: primary neuroectodermal tumor; DGN: deep gray nuclei; WI: weighted imaging.

- necrosis, which is not isointense with the CSF, usually seen on FLAIR imaging, is rarer than in adults, as the tumors tend to be more hypercellular than necrotic. Embryonic tumors and ependymomas of the posterior fossa however are often necrotic. Necrosis, however, may be present in benign tumors (pilocytic astrocytomas) and completely absent from high grade hypercellular tumors.

Diffusion-weighted MR

Diffusion-weighted imaging should be used routinely as this is sensitive to tumor cellularity. The ADC is inversely proportional to cell density and therefore the tumor grade [15]. All embryonic tumors (medulloblastoma, rhabdoid tumor and PNET) and the pineoblastoma and germ cell tumors, mostly seen in children, are malignant (grade 4), and have a very low ADC. Diffusion-weighted imaging, therefore, distinguishes high from low grade tumors in children in the absence of hemorrhage [16], both on a supra- and

infratentorial level and there is no cutoff value between grades or between the main tumors [12,16–19]. All tumors combined, the DNET has the highest value and embryonic tumors have the lowest value.

Perfusion MR

Dynamic contrast-enhanced T2*-weighted perfusion MRI indirectly reflects the density and vascular proliferation within the tumor from measurement of the rCBV (relative cerebral blood volume). The rCBV correlates with grade of glial tumor in adults, high grade tumors having a higher rCBV than low grade tumors [20]. These findings cannot perhaps be extrapolated to children because of histological differences between the tumors. There are few MR perfusion studies on cerebral tumors in specific pediatric populations [21,22], probably because of technical injection problems. It is, however, entirely possible to perform a perfusion MR in young children using small catheters and injection rates of 0.8 to 1 ml/s [22], or using manual injection [23].

An alternative MR perfusion-weighted technique (arterial spin labeling) is reported to distinguish high from low grade brain tumors in children, but not the different histological types [24]. Maximum rTBF (relative tumor blood flow) is higher in high grade compared to low grade tumors.

MR spectroscopy

MR spectroscopy (MRS) is used to study tumor metabolism. The main features of malignant tumors are elevated choline (Cho) levels, attributed to cell membrane proliferation and reduced *N*-acetylaspartate (NAA) and creatine (Cr) levels attributed to edema and necrosis [25]. A lactate (Lac) signal is often seen in malignant rather than low grade lesions [26], due to metabolic acidosis within tumor tissue. Necrosis is reflected by an increase in Lac/Lip (lipid) resonance associated with a fall in all of the other metabolites [27]. The highest ratios of Cho/Cr and Cho/NAA are seen in aggressive tumors and diffuse brain stem gliomas.

Some spectroscopic features of different pediatric brain tumors are seen. The benign pilocytic astrocytoma has a high Lac level as a result of still poorly understood biochemical metabolism. The Cho/NAA and Cho/Cr ratios are raised despite the fact that these lesions are benign [4,28,29]. In children, therefore, raised choline is not synonymous with a malignant tumor. In type 1 neurofibromatosis, the Cho/Cr ratio helps to distinguish between local intraparenchymal involvement, which is hyperintense on T2 imaging, from glial tumors and healthy parenchyma [30] in children. The glioblastomas have the highest alanine glutamine-glutamate and glycine concentrations [31]. Myoinositol levels have been shown to be raised in choroid plexus papilloma [29,32]. This is a benign lesion, particularly compared to the malignant choroid plexus carcinoma, in which choline is increased [33]. Myoinositol is also raised in ependymoma [4]. Taurine concentrations are increased in primary neuroectodermal tumors (PNET) [18,33] and citrate is found in malignant infiltrating pontine astrocytomas [4]. Germ cell tumors have high levels of glutamine-glutamate.

Taken in isolation, each technique appears to be limited in distinguishing and grading tumors. A multimodal

MR approach therefore appears to be essential in the preoperative assessment of a tumor [34] in order to improve its characterization. Unlike in adults, few large pediatric series are available [18,21] as these tumors are less common and are histologically more diverse.

Gliomas

Pilocytic astrocytoma

This is the commonest tumor in children and accounts for 30% of posterior fossa tumors [35]. It is a benign (grade 1), clearly circumscribed and slow-growing, often small tumor, found between the ages of 5 and 13 years old [36]. Surgery is curative without recurrence if completely excised. Sixty to 80% of pilocytic astrocytomas are located in the cerebellum, usually in the cerebellar hemispheres or in the vermis. The other sites involved are the hypothalamic-pituitary region, thalamus, brain stem and, far less commonly, the cerebral hemispheres and ventricles [37]. Its most typical appearance is a cyst with an intensely enhancing mural nodule, which is only seen in 50% of cases (Fig. 1). This is not specific for a pilocytic astrocytoma and may be seen in gangliogliomas and pleomorphic xanthoastrocytomas. PA may present as a solid lesion with a cystic-necrotic center in 40 to 45% of cases, or as a purely solid lesion in 10% of cases [36]. The solid portion of the tumor is hypodense on CT and occasionally calcified. It is iso-/hypointense on T1WI and hyperintense on T2WI. It enhances intensely with contrast, occasionally heterogeneously on MR. On diffusion-weighted imaging the ADC is increased as the tumor is paucicellular. On perfusion imaging the rCBV is low, with a curve rising above the baseline, reflecting blood-brain barrier disruption [38]. This cyst has occasionally a high protein or hemorrhagic content and the cyst wall occasionally enhances homogeneously, often if the adjacent cerebral parenchyma is infiltrated or if hemorrhage has occurred within the cyst [36].

The optic tract glioma is a pilocytic astrocytoma, which develops in the region of the hypothalamus, optic chiasma or optic nerves and accounts for 5% of childhood tumors [24]. Fifteen percent of patients with NF1 develop an optic tract glioma (OTG) and a third of patients with optic nerve pilocytic astrocytoma have NF1 [39], whereas posterior visual tract involvement is usually seen outside of NF1. OTG is characterized by visual and endocrine disturbances (early puberty). It presents as a multilobulated oval or round mass, with superior extension into the third ventricle, inferior ventricle or sella turcica, posteriorly into the interpeduncular fossa and anteriorly into the anterior cerebral fossa. It develops along the anterior and posterior optic tract but it is occasionally located in the hypothalamic-chiasmal region. Meningeal spread is rare and usually occurs after surgery. Contrast enhancement varies and is occasionally only seen on the peripheral part of the lesion.

The pilomyxoid astrocytoma is a more aggressive form of disease (grade 2) [40] and is typically found in the suprasellar region in young children. It should be considered in infants or young children, particularly if the lesion is

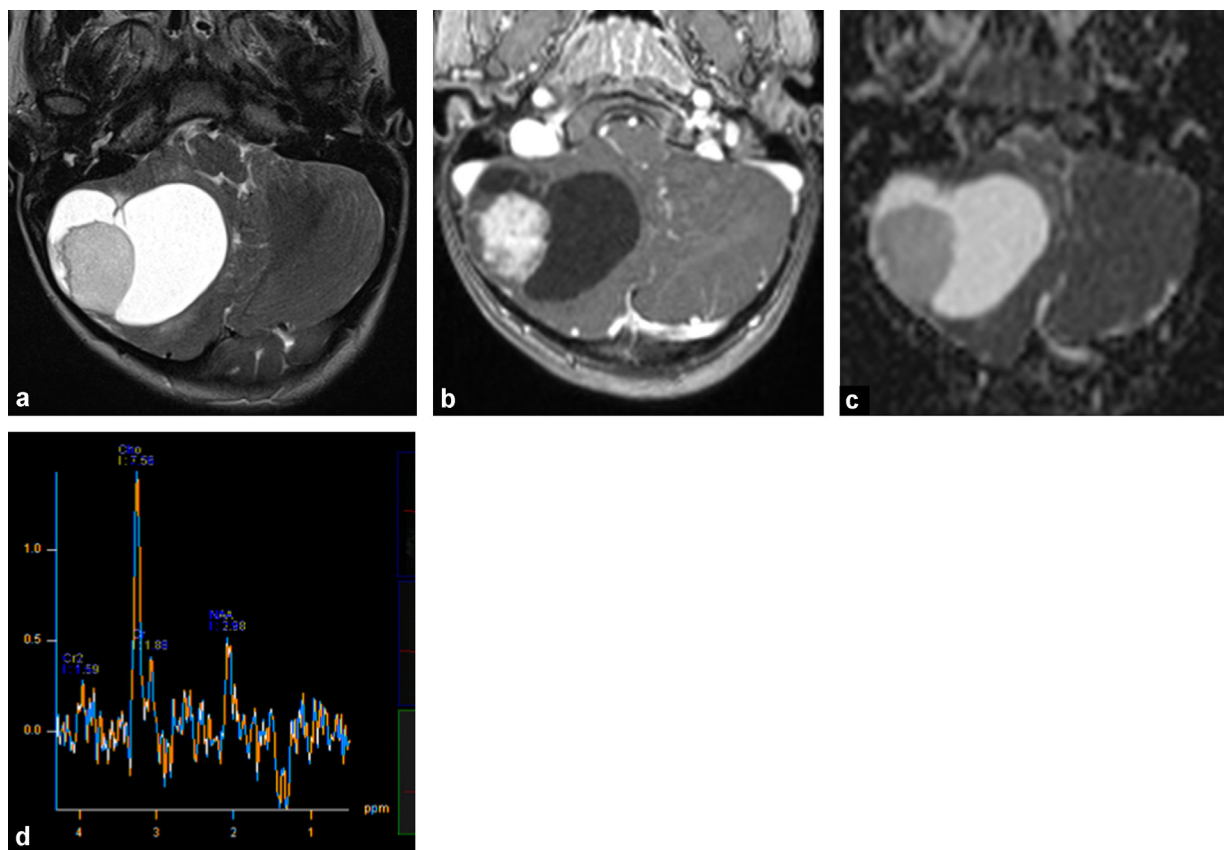


Figure 1. Pilocytic astrocytoma in a 6-year-old child; a: axial T2-weighted image: cystic tumor lesion with a wall tissue nodule in the right cerebellar hemisphere; b: axial T1-weighted image after gadolinium injection: intense enhancement in the solid part of the lesion; c: axial ADC map: the tissular part of the lesion has a high ADC value; d: monovoxel MR spectroscopy (PRESS, TE 135): raised choline, low NAA, and lactate present.

hemorrhagic (25% of cases) with intense contrast enhancement [7].

Other gliomas

Pleomorphic xanthoastrocytoma is a grade 2 tumor, which is occasionally anaplastic (grade 3), and is usually seen in adolescents and young adults over 20 years old. It is a cortical tumor which is located in the temporal (49%) parietal, frontal or occipital regions [7] and causes epilepsy. It is seen as a cyst with a peripheral wall nodule and meningeal or dural attachment in 2/3 of cases or occasionally as a purely solid lesion [41]. Calcifications are rare. The adjacent cranial vault may become eroded. The solid peripheral portions of the tumor are isointense on T2WI and enhance with gadolinium (the enhancement is described as "meningocerebral").

Grade 2 glioma (diffuse fibrillary astrocytoma)

Grade 2 glioma (diffuse fibrillary astrocytoma) (Fig. 2) mostly develops in the frontal and temporal lobes and is a homogeneous infiltrative lesion without surrounding edema or hemorrhage. It is hypodense on CT, hypointense on T1WI, and hyperintense on T2WI, and does not enhance with gadolinium.

Anaplastic gliomas (astrocytomas, oligoastrocytomas)

Anaplastic gliomas (astrocytomas, oligoastrocytomas) are grade 3, and have the same appearance, although with a more heterogeneous image. Myoinositol appears lower than in the low grade gliomas on MRS [4] (Fig. 3).

Glioblastomas (grade 4)

Glioblastomas (grade 4) are rare in children. This is a hemorrhagic necrotic lesion, which is enhanced after gadolinium injection. Choline is increased, lactate and lipids are present, the rCBV is increased and the ADC is low [42]. These malignant gliomas are also seen in the deep gray nuclei. Focal anaplastic sites are suggested by a hemorrhagic area or by contrast enhancement [43]. It is occasionally difficult to formally distinguish the different grades of gliomas on MR.

Oligodendrogliomas

The oligodendrogliomas are rare grade 2 tumors accounting for under 2.5% of childhood tumors [44], which are seen around adolescence. More aggressive anaplastic forms of the tumor (grade 3) are seen. They are located in the frontal (50–85%) and temporal [2] regions in the cortex and adjacent white matter, or in the central gray nuclei and thalamus, and they carry a poorer prognosis. They

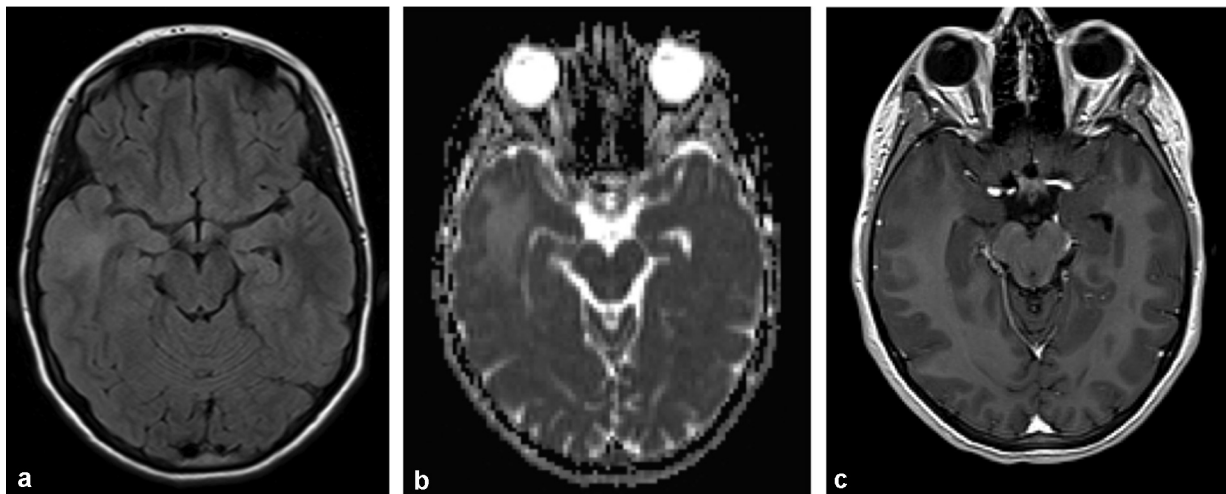


Figure 2. Diffuse grade 2 astrocytoma in a 13-year-old child; a: axial FLAIR image: infiltrating temporal lobe white matter lesion, hyperintense on FLAIR; b: axial ADC map: increased ADC within the lesion; c: T1-weighted axial image after gadolinium injection: the lesion is hypointense and does not enhance with gadolinium.

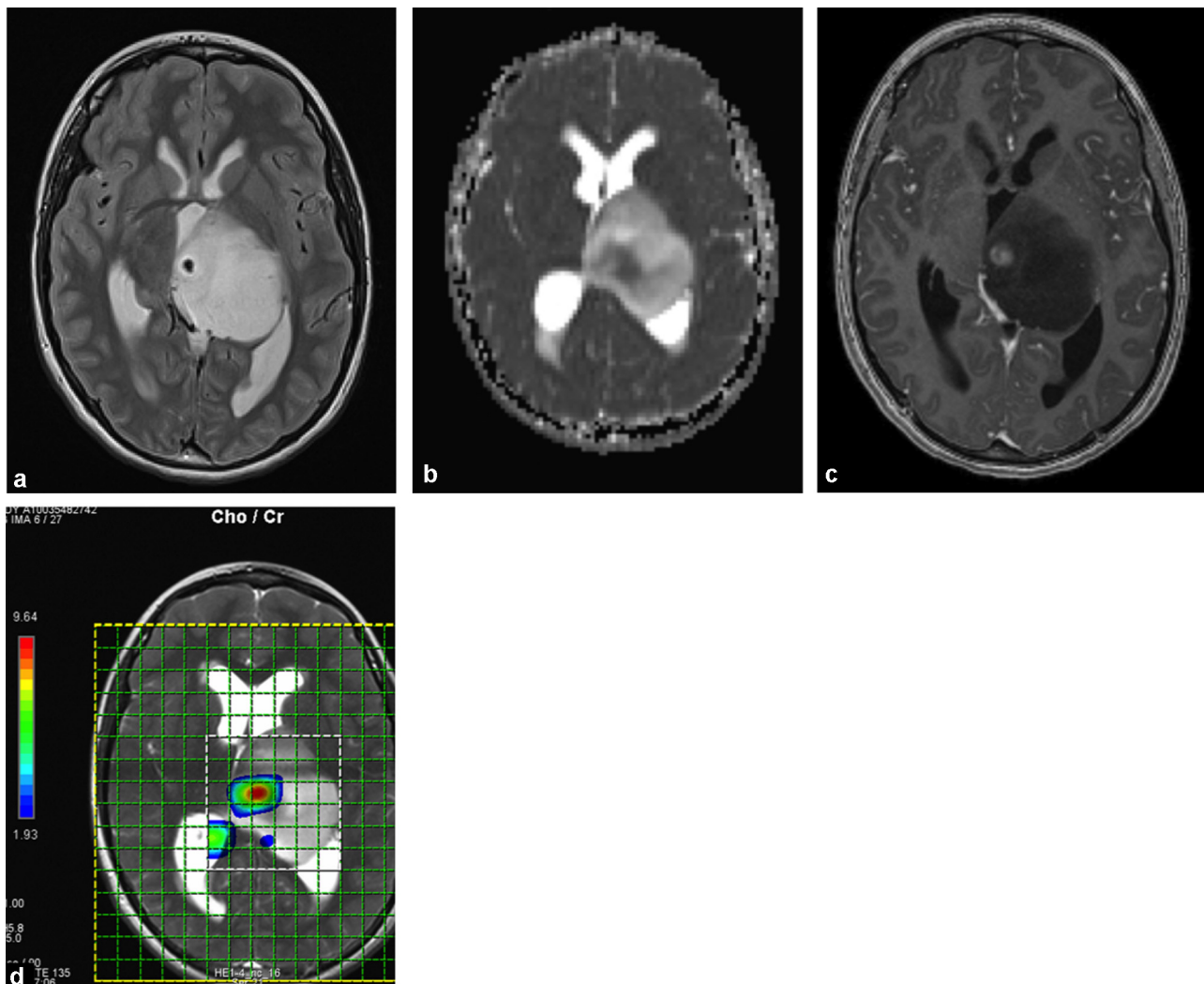


Figure 3. Grade 3 anaplastic astrocytoma in a 12-year-old child; a: axial T2-weighted image: hyperintense lesion in the left thalamus on T2-weighted imaging; small hypointense area of hemorrhage on T2, suggesting an anaplastic component; b: axial ADC map: area of reduced ADC within the lesion which, overall, displays a high ADC suggesting an anaplastic area; c: T1-weighted axial image after gadolinium injection: contrast enhancement around the area of hemorrhage; d: spectroscopic imaging (CSI, TE 135): the contrast-enhanced area has a large increase in the choline/creatine ratio.

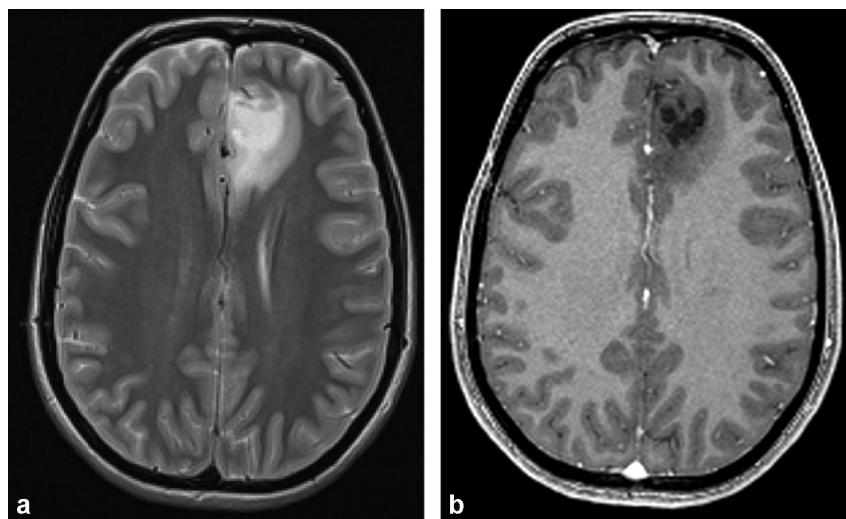


Figure 4. Grade 2 left frontal oligodendroglioma in a 17-year-old child; a: axial T2-weighted image: infiltrating lesion in the cortex and underlying white matter, hyperintense on T2-weighted imaging; b: axial T1-weighted image after gadolinium injection: the lesion does not enhance and contains small cysts.

are characterized by epilepsy, RICP and neurological deficit [44]. The lesion is hypodense with calcifications in 40% of cases (fewer than in adults) [45]. The signal is hypointense on T1-weighted and hyperintense on T2-weighted imaging, occasionally with small cysts (Fig. 4). Contrast enhancement is rare in children, as is the mass effect or the edema [46], apart from anaplastic tumors. The ADC is increased. The differential diagnosis is with dysplasias and DNET.

Angiocentric glioma

Angiocentric glioma is a cortico-subcortical lesion, which is characterized by partial refractory partial epilepsy in children between 2 and 14 years old. It is located in the temporal region in 38% of cases, frontal in 25%, parietal in 10% and occipital in 8% [47]. The grade 1 lesion may remain stable or increase in size slowly. It is clearly delineated, does not

enhance and is located in the cortex with extension towards the ventricle. The tumor is hypointense on T1WI and hyperintense on T2WI and FLAIR, without contrast enhancement. The most characteristic features are reported to be extension of the lesion towards the lateral ventricles on T2WI and FLAIR and a hyperintense cortical rim on T1WI (Fig. 5) [48].

Giant cell astrocytoma

Giant cell astrocytoma is a benign grade 1 intraventricular lesion which develops close to the foramen of Monro, and may produce acute obstructive hydrocephalus. It is almost only seen in tuberous sclerosis, in 15% of patients under the age of 20 years old [45]. Other stigmata of the disease include: cortical tubers, subependymal nodules and radial migration lines in the white matter. It presents as an occasional multifocal mass containing calcifications which

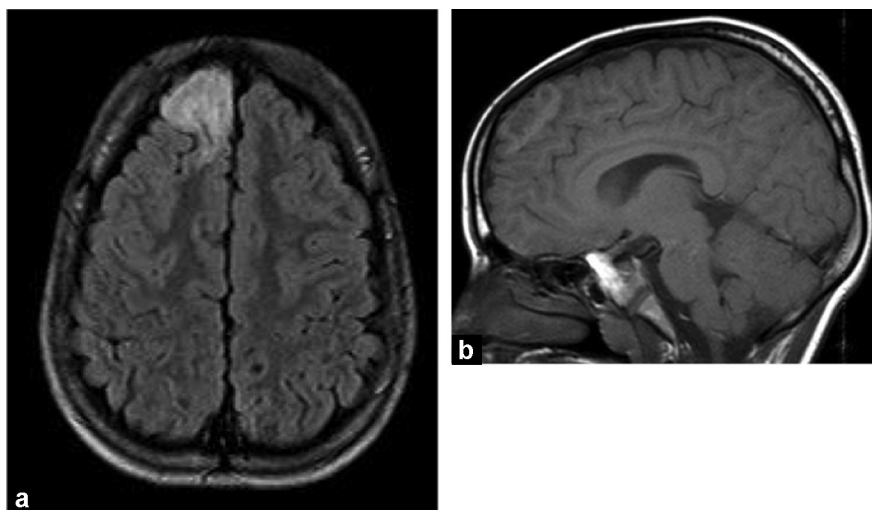


Figure 5. Angiocentric glioma in a 10-year-old child; a: FLAIR axial image: right frontal lesion involving the cortex and white matter, hyperintense on FLAIR imaging; b: sagittal T1-weighted image without contrast injection: the lesion displays a characteristic peripheral ring, hyperintense on T1-weighted imaging.

are hypo-/isointense on T1 and iso-/hyperintense on T2-weighted imaging, enhancing strongly with gadolinium. It should be distinguished from a subependymal nodule, from which it may develop, from its size, which is over 10/12 mm. Tumor size can be reduced by treatment with everolimus [49].

Brain stem glioma

Brain stem glioma accounts for 10 to 20% of childhood cerebral tumors and 20 to 25% of infratentorial tumors. Two types of presentation are found on MR, which carry a different prognosis: diffuse (the commonest) and focal (better prognosis) [50].

Diffuse intrinsic pontine glioma

The diffuse intrinsic pontine glioma accounts for 60 to 80% of pontine tumors and 75% are found before the age of 10 years old [35]. These have a very poor prognosis, with a life expectancy of no more than one year. Clinical signs are a combination of cranial nerve involvement, particularly VI and VII, a pyramidal syndrome and ataxia. These are, in general, grade 2 fibrillary gliomas, although the tumors are rarely biopsied because of the risk of the complications. They are difficult to see on CT and are only indicated by the mass effect, which they produce on the fourth ventricle. On MR, they appear as a large infiltrating pontine lesion (Fig. 6), which may extend vertically, into the cerebellum and anteriorly, surrounding the basilar artery. They are hypo-/isointense on T1 and hyperintense on T2 and FLAIR imaging, show little or no enhancement with gadolinium [51], and have a high ADC [51]. Transformation into a higher grade (3 and 4) is reflected by development of contrast enhancement, hemorrhage, an area of reduced ADC, raised rCBV [22], or on MRS by an increase in choline and the presence of lipids.

Focal tumors

Focal tumors are located in the mesencephalon, medulla or cervico-medullary junction. They are usually low grade tumors (pilocytic astrocytoma or ganglioglioma), which are identical in appearance to those in other sites. They may be intrinsic or dorsal exophytic in the fourth ventricle or

perimedullary cisterns, or may be anterolateral [50,52]. Surgery is often partial although it still has a good prognosis.

Glioneuronal tumors

Ganglioglioma

This is an, often benign, grade 1 tumor accounting for 1 to 4% of childhood brain tumors and 40% of tumors causing epilepsy. The most common clinical presentation is refractory complex partial epilepsy and the tumor is 10 times more common in children than in adults [53]. The majority of cases are seen between the ages of 10 and 20 years old [54]. They are located in the cortex, in 80% of cases in the temporal regions and in 12% of cases in the frontal regions and present as a solid mass (43%), cyst (5%), or mixed lesion (52%) (Fig. 7) [53]. Highly suggestive calcifications are seen in 50% of cases. Scalloping of the adjacent cranial vault may occur, although no edema is seen around the lesion. CT appearances vary and on MR the lesion enlarges a gyrus and affects the cortex. The solid part of the lesion appears iso-/hypointense on T1 and hypointense on T2WI. Contrast enhancement is variable in intensity, and occasionally peripheral, and is seen in 60% of cases. The ADC is increased and the rCBV appears higher than in low grade gliomas [45]. Multimodal MRI is required in refractory epilepsy to distinguish a ganglioglioma from a DNET and local cortical dysplasia because of its potential for anaplastic transformation (grade 3). rCBF, rCBV and choline are higher in gangliogliomas and the creatine is lower [55].

Desmoplastic infantile ganglioglioma

Desmoplastic infantile ganglioglioma is a specific form of ganglioglioma seen in children under 6 months/1 year old. It causes macrocrania, epilepsy and focal neurological signs. It is usually located in the frontal or parietal region and presents as a large, solid cystic lesion. The solid portion of the lesion is attached to the dura and is hypointense on T2WI with a low ADC, enhancing intensely with contrast. Edema may be present, although calcifications are not seen. It has a good prognosis.

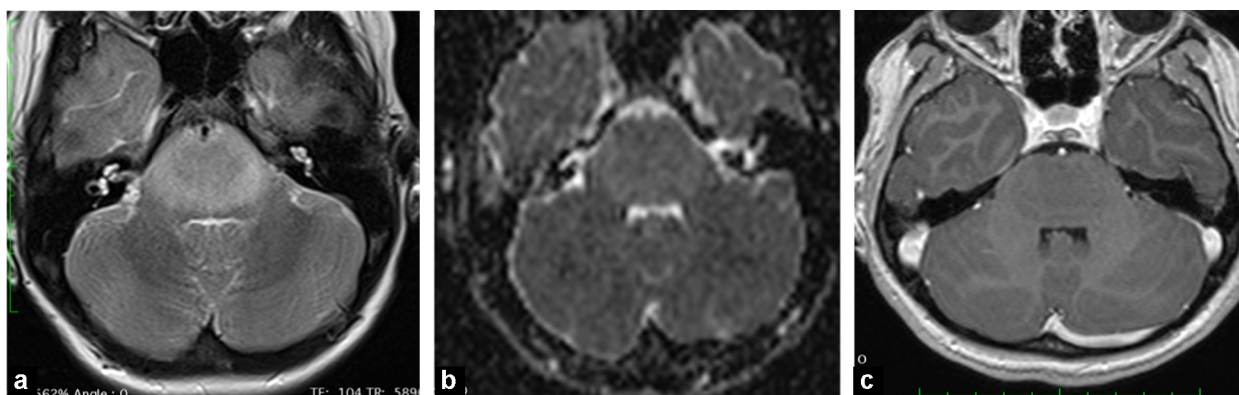


Figure 6. Anaplastic brain stem glioma in a 15-year-old child; a: T2-weighted sagittal image: infiltrating lesion in the pons which is hyperintense of T2-weighted imaging, causing compression of the fourth ventricle; b: axial ADC map: the ADC is increased due to the paucicellular nature of the lesion; c: axial T1-weighted axial image after gadolinium injection: the lesion does not enhance.

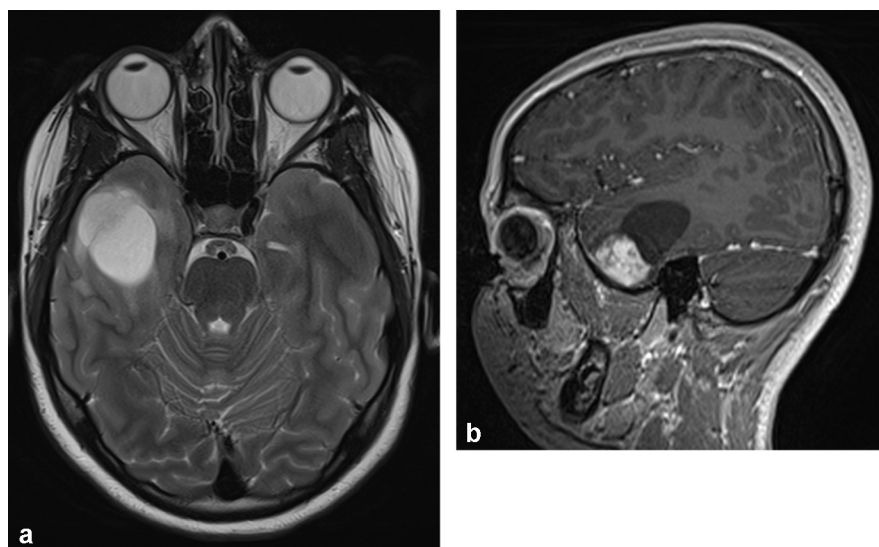


Figure 7. Right temporal ganglioglioma in a 13-year-old child; a: axial T2-weighted image: right temporal solid cystic lesion with a peripheral tissue portion, which is hyperintense on T2-weighted imaging; b: sagittal T1-weighted image after gadolinium injection: intense contrast enhancement in the tissular part of the lesion.

Dysembryoplastic neuronal tumor (DNET)

This is a grade 1 neuroglial tumor, which accounts for 14% of epilepsy-related tumors and is seen before the age of 20 years old. Surgery can cure the epilepsy and recurrences are rare. It is located in the cortical/subcortical, temporal (62%) or frontal (31%) regions [54]. Occasional intraventricular tumors are seen, together with those which develop in the deep gray nuclei and thalamus. The most typical histological appearance is the specific glioneuronal component. According to the Daumas-Duport classification, there are three histological types of the tumor: simple, complex and non-specific. The simple form is associated with pseudocystic appearances, the complex form with pseudomulticystic appearances, and the non-specific form with homogeneous or heterogeneous nodules or pseudodysplasia [56]. The tumor is clearly delineated, triangular in shape, with no edema and occasionally produces scalloping on the cranial vault (Fig. 8) [57]. It appears multilobulated with

septa made up of multiple pseudocysts. Calcifications are seen in a third of cases [54]. The image is hypointense on T1, and hyperintense on T2WI and FLAIR images, with no edema or space-occupying effect apart from local expansion of a gyrus. Contrast enhancement is seen in 21 to 50% of cases and is nodular, annular or heterogeneous. A raised myoinositol and insignificant fall in NAA have been reported on MRS [58]. The images of DNET or contrast enhancement may change spontaneously over time [53].

Disseminated glioneuronal tumor

This is a rare tumor with fewer than 30 cases described in the literature, grouped under the term glioneuronal tumor or diffuse meningeal neuroepithelial tumor [59–61]. It is made up of glioneuronal oligodendrocytes-like cells which infiltrate the meninges and is believed to be a lesion related to the glioneuronal tumors (not yet listed in the WHO classification). MR appearances are very characteristic, with

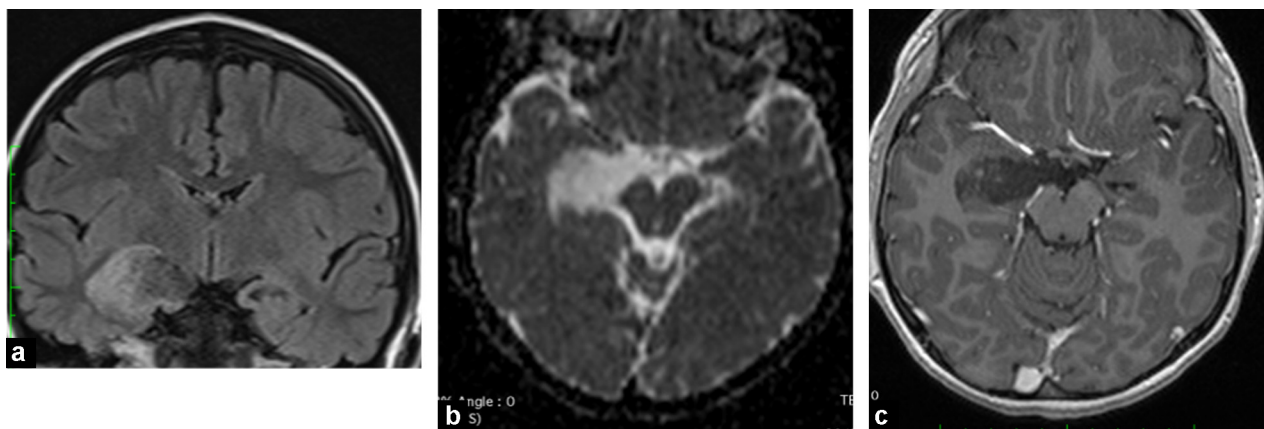


Figure 8. Right internal temporal DNET in a 15-year old child; a: coronal FLAIR image: the lesion displays a hyper- and hypointense image with microcysts present; b: axial ADC map: the ADC of the lesion is extremely high; c: axial T1-weighted image after gadolinium: slight micronodular contrast enhancement in the lesion.

diffuse cerebral and spinal meningeal contrast enhancement associated with multiple cystic lesions within the ventricles and in the cerebral meningeal, particularly cerebellar and spinal, spaces. It often presents with tetraventricular hydrocephalus and headaches, RICP and ataxia. Initial biopsies are often negative with no abnormal cells on lumbar puncture. It grows slowly with a gradual increase in cyst size. Treatment is with a combination of chemotherapy and radiotherapy.

Embryonic tumors

Medulloblastoma

Medulloblastoma is the commonest malignant childhood brain tumor (grade 4) (20–30%) [62], and accounts for 40%

of posterior fossa tumors. Together with PNET and rhabdoid tumors it is one of the highly aggressive embryonic tumors. It is usually seen before the age of 7 years old, with a peak incidence at 4 years old, and is more common in boys.

It is located in the cerebellum in 94.4% of cases, on the midline in 75%. It is a clearly delineated tumor, which develops in the fourth ventricle from the inferior vermis, causing hydrocephalus (Fig. 9) [62]. It may occasionally extend into the foramen of Luschka and the cerebellopontine angle, like an ependymoma. Spinal meningeal tumors are found in 30 to 40% of cases at the time of diagnosis. Five histological subtypes are seen, with or without N-Myc amplification, which carry different prognoses: classical (65–80% of cases), large cell anaplastic (4–5%, very aggressive and

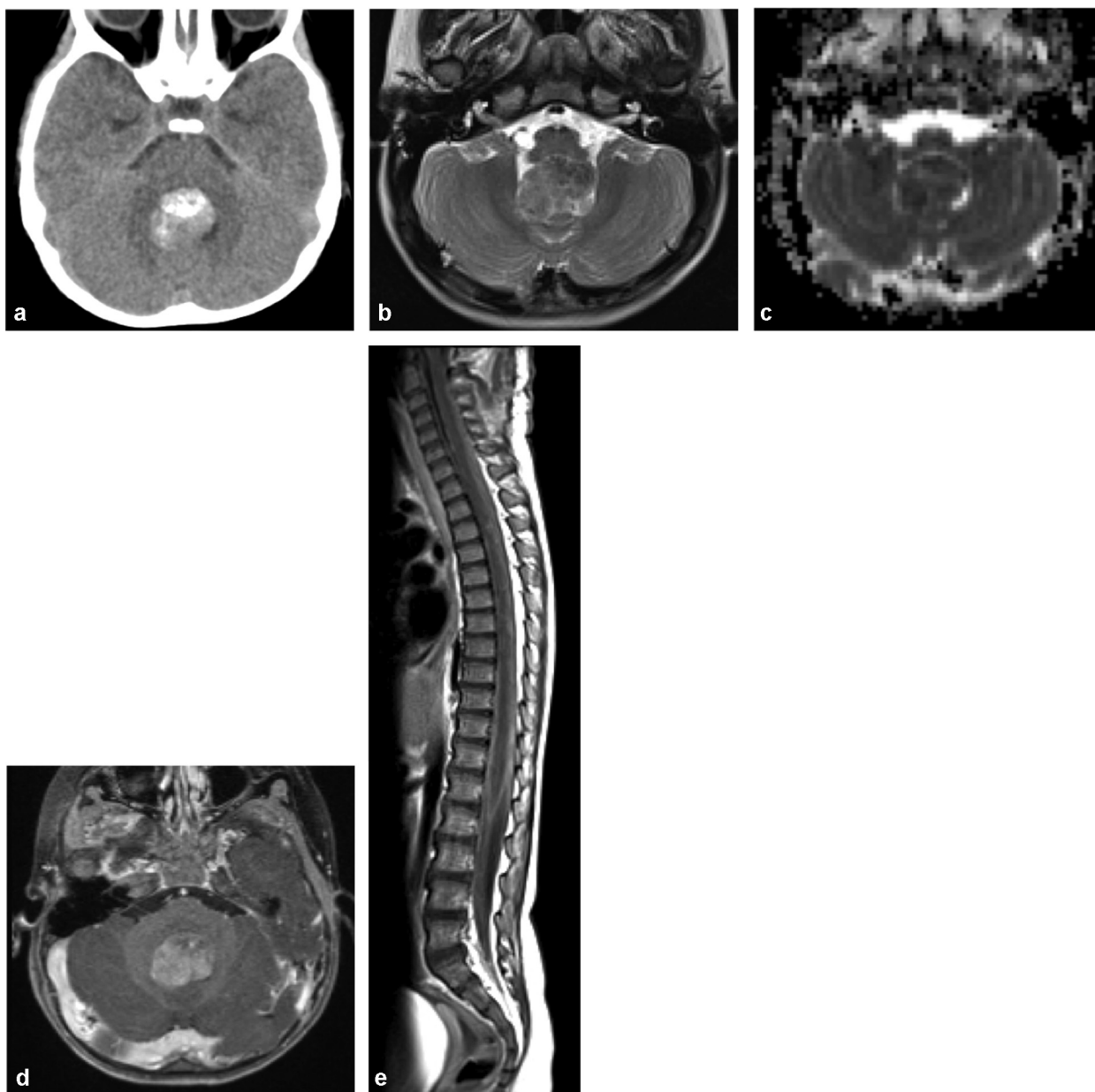


Figure 9. Medulloblastoma in a 2-year-old child; a: axial CT image: hyperdense, partially calcified tumor mass within the fourth ventricle; b: axial T2-weighted MR: the mass displays an iso-hypointense signal to gray matter; c: axial ADC map: the ADC is low due to the hypercellularity of the tumor; d: axial T1-weighted image after gadolinium injection: the lesion enhances intensely; e: spinal MR, sagittal T1-weighted image after gadolinium injection: contrast enhancement around the spinal cord representing leptomeningeal metastases.

metastatic), extensive nodularity (5%, before 1–3 years old with multinodular appearance), and desmoplastic (15–25%, hemispheric, occurring in adolescence and carrying a good prognosis) [35,36].

Medulloblastomas are hyperdense tissue masses on CT because of their hypercellularity [28] and calcifications are seen in 20 to 40% of cases. On MR the lesion appears hypointense on T1-weighted and iso-/hypointense on T2-weighted and FLAIR imaging. Small peripheral cysts (50%) and necrotic areas are commonly seen. Cysts are more common in classical medulloblastoma and desmoplastic medulloblastoma [63]. These enhance strongly with contrast, although the enhancement is occasionally heterogeneous or absent. The most reliable indicator in the diagnosis is the ADC, which is low for both tumor and related meningeal metastases because of the tumor hypercellularity. Occasional cases are seen with a high ADC [64]. MRS shows features of an aggressive tumor, with high choline, lipids and taurine resonances [18]. The rCBV is high on perfusion MR.

Rhabdoid tumor

Rhabdoid tumors or ATRT (atypical teratoid rhabdoid tumors) are rare tumors with a very poor prognosis which are found in children under 3 years old [65]. A characteristic mutation in the *SMARCB1/INI1* gene is seen in 76% of cases, which predisposes to the formation of intra- and extracerebral rhabdoid tumors [5]. They may be found in the posterior fossa (60–70%) and/or in the supratentorial region (hemispheres, pineal region), may be intra- or extra-axial and are often multifocal. These are large calcified, hemorrhagic, necrotic tumors surrounded by extensive edema, with peripheral cysts and meningeal metastases [66]. The commonest site is intra-axial, away from the midline. In extra-axial tumors, the cerebellopontine angle is often involved (Fig. 10). They are heterogeneous tumors which are hyperdense on CT, and hypointense on T2-weighted MR, with variable contrast enhancement. Rhabdoid tumors and medulloblastomas have the same appearances on MR, with a low ADC, although rhabdoid tumors occur in younger children (under 3 years old), affect the cerebellopontine angle

and often contain hemorrhage [67]. Treatment is a combination of surgery and radiotherapy.

Supratentorial tumors are larger in size, more contain cysts, and have central necrosis with a thick layer of peripheral contrast enhancement. These tumors are found predominantly in the frontal lobes and are also less prone to metastasize [68].

Central nervous system (CNS) PNET tumors

These are rare, very aggressive, poor prognosis tumors seen in children under 5 years old. They are located in the supratentorial region or in the brain stem. Five subtypes of PNET are recognized: classical, CNS neuroblastoma, CNS ganglioneuroblastoma, medulloepithelioma and ependymoblastoma [3]. They are large locally invasive tumors and are associated with leptomeningeal metastases in 40% of cases [45]. They are hyperdense and contain calcifications, necrosis, hemorrhage and enhance variably with contrast. The ADC is low and MRS shows signs of an aggressive tumor.

ETANTR (embryonal tumor with abundant neuropil and true rosettes)

ETANTR (embryonal tumor with abundant neuropil and true rosettes) is a specific form of PNET occurring before the age of 4 years old (particularly under two years old) and more commonly in girls. It is predominantly found in the supratentorial region, in the frontal and temporal lobes. They are large, solid, clearly delineated tumors adhering to the dura mater [69], with limited peritumoral edema, although a space-occupying effect is seen. Hemorrhage does not occur, although microcalcifications, cysts and blood vessels are found in the tumor. They do not enhance particularly with contrast, the ADC is low and meningeal metastases are common [70].

Ependymoma

Ependymomas account for 6 to 10% of all tumors and 15% of posterior fossa tumors [71]. Seventy percent of the tumors

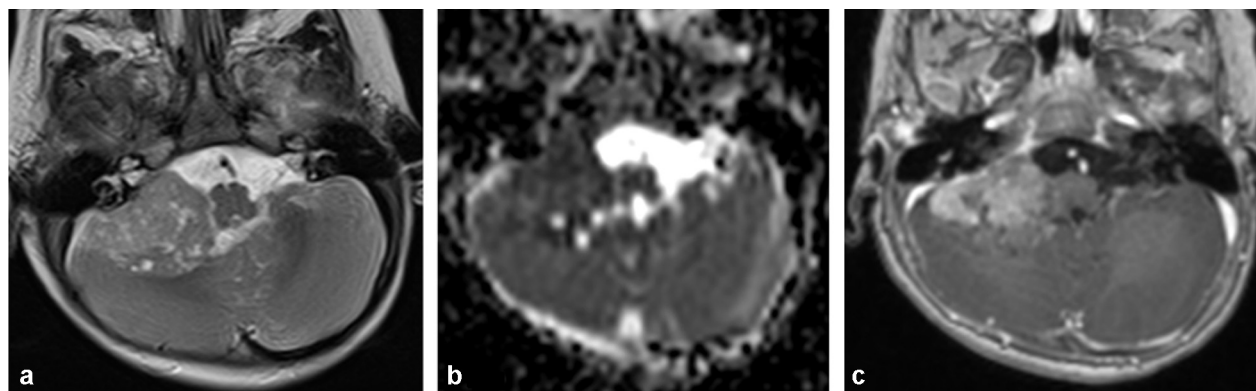


Figure 10. Rhabdoid tumor (ATRT) in a 9-month-old child; a: axial T2-weighted image: heterogeneous tumor lesion in the right cerebellopontine angle, which is iso- and hyperintense on T2-weighted imaging, containing areas of necrosis and cysts within the tumor; b: axial ADC map: the ADC is reduced within the lesion, reflecting its hypercellularity and high grade; c: axial T1-weighted image after gadolinium: heterogeneous enhancement of the lesion.

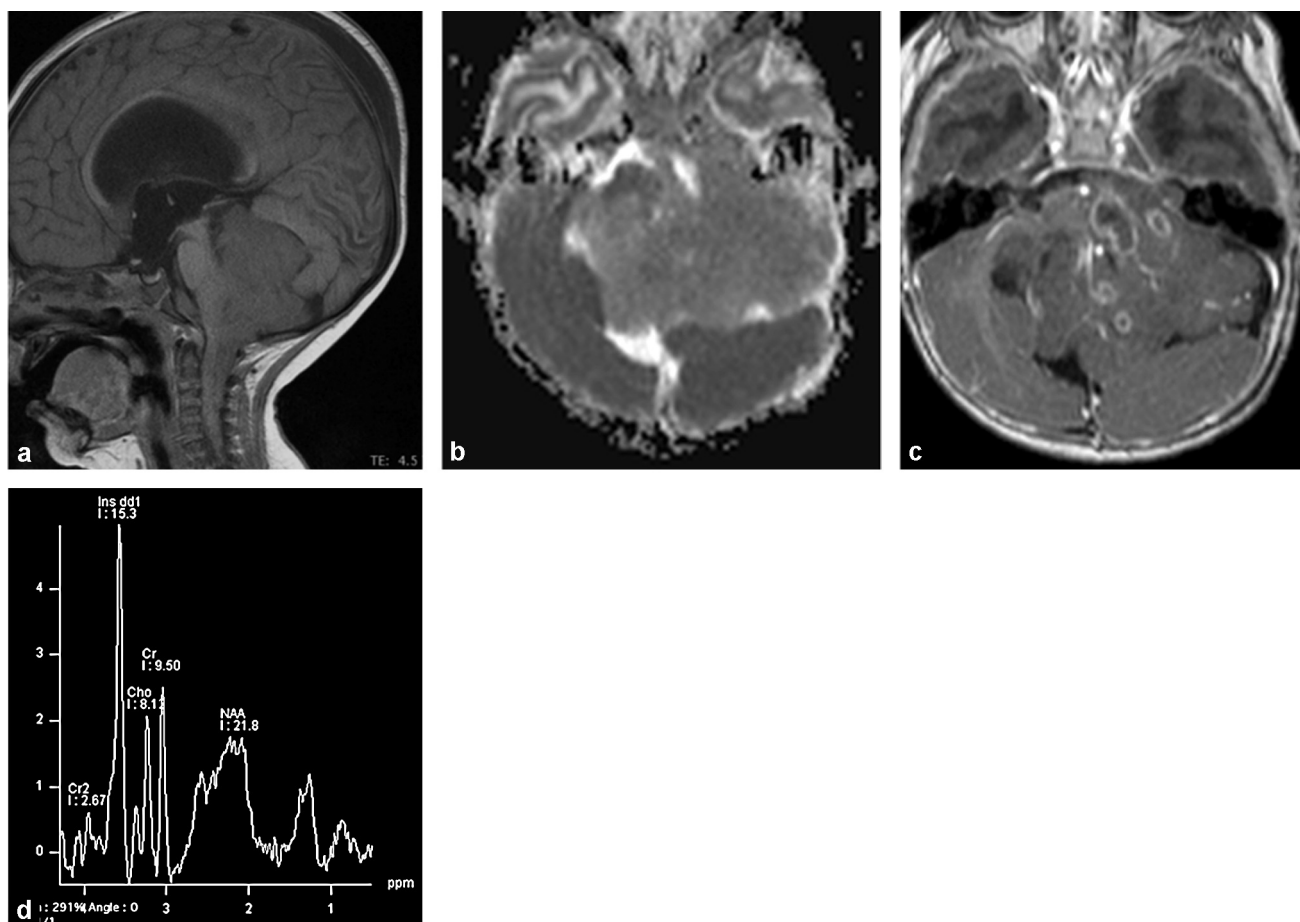


Figure 11. Anaplastic ependymoma in a 4-year-old child; a: sagittal T1-weighted image: tumor mass in the fourth ventricle extending towards the posterior surface of the cervical cord; b: axial ADC map: the tumor mass extends into the left cerebellopontine angle, through the foramen of Luschka. The ADC is intermediary; c: axial T1-weighted image after gadolinium injection: heterogeneous enhancement of the lesion with annular contrast uptake; d: monovoxel MR spectroscopy (PRESS, TE 30): raised myoinositol, low NAA, and lactate present.

are infratentorial [72], in which case, they are malignant (grade 3, anaplastic) and are seen particularly in children under 3 years old. They arise from the ependymal cells of the fourth ventricle, the foramen of Luschka, or from the lateral ventricles. The most characteristic appearance of the tumor is its morphology and extension (Fig. 11), showing a mass growing inside the fourth ventricle extending into the foramina of Magendie and Luschka towards the cerebellopontine angles, the cisterna magna and the spinal canal, surrounding vessels and nerves. Less commonly, the tumor grows from the foramen of Luschka into the cerebellopontine angle. On CT the tumor is iso-/hyperdense. Punctiform calcifications are seen in 50% of cases, with hemorrhages in 10% [72]. MR appearances vary and are heterogeneous, iso-/hypointense on T1-weighted, and hyperintense on T2-weighted and FLAIR imaging, and contain cysts and necrosis. Contrast enhancement is heterogeneous but may be absent [72]. The ADC varies between that of a medulloblastoma and a pilocytic astrocytoma [18]. The rCBV also varies. Choline, myoinositol, glutamate-glutamine and lactate are increased, and NAA is reduced on MRS. Myoinositol is reported to be higher in anaplastic (grade 3) ependymomas [4]. Meningeal metastases are only seen in 10 to 12% of cases at the time of

diagnosis, particularly with anaplastic tumors [36] but are found more commonly during follow-up over time.

Ependymomas are located in the supratentorial region in 30% of cases, when they are often benign (grade 2). Intraventricular localizations (30%) are less common than intraparenchymal tumors (70%), in the parietal or temporo-parietal lobe alongside the ventricular trigone. This is a well-delineated tumor with heterogeneous appearances and cysts. Calcifications are present in a third to 50% of cases [72].

Germ cell tumors

Germinoma

This grade 4 tumor accounts for 1 to 2% of childhood brain tumors [73,74]. The most common site is pineal (57%), and suprasellar (32%), followed by the deep gray nuclei (9%). It tends to affect boys between 10 and 18 years old. Bifocal suprasellar and pineal tumors are seen in 15% of cases. It presents with signs of raised intracranial pressure, visual disturbance or ataxia. CT shows a hyperdense calcified mass.

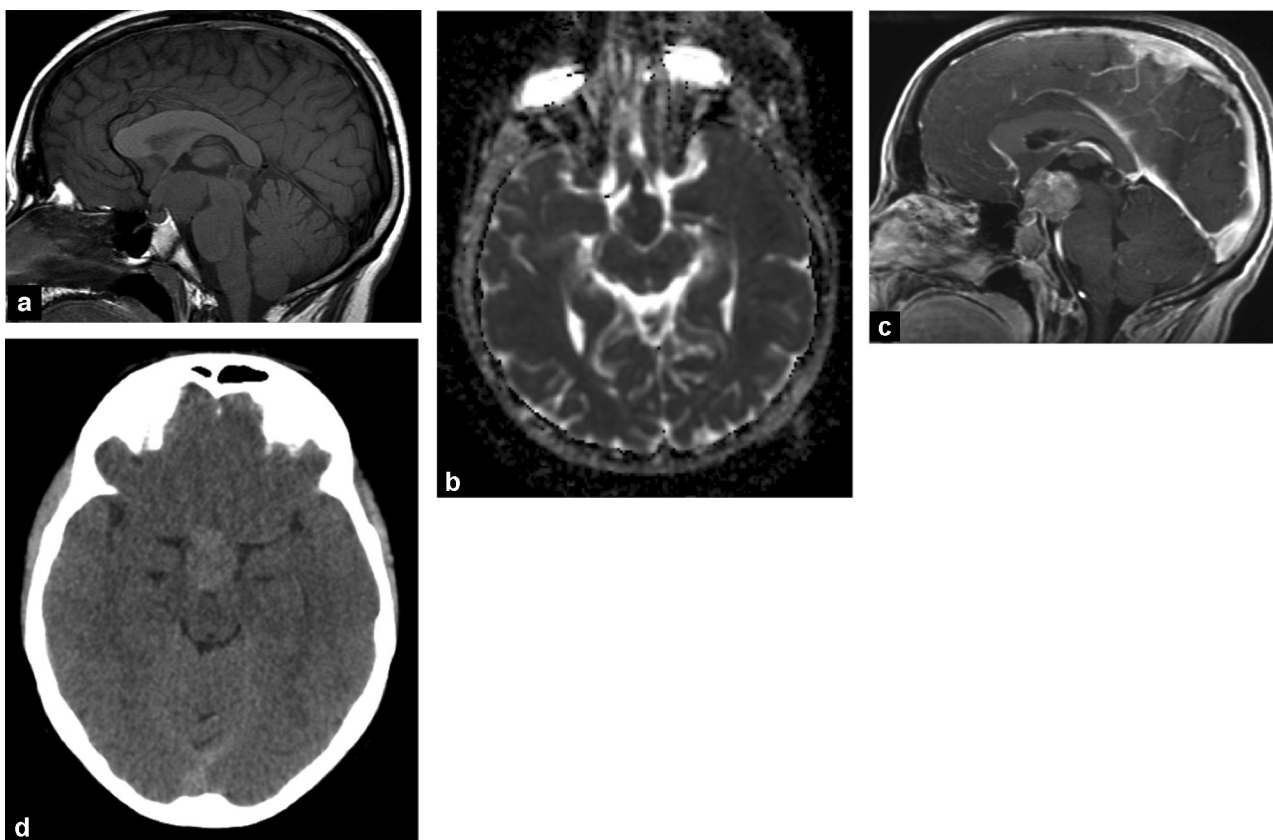


Figure 12. Suprasellar germinoma in a 16-year-old child; a: sagittal T1-weighted image: the tissular mass has developed in the suprasellar region and interpeduncular cistern. It is isointense on T1-weighted imaging. The normal T1 signal hyperintensity in the posterior pituitary is not seen; b: axial ADC map: The ADC is low, unlike the pilocytic astrocytoma, which has a high ADC; c: sagittal T1-weighted image after gadolinium injection: heterogeneous enhancement in the lesion; d: unenhanced CT: the lesion is hyperdense on the unenhanced image because of its hypercellularity.

Appearances are isointense/hypointense on T1-weighted, and iso-/hyperintense on T2-weighted imaging. Cystic or necrotic hemorrhagic areas may be seen, particularly in large tumors. The ADC of the soft tissue component varies and is the same (55%), less (36%), or greater (9%) than that of the normal cerebral parenchyma [75]. They generally enhance intensely with contrast. Meningeal metastases may occur and spinal investigations are therefore required. They have a very good prognosis because of their excellent radio- and chemo-sensitivity. Treatment includes chemotherapy, which is very effective, followed by radiotherapy for any residual tumor.

Suprasellar germinoma (Fig. 12) presents with diabetes insipidus, and visual and pubertal disturbances. The tumor is only found 3 to 6 months after the onset of diabetes insipidus, when MR is normal or shows only slight thickening of the pituitary stalk. Hemorrhage and necrosis in the tumor occur more often than with the pineal tumors, and no calcifications are present [73]. The low ADC and iso-/hypointense T2-weighted appearances of the lesion distinguish it from other suprasellar tumors.

The tumor is located more often in the deep gray nuclei in the Asian population [2,76]. It develops in the caudate and lenticulate nuclei and thalami, although this occasionally reflects thalamic extension of a pineal tumor through the walls of the third ventricle [43]. It carries a poorer prognosis than other sites and is an initially

infiltrating tumor, the larger ones of which are more circumscribed.

Non-germinomatous germ cell tumors

These are the teratomas (10%), choriocarcinomas, endodermal sinus tumors and embryonic carcinomas or mixed tumors [2].

Teratomas

Teratomas are seen in children under 10 years old. This is also the most common neonatal tumor (over 50%) and is located in the pineal region, although may also be seen in the hypothalamic or suprasellar regions, or in the cerebral hemispheres [77]. They range from grade 0 tumors (mature tissue) to grade 3 (immature tissue). The mature forms contain fat, cysts, calcifications and soft tissue components. They enhance heterogeneously with contrast, particularly in the tissular parts of the tumor. The ADC is increased and lipids are seen on MRS [78]. The immature, malignant forms of the tumor have less clearly demarcated edges and edema around the tumor, although are impossible to distinguish formally from mature tumors, unless meningeal spread is present [5].

Germ cell tumors may secrete β -hCG and alpha-fetoprotein into blood and CSF, allowing a positive,

unequivocal diagnosis to be made, which occasionally avoids surgery in order to obtain histology [74].

Pineal tumors

These account for 3 to 11% of supratentorial childhood tumors [2]. Germ cell (50–80%) (cf. section “Germ cell tumors”), pineal parenchymal (15%) and neuroepithelial tumors are seen [79]. They present with headaches and hydrocephalus.

Pineoblastoma

Pineoblastoma accounts for 40% of pineal parenchymal tumors. This is a grade 4 tumor often associated with meningeal metastases and may coexist with retinoblastoma. The appearances of these lesions on imaging are similar to the PNET [2], with a very low ADC, distinguishing them from a germinoma, which has a comparatively higher ADC [73].

Papillary pineal gland tumor

Papillary pineal gland tumor is difficult to distinguish from a pineoblastoma, although it is reported to have a higher signal intensity on T2WI [74].

Suprasellar tumors

These account for 40% of all intracranial tumors and generally present with visual and endocrine disturbances. They include craniopharyngiomas, optic tract gliomas (cf. section “Pilocytic astrocytoma”) and germinomas (cf. section “Germ cell tumors”). All of these tumors may have mixed solid and cystic appearances on imaging. The ADC and MRS may help to distinguish them.

Craniopharyngioma

This is a grade 1 tumor seen around the age of 4/5 years old in children and accounts for 5 to 10% of childhood brain tumors and 50% of suprasellar masses [35,80]. It presents with visual disturbance, headaches and endocrine abnormalities (growth and weight retardation) and develops from residues of the Rathke’s pouch or craniopharyngeal canal [81]. It is found particularly in the suprasellar region and is less commonly intrasellar in the region of the hypothalamus and optic chiasma, and may extend to the anterior and middle cerebral fossae. The most typical tumor in children is the adamantinomatous form, whereas in adults the squamous papillary form is more common. The most typical appearances are those of a calcified, suprasellar cyst. Appearances are hypointense or hyperintense on T1-weighted and hypointense in T2-weighted imaging (lipids, proteins or blood) and hyperintense on FLAIR imaging, the wall enhancing with contrast. A tissue component may be present, which enhances heterogeneously. The tumor may surround the adjacent blood vessels. On MRS, lipids and lactate are found inside the cyst, distinguishing it from an optic tract astrocytoma. The ADC is

high. It is important to recognize cystic tumors, which are treated locally (for example, with bleomycin). Solid tumors are rarer and mixed tumors are common. Surgery is offered when the lesion is distant to the hypothalamus (type 0), or in contact with the hypothalamus (type 1). If the tumor and hypothalamus are indissociable (type 2), partial resection is offered, combined with radiotherapy, in children over 5 years old [82]. Recurrences are common.

Hypothalamic hamartoma

This is a malformation consisting of well-differentiated neurones and glial cells, similar to normal hypothalamic tissue [24]. It is characterized by attacks of gelastic seizures or precocious puberty. Two forms are seen: intra-hypothalamic (sessile, invading the mamillary bodies and the tuber cinereum, epilepsy) or para-hypothalamic (pedunculated and inserted beneath the roof of the 3rd ventricle, associated with precocious puberty) [24]. Appearances are the same as those of the gray matter on T1WI, and are slightly hyperintense on T2WI, occasionally with cysts but not enhancing with contrast.

Choroid plexus tumors

Choroid plexus papilloma is a benign tumor which develops from the choroid plexuses and is seen in the first two years of life. It is located in the lateral ventricles (trigone) in 80% of cases (Fig. 13), in the 4th ventricle in 16% of cases and in the 3rd ventricle in 4% of cases [5]. Sites close to the foramen of Monro are only seen in 4% of cases [83]. CSF production by

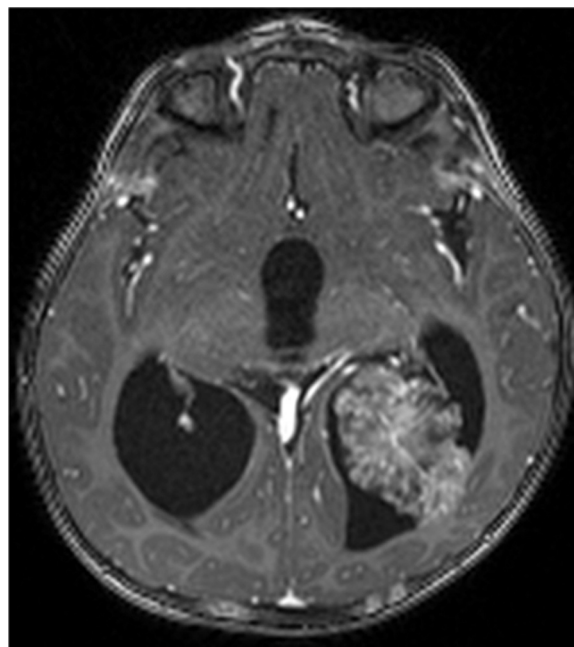


Figure 13. Choroid plexus papilloma in a 6-year-old child. Axial T1-weighted image after gadolinium: lobulated lesion at the trigone of the left ventricle, enhancing with gadolinium, and containing multiple vascular structures. Hydrocephalus is present due to the overproduction of CSF by the tumor.

the tumor explains the hydrocephalus which occurs. Choroid plexus papillomas, which are benign grade 1 tumors, are five times more common than the atypical grade 2 papillomas and grade 3 carcinomas [84]. The papilloma appears as a multilobular iso-/hyperdense mass on CT. It is isointense on T1-weighted and iso-/hypointense on T2-weighted MR, occasionally with a more heterogeneous appearance due to the presence of necrotic or hemorrhagic cystic areas [5]. It enhances intensely with gadolinium. Ectatic vessels are seen inside the tumor. The ADC is slightly reduced and the rCBV is high, occasionally with blood-brain barrier disruption [45]. Carcinomas invade the neighboring parenchyma and are associated with peritumoral edema, which may, however, also be present in a benign papilloma [85].

Conclusion

There is a very wide histological range of childhood brain tumors, although the most common are medulloblastomas, astrocytomas and ependymomas. Analysis of the site of the lesion and its CT and MR appearances can provide a preoperative diagnosis of the type and grade in the majority of cases. The histological classification of tumors has been constantly evolving since it was created. Future classifications will incorporate genetic and molecular findings, distinguishing subtypes associated with different prognoses, with more targeted individualized treatments. Imaging, particularly using advanced techniques, will have an important role to play to try and identify new biomarkers, which correlate with these biological subtypes.

TAKE-HOME MESSAGES

- Childhood brain tumors are primary intra-axial tumors, which are equally common in the posterior fossa and supratentorial regions.
- Histological findings are extremely varied but 80% are accounted for by gliomas (pilocytic astrocytoma+++), medulloblastoma and ependymoma.
- Diagnosis is based on the child's age, site of the tumor and its CT and MR appearances.
- Imaging: an urgent cerebral CT is performed if signs of raised intracranial pressure are present in order to plan surgery (with and without enhancement, diagnosis of a mass, calcification, hemorrhage and density).
- Imaging: cerebral MR is part of the preoperative assessment and should include the spinal canal in posterior fossa tumors, or those close to the midline, looking for meningeal spread.
- Cerebral MR should include T1, T2, T2*, diffusion-weighted, FLAIR, T13D gadolinium (or 2 planes), and, if possible, MR spectroscopy and perfusion MR.

Clinical case

A 4-year-old child presented with left VI nerve palsy. The child's cerebral MR images are shown on Figs. 14–16.

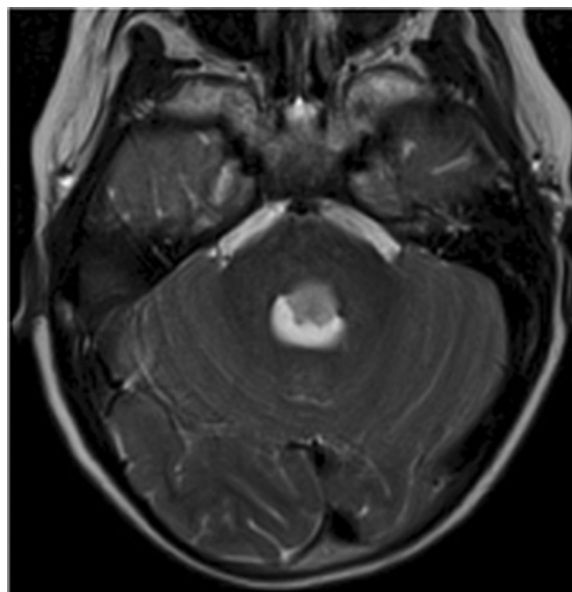


Figure 14. DP-T2SE axial image.

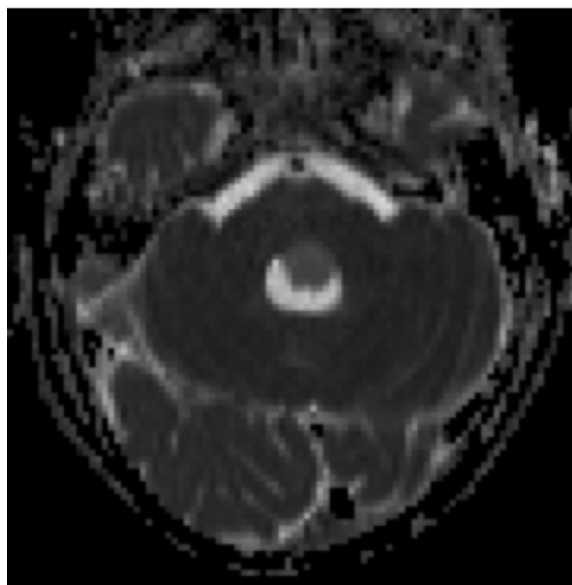


Figure 15. Axial image, ADC mapping.

Questions

1. What diagnosis would you consider for the 4th ventricle tumor?
2. What are the distinctive features of each of these diagnoses?
3. Which tumors carry risks of meningeal spread?
4. What is the specific feature of the tumor in this child and what is your diagnosis?
5. What are its MR spectroscopy and perfusion MR appearances?

Answers

1. Medulloblastoma, pilocytic astrocytoma, ependymoma.

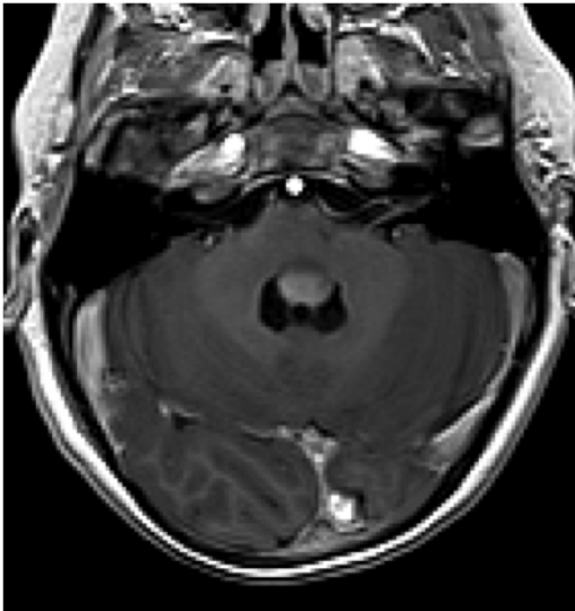


Figure 16. Axial T1-weighted image after gadolinium injection.

2. Medulloblastoma: a solid hyperdense tumor on CT, iso-/hypointense on T2WI, with a very low ADC, reflecting its hypercellular nature and, therefore, high grade. Pilocytic astrocytoma: a solid cystic mass with enhancement of its tissular component. The tissue area appears hypodense on CT and hyperintense on T2WI with a high ADC, reflecting its hypocellularity and, therefore, low grade. Ependymoma: a tumor with characteristic extension into the foramina of Magendie and Luschka, sheathing the vessels and nerves.
3. Ependymoma and medulloblastoma.
4. This tumor is located within the posterior wall of the pons. It is hyperintense on FLAIR imaging, enhances with gadolinium and has a raised ADC. This is an exophytic pilocytic astrocytoma of the brain stem in the fourth ventricle.
5. The perfusion MR of the pilocytic astrocytoma shows a low rCBV and the perfusion curve goes over the baseline reflecting blood-brain barrier disruption. On MR spectroscopy, choline is increased with reduced NAA and lactate present, simulating a malignant tumor.

Disclosure of interest

M. Koob declares that she has no conflicts of interest concerning this article.

N. Girard: consulting with OLEAMEDICAL.

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