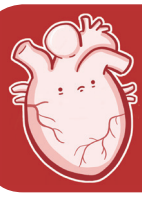

NOTES
VASCULAR TUMORS

GENERALLY, WHAT ARE THEY?

PATHOLOGY & CAUSES

- Abnormal growths of blood/lymph vessels
- Can be benign/malignant, can occur anywhere in body
- Vascular tumors are rare, but most commonly found in neonates (e.g. hemangiomas), HIV-positive individuals (e.g. Kaposi's sarcoma)

COMPLICATIONS

- Metastasis
- Complications from chemo/radiation therapy

SIGNS & SYMPTOMS

- See individual disorders

DIAGNOSIS

DIAGNOSTIC IMAGING

- Visual identification, imaging studies (MRI, CT scan, ultrasound with Doppler, biopsy)
  - Determine location, tumor size, extent of spread

LAB RESULTS

- Biopsy for definitive diagnosis

TREATMENT

- Depends on type, location, severity, malignancy; see individual disorders

ANGIOSARCOMA

osms.it/angiosarcoma

PATHOLOGY & CAUSES

- Rare blood vessel malignancy involving blood vessel endothelial lining
- Aggressive, rapidly proliferating → poor prognosis
- Can occur anywhere; usually occurs in sun-exposed areas (head, neck, breast)
  - Cutaneous angiosarcomas (occur beneath skin's surface) most common
- Can affect liver blood vessels

CAUSES

- Most likely due to lymphedema (fluid buildup causing sweeling), radiation exposure, carcinogens

RISK FACTORS

- Biologically male (twice as likely), elderly, sun-exposure, radiation therapy, chronic post-mastectomy lymphedema, frequent exposure to vinyl chloride monomer gas in PVC manufacturing/arsenic insecticides
  - *High-grade:* aggressive, fast-growing

- *Low-grade:* less aggressive, slow-growing

COMPLICATIONS

- High chance of metastasis, poor prognosis. Better prognosis for individuals with smaller tumors with clearly delineated margins. Low grade breast angiosarcoma has better prognosis than tumors with poorly-defined borders

SIGNS & SYMPTOMS

- Lesion resembling non-healing bruise/wound
- Violet color
- Soft, visible, tactile lump/swelling
- Can form irregular vascular channels that disrupt tissue planes
- Fatigue
- Bone pain
- Anemia

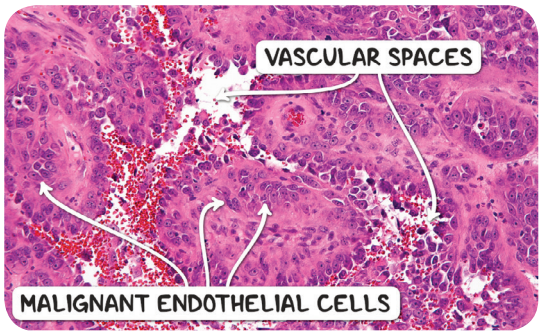

Figure 24.2 Histological appearance of an angiosarcoma composed of malignant endothelial cells with vascular spaces containing red blood cells.

DIAGNOSIS

LAB RESULTS

- Biopsy, usually diagnosed late after the disease has spread

TREATMENT

MEDICATIONS

- Chemotherapy

SURGERY

- Difficult to resect due to delay in diagnosis

OTHER INTERVENTIONS

- Radiation

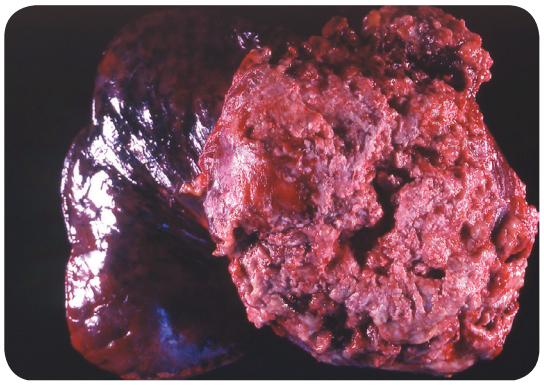

Figure 24.1 A surgically excised angiosarcoma.

GLOMUS TUMOR

osms.it/glomus-tumor

PATHOLOGY & CAUSES

- Benign tumor arising from modified smooth muscle cells of skin's thermoregulatory glomus bodies
- Derives from small vessels/arteriovenous anastomoses in glomus bodies
- Malignancy, metastasis rare
- Etiology includes loss-of-function mutation of protein glomulin in familial glomangiomas

RISK FACTORS

- *Adults:* 20–40 years old
- Most lesions solitary, localized
- Autosomal dominant inheritance pattern

COMPLICATIONS

- Good prognosis, low recurrence rate after resection
- Malignant glomus tumors rare, have good prognosis when treated with wide excision
- Metastasis associated with poor prognosis

SIGNS & SYMPTOMS

- Painful, small, red-blue growths
  - Pain associated with solitary lesions
  - Younger individuals: multiple tumors, usually asymptomatic
- Usually found on distal extremities
- Paroxysmal pain depending on temperature, pressure changes
  - Cold, pressure worsens pain

DIAGNOSIS

OTHER DIAGNOSTICS

- Visual inspection
- Occasional imaging

TREATMENT

SURGERY

- Resection

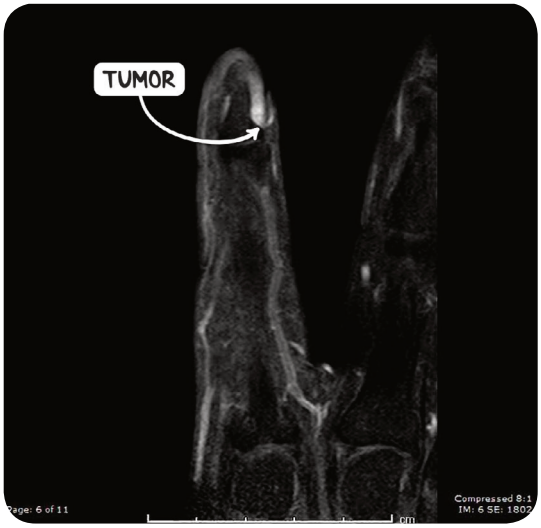

Figure 24.3 Homogenous enhancement of a glomus tumor of the nail bed at the ulnar aspect of the left index finger.

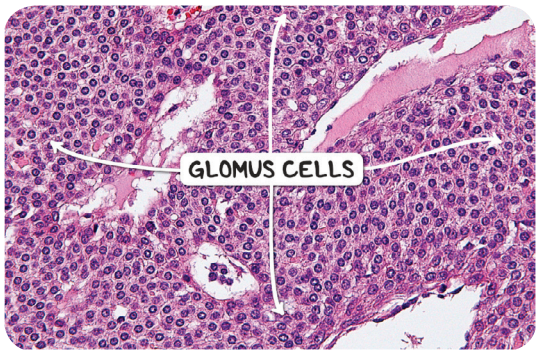

Figure 24.4 Histological appearance of a glomus tumor.

KAPOSI SARCOMA

osms.it/kaposi

PATHOLOGY & CAUSES

- Malignant vascular tumor/lesions of blood vessel endothelial cells
- Due to *human herpesvirus 8*
- Virus penetrates cells, causing uncontrollable replication
- May involve visceral organs
- Progression, severity of tumor depends on underlying factor
  - *Genetic:* usually seen in older Eastern European males; tumor localized to skin
  - *AIDs:* tumor spreads (see staging below)
  - *Organ transplant recipients:* tumor spreads

TYPES

AIDS-related

- Most common malignancy in AIDS

Immunocompromised & iatrogenic-related

Classic/sporadic

Endemic (African)

- Burkitt's lymphoma due to Epstein-Barr virus

RISK FACTORS

- Immunocompromised individuals
  - AIDS
  - Kaposi's sarcoma associated *human herpesvirus-8* (*HHV-8*)
  - Organ transplant
- Biologically male
- Eastern European
- *Higher risk:* biologically-male individuals engaging in same-sex sexual acts ("MSM")

STAGING

AIDS-related Kaposi's sarcoma

- Takes three factors into account
- Extent/severity of tumor
  - *T0:* localized tumor
  - *T1:* widespread, multiple lesions that spread to other organs
- CD4 cell count (immune status)
  - *I0:* CD4 count above 150 cells/mm^3
  - *I1:* CD4 count less than 150 cells/mm^3
- Presence/absence of systemic illness
  - *S0:* no systemic illness/opportunistic infections, and/or B symptoms. B symptoms: systemic fever symptoms, night sweats, weight loss, diarrhea
  - *S1:* presence of systemic illness, opportunistic infections, and/or B symptoms

COMPLICATIONS

- Lymphedema
- Bleeding
- Infection
- Long term hyperpigmentation
- Prognosis depends on individual's immune status, viral load (amount of HIV virus in blood)

SIGNS & SYMPTOMS

- Most common symptoms affect skin, also affect mouth, GI tract, respiratory tract
  - Progresses from flat lesion → plaque → ulcerating nodule
  - Purple, red lesion similar to bruise that does not blanch
  - Lesion starts off flat, may become raised, more painful

- Lesions in other tissues (e.g. mouth, nose, throat, lymph nodes, lungs, gastrointestinal tract); Commonly found in mucous membranes (esp. hard palate)
- *Pulmonary symptoms:* pulmonary Kaposi's sarcoma
  - Coughing (possibly bloody cough)
  - Dyspnea

DIAGNOSIS

DIAGNOSTIC IMAGING

- Bronchoscopy/endoscopy

LAB RESULTS

- Biopsy

TREATMENT

- Depends on
  - Severity of immunosuppression
  - Number, location of tumors
  - Symptoms

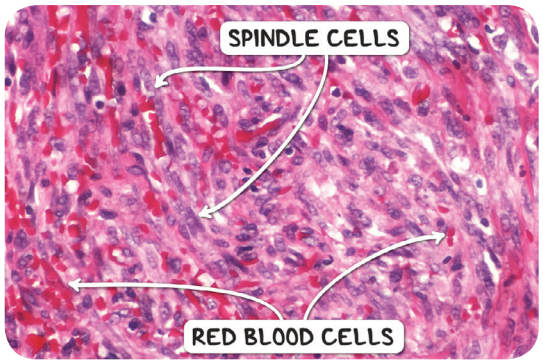

Figure 24.6 A Kaposi sarcoma composed of spindle cells which form slits filled with erythrocytes.

MEDICATIONS

- HIV/AIDS management with antivirals
  - Control HIV/AIDS → lesions shrink
- Removal of drugs (e.g. corticosteroids) allows immune system to recover
  - Treatment more difficult in immunocompromised individuals
- Chemotherapy

SURGERY

- Surgically remove affected skin

OTHER INTERVENTIONS

- Cryotherapy → freeze affected skin
- Radiation

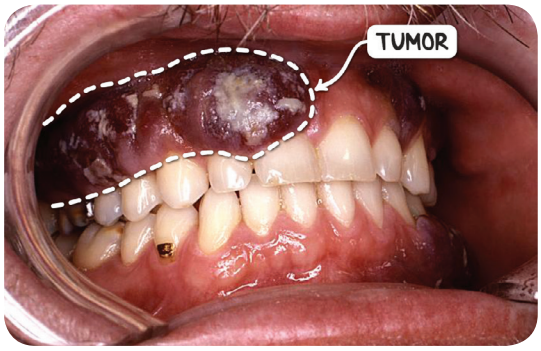

Figure 24.5 Kaposi sarcoma of the gingiva in a HIV positive individual. The tumor has replaced the gingiva of the upper right side of the jaw. There is overlying oral candidiasis.