TUMORS OF BLOOD VESSELS

• Tumors of blood vessels and lymphatics include:

- common and benign tumors \rightarrow hemangioma
- borderline (locally aggressive but metastasize infrequently) \rightarrow kaposi sarcoma
- rare, highly malignant \rightarrow angiosarcoma
- Benign tumors usually contain vascular channels lined by normal-appearing endothelial cells.
- Malignant tumors are more cellular, show cytologic atypia, are proliferative, and usually do not form well-organized vessels

HEMANGIOMAS

- are very common tumors composed of blood-filled vessels.
- Most common in infancy and childhood
- most are present from birth and initially increase in size, but many eventually regress spontaneously.
- Most common in the head and neck
- can arise internally (1/3 → liver)
- Malignant transformation is rare

HEMANGIOMA



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HISTOLOGIC AND CLINICAL VARIANTS:

- *1- Capillary hemangiomas* : most common type; occur in the skin, subcutaneous tissues, and mucous membranes of the oral cavities and lips
- 2- *Juvenile hemangiomas* (so-called strawberry hemangiomas) of the newborn skin
- 3- **Pyogenic granulomas** are rapidly growing red pedunculated lesions on the skin, gingival, or oral mucosa (¼→history of trauma)
- 4- *Cavernous hemangiomas* : composed of large, dilated vascular channels; frequently involve deep structures, and do not spontaneously regress

INTERMEDIATE-GRADE (BORDERLINE) TUMORS

<u>o Kaposi Sarcoma (KS)</u>

• a vascular neoplasm caused by *a herpesvirus* (KSHV= human herpesvirus-8 = **HHV-8**).

- most common in patients with AIDS
- its presence is used as a criterion for the diagnosis of AIDS.
- multiple red-purple skin plaques or nodules, usually on the distal lower extremities; progressively increase in size and number and spread proximally

PATHOGENESIS

- KS is the most common HIV-related malignancy
- AIDS-associated KS often involves lymph nodes and disseminates widely to viscera early in its course.
- transmitted both through sexual contact and by poorly understood nonsexual routes (oral secretions and cutaneous exposures.
- KSHV and altered T cell immunity probably are required for KS development
- KSHV-encoded proteins disrupt normal cellular proliferation controls (a viral homologue of cyclin D) and prevent apoptosis by inhibiting p53.

A, Characteristic coalescent cutaneous redpurple macules and plaques.
B, Histologic view of the nodular stage, demonstrating sheets of plump, proliferating spindle cells and slitlike vascular spaces



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MALIGNANT TUMORS

• Angiosarcomas :

- o malignant endothelial neoplasms
- Older adults are more commonly affected.
- There is no gender bias
- lesions can occur at any site, but most often involve the **skin**, soft tissue, breast, and liver.
- Pathogenesis= carcinogens

• A latent period of years between exposure and subsequent tumor development is typical.

RISK FACTORS OF ANGIOSARCOMAS

- Chemical carcinogens \rightarrow liver angiosarcoma
- Irradiation
- Lymphedema→ ipsilateral upper extremity several years after radical mastectomy (i.e., with lymph node resection) for breast cancer
- long-term (years) indwelling foreign bodies (e.g., catheters).

CARDIAC TUMORS

<u>Metastatic Neoplasms :</u>

- the most common malignancy of the heart
- occur in 5% of patients dying of cancer.
- certain tumors have a higher predilection for cardiac metastases. In descending order these are:
- lung cancer \rightarrow most common primary
- lymphoma
- breast cancer
- Leukemia
- Melanoma
- hepatocellular carcinoma
- colon cancer.

PRIMARY CARDIAC TUMORS

- o uncommon
- most are benign (80% to 90% of all primary heart tumors).
- The five most common in descending order of frequency: **Myxoma**; Fibromas; Lipomas; papillary fibroelastomas; rhabdomyomas.
- Angiosarcomas constitute the most common **primary** *malignant* tumor of the heart.

Мухома

- the most common **primary** tumors of the **adult** heart
- 90% \rightarrow atrium (left atrium >80%)
- pedunculated lesion arises from the atrial wall with a gelatinous appearance
- The cells are embedded in an abundant acid mucopolysaccharide ground substance

o Rhabdomyoma

- the most frequent **primary** tumors of the heart in infants and **children**
- often regress spontaneously for unknown reasons
- Morphology: characteristic large cells containing numerous glycogen vacuoles separated by strands of cytoplasm running from the plasma membrane to the centrally located nucleus, so-called **spider cells**

CLINICAL FEATURES AND SIGNIFICANCE

- 1- valvular "ball-valve" obstruction
- 2- Embolization
- 3- constitutional signs and symptoms (fever and malaise)→ attributable to tumor elaboration of the cytokine **interleukin-6**, a major mediator of the acute-phase response.
- Diagnosis: Echocardiography
- Treatment: surgical resection is almost uniformly curative in benign tumors.