Small Round Blue Cell Tumors

Lymphoma Tumor Board
November 6, 2015
Small round blue cell tumors

- Lymphoma
- Neuroblastoma
- Rhabdomyosarcoma
- Ewing sarcoma/primitive neuroectodermal tumor (PNET)
- Desmoplastic small round cell tumor
- Poorly differentiated synovial sarcoma
- Small cell osteosarcoma
- **DDx: Wilm’s tumor**
## Small round blue cell tumors - Immunohistochemistry

<table>
<thead>
<tr>
<th>Tumor type</th>
<th>Immunohistochemical markers</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lymphoma</td>
<td>CD45; CD30 (HD, ALCL); CD20 (B-NHL); CD3 (T-NHL)</td>
</tr>
<tr>
<td>Neuroblastoma</td>
<td>NSE (neuron-specific enolase); S100</td>
</tr>
<tr>
<td>Rhabdomyosarcoma</td>
<td>Desmin, myosin, MyoD</td>
</tr>
<tr>
<td>Ewing sarcoma/PNET</td>
<td>PAS+ (Glycogen); NSE; CD99</td>
</tr>
</tbody>
</table>
# Small round blue cell tumors - Cytogenetics

<table>
<thead>
<tr>
<th>Tumor type</th>
<th>Cytogenetic/FISH/molecular markers</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lymphoma</td>
<td><em>e.g.</em>, t(8;14) MYC/IGH; t(2;5) NPM/ALK</td>
</tr>
<tr>
<td>Neuroblastoma</td>
<td>MYCN amplification; added material 17q</td>
</tr>
<tr>
<td>Rhabdomyosarcoma</td>
<td>Chromosomal translocations involving 13</td>
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<tr>
<td>Ewing sarcoma/PNET</td>
<td><em>EWSR1</em> gene rearrangement, <em>e.g.</em> <em>EWSR1</em>/FLI1 gene fusion (90%)</td>
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</tbody>
</table>
Age-Adjusted and Age-Specific Cancer Incidence Rates for Patients 0-19 Years of Age (SEER 2005-2009)

**0-14 Years**
- 31.1% Leukemia
- 25.4% CNS
- 6.6% Soft tissue
- 4.4% Bone tumors
- 4.0% Carcinomas and Melanoma
- 3.4% Germ cell
- 2.5% Retinoblastoma
- 1.5% Hepatic
- 10.0% Lymphomas
- 0.3% Other

**15-19 Years**
- 19.6% Carcinomas and Melanoma
- 20.7% Lymphomas
- 13.8% Leukemia
- 12.3% Germ cell
- 6.8% Soft tissue
- 6.2% Bone tumors
- 0.6% Hepatic
- 0.7% Renal
- 0.4% Neuroblastoma
- 0.3% Other
- 18.7% CNS
Geographic origin of neuroblastoma