Vascular Neoplasms

Hemangiomas

Benign tumors composed of hyperplastic blood vessels. Usually present as flat, raised, red or purplish lesions that do not enlarge. Common hemangiomas include port wine stains, strawberry marks, cavernous hemangiomas, and vascular spiders. Generally these are not clinically relevant and do not require treatment except for cosmetic reasons, though when present inside organs they can cause problems by rupturing, disrupting normal blood flow, or compressing other structures (eg sphincters, the eye).

Kaposi's Sarcoma

Etiology: malignant neoplasm due to human herpes virus-8 (Kaposi sarcoma-associated virus). Actually now known to be a tumor of lymphatic epithelium and not a sarcoma, causing vascular channels in the lymphatics that fill with blood and discolor the typical skin lesions.. Also known as Kaposi's lymphohemangioma.

Risk factors: unclear route of transmission but appears to be spread (at least in North America) by saliva predominantly. Immunosuppression is a major factor in the cancer forming, being seen primarily in organ transplant recipients and AIDS patients (the latter rarely seen now with effective antiretroviral drug cocktails available). Areas of high endemic transmission of the virus are Africa (up to 50% of population infected) and Mediterranean rim.

Complications: high rate of mortality in AIDS and transplant patients, as well as in endemic form in Africa. Fairly indolent course in the classic form.

Clinical characteristics: Malignant neoplasm seen as reddish-purple to dark-blue cutaneous macules, plaques, or nodules. Five forms exist globally: AIDS-associated KS, Immunosuppression-associated KS, classic KS (seen in older men from Mediterranean region primarily), and two endemic African forms (cutaneous and lymphadenopathic).

Infectious Vascular Diseases

Bacterial Endocarditis

See Endocarditis

Chagas Disease

Etiology: infection with *Trypanosoma cruzi*. Vector: triatomine (reduviid, kissing, or assassin) bugs, found throughout the Americas but primarily in South. Reservoir: dogs, cats, opossums, rats, and other mammals. Infection can also be transmitted by blood transfusion or transplacentally.

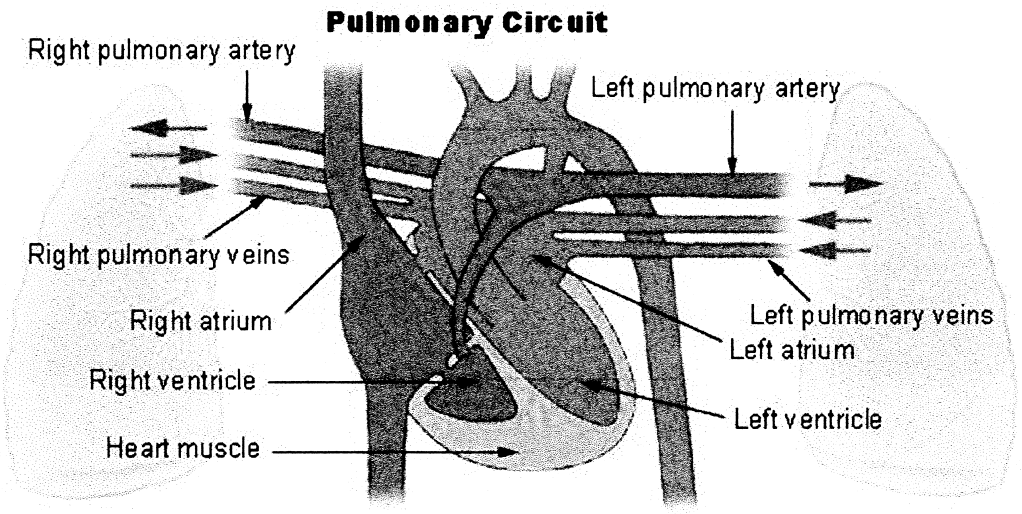
Infected kissing bugs deposit feces containing trypomastigotes on the skin while biting, which enter through wounds and penetrate mucous membranes. The parasites invade macrophages, where they transform into amastigotes, multiply, and are released as trypomastigotes into the blood and tissue space. Other cells are then infected, particularly cells of the nervous system, and reticuloendothelial, myocardial, and muscle cells.

Risk factors: residence in endemic area, immune suppression

Complications: cardiomyopathy, apical aneurysms, heart failure, Stokes-Adams attacks, thromboembolism, megaesophagus, or megacolon from chronic infection

Clinical characteristics: The initial infection is typically asymptomatic. An acute phase may present 1–2 weeks later with fever, hepatomegaly and lymphadenopathy. See complications for chronic symptoms. Diagnosis is by blood or lymph node culture or PCR.

Figure 6.23: Cardiopulmonary Blood Flow



Lyme Disease

Caused by infection with *Borrelia burgdorferi*, a loosely coiled, motile spirochete that can be visualized with Giemsa stain or dark field microscopy. Routine cultures from humans are generally negative, whereas cultures from ticks are usually positive.

Clinical characteristics: The organism gains entrance to the body through a tick bite. Once in the skin, it spreads by blood to various organ systems (e.g., heart, synovial joints, CNS).

First stage: often a spreading circular red rash with a white center known as erythema migrans. Nonspecific flu-like symptoms may be present. This occurs 3–32 days after the tick bite. Regional lymphadenopathy may occur.

Second stage: occurs months after the primary lesion has cleared. This stage is characterized by either cardiovascular or neurological symptoms, such as heart blocks, Bell's palsy or peripheral neuropathies.

Final stage: characterized by arthritis of the large joints, particularly the knee joint, and a myriad of other systemic, confusing, complicated symptoms.

Diagnosis: Because cultures of human samples are rarely positive, diagnosis relies on serological testing for either an IgM or an IgG antibody to the spirochete. PCR tests are available to test for the organism's DNA.

Rocky Mountain Spotted Fever

Etiology: infection with *Rickettsia rickettsii*. Vector: *Dermacentor* ticks. Found throughout the Americas. Causes widespread vasculitis, which affects coronary blood vessels.

Risk factors: tick bite, presence in area with endemic ticks and microbes

Complications: can be rapidly lethal without antibiotic treatment

Clinical characteristics: acute flu-like illness, purpural rash, petechiae, GI pain, arthralgias. Rashes start on limbs and move toward trunk.

Viral Hemorrhagic Fever

Most are rare in the US (e.g. Ebola hemorrhagic fever, Crimean-Congo hemorrhagic fever). In all these diseases, the viruses trigger widespread vascular damage thus multiorgan damage or failure and bleeding from any of a number of sites. All causative viruses require an animal host (reservoir), and tend to remain localized as they are dependent on the reservoir to stay alive. All causative viruses are enveloped RNA viruses. In all known cases, humans are infected incidentally and are not required for the life cycle of the viruses.

Yellow fever: Flavivirus genus transmitted by female *A. aegypti* mosquito mainly in South America and Africa. Presents as mild, self-limiting illness in majority of patients with fever, chills, headache, anorexia, nausea, and muscle pain, which generally subsides after several days. More serious illness in the minority with vomiting, renal failure, and hemorrhage. End stage marked by delirium, stupor, and coma.

Dengue fever: transmitted by the *A. aegypti* mosquito found in the Middle East, Africa, Central America, and the Caribbean. Classic form starts influenza-like and progresses to severe pain in muscles and joints, enlarged lymph nodes, maculopapular rash, and leukopenia. The more severe form, dengue hemorrhagic fever, results in shock and hemorrhaging that progresses, sometimes to death.

Filoviruses: only two members of this family identified: Marburg virus and Ebola virus. Symptoms begin with influenza–type stage, followed by respiratory and CNS system involvement. Development of hemorrhagic symptoms indicate poor prognosis. Death occurs due to multiple organ dysfunction syndrome.

Cardiopulmonary Relationship

The cardiovascular system is responsible for oxygenation of the body's tissues, often under conditions of great variability. For this reason, the pulmonary and cardiovascular systems work closely together via complex monitoring and control functions. The most basic aspect of the relationship involves blood flow. The cycle begins with deoxygenated blood flowing out of the right ventricle into the pulmonary arteries and finally into the lungs to be oxygenated in the alveolar beds. Oxygenated blood then flows back through the pulmonary veins to the left atrium and then the left ventricle, where it is pumped throughout the rest of the body. There is a separate circulation branching ultimately from the aorta, the bronchial arteries, that provide arterial blood to cells in the lungs.