lupus vasculitis and antiphospholipid antibody syndrome can be morphologically and clinically similar, immunosuppressive therapy is required in the former, and anticoagulant therapy is indicated in the latter.

Infectious Vasculitis

Arteritis can be caused by the direct invasion of infectious agents, usually bacteria (*Pseudomonas* being the classic example) or fungi, in particular *Aspergillus* and *Mucor* species. Vascular invasion can be part of a localized tissue infection (e.g., bacterial pneumonia or adjacent to abscesses), or—less commonly—can arise from hematogenous spread of microorganisms during septicemia or embolization from infective endocarditis.

Vascular infections can weaken arterial walls and culminate in *mycotic aneurysms* (see earlier), or can induce thrombosis and infarction. Thus, inflammation-induced thrombosis of meningeal vessels in bacterial meningitis can eventually cause infarction of the underlying brain.

KEY CONCEPTS

Vasculitis

- Vasculitis is defined as inflammation of vessel walls; it is frequently associated with systemic manifestations (including fever, malaise, myalgias, and arthralgias) and organ dysfunction that depends on the pattern of vascular involvement.
- Vasculitis can result from infections, but more commonly has an immunologic basis, including immune complex deposition, formation of anti-neutrophil antibodies (ANCA), or T cell responses to vascular wall antigens.
- Different forms of vasculitis tend to specifically affect vessels of a particular caliber and location, and the clinical manifestations depend on the pattern of vessel involvement.

Disorders of Blood Vessel Hyperreactivity

Several disorders are characterized by inappropriate or exaggerated vasoconstriction of blood vessels.

Raynaud Phenomenon

Raynaud phenomenon results from exaggerated vasoconstriction of arteries and arterioles in the extremities, particularly the fingers and toes, but also occasionally the nose, earlobes, or lips. The restricted blood flow induces paroxysmal pallor, and even cyanosis in severe cases; involved digits classically show "red, white, and blue" color changes from most proximal to most distal, correlating with proximal vasodilation, central vasoconstriction, and more distal cyanosis (Fig. 11-29). Raynaud phenomenon can be a primary entity or secondary to other disorders.

Primary Raynaud phenomenon (previously called Raynaud disease) is caused by exaggerated central and local vasomotor responses to cold or emotion; it affects 3% to 5% of the general population and has a predilection for young women. It tends to symmetrically affect the extremities, and the severity and extent of involvement typically remains static over time. Structural changes in the arterial walls are absent except in long-standing disease, when intimal thickening can develop. The course is usually benign, but chronicity can lead to atrophy of the skin, subcutaneous tissues, and muscles. Ulceration and ischemic gangrene are rare.

Secondary Raynaud phenomenon refers to vascular insufficiency due to arterial disease caused by other entities including SLE, scleroderma, Buerger disease, or even atherosclerosis. Clinically, secondary Raynaud phenomenon tends to have asymmetric involvement of the extremities and progressively worsens in extent and severity over time.

Since Raynaud phenomenon may be the first manifestation of immune-mediated vasculitides, any patient with

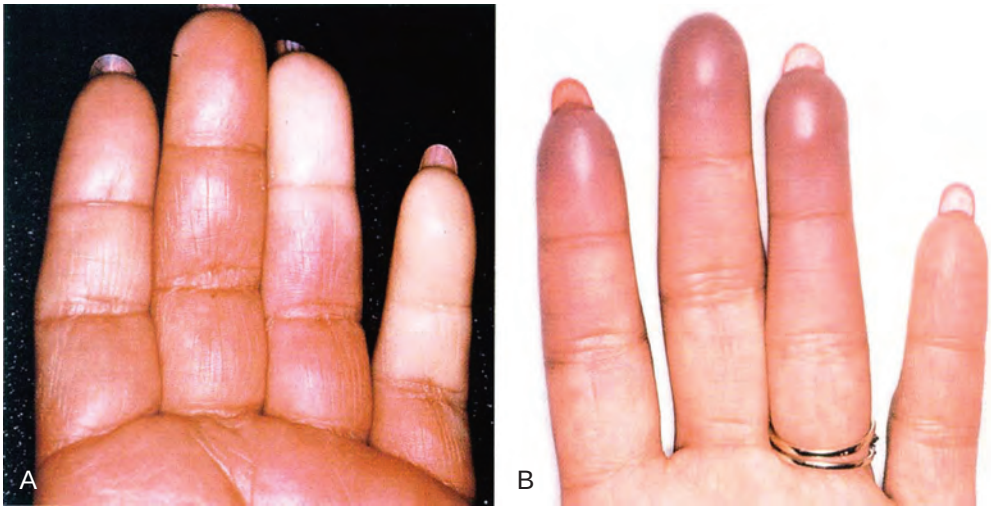

Figure 11-29 Raynaud phenomenon. **A,** Sharply demarcated pallor of the distal fingers resulting from spasm of the digital arteries. **B,** Cyanosis of the fingertips. (Reproduced from Salvarani C, et al: Polymyalgia rheumatica and giant-cell arteritis. N Engl J Med 347:261, 2002.)