Pediatric Brain Tumors

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Overview

- Pediatric brain tumors (PBT) are 15-20% of all brain tumors.
- Second most common pediatric tumor. Only leukemia more common.
- Overall, supratentorial and infratentorial tumors occur in equal frequency.
  - Supratentorial more common <2yrs; infratentorial more common 4-10yrs; equally common after 10yrs age.
- Clinical Presentation:
  - Infants-Increasing head circumference, lethargy, nausea and vomiting.
  - Children-Also may have headaches, ↓visual acuity, seizures, cranial nerve palsies, endocrine dysfunction.
Imaging

- CT can be used for initial screening.

- MRI is superior and essential if CT finds abnormality or inconclusive.
  - MR spectroscopy can be useful: Elevated Choline and depressed NAA peaks (higher choline, more likely tumor to be high grade).
Classification

- Posterior fossa

- Supratentorial
  - Intraparenchymal
  - Sellar/Suprasellar
  - Extra-axial
Intraventricular

Cerebral hemisphere

Sella/
Suprasellar

Post. fossa

Brain stem
Classification

Posterior fossa

- Astrocytoma
- Medulloblastoma
- Ependymoma
- Brainstem glioma

Dermoid/Epidermoid

Schwannoma/Meningioma (NF2)
Classification

Supratentorial

- Intraparenchymal
  - Astrocytoma
  - Ependymoma
  - Desmoplastic neuroepithelial tumor (DIG)
  - Dysembryoplastic neuroepithelial tumor
  - Ganglioglioma/Gangliocytoma
  - Teratoma
  - Primitive Neuroectodermal Tumor (PNET)
  - Atypical teratoid/rhabdoid tumors
Classification

Supratentorial

- Extra-axial
  - Choroid plexus papilloma/carcinoma
  - Langerhans cell histiocytyosis
  - Epidermoid/Dermoid
  - Arachnoid cyst
  - Metastasis
Classification

- Supratentorial
- Sellar/Suprasellar
  - Craniopharyngioma
  - Astrocytoma
  - Rathke cleft cyst
  - Germ cell tumors
  - Hypothalamic hamartoma
  - Langerhans cell histiocytosis
  - Pituitary adenoma
Infratentorial Tumors

Astrocytoma (Juvenile Pilocytic)

- Most common pediatric brain tumor: 40-50% of intracranial neoplasms
  - 60% located in posterior fossa
  - M=F
  - Typically occurs between 5-15 yrs of age.
  - Clinical findings: Early morning headaches, vomiting (worsens over time). Ataxia, papilledema
  - 25yr survival rate of 90% following successful surgery
Infratentorial Tumors

Astrocytomas (Juvenile Pilocytic)

Imaging findings

- Cystic lesion with enhancing mural nodule
- Heterogeneous enhancement in more solid tumors
- Calcification in 20%
- Location: Typically cerebellar hemisphere
- Hydrocephalus
- Paradoxical MR spectroscopy findings: appears aggressive although low grade
Juvenile pilocytic astrocytoma
Juvenile pilocytic astrocytoma

Post contrast T1

Post contrast T1
Infratentorial Tumors
Medulloblastoma (PNET)

- Highly malignant, undifferentiated tumor
- 15-20% of pediatric brain tumors
- Occur in 4-11yr age group and peaks at age 5
- Males 2-4 times more likely to be affected
- 1/3 have subarachnoid mets; drop mets to spine (40%)
- Intraventricular tumor from roof of 4th ventricle (superior medullary velum), vermis
- Midline in children; Laterally located in older children, adults
Infratentorial Tumors
Medulloblastoma (PNET)

- Clinical findings: Short duration of symptoms with nausea, vomiting, headaches, ataxia
- Increasing head size, lethargy in children less than 1 yr of age
- Nondisseminated: 60-70% 5yr survival
- Disseminated: 36% 5yr survival
Infratentorial Tumors
Medulloblastoma (PNET)

- Imaging findings:
  - CT: Midline hyperdense mass of cerebellar vermis extending to 4th ventricle.
  - CT/MRI: Heterogeneous enhancement.
  - Calcifications in 20%.
  - Hydrocephalus.
  - Essential to image brain and spine for mets!!
Medulloblastoma
Medulloblastoma

T2

FLAIR

Medulloblastoma

T1
Medulloblastoma
Infratentorial Tumors
Ependymoma

- ~10% of primary brain tumors in children
- 70% are infratentorial/30% supratentorial
- Slight increased incidence in males
- 2 age peaks: 1-5yrs and ~35yrs
- Location: Vermis, floor of 4\textsuperscript{th} ventricle extending into ventricle
- “Plastic” tumor that conforms to ventricle and extrudes through Foramen of Luschka
- Clinical findings: Headaches, vomiting, ataxia
- 60-70% 5yr survival
Infratentorial Tumors
Ependymoma

Imaging findings:
• 4th ventricle tumor that may extend through FOL
• Calcifications common (50%)
• Hydrocephalus
• CT/MRI: Heterogeneous enhancement
• Gradient echo images demonstrate blooming from calcifications
Ependymoma

T1  T2

Ependymoma
Post contrast T1

Ependymoma
Infratentorial Tumors

Brainstem glioma

- 15% of pediatric CNS tumors and 20-30% of infratentorial tumors
- 80% high grade: 20% low grade
- M=F
- Typically between 3-10yrs of age
- Location: Pons>>midbrain>medulla
- 4 types: Diffuse, focal, exophytic, cervicomedullary
  - Prognosis and treatment depend on type with diffuse having poor prognosis, focal better prognosis
- BG can be confused with brainstem encephalitis
Infratentorial Tumors

Brainstem gliomas

- Tectal gliomas are subtype with better prognosis
- Brainstem “tumors” related to NF1 nonaggressive
- Clinical: Hydrocephalus, cranial nerve VI and VII palsies
Infratentorial Tumor
Brainstem gliomas

Imaging findings:

- Enlargement of brainstem, posterior displacement 4th vent
- T2, FLAIR hyperintensity
- 50% of BG enhance; heterogeneous enhancement
Brainstem glioma
MR spectroscopy

T1  T2  PG T1

Normal control voxel  Lesion voxel

Courtesy of G. Barest, M.D.
Brainstem glioma
Post contrast T1
Supratentorial Tumors

Astrocytoma

- 30% of hemispheric tumors. Most common cerebral hemispheric tumor
- Peak incidence at 7-8 yrs age
- Slight male predominance
- Low grade astrocytomas more common
- Glioblastoma Multiforme (GBM) – WHO IV/IV ~ 20%
- Typically involve basal ganglia, thalamus
- Can be multi-centric
- Clinical: Seizures, focal neurologic deficits, headaches
Supratentorial Tumors
Astrocytomas

Imaging findings:

- Can appear like JPA.
- Solid tumors have variable degrees of enhancement.
- Pilocytic astrocytomas on T2, FLAIR signal but minimal surrounding edema.
- Low grade: Little to no enhancement.
- High grade: Heterogeneous with areas of necrosis.
Astrocytoma

Post contrast
T1

Astrocytoma
Supratentorial Tumors

Other less common astrocytomas:

- **Subependymal Giant cell (SGCA):**
  - Associated with tuberous sclerosis
  - Low grade
  - Arises near foramen of Monro
  - Hydrocephalus

- **Pleomorphic Xanthoastrocytoma (PXA):**
  - Similar to Desmoplastic Infantile Ganglioglioma (DIG); low grade
  - Peripheral cerebral hemisphere
  - Typically in adolescents, young adults
Tuberous Sclerosis

T2

Post contrast T1
Supratentorial Tumors

Ependymoma

- 30% of ependymomas
- Peak incidence between 1-5 yrs age
- Histologically similar to infratentorial ependymomas
- Typically in periventricular white matter and NOT intraventricular (metastatic spread uncommon)
- Clinical: Increased intracranial pressure, ataxia, seizures
- 5 yr survival: 80% w/total resection
  40-60% w/subtotal resection
Supratentorial Tumors

Ependymoma

Imaging findings:

- Variable appearance on CT or MRI
- Slightly hyperdense on CT with calcifications (50%)
- Heterogeneous enhancement with cystic areas
- Typically seen in frontal lobe
Supratentorial Tumors
Desmoplastic Infantile Ganglioglioma

• Arises from subpial astrocytes
• Found between 1-24 months age w/peak at 3-6 months. Occasionally seen from 5-17yrs
• M/F: 1.7/1
• Cortically based tumor nodule
• Clinical: Head size, bulging fontanels, seizures
• Greater then 75% survival after 15yrs w/complete resection
Supratentorial Tumors
Desmoplastic Infantile Gangliogioma

Imaging findings:

- Large cyst with cortically based enhancing tumor nodule
- Enhancement of adjacent dura
- Occupies majority of cerebral hemisphere
- Looks worse than it is!!
Desmoplastic infantile ganglioglioma
Supratentorial Tumors

Dysembryoplastic Neuroepithelial Tumor (DNET)

• Benign tumor of cerebral cortex
• Cause of 20% cases of medically refractory epilepsy
• 60% in temporal lobe, 30% frontal lobe
• Solid and cystic tumors
• Scalloping of inner table skull
• Associated w/cortical dysplasia
• Slow to No growth!
Supratentorial Tumors

DNET

Imaging findings:

- Multicystic cortically based lesion
- Solid components
- Temporal and frontal lobes most common
- Approx. 25% have calcifications
- Enhancement in 20-40%
- Characteristic “bright rim” on FLAIR
- Wedge shaped appearance
Supratentorial Tumors
Ganglioglioma/Gangliocytoma

- Mixed neuronal-glial tumors
- 3% brain tumors in children
- Found in adolescents
- Associated with mesial temporal sclerosis
- Most common in temporal, parietal, frontal lobes
- Difference between GG and GC is histological
- Clinical: Partial complex seizures
Supratentorial Tumors
Ganglioglioma/Gangliocytoma

Imaging findings:
- Solid or cystic or cyst
  w/mural nodule
- Variable enhancement
- 35% w/calcifications
- If peripheral location,
  then scalloping of
  adjacent calvarium
Ganglioglioma

GRE

Post contrast T1

Ganglioglioma
Ganglioglioma
Supratentorial Tumors

Teratoma

- 2-5% of tumors in children less than 15
- Midline lesion typically in pineal gland, third ventricle
- More common in males
- Most are benign
- Clinical: Hydrocephalus
Supratentorial Tumors

Teratomas

Imaging findings:

• Midline mass with calcifications and fat
• Enhancement of soft tissue components
• Malignant teratomas have more vasogenic edema, irregular, less well defined
CT

Teratoma
Teratoma

T1

T2

Post contrast T1

Teratoma
Supratentorial Tumors

Extra-axial

- Choroid plexus papilloma/carcinoma
- Langerhans cell histiocytosis
- Epidermoid/Dermoid
- Arachnoid cyst
- Metastasis
Supratentorial Tumors
Choroid plexus papilloma/carcinoma

- Arise from epithelium of choroid plexus
- 5% of supratentorial tumors
- Typically age 1-5yrs
- Male predominance
- CP carcinomas are 30-40% choroid plexus tumors
- Most common in trigone of left lateral ventricle
- CPC more irregular and invasive then CPP but diagnosis is histological
- Clinical: Hydrocephalus
Supratentorial Tumors
Choroid plexus papilloma/carcinoma

Imaging:
- CPP is lobulated, homogeneous mass
- Punctate calcifications, hyperdense on CT
- Intense enhancement on CT/MRI
- CPC irregular, heterogeneous w/cystic necrosis, invasive and vasogenic edema
Choroid plexus papilloma
Choroid plexus papilloma

CT

CT with contrast

Choroid plexus papilloma
Choroid plexus papilloma

T2 Post contrast T1
Choroid plexus carcinoma

Post contrast T1
Supratentorial Tumors
Langerhans cell histiocytosis

- LCH disorder of reticuloendothelial system
- Rarely involves CNS
- In calvarium, expansile erosive soft tissue mass w/”beveled edges” and intense enhancement
- Intracranially, similar to gray matter?????
- MRI: hypointense on FLAIR and T2; intense enhancement
Supratentorial tumors
Langerhans cell histiocytosis
Imaging findings:

CT
Langerhans cell histiocytosis
Supratentorial Tumors
Epidermoid/Dermoid

T2
FLAIR
Post contrast T1
Dermoid
Supratentorial Tumors

Sellar/Suprasellar

- Craniopharyngioma
- Astrocytoma
- Rathke cleft cyst
- Germ cell tumor
- Hypothalamic hamartoma
- Langerhans cell histiocytosis
- Pituitary adenoma
Common Presentations of Hypothalamic and Pituitary Lesions

- Hypopituitarism – craniopharyngioma
- Diabetes insipidus – LCH, GCT, cranio
- Precocious puberty – hamartoma of tuber cinereum, hypothalamic glioma
- Amenorrhea – pituitary adenoma, Rathke cleft cyst
Supratentorial Tumors

Craniopharyngiomas

- Thought to arise from remnant of craniopharyngeal duct
- Adamantinomatous (children) and papillary (adults) types
- 15% supratentorial tumors, 50% suprasellar tumors
- 2 peaks: 10-14 yrs age; 4th to 6th decade of life
- Most common in males
- Clinical: Headaches, visual disturbances, diabetes insipidus
- Diff. Dx.: Rathke cleft cysts, hemorrhagic pituitary adenomas
Supratentorial Tumors
Craniopharyngiomas

**Imaging findings:**

- Suprasellar
- Cystic and solid; 90% are calcified
- Rim enhancement of cysts; heterogeneous enhancement solid portions
Craniopharyngioma
Supratentorial Tumors

Rathke Cleft cyst

- Benign epithelial lined cyst in sella
- Arises from remnants of Rathke pouch
- Arises in pituitary gland with frequent suprasellar extension
- Rare in children

Clinical:
- Typically asymptomatic
- Symptomatic patients present with headaches, pituitary dysfunction
Supratentorial Tumors
Rathke cleft cyst

Imaging findings:

- CT: Cystic, nonenhancing, noncalcified mass
- MRI: T1 or T2 hyperintense. No enhancement
Supratentorial Tumors

Astrocytoma

- Suprasellar type arises from optic chiasm or hypothalamus
- Many tumors in optic nerve are JPA
- M=F
- Presents from 2-4yrs age
- 30-50% have family history of NF1
- Clinical: Hydrocephalus, decreased vision, pituitary dysfunction (short stature)
Supratentorial Tumors

Astrocytoma

Imaging findings:

- MR is imaging tool of choice. T2, FLAIR hyperintense
- Fusiform or lobulated enlargement optic nerves
- Heterogeneous enhancement
- Gliomas in patients without NF1 have cystic components with heterogeneous enhancement
Astrocytoma
NF1, Optic glioma
Post contrast T1

NF1, Optic glioma
Supratentorial Tumors
Germ cell tumors

- Originate in hypothalamus and extend into infundibulum
- Germinoma most common histologic type.
- 35% suprasellar; 60% pineal
- M=F in suprasellar germinomas (10:1 in pineal region tumors)
- Clinical: Typically present w/diabetes insipidus
  - If child presents w/DI but imaging negative, 2 repeat studies at 6 month intervals should be performed!!!! Initial germinoma may be too small for visualization!!!!
Supratentorial Tumors
Germ cell tumors

**Imaging findings:**

- MR: Infundibular thickening w/uniform enhancement
- Iso to hypointense on T1, T2 and FLAIR
- When large, can have areas of cystic necrosis
- *REMEMBER: HIGH ASSOCIATION W/DIABETES INSIPIDUS!!.*
Post contrast T1

Germ cell tumor
Supratentorial Tumors
Hypothalamic Hamartoma

- Heterotopic gray matter generally located in tuber cinereum
- Can originate from floor third ventricle, mamillary bodies
- Can be sessile or pedunculated
- Presents between 1-3 yrs age; M=F
- Large lesions cause gelastic seizures; small lesions have precocious puberty
  - Found in 33% of patients w/precocious puberty
  - Treatment is hormonal therapy
Supratentorial Tumors
Hypothalamic Hamartomas

Imaging findings:
• MR: Isointense on T1, slightly hyperintense on T2
• Non-enhancing
• Can be 2mm to 4cm in size
Post contrast T1

Hypothalamic hamartoma
Supratentorial Tumors
Langerhans cell histiocytosis

- Most common manifestation of LCH
- CNS presentation is more common in patients with multi-systemic disease
- Granulomas in the subarachnoid space which infiltrate hypothalamus/infundibulum
- Clinical: Diabetes insipidus (5% of patients on diagnosis and up to 50% on follow-up exams)
Supratentorial Tumors
Langerhans cell histiocytosis

Imaging findings:
- MR: Mild thickening of infundibulum to large hypothalamic mass
- Intense enhancement with contrast

Post contrast T1
References

• Osborn AG. In: Diagnostic Imaging Brain. Ed. Elsevier Saunders, 2004
Thank You.