Glandular Lesions and Tumors in Uropathology

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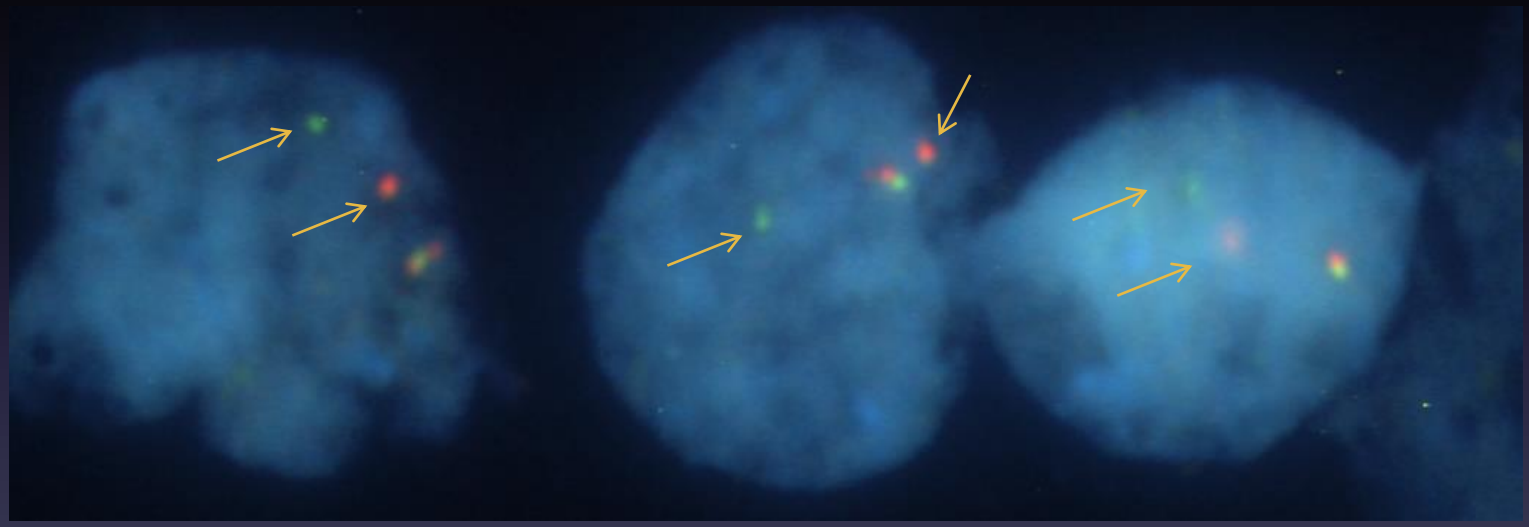
– *Videomicroscopy Case #5* –

History

- 64 year-old male patient
- Complaint of unilateral testicular enlargement
- Physical Exam: Swelling of right testis
- Scrotal USG:
  - 6 cm mass lesion occupying right testis
- Past Medical History: Not significant
- Inguinal orchiectomy
  - 5.8 cm tumor in the largest diameter
    - Intratesticular
    - Smooth contoured with pseudocapsule
    - Cream to pale yellow colored
    - Areas of hemorrhage

FISH

(18q11.2) Dual Color, break Apart Rearrangement Probe



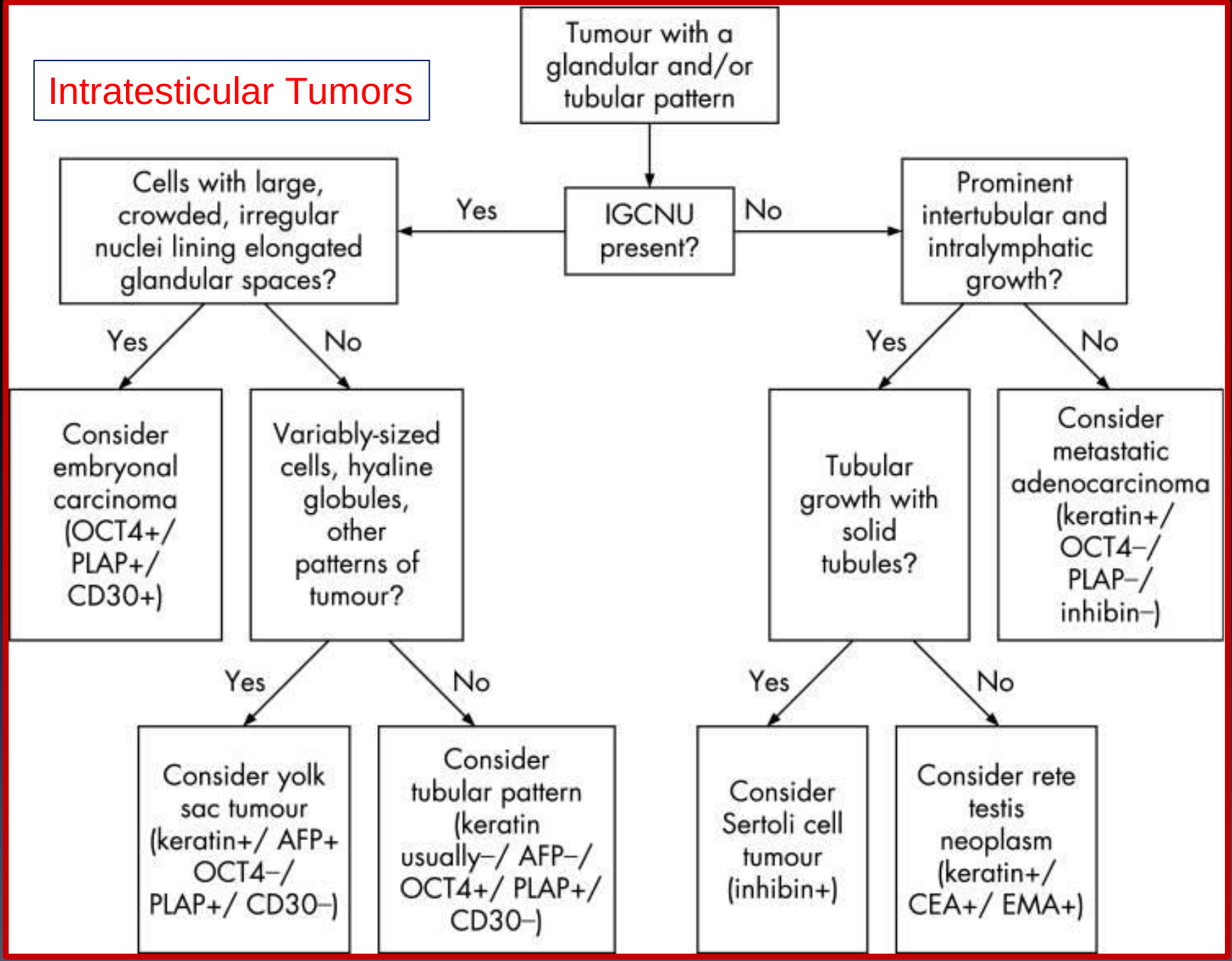
break apart of SYT gene

Glandular Lesions of Testis

- Intratesticular
  - Sertoli cell lesions (hamartomas or tumors)
  - Germ cell tumors with glandular pattern
    - Yolk sac tm, Embryonal carcinoma, Teratomas
  - Metastases
- Paratesticular
  - Rete testis proliferations
  - Epididymal lesions
  - Ovarian type surface epithelial lesions
  - Mesothelial proliferations
  - Metastases

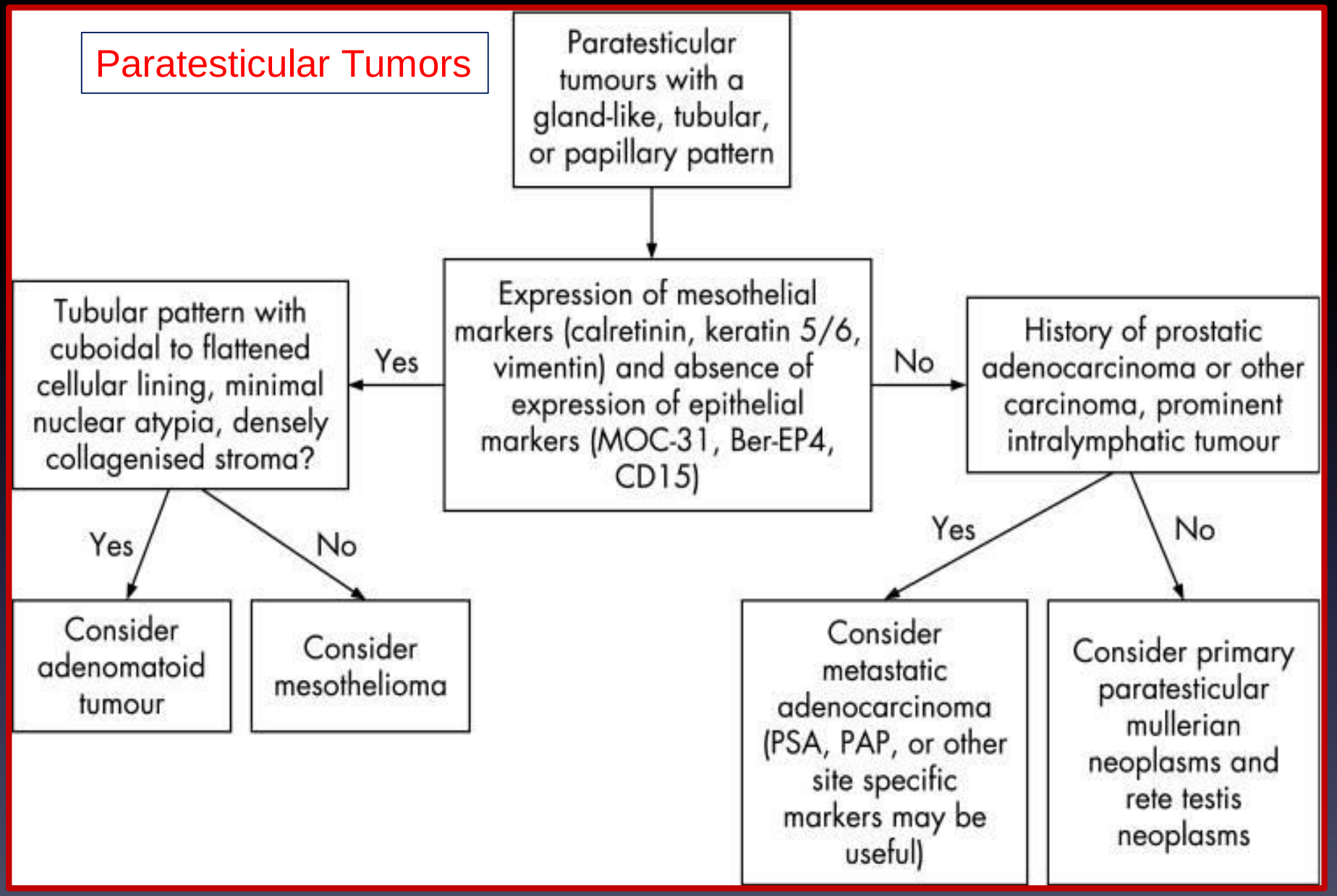
Emerson & Ulbright: J Clin Pathol. 2007 Aug; 60(8): 866–880.

Intratesticular Tumors



Emerson & Ulbright: J Clin Pathol. 2007 Aug; 60(8): 866–880.

Paratesticular Tumors



Sarcomas with True Epithelial Differentiation

- Epithelioid Sarcoma
- Synovial Sarcoma

Synovial Sarcoma

- 5% to 10% of all soft tissue sarcomas
- Malignant soft tissue tumor of uncertain type
- Uncertain histogenesis
- No known normal tissue counterpart

Synovial Sarcoma

- Age: Newborn to 82 years (mean 34)
- Most prevalent: 15 to 40 years of age
- M/F: 1.2
- Palpable, deep seated swelling or mass
- Pain or tenderness ~50% of the cases

Synovial Sarcoma

- Extremely uncommon in joint cavities
- Encountered in areas with no apparent relation to synovial structures
- Extremities: 85-95%
  - primarily in the paraarticular regions, usually in close association with tendon sheaths, bursae, and joint capsules
  - tend to arise in the vicinity of large joints, especially knee
- Head &neck: 10-15%
  - paravertebral connective tissue spaces
- Trunk: 5% (chest wall and abdominal wall)
- Virtually every anatomic site
  - heart, pleuropulmonary region, kidney, prostate,retroperitoneum,GIS, peripheral nerve

ANATOMIC LOCATION	NO. OF CASES
Head-Neck	31 (9.0%)
Neck	12
Pharynx	7
Larynx	7
Other	5
Trunk	28 (8.1%)
Chest	10
Abdominal wall	9
Other	9
Upper Extremities	80 (23.2%)
Forearm-wrist	24
Shoulder	22
Elbow-upper arm	20
Hand	14
Lower Extremities	206 (59.7%)
Thigh-knee	102
Foot	45
Lower leg-ankle	33
Hip-groin	22
Other	4
Total	345 (100.0%)

AFIP, Armed Forces Institute of Pathology.

Enzinger and Weiss's Soft Tissue Tumors

Single case in literature :

Cancer Biol Med 2012; 9: 274-276 doi: 10.7497/j.issn.2095-3941.2012.04.010

Case Report

Testicular Synovial Sarcoma: A Case Report

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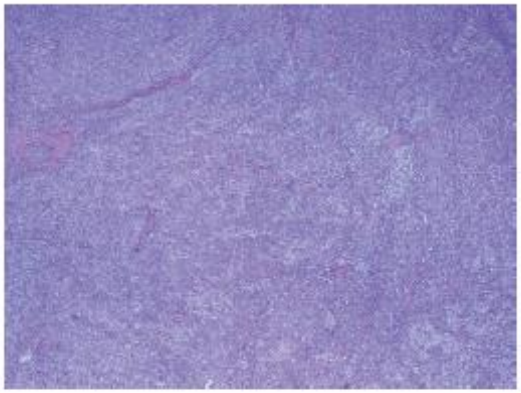

Figure 1. Monophasic poorly differentiated synovial saroma (H&E staining, ×20).

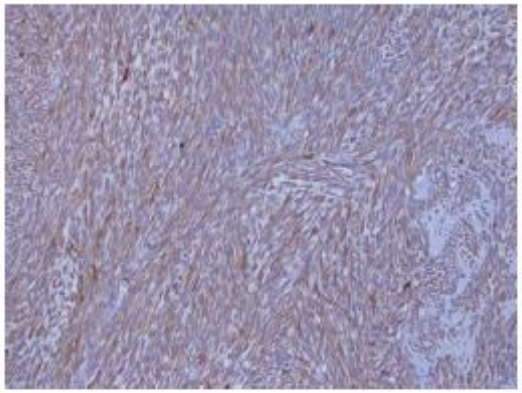

Figure 2. Pancytokeratine positive staining (IHC staining, ×40).

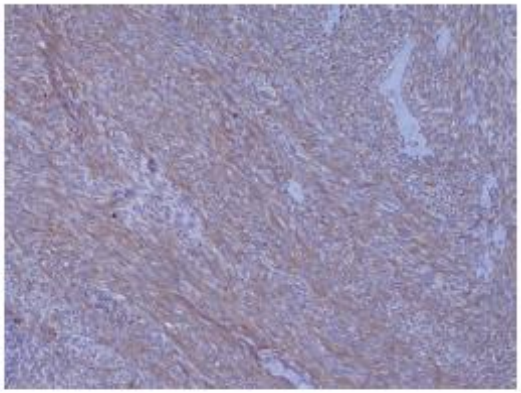

Figure 3. EMA positive staining (IHC staining, ×40).

- 24-year-old man
- Painless left testicular swelling
- Firm mobile mass, 66 × 34 mm
- Normal b-hCG and AFP
- Histology: Monophasic synovial sarcoma, poorly differentiated; grade III
- 1 yr later: recurrent scrotal mass, multiple inguinal l.nodes, pleural met.s
- DOD despite chemo 19 mo.s after diagnosis

SS - Macroscopy

Slowly growing tumors:

- be sharply circumscribed, round, or multilobular
- completely or partially invested by a pseudocapsule
- cyst formation may be prominent, occasional multicystic lesions
- yellow to gray-white
- most 3 and 6 cm

Rapidly growing tumors:

- poorly circumscribed
- variegated, friable or shaggy appearance
- multiple areas of hemorrhage, necrosis, and cyst formation

Histologic Types of SS

- Biphasic
- Monophasic
  - Fibrous type (most common subtype of SS)
  - Epithelial type (difficult to differentiate from adenoca)
- Poorly differentiated (round cell)

Biphasic SS - Microscopy

Coexistence of :

- epithelial cells
  - cuboidal - columnar cells arranged in cords, nests, or glands
  - large, round or oval, vesicular nuclei
  - abundant pale-staining cytoplasm.
  - granular or homogeneous eosinophilic secretions
  - +/- focal squamous metaplasia
- fibroblast-like spindle cells
  - well-oriented, plump, uniform spindle cells
  - narrow indistinct cytoplasm
  - oval dark-staining nuclei
  - solid compact sheets

IHC

- EMA 97 %
- AE1/AE3 90 %
- CK7 85 %
- CK19 85 %
- S-100 protein 30 %
- CD99 60 %
- TLE1 95 %
- β-Catenin 84%

TLE1 (9q21.32)

- Transducin-like enhancer of split 1
- Nuclear protein
- Transcriptional repressor of wnt/β-catenin signaling
- One of the most consistent synovial sarcoma-associated genes
- Sensitive marker of SS including cytokeratin-negative tumors

Molecular Background

- t(X;18)(p11.2;q11.2)
  - *SS18-SSX1 (65%)*

    (Strongly correlates with epithelial differentiation)
  - *SS18-SSX2 (35%)*
  - *SS18-SSX4 (<1%)*

Prognosis of SS

- considered to be a high grade malignancy
- 5-year overall survival : 64% to 76%
  - Lower rates in patients with metastases at the time of diagnosis.
- clinical factors associated with a more favorable clinical outcome
  - age of the patient (15 years or younger)
  - tumor size <5 cm,
  - distal extremity location
- histologic features of prognostic value
  - biphasic synovial sarcomas
  - extensively calcified synovial sarcomas
  - fusion subtype *SS18-SSX2*
  - poorly differentiated
  - rhabdoid cells
  - extensive tumor necrosis
  - high mitotic index (greater than 10 MF/ 10 HPF)
  - high nuclear grade

Therapy of SS

- extensive surgery - radical local excision
- adjunctive radiotherapy
- chemotherapy
  - ifosfamide and doxorubicin or epirubicin
- Recurrence rate with adequate excision and adjuvant therapy: <40%

Follow-up

- 4 courses of chemotherapy

  (Ifosfamide/Mesna/Adriamycin)
- Alive without disease for 4 years

Summary

When tumors arising in unusual sites:

- Definitive recognition is more difficult
- Algorithms are useful, but they are not all inclusive
- Spectrum of differential diagnoses needs to be kept broad
- Often confirmation by molecular - genetic techniques is required

Thank you