

TUMORS OF BLOOD VESSELS

- Tumors of blood vessels and lymphatics include:
 - common and benign tumors → hemangioma
 - borderline (locally aggressive but metastasize infrequently) → kaposi sarcoma
 - rare, highly malignant → 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

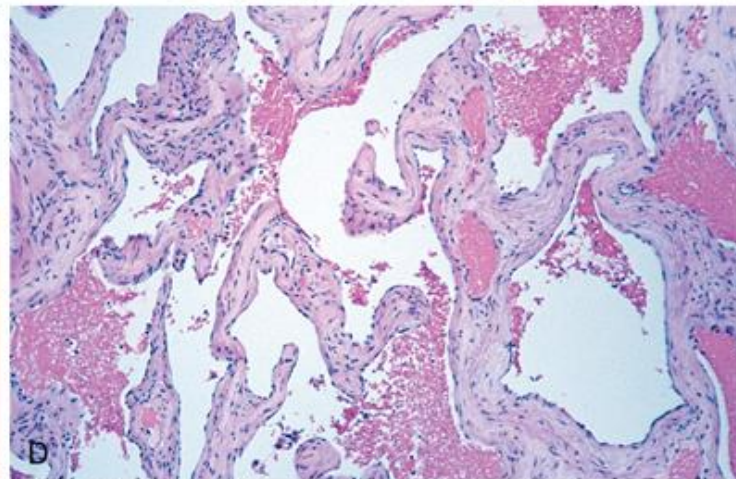
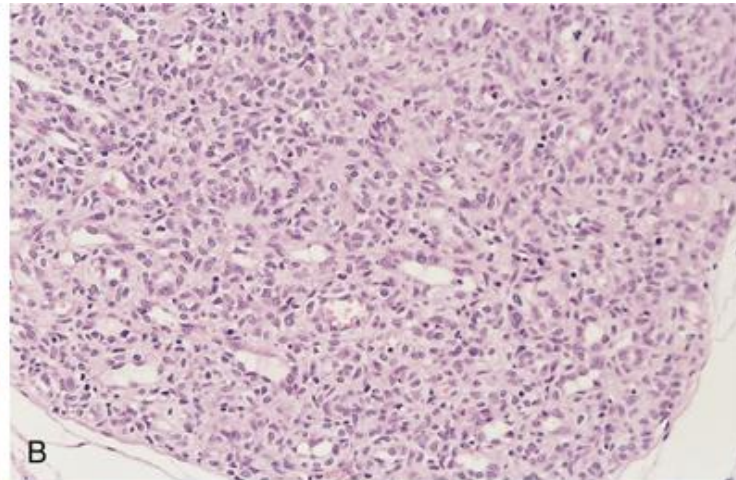


HEMANGIOMAS

- 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 ($\frac{1}{4}$ → 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

- *Kaposi Sarcoma (KS)*
- 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



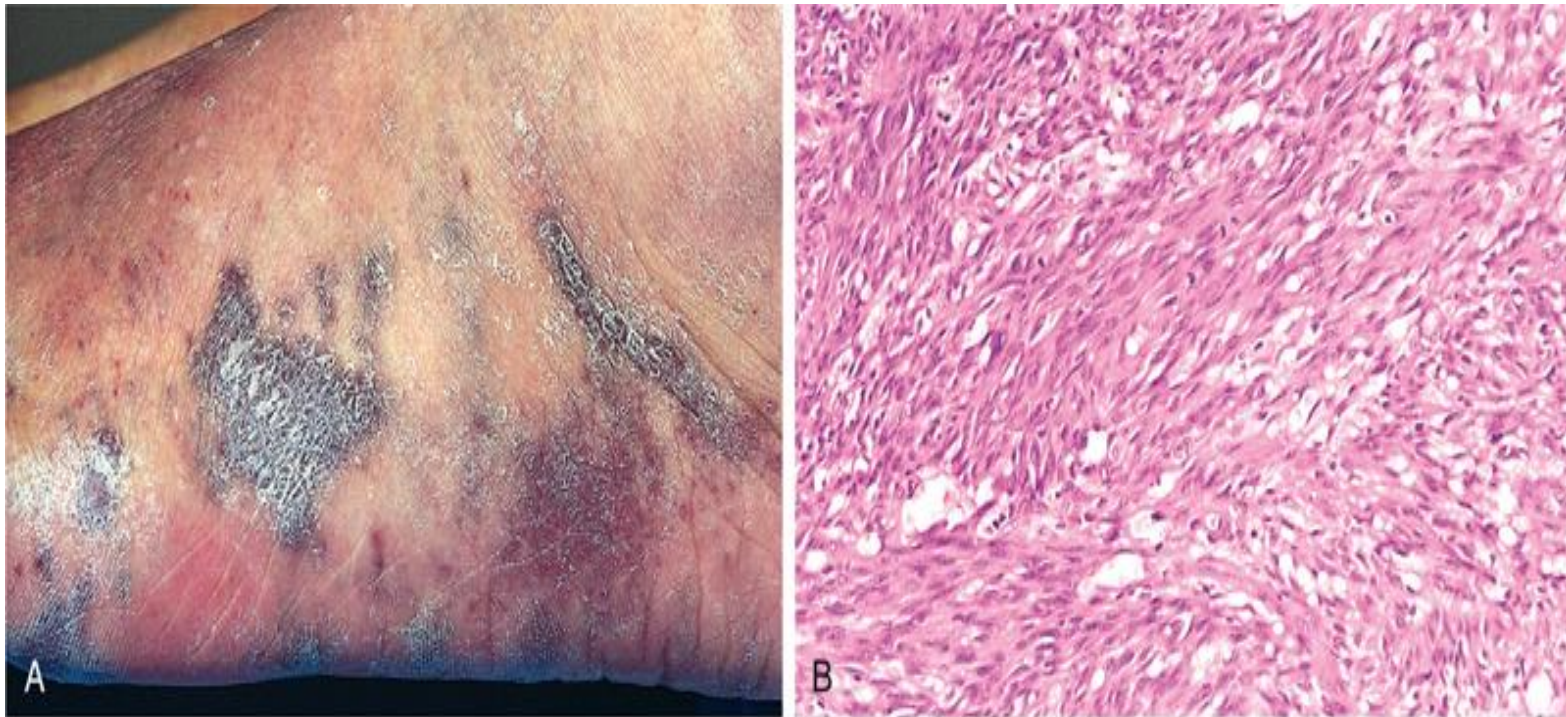
PATHOGENESIS

- *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 red-purple 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* :
- 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 → 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

Metastatic Neoplasms :

- *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** → **most common primary**
 - lymphoma
 - breast cancer
 - Leukemia
 - Melanoma
 - hepatocellular carcinoma
 - colon cancer.



PRIMARY CARDIAC TUMORS

- *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.



MYXOMA

- the most common **primary** tumors of the **adult** heart
- 90% → 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

- ***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.

