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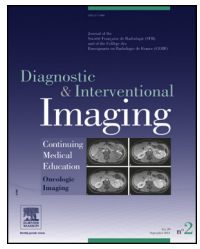

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Cerebral tumors: Specific features in children

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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.

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	Embryonic tumor
		Malignant glioma
		Ependymoma
Deep gray nuclei		Malignant glioma
		Germinoma
Intraventricular		Choroid plexus papilloma
		Subependymal giant cell astrocytoma
Suprasellar		Craniopharyngioma
		Optic tract glioma
		Germinoma
		Hypothalamic hamartoma
Pineal		Germinoma
		Pineoblastoma
		Papillary pineal gland tumor

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