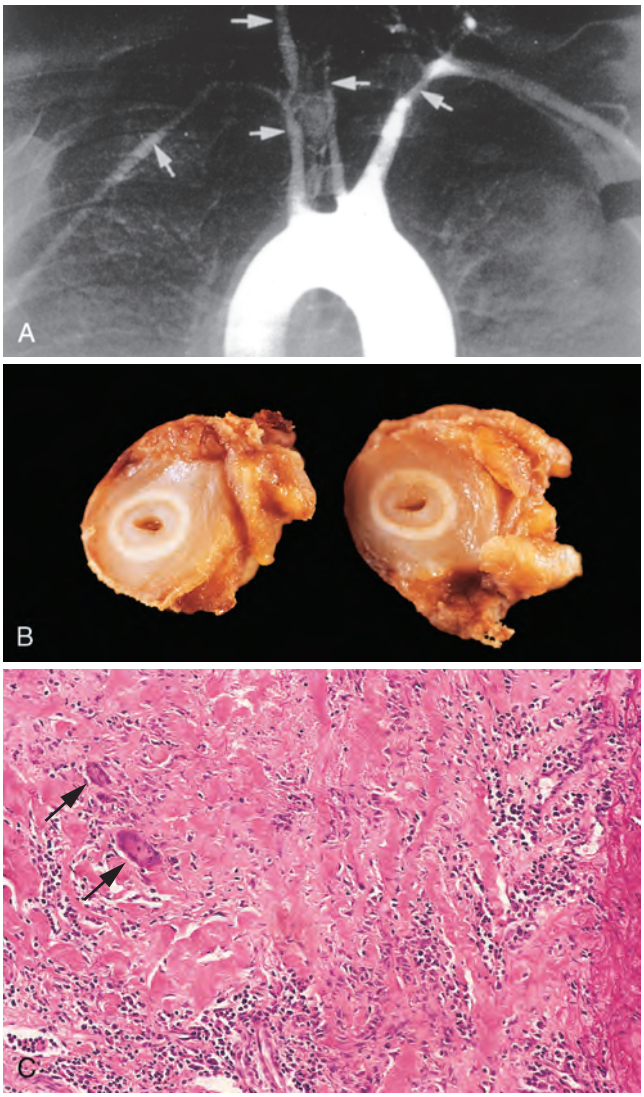

Figure 11-25 Takayasu arteritis. **A,** Aortic arch angiogram showing narrowing of brachiocephalic, carotid, and subclavian arteries *(arrows).* **B,** Gross photograph of two cross-sections of the right carotid artery taken at autopsy of the patient shown in **A,** demonstrating marked intimal thickening and adventitial fibrosis with minimal residual lumen. **C,** Histologic appearance in active Takayasu aortitis, illustrating destruction and fibrosis of the arterial media associated with mononuclear infiltrates and inflammatory giant cells *(arrows).*

aortitis. Although historically associated with the Japanese population and a subset of HLA haplotypes, Takayasu aortitis has a global distribution. An autoimmune etiology is likely.

MORPHOLOGY

Takayasu arteritis classically involves the aortic arch. In a third of patients, it also affects the remainder of the aorta and its branches, with **pulmonary artery** involvement in half the cases; **coronary and renal arteries** may be similarly affected. There is irregular thickening of the vessel wall with intimal hyperplasia; when the aortic arch is involved, the great vessel lumina can be markedly narrowed or even obliterated (Fig. 11-25*A* and *B*). Histologically, the changes range from adventitial mononuclear infiltrates with perivascular cuffing of the vasa vasorum, to intense mononuclear inflammation in the media, to granulomatous inflammation, replete with giant cells and patchy medial necrosis. The histology (Fig. 11-25*C*) is indistinguishable from giant cell (temporal) arteritis. As the disease progresses, collagenous scarring, with admixed chronic inflammatory infiltrates, occurs in all three layers of the vessel wall. Occasionally, aortic root involvement causes dilation and aortic valve insufficiency.

Clinical Features. Initial symptoms are usually nonspecific, including fatigue, weight loss, and fever. With progression, vascular symptoms appear and dominate the clinical picture, including reduced blood pressure and weak pulses in the carotids and the upper extremities; ocular disturbances, including visual defects, retinal hemorrhages, and total blindness; and neurologic deficits. Involvement of the more distal aorta may lead to claudication of the legs; pulmonary artery involvement can cause pulmonary hypertension. Narrowing of the coronary ostia may lead to myocardial infarction, and involvement of the renal arteries leads to systemic hypertension in roughly half of patients. The course of the disease is variable. In some there is rapid progression, while others enter a quiescent stage after 1 to 2 years, permitting long-term survival, albeit with visual or neurologic deficits.

Polyarteritis Nodosa

Polyarteritis nodosa (PAN) is a systemic vasculitis of small- or medium-sized muscular arteries, typically involving renal and visceral vessels but sparing the pulmonary circulation. There is no association with ANCA, but about 30% of patients with PAN have chronic hepatitis B and deposits containing HBsAg-HBsAb complexes in affected vessels, indicating an immune complex–mediated etiology in that subset. The cause remains unknown in the remaining cases; there may be etiologic and clinical distinctions between classic idiopathic PAN, the cutaneous forms of PAN, and the PAN associated with chronic hepatitis.

MORPHOLOGY

Classic polyarteritis nodosa is characterized by **segmental transmural necrotizing inflammation of small- to medium-sized arteries**. Vessels of the kidneys, heart, liver, and gastrointestinal tract are involved in descending order of frequency. Lesions usually involve only part of the vessel circumference with a predilection for branch points. The inflammatory process weakens the arterial wall and can lead to aneurysms or even rupture. Impaired perfusion with ulcerations, infarcts, ischemic atrophy, or hemorrhages may be the first sign of disease.

During the acute phase, there is transmural inflammation of the arterial wall with a mixed infiltrate of neutrophils, eosinophils, and mononuclear cells, frequently accompanied by **fibrinoid necrosis** (Fig. 11-26). Luminal thrombosis can occur. Later, the acute inflammatory infiltrate is replaced by fibrous (occasionally nodular) thickening of the vessel wall that can extend into the adventitia. Characteristically, all stages of activity (from early to late) may coexist in different vessels or even within the same vessel, suggesting ongoing and recurrent insults.