Rhabdomyosarcoma

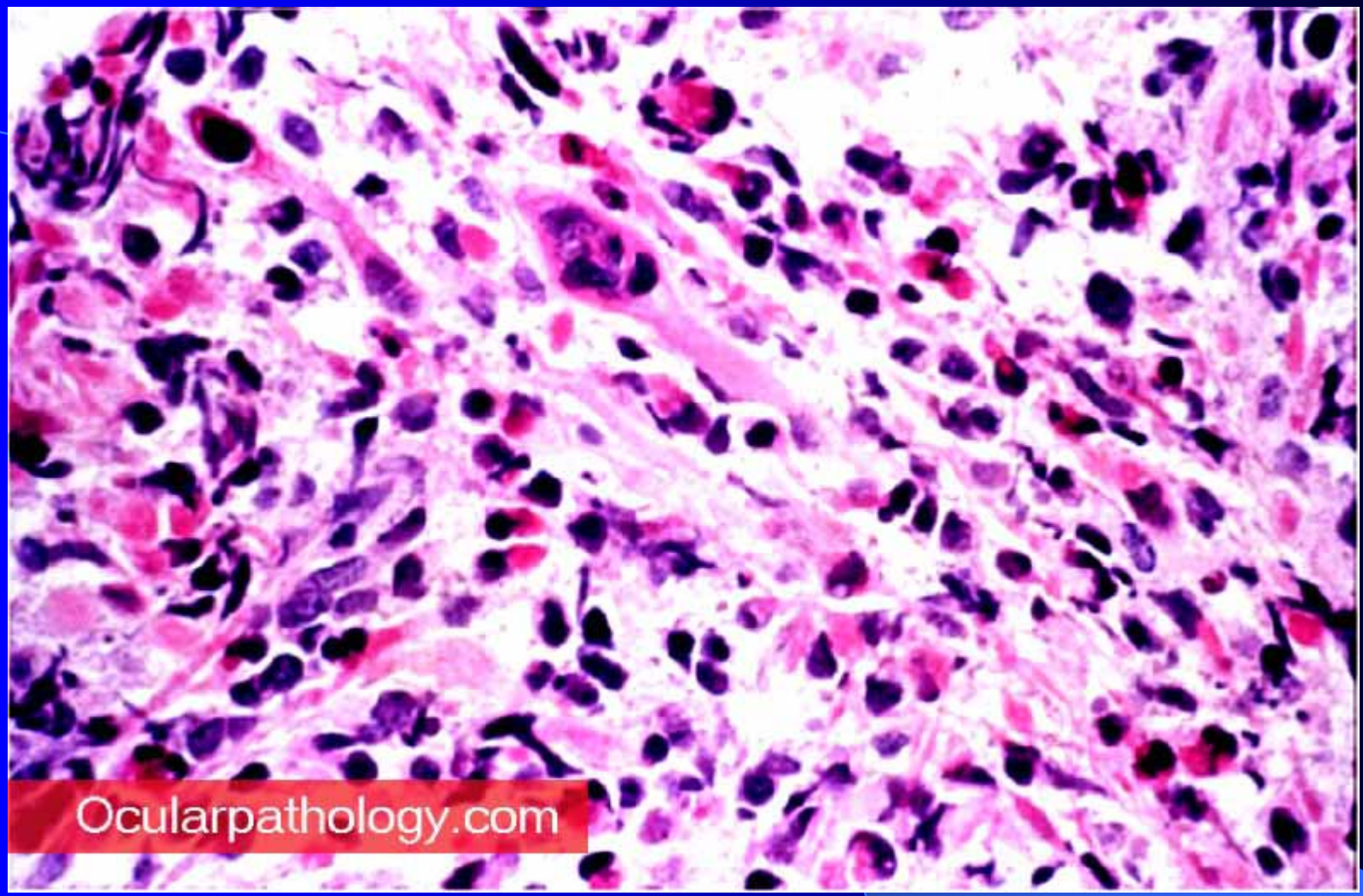
By Andrew Pelser

Background

- Most common soft tissue sarcoma in childhood.
- USA 250 Dx per year.
- Arise from rhabdomyoblast.
- Located through out body
- Four major sites: H+N 35-40%
- GU 20%
- Extremities 15-20%
- Trunk 10-15%

Histological Classification

- **Embryonal-** Most common type 60-70%; H+N and GU tract; most treatable form; >6years.
- **Botryoid type-** Variant of Embryonal; 10%; grape like lesions of mucosal lined hollow organs; vagina and bladder.
- **Alveolar type-** More aggressive; 20%; muscles of extremities; older children.
- **Pleomorphic type-** Adults; extremities; in children called anaplastic


Ocularpathology.com

Causes???

No recognised predisposing factor or risk factor- small proportion associated with genetic conditions.

Rhabdomyoblast- primitive cells in foetus that develop into striated muscle cells.

Embryonal- abnormality chromosome 11

Alveolar- rearrangement of chromosome material between chr 2 and chr 13 better known as a "fusion transcript". Two genes involved PAX3 and FKHR. Also mention of PAX7

More commom in children with neurofibromatosis/ Li-Fraumeni syndrome, Costello syndrome.

No association with environmental exposures.

Symptoms

- Location dependant
- Size dependant
- May include: Tumour or mass felt or seen.
  Bleeding.
  Tingling, numbness, pain if nerve compression.
  Protrusion of overlying structure.

Diagnosis

- Medical Hx and Physical Ex.
- Bloods / Urine / Sputum.
- Biopsy.
- Multiple imaging: Most useful MRI and bone scan for bony metastasis.
- Bone marrow aspiration.
- Lumbar puncture: parameningeal tumours
- Goal: Staging and Classification

TNM Staging System

- Stage I: Area near eye, H+N, GU(not prostate and bladder) and localized.
- Stage II: < 5cm any site not stage I, no lymph nodes.
- Stage III: >5cm any site not stage I and/or to lymph nodes.
- Stage IV: Metastasis to other areas.

Staging

- Complex
- **1 Grouping System** (I-IV): Status postsurgical resection/biopsy
- **2 Staging System**: Sites of primary, Tumour size,Grouping, Regional LN, Distant mets (1-4)
- **3 Risk Group**:(Low, intermediate, high) Grouping, Staging and Histology

Treatment

- Guide lines: Intergroup Rhabdomyosarcoma Study(IRS).
- Surgery- Removal if total is possible.
- ChemoTx- Alone or in conjunction
- RadioTx- Brachytherapy new development
- Stemcell Transplant: Allogenic/ Autologous

Surgical H+N

- **Parameningeal tumors**:? Skull base ?Skull erosion ? Transdural extension= MRI/CT/CSF
- **Superficial and nonorbital**: Wide excision and ipsilateral neck dissection.
- **More complex/recurrent locoregional Dx**: Multidisciplinary surgical team AFTER chemo/radio
- **Unresectable/orbital**: Chemo Tx Radio Tx

Prognosis

- **Age**: <1 poor, 1-9 good
- **Site of origin**: Orbit, nonparameningeal H+N, paratestis, vagina
- **Resectability**: <5cm
- **Presence of metastases and sites**
- **Lymph node involvement**
- **Histopathology**: Embryonal good, Alveolar poor