Myogenic Embryonal Neoplasms in a Child/Adolescent

If very bland with large cells, could this be rhabdomyoma?

Desmin / Myogenin +

But also think of other tumors with myogenic cells: MPNST, Pleomorphic sarcoma, Epithelioid sarcoma, Etc...

Rhabdomyosarcoma

+ Cyto-Molecular Genetics FOXO1 (FISH, RT-PCR) and or aCGH −

Fusion-positive Alveolar Rhabdomyosarcoma

- PAX7-FOXO1 t(1;13) 20-25%
  - COG data suggests slightly improved survival with this fusion partner
- PAX3-FOXO1 t(2;13) 55-65%
  - COG data suggests worse overall survival with this fusion partner

OUTCOMES

87% overall survival	64% overall survival

Fusion-negative RMS - Morphologic patterns

Polygonal / Irregular	Solid, uniform round cells	Round cells within hyalinized stroma	Thin cells & stromal desmoplasia
Embryonal	Alveolar	Sclerosing	Spindle

Maybe these should just be called "Fusion-negative RMS"?

- Dense
- Solid
- Alveolar spaces

These might be real Fusion-negative RMS subtypes

- Infants: VGLL2-fusion
- Micro-alveolar & organoid growth - Hyaline stroma
- Spindled fibromatosis-like or Fibrosarcoma-like pattern
- Non-infants: MYOD-1 or PI3KCA mutation

MIXED HISTOLOGIES

OUTCOMES

Generally better than fusion+ RMS Certain sites (paratestis, distal extremities) carry better treatment options for complete surgical resection -> better EFS	Infants with VGLL2 ScS-RMS have very good prognosis Older kids with MYOD-1 mutated Sc-S RMS have poor outcomes