“Small Round Blue Cell Tumors”
(in kids)  Prepared by Kurt Schaberg

Leukemia/Lymphoma

Always a consideration! Do several heme markers and show a hematopathologist.

Ewing Sarcoma

Malignant tumor of neuroectodermal differentiation that is often arises in the bone (but can see in many organs; Chest wall = Askin tumor)

Often have EWSR1 translocation (with FLI-1 or ERG)  t(11;22)

Usually uniform, small, round, blue cells with sheet-like to lobular, growth pattern with variable necrosis

Strong, membranous CD99 staining
(Sensitive, but not Specific staining)

Cytoplasmic glycogen stains with PAS

Rhabdomyosarcoma

Malignant tumor with primary skeletal muscle differentiation, several types

Stain with Desmin, MyoD1, Myogenin

Embryonal Rhabdo:

Variable numbers of round ("rhabdoid"), strap-, or tadpole-shaped eosinophilic rhabdomyoblasts in a myxoid stroma

Can see cytoplasmic cross striations

Alveolar Rhabdo:

Larger, more rounded undifferentiated cells with only occasional rhabdomyoblasts

Often arranged in an alveolar (nested) pattern

Distinctively strong and diffuse myogenin positivity

Characteristic FOXO1 translocations

Wilms Tumor  aka nephroblastoma

Malignant tumor originating in Kidney

3 key elements: 1) Primitive epithelial tubules, 2) Blastema (sheets of small high N:C ratio cells), 3) Stroma

In some cases may only see 2 (or possibly even 1) element

3 component stain differently with IHC stains

Epithelium: ⊕ WT-1, CK
Blastema: ⊕ WT-1, Desmin
Stroma: Weak WT-1, (plus heterologous elements)
For all pediatric tumors, consider in addition to Formalin-Fixed Tissue:
1) Flow cytometry, 2) Cytogenetics, 3) Freezing some (depends on quantity, etc..)

### Neuroblastoma
- Most primitive/aggressive
- Malignant
- SRBCT +/- rosettes, neurofibrillary matrix

Peripheral neuroblastic tumors derive from the sympathetic nervous system (therefore develop anywhere along the distribution of the sympathoadrenal neuroendocrine system)
Positive stains: synaptophysin, chromogranin, CD56, NB84, and neuron-specific enolase
Staining for S-100 protein has been used to identify cytodifferentiated cells such as Schwann cells
MYCN amplification → Poor prognosis!

### Desmoplastic Round Cell Tumor
- Malignant tumor of uncertain histogenesis often found in the peritoneal cavity; often in young men
- Characteristic EWSR1 – WT-1 translocation
- Basaloid nests of tumor that are surrounded by cellular desmoplastic stroma
- Stains: Positive CK, Desmin, WT-1 (but C-terminus—opposite of Wilms!)

### Ganglioneuroblastoma
**Most primitive/aggressive**
**Malignant**
**SRBCT +/- rosettes, neurofibrillary matrix**

Intermediate differentiation
Neuroblastoma + Ganglion cells

Most mature; Benign Ganglion cells set in fibrillary stroma
**NO** neuroblastoma

### Ganglioneuroma
**Most mature; Benign**
Ganglion cells set in fibrillary stroma

### Table

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