Rhabdomyosarcoma

- Skeletal muscle neoplasms are almost all malignant
- rhabdomyoma is the only benign skeletal muscle neoplasms
- more frequent in individuals with tuberous sclerosis
- Rhabdomyosarcoma is a malignant mesenchymal tumor with skeletal muscle differentiation.

Three main subtypes are of Rhabdomyosarcoma

- Alveolar (**20%**), Embryonal (**60%**), and Pleomorphic (**20%**).
- alveolar and embryonal most common soft tissue of childhood and adolescence, before age 20
- Pleomorphic rhabdomyosarcoma is seen predominantly in **adults**
- The pediatric forms (alveolar / embryonal) arise in sinuses, head and neck, and genitourinary tract

Embryonal rhabdomyosarcoma

- soft gray infiltrative mass. The tumor cells mimic skeletal muscle at various stages of differentiation
- Rhabdomyoblasts with straplike cytoplasm and visible cross-striations may be present.

Sarcoma botryoides

- develops in the walls of hollow viscera such as the urinary bladder and vagina.
- Best Prognosis , Death due to Direct Extension

alveolar rhabdomyosarcoma

- a network of fibrous septae creating a crude resemblance to pulmonary alveoli
- fusions of the FOXO1 gene to either the PAX3 or the PAX7 gene, rearrangements marked by the presence of (2;13) or (1;13) translocations, respectively
- PAX3 is a transcription factor that initiates skeletal muscle differentiation

Pleomorphic rhabdomyosarcoma

- characterized by numerous large, sometimes multinucleated, bizarre eosinophilic tumor cells that can resemble other pleomorphic sarcomas
- Rhabdomyosarcomas are aggressive neoplasms / pleomorphic subtype is often fatal.
- The botryoid variant of embryonal rhabdomyosarcoma has the best prognosis (highest chance of survival)

Synovial Sarcoma

- these tumors can present in locations that lack synovium.
- this name is a misnomer, as these tumors can present in locations that lack synovium and their morphologic features are inconsistent with an origin from synoviocytes.
- Most occur in people in their 20s to 40s.
- Patients usually present with a deep-seated mass (present in the body before symptoms) that has been present for several years.
- chromosomal translocation producing fusion genes composed of portions of the SS18 gene and one of three SSX genes
- lung and regional lymph nodes are site of metastasis

MORPHOLOGY

Monophasic synovial sarcoma

- consists of uniform spindle cells with scant cytoplasm and dense chromatin growing in short, tightly packed fascicles.
- Hemangiopericytic vessel/ staghorn, dilated vessels

The biphasic

- glandlike structures composed of cuboidal to columnar epithelioid cells in addition to the aforementioned spindle cell component.
- Immunohistochemistry to identifying these tumors
- the biphasic type in Immunohistochemistry are positive for epithelial antigens (e.g., keratins)