younger than 45 years of age. The mechanism is unknown, but all patients have a history of heavy tobacco addiction. The presenting symptom is claudication of the feet, legs, hands, or arms. Multiple-limb involvement and superficial thrombophlebitis are common. The C-reactive protein level and erythrocyte sedimentation rate (Westergren method) typically are normal, and a search for serologic markers for connective tissue disease (e.g., antinuclear antibody, rheumatoid factor, antiphospholipid antibody) is negative. The diagnosis is based on the typical clinical presentation. If the presentation is atypical, then biopsy is needed to make the diagnosis. The histologic hallmark is inflammatory intramural thrombi within the arteries and veins with sparing of internal elastic lamina and other arterial wall structures. The most effective treatment for Buerger's disease is complete tobacco abstinence. The prostacyclin analogue iloprost constitutes adjunctive therapy to reduce limb ischemia and improve wound healing.

Raynaud's Phenomenon

Raynaud's phenomenon is a vasospastic disease of the small arteries of mainly the fingers and toes. Primary (idiopathic) Raynaud's phenomenon occurs in the absence of underlying disorders. Secondary Raynaud's phenomenon occurs in association with connective tissue diseases (e.g., scleroderma, polymyositis, rheumatoid arthritis, systemic lupus erythematosus) repeated mild physical trauma (e.g., use of jack hammers), use of certain drugs (e.g., antineoplastic chemotherapeutic agents, interferon, monamine-reuptake inhibitors such as tricyclic antidepressants, serotonin agonists), and Buerger's disease. Patients usually complain of recurrent episodes of ischemia of the digits, which exhibit a characteristic white-blue-red color sequence: Pallor is followed by cyanosis if ischemia is prolonged and then by erythema (reactive hyperemia) when the episode resolves. Episodes are precipitated by cold temperature or emotional stress. The physical examination can be entirely normal between attacks with normal radial, ulnar, and pedal pulses. Some patients have digital ulcers or thickening of the fat pad (sclerodactyly). Patients should be instructed to avoid cold temperatures and dress warmly. Calcium-channel blockers (CCBs) reduce the frequency and severity of vasospastic episodes.

Giant-Cell Arteritis

Giant-cell arteritis is an immune-mediated vasculitis that predominantly involves medium-sized and large arteries such as the subclavian artery, axillary artery, and aorta of older adults, with a strong male predominance. Approximately 40% of patients with giant-cell arteritis also have polymyalgia rheumatica, a syndrome characterized by severe stiffness and pain originating in the muscles of the shoulders and pelvic girdle. Patients may exhibit headache from temporal arteritis, jaw claudication from ischemia of the masseter muscles, or visual loss from involvement of the ophthalmic artery. Chest pain suggests the coexistence of aortic aneurysm or dissection. Physical findings include low-grade fever, scalp tenderness in the temporal area, pale and edematous fundi, or a diastolic murmur of aortic regurgitation. A BP difference of more than 15 mm Hg between the arms suggests subclavian artery stenosis. Laboratory findings include a significantly elevated C-reactive protein level and Westergren sedimentation rate plus anemia. The diagnosis is confirmed by histologic examination of the arterial tissue (frequently obtained by temporal artery biopsy) showing infiltration of lymphocytes and macrophages (i.e., giant cells) in all layers of the vascular wall. High-dose corticosteroids are highly effective. To minimize complications from long-term corticosteroid administration, the steroid dose should be tapered to find the lowest dose needed to suppress symptoms, which often wane. Every attempt should be made to discontinue corticosteroids over time.

Takayasu's Arteritis

Takayasu's arteritis is an idiopathic granulomatous vasculitis of the aorta, its main branches, and the pulmonary artery. This condition is particularly common in young women of Asian descent, but it also occurs in non-Asian women and in men. The inflammatory process in the vascular wall can lead to stenosis or aneurysm formation or both. Hypertension, as a result of renal artery stenosis or aortic coarctation, is the most common manifestation and is present in as many as 80% of affected individuals. Because the vascular involvement is so widespread, patients may have symptoms and signs of coronary ischemia, congestive heart failure, stroke, vertebrobasilar insufficiency, or intermittent claudication. Physical findings include bruits over the subclavian arteries or aorta with diminished brachial pulses and therefore a low brachial artery BP. The diagnosis is based primarily on this clinical presentation. First-line treatment is with corticosteroids. Other immunosuppressive agents such as methotrexate or cyclophosphamide are often added to prevent disease progression and relapse. Immunosuppressive therapy does not cause regression of preexisting vascular stenoses or aneurysms. For this reason, percutaneous or surgical revascularization is usually required.

Arteriovenous Fistula

Arteriovenous (AV) fistulas are abnormal vascular communications that shunt blood flow from the arterial system directly into the venous system, bypassing the capillary beds that normally ensure optimal tissue perfusion and nutrient exchange. AV fistulas may be congenital, as in AV malformation, or acquired. The main causes of acquired AV fistula are penetrating trauma (e.g., gunshot, knife wound) and shunts created surgically for hemodialysis access. Patients may exhibit a pulsatile mass, symptoms related to compression of an adjacent organ, or bleeding from spontaneous rupture of an AV malformation. Systolic and diastolic bruits or thrills may be detectable over the fistula or malformation. An AV malformation in skeletal muscle may lead to bone malformation or a pathologic fracture, whereas an AV malformation in the brain may result in neurologic deficits or seizures. High-output heart failure is another complication from a large AV malformation or fistula. MR angiography, CT angiography, or conventional angiography confirms the diagnosis. Depending on the size and location of the lesion, treatment options include surgical resection, transcatheter embolization, or pulse laser irradiation. Patients with acquired AV fistulas from trauma usually need surgical closure.

PULMONARY VASCULAR DISEASE

Pulmonary hypertension is characterized by elevated mean pulmonary artery pressure of greater than 25 mm Hg at rest or