

“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

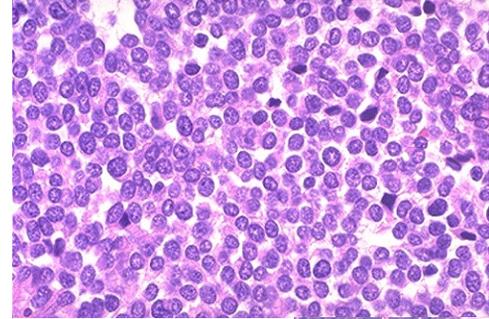
Malignant tumor of neuroectodermal differentiation that is often arises in the bone (but can see in many organs; Chest wall = *Askin tumor*)

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

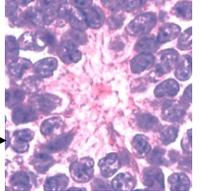
Strong, membranous CD99 staining

(Sensitive, but not Specific staining)

Cytoplasmic glycogen stains with PAS



Can see pseudorosettes

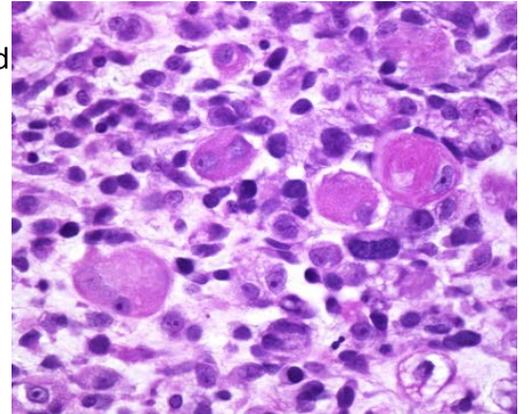


Rhabdomyosarcoma

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

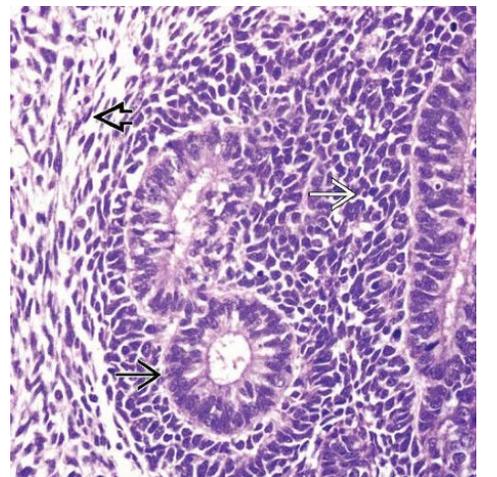
In some cases may only see 2 (or possibly even 1) element

3 component stain differently with IHC stains

Epithelium: ⊕ WT-1, CK

Blastema: ⊕ WT-1, Desmin

Stroma: Weak WT-1, (plus heterologous elements)



Neuroblastoma

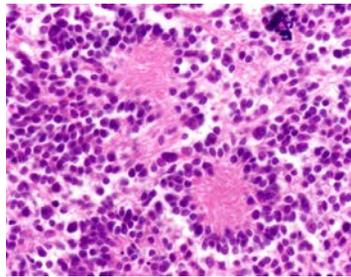
Maturing →

Ganglioneuroblastoma

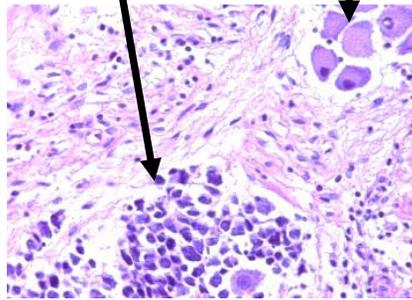
Maturing →

Ganglioneuroma

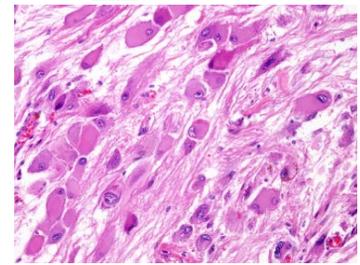
Most primitive/aggressive
Malignant
SRBCT +/- rosettes,
neurofibrillary matrix



Intermediate differentiation
Neuroblastoma + Ganglion cells



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



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!

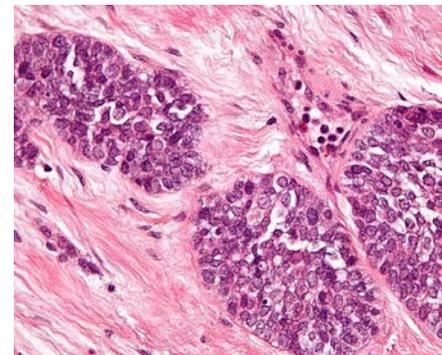
Desmoplastic Round Cell Tumor

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!)



For all pediatric tumors, consider in addition to Formalin-Fixed Tissue:

1) Flow cytometry, 2) Cytogenetics, 3) Freezing some (depends on quantity, etc..)

	CD45	TdT	CK AE1/AE3	Desmin	MyoD1 Myogenin	Synapto. Chromo	CD99	WT-1
Leukemia/ Lymphoma	+	+/-	-	-	-	-	-/+	-
Ewing Sarcoma	-	-	Usu. -	-	-	-	+	-
Rhabdomyosarcoma	-	-	-/+	+	+	Rare	-/+	-
Wilms' Tumor	-	-	+	-/+	-	-/+	-	+
Neuroblastoma	-	-	-	-	-	+	-	-
Desmoplastic Round Cell Tumor	-	-	+	+	-	-	-/+	C-terminus