Infratentorial tumors diagnosed by MRI in pediatric patients

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Learning objectives

To describe the most common infratentorial tumors in children, primarily the posterior fossa tumors, according to the epidemiology, and morphological features underlining specific imaging findings to have the best diagnostic clue for each differential diagnosis: Pilocytic Astrocytoma, Medulloblastoma, Ependymoma and Stem glioma.

Background

The MRI is considered, by far, the best image tool to describe and have the best characterization of the pediatric neuropathology. In dispite of the high cost, and longer times this diagnostic method continue being preferred because of non radiation exposure for young patients.

As we know central nervous system neoplasms are the leading cause of cancer death in children, in which solid tumors are describe as more frequent.

This group of tumors are much more heterogeneous that the ones described in adults, that is why the importance of recognize, and describe each type on early stage take palace on timely diagnosis and treatment. Pilocytic astrocytoma, medulloblastoma, ependymoma, stem glioma are described in this article.

PILOCYTIC ASTROCITOMA: (PAs)

Is the most common in pediatric patients. Sometimes is also known as "juvenile pilocytic astrocytoma", is a well-circumscribed, typically slow-growing glioma. More than 80% occur in patients under 20 and there is no gender predilection.

There is a syndromic association with neurofibromatosis type 1 (NF1), 15% of NF1 develop PAs.

The cerebelum is the most common localization nearly 60%, but may arise anywhere. The second most common site is in anda round the optic nerve, chiasma and hipotalamus.

Typically are well delineated soft tumors that often form intramural cysts, and neoplastic elemente confined to the mural tumor nodule.

PA is a WHO grade 1 tumor, dissemination is rare.

Imaging finding varies with the PA location, the most common is cerebellar cyst with mural nodule, may compare the nodule to a bubblegum center surrounded by caramel.
MR Findings: Cystic PA are usually well delineated and appear slightly hyperintense on T1 and T2W1. Do not suppress completely on FLAIR. The mural node is iso/hypointense on T1W1 and iso/hyperintense on T2W1. Solid PAs appear iso- or o hypointense to parenchyma on T1W1 and hypeintense on T2 FLAIR. They may show striking enhancement following contrast administration, intense but heterogeneous enhancement of the solid potion. Cyst walls occasionally enhances. A varianty pattern is a solid mass with central necrosis and thick peripherally enhancing of tumor. MRS agressive appearing metabolite pattern: high choline, low NAA, high lactate.

MEDULOBLSTOMA: (MB)

Is the most malignant CNS neoplasma of childhood and the second most common overall peditric brain tumor.

There are four main molecular subgroups, each arising from different cytogenetic pathways. Group 3 is the largest, are he most common in infant, exhibit nearly 2:1 M:F predominante, frecuently metastasize, and have the worst outcome of all.

Classic MB is a primitive neuroectodermal tumor (PNET) that arise in the cerebellum. The 85% arise in the midline, they are located within the forth ventricle and focally infiltrate the dorsal brainstem. Posterior extention into the cisterna magna is common.

CMB have reatively defined margins on imaging studies. Despite this appearance 40-50% have CSF dissemination at the time of inicial diagnosis, thats why the MR Imaging of entire neuraxis is recommended. Up to a tirad have subarachnoid metastático disease at presentation. Image pre op to avoid false (+) post op: blood in spinal canal may mimic or mask metastases.

MR Findings: Almost all CMB are hypointense relative to gray matter on T1W1 and hyperintense on T2W1. Peritumoral edema is present in one-third of the cases. Obstructive hydrocephalus with transependymyal CSF migration is common and best delineated on FLAIR.

Enhancement patterns show striking variation, ranking from minimal to patchy to marked. Because of their dense cellularity this tumors often show moderate restriction. T1C > 90% enhance often heterogeneous. Contrast-enhanced MR of spine (entire neuraxis). MRS NAA very low, high elevation choline, lactate usually present.

EPENDYMOMA

They are solitary neoplasms. Size varies, but most supratentorial ependymonas are large bulky neoplasms, lobulated masses that extrude through the lateral recesses of fourth ventricle. They are described as "plastic" tumors like "taffy look "appearance. Obstructive
hydrocephalus is frequent accompanying feature, extracellular fluid often accumulates around the ventricles living the appearance of "blurred" margins.

Approximately 60-70% are infratentorial. 95% are found in fourth ventricle.

Ependymomas are WHO grade II neoplasms.

This kind of tumor accounts for approximately 10% of CNS neoplasms in children and 30% of all brain tumors in children under the age of three years. Ependymoma is the tirad most common posterior fossa tumor of childhood.

The mean age at presentation is between four and six years, and there is male predominante of 57%.

Dissemination is a key factor in staging, prognosis and treatment. The Imaging of the entire cranial- spinal axis should be performed in any child with a posterior fossa neoplasms, especially if medulloblastoma or ependymoma is suspected, thus the preoperative imagering predictor of a patient outcome is evidence of tumor spread.

MR findings: Ependymoma are generally heterogenously relative to brain parenchyma. T1 Heterogeneous usually iso to hypointense, cystic foci slightly hyperintense to CSF. Hyperintense Ca ++, and blood products.

T2W1 heterogenous, usually iso to hyperintense. Hyperintense cyst foci, hypointense Ca ++, blood. FLAIR can show sharp interface between the tumor and CSF, tumor cysts very hyperintense to CSF. T2*GRE "Blooming" of hypointense Ca ++ foci and old hemorrhage. DWI most ependymomas do not restrict, in some cases foci of restrict difusión can be identified.

T1 C most ependymomas enhance moderate heterogenous MRS NAA reduce, choline high, lactate peak are common as in many other tumos. Perfusion MR generally demonstrates markedly elevated cerebral blood volumen with poor return to baseline, spectroscopy alone does not reliably for differential diagnosis from astrocytoma.

STEM GLIOMA

They constitute between 10-15% of all chilhood brain tumors and are the main cause of death in this group. Can involve the midbrain, pons or medulla. This kind of tumor are astrocytomas but the histologic subtypes and patient outcome is remarkably variable.

The stem glioma is an infiltrative neoplasms generelly of poor prognosis, may as low grade or anaplastic (WHO grade II or even IV).

Imaging features vary on each type of tumor as in location.
The most representative used to be expansive involving the pons with compression but not invading the fourth ventricle. They often anteriorly exophytic engulf the basilar artery territory. T1 low signal, T2W1 are variably hyperintense, FLAIR high signal. T1C variable enhancement. MRS NAA levels are higher in diffuse pontine glioma in patients with NF1 than without, this association course with less malignant prognosis. The intensity of the enhancement and the restriction on DWI is a predictive factor of poor prognosis.

**Images for this section:**

![MR images on T1WI, T2WI, FLAIR, T1 post-gadolinium shows a large medulloblastoma expanding the fourth ventricle (a-e). DWI (f) with characteristic restricted diffusion.](image)

**Fig. 1:** MR images on T1WI, T2WI, FLAIR, T1 post-gadolinium shows a large medulloblastoma expanding the fourth ventricle (a-e). DWI (f) with characteristic restricted diffusion.
Fig. 2: Spinal subarachnoid metastatic disease of medulloblastoma secondary to CFS dissemination. T1WI post gadolinium (a-c)
**Fig. 3:** Classic ependymoma. MR images on T1WI, T2WI, FLAIR, T1 post gadolinium (a-d) with a heterogeneous mass arising from the floor of the fourth ventricle, expanding through Lushka foramina into cerebellopontine angle. DWI (e) shows no restriction. Maximum slope of decrease perfusion color map (f) with marked increased in rCBV.
Fig. 4: Pilocytic Astrocytoma. MR images on T1WI, T2WI, FLAIR, T1 post gadolinium (a-d) shows a well-circumscribed cystic cerebellar mass with enhancing mural nodule. Content of cyst is hyperintense on FLAIR. Perfusion color map (e) shows moderate elevation in rCBV. MRS (f) shows aggressive-appearing metabolite pattern, despite a low histological grade (WHO I).
Fig. 5: Diffuse pontine glioma. MRI shows pontine enlargement that compress fourth ventricle with exofitic ventral component that partially engulfs basilar artery. Some central necrotic areas with faint enhancing are seen.
Findings and procedure details

GE Signa 1.5 T TwinSpeed Excite (GE Medical Systems, Milwaukee, WI, USA) scanner using an 8 element phased array surface coil. For tissue characteritation protocol included sistematically a axial, sagital and coronal T1SE, T2FSE, STIR and fat saturation T1SE post gadolinium injection.

Conclusion

Learning about the most common infratentorial tumors in children, their epiemiology, and natural history enable us a better understanding and correlation of the imaging findings. The importance of early diagnosis of the posterior fossa tumors take place on timely diagnosis and treatment.

The neuroimaging assessments, in this escenario MR, performed with the full spectrum modalities, play an important role in the characterization and description of these neoplasms, allowing an accurate diferencial diagnosis.

Personal information

References