CHAPTER e40

Atlas of the Vasculitic Syndromes

Carol A. Langford
Anthony S. Fauci

Diagnosis of the vasculitic syndromes is usually based upon characteristic histologic or arteriographic findings in a patient who has clinically compatible features. The images provided in this atlas highlight some of the characteristic histologic and radiographic findings that may be seen in the vasculitic diseases. These images demonstrate the importance that tissue histology may have in securing the diagnosis of vasculitis, the utility of diagnostic imaging in the vasculitic diseases, and the improvements in the care of vasculitis patients that have resulted from radiologic innovations.

Tissue biopsies represent vital information in many patients with a suspected vasculitic syndrome, not only in confirming the presence of vasculitis and other characteristic histologic features, but also in ruling out other diseases that can have similar clinical presentations. The determination of where biopsies should be performed is based upon the presence of clinical disease in an affected organ, the likelihood of a positive diagnostic yield from data contained in the published literature, and the risk of performing a biopsy in an affected site. Common sites where biopsies may be performed include the lung, kidney, and skin. Other sites such as sural nerve, brain, testicle, and gastrointestinal tissues may also demonstrate features of vasculitis and be appropriate locations for biopsy when clinically affected.

Surgical biopsies of radiographically abnormal pulmonary parenchyma, have a diagnostic yield of 90% in patients with granulomatosis with polyangiitis (Wegener's), and play an important role in ruling out infection or malignancy. The yield of lung biopsies is highly associated with amount of tissue that can be obtained, and transbronchial biopsies, while less invasive, have a yield of only 7%. Lung biopsies also play an important role in microscopic polyangiitis, Churg-Strauss syndrome, and in any vasculitic disease where an immunosuppressed patient has pulmonary disease that is suspected to be an infection.

Kidney biopsy findings of a focal, segmental, crescentic, necrotizing glomerulonephritis with few to no immune complexes (pauci-immune glomerulonephritis) are characteristic in patients with granulomatosis with polyangiitis (Wegener's), microscopic polyangiitis, or Churg-Strauss syndrome, who have active renal disease. These findings not only distinguish these entities from other causes of glomerulonephritis, they can confirm the presence of active glomerulonephritis that requires treatment. Because of this, renal biopsies can also be helpful to guide management decisions in these diseases when an established patient has worsening renal function and an inactive or equivocal urine sediment. Cryoglobulinemic vasculitis and Henoch-Schönlein purpura are other vasculitides where renal involvement may occur and where biopsy may be important in diagnosis or prognosis.

Biopsies of the skin are commonly performed and are well tolerated. As not all purpuric or ulcerative lesions are due to vasculitis, skin biopsy plays an important role to confirm the presence of vasculitis as the cause of the manifestation. Cutaneous vasculitis represents the most common vasculitic feature that affects people and can be seen in a broad spectrum of settings including infections, medications, malignancies, and connective tissue diseases. Because of this, for systemic vasculitides that will require aggressive immunosuppressive treatment, a skin biopsy may not represent sufficient evidence to secure the diagnosis.

Diagnostic imaging represents a critical assessment tool in patients who are known or suspected to have a systemic vasculitic disease. Imaging contributes unique information about the patient that, when taken together with the history, physical examination, and laboratory determinations, can guide the differential diagnosis and the subsequent assessment or treatment plan. A diverse range of imaging techniques are utilized in the assessment of vasculitis including plain radiography, ultrasonography, CT, MRI, positron emission tomography, and catheter-directed dye arteriography. These procedures have specific utilities that can allow differing perspectives on the spectrum and severity of vasculitis.

For vasculitic diseases that involve the large- or medium-sized blood vessels, arteriography provides information regarding blood vessel stenoses or aneurysms that can support the diagnosis. Catheter-directed dye arteriography offers the most precise information regarding the vessel lumen but carries risks related to dye exposure and the invasive nature of the procedure. Advancements in MR and CT arteriography have brought about noninvasive options to view the lumen and vessel wall, thus enhancing the ability to perform serial studies for patient monitoring.

Although vasculitis involving the small blood vessels cannot be directly visualized, diagnostic imaging plays an essential role in detecting tissue injury that occurs as result of blood vessel and tissue inflammation. In granulomatosis with polyangiitis (Wegener's), 80% of patients may have pulmonary involvement during their disease course. Chest imaging should be obtained whenever active disease is suspected, as up to one-third of patients with radiographic abnormalities are asymptomatic. Pulmonary imaging is also important to detect complications of vasculitis therapy such as opportunistic pneumonias as well as medication-related pneumonitis.

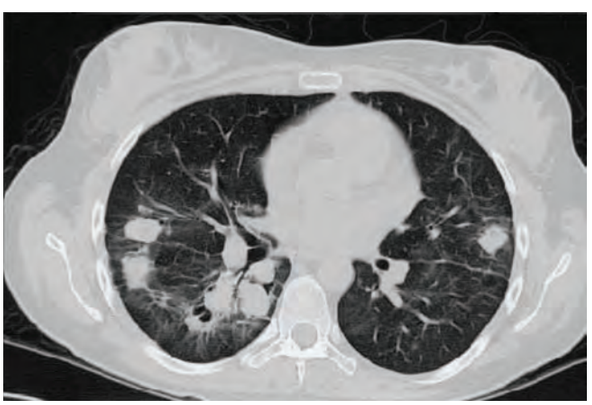

Figure e40-1 **Bilateral nodular infiltrates** seen on computed tomography of the chest in a 40-year-old woman with granulomatosis with polyangiitis (Wegener's).

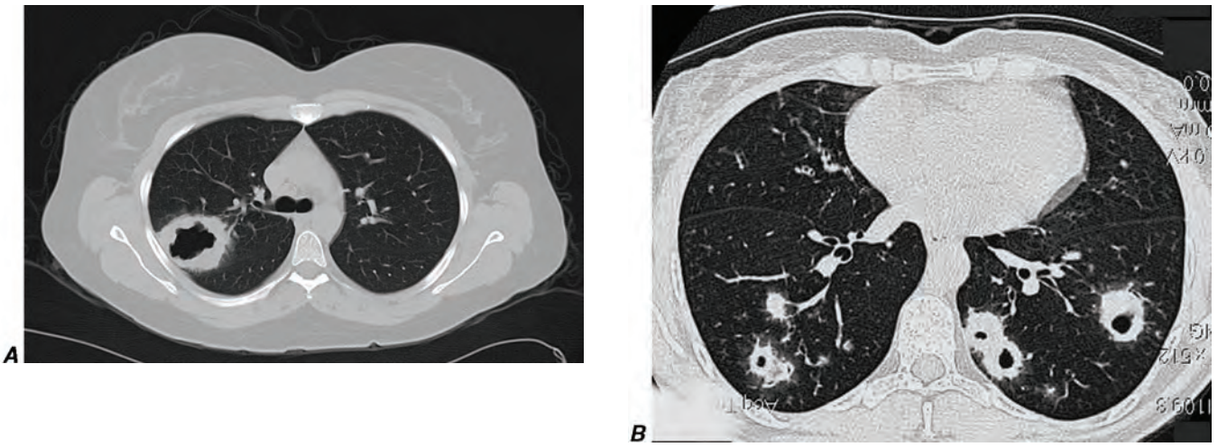

Figure e40-2 **Computed tomography of the chest** in two patients with granulomatosis with polyangiitis (Wegener's) demonstrating (***A***) single and (***B***) multiple cavitary lung lesions.

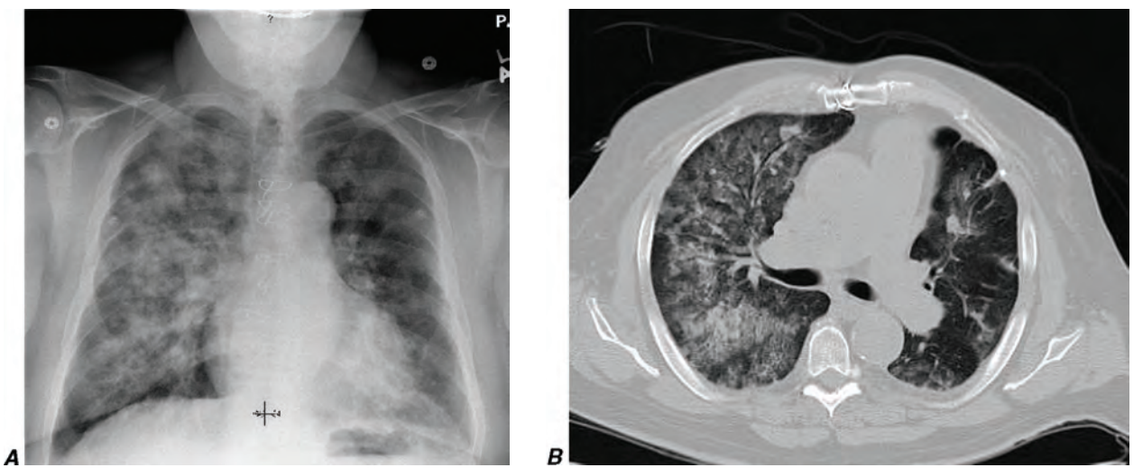

Figure e40-3 **Bilateral ground-glass infiltrates due to alveolar hemorrhage from pulmonary capillaritis as seen in the same patient by (*A*) chest radiograph and (*B*) computed tomography.** This manifestation can occur in granulomatosis with polyangiitis (Wegener's) or microscopic polyangiitis.

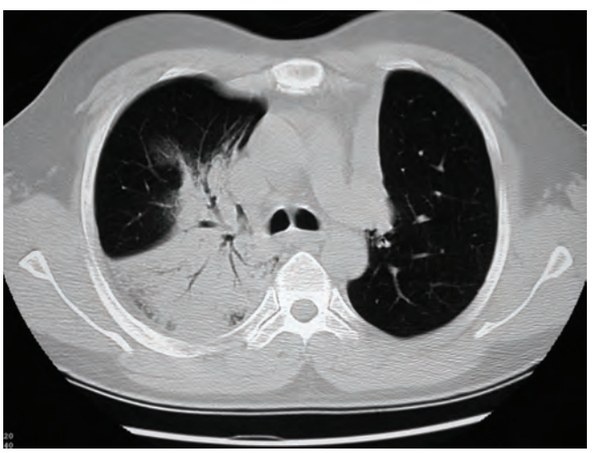

Figure e40-4 **Computed tomography of the chest demonstrating a dense infiltrate with air bronchograms involving a segment of the right upper lobe due to bacterial pneumonia in an immunosuppressed patient with granulomatosis with polyangiitis (Wegener's).** Collapse of the left upper lobe secondary to endobronchial stenosis from granulomatosis with polyangiitis (Wegener's) also is seen on this image.

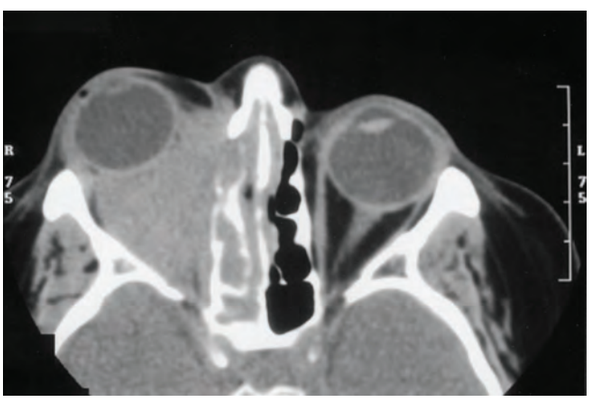

Figure e40-5 **Computed tomography of the orbits in a patient with granulomatosis with polyangiitis (Wegener's), who presented with right-eye proptosis.** The image demonstrates inflammatory tissue extending from the ethmoid sinus through the lamina papyracea and filling the orbital space.

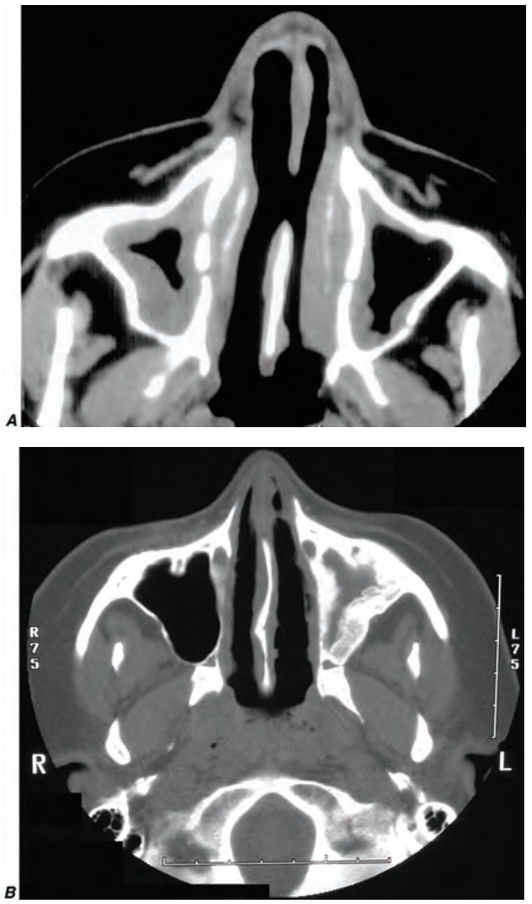

Figure e40-6 Computed tomography of the sinuses in two patients with granulomatosis with polyangiitis (Wegener's) (***A***) Mucosal thickening of the bilateral maxillary sinuses and a perforation of the nasal septum. (***B***) Osteitis with obliteration of the left maxillary sinus in a patient with long-standing sinus disease.

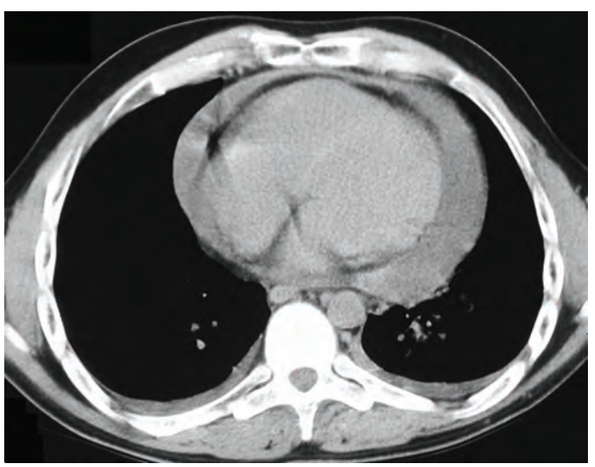

Figure e40-7 Computed tomography of the chest demonstrating a large pericardial effusion in a patient with Churg-Strauss syndrome. Cardiac involvement is an important cause of morbidity and mortality in Churg-Strauss syndrome and can include myocarditis, endocarditis, and pericarditis.

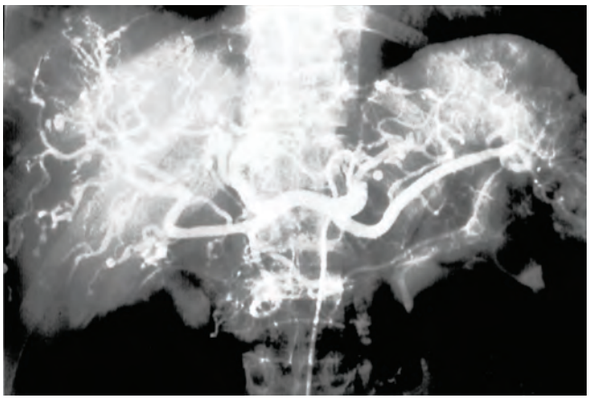

Figure e40-8 Arteriogram of a 40-year-old man with polyarteritis nodosa demonstrating microaneurysms in the hepatic circulation.

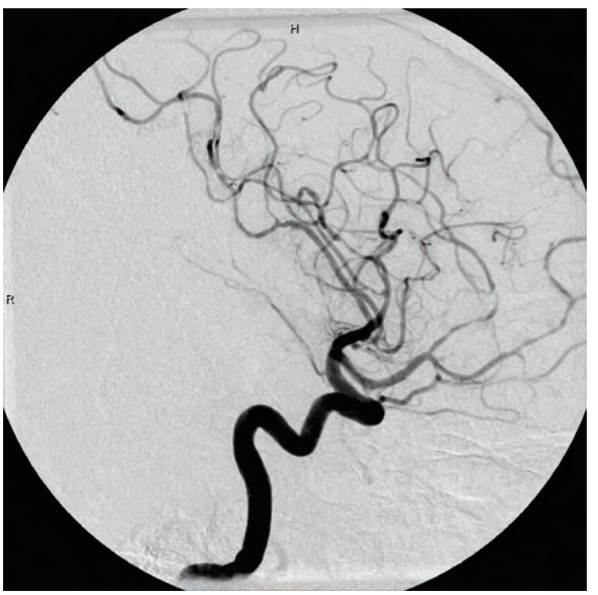

Figure e40-9 Cerebral arteriogram demonstrating beading along branches of the internal carotid artery in a patient with isolated central nervous system vasculitis.

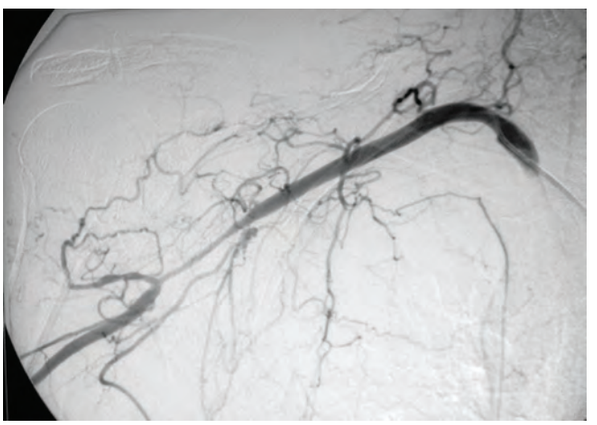

Figure e40-10 Upper-extremity arteriogram demonstrating a long stenotic lesion of the axillary artery in a 75-year-old female with giant cell arteritis.

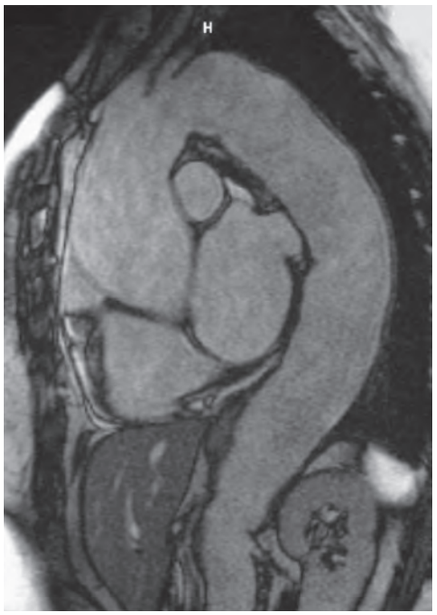

Figure e40-11 Magnetic resonance imaging demonstrating extensive aneurysmal disease of the thoracic aorta in an 80-year-old female. The patient had been diagnosed with biopsy-proven giant cell arteritis 10 years prior to presenting with this aneurysm.

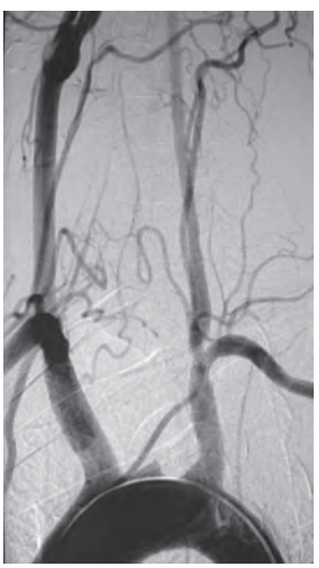

Figure e40-12 Arteriogram of the aortic arch demonstrating complete occlusion of the left common carotid artery just after its origin from the aorta. This 20-year-old female presented with syncope and was subsequently diagnosed with Takayasu's arteritis.

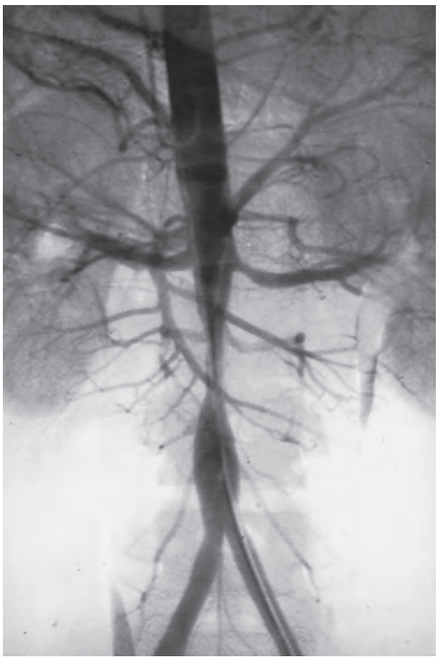

Figure e40-13 Arteriogram demonstrating stenosis of the abdominal aorta in a 25-year-old female with Takayasu's arteritis.

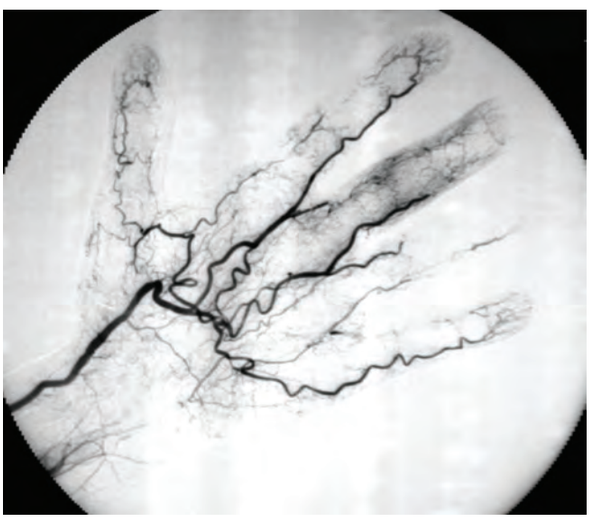

Figure e40-14 Arteriogram of the hand demonstrating arterial skip lesions and vessel cutoffs in a patient with cryoglobulinemia due to multiple myeloma.

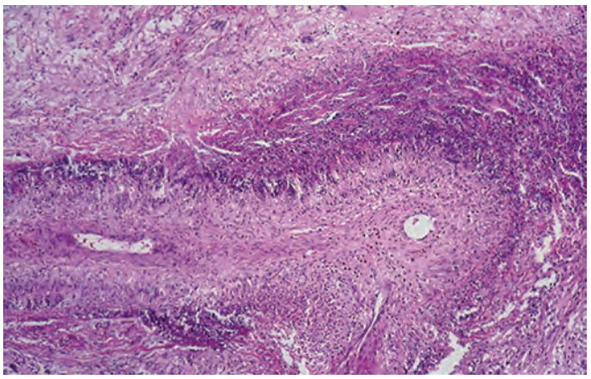

Figure e40-15 Lung histology in granulomatosis with polyangiitis (Wegener's). This lung biopsy shows areas of geographic necrosis with a border of histiocytes and giant cells. There is also vasculitis with neutrophils, lymphocytes, and giant cells infiltrating the wall of an artery.

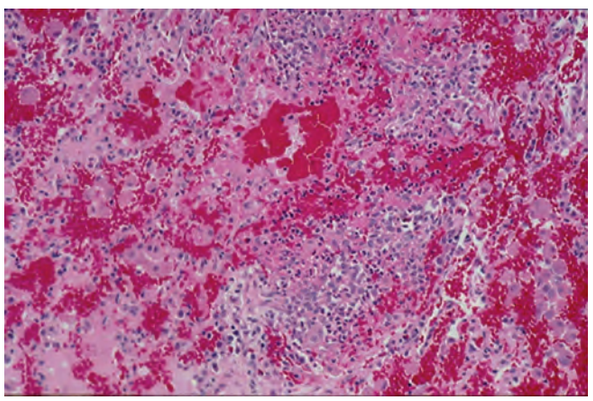

Figure e40-16 Lung histology in microscopic polyangiitis. This lung biopsy demonstrates hemorrhage in the alveolar spaces due to capillaritis in a patient with microscopic polyangiitis. Similar findings can also be seen in granulomatosis with polyangiitis (Wegener's) and less commonly in Churg-Strauss syndrome.

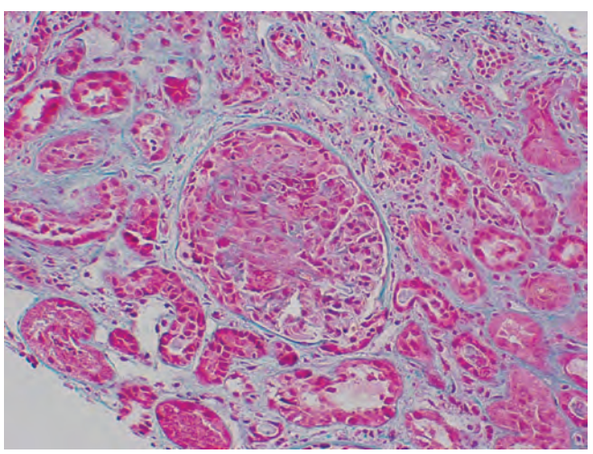

Figure e40-17 Kidney biopsy in granulomatosis with polyangiitis (Wegener's). This renal biopsy shows a crescentic and necrotizing glomerulonephritis. These findings were focal and segmental with normal and scarred glomeruli also being found in the biopsy. By immunofluorescence and electron microscopy, no immune deposits were present, indicative of a pauci-immune glomerulonephritis. Similar findings can also be seen in microscopic polyangiitis and Churg-Strauss syndrome.

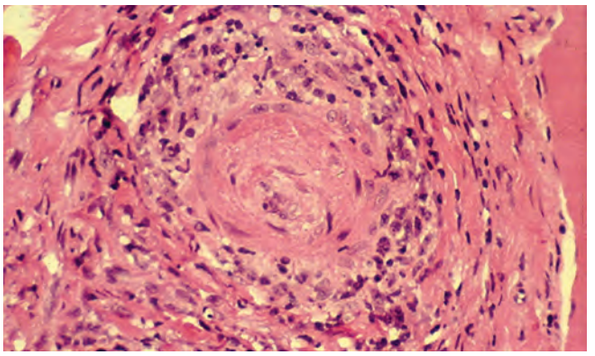

Figure e40-18 Sural nerve biopsy in polyarteritis nodosa. This sural nerve biopsy was performed in a patient with polyarteritis nodosa, who had presented with a mononeuritis multiplex. Neutrophils are seen infiltrating all layers of this medium-sized vessel, which resulted in vessel occlusion and nerve infraction.

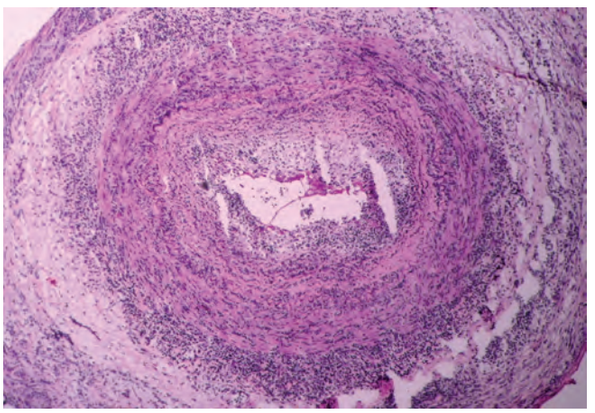

Figure e40-19 Temporal artery biopsy in giant cell arteritis. This temporal artery biopsy demonstrates a panmural infiltration of mononuclear cells and lymphocytes that are particularly seen in the media and adventitia. Scattered giant cells are also present.

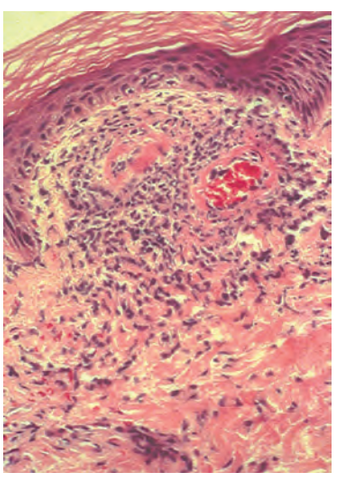

Figure e40-20 Cutaneous vasculitis. This skin biopsy reveals two arterioles beneath the dermis with a neutrophilic inflammatory infiltrate in and around the vessel wall with leukocytoclasis (nuclear debris). While such features are diagnostic of vasculitis, they can be seen in a variety of settings and are not specific for any single disease.

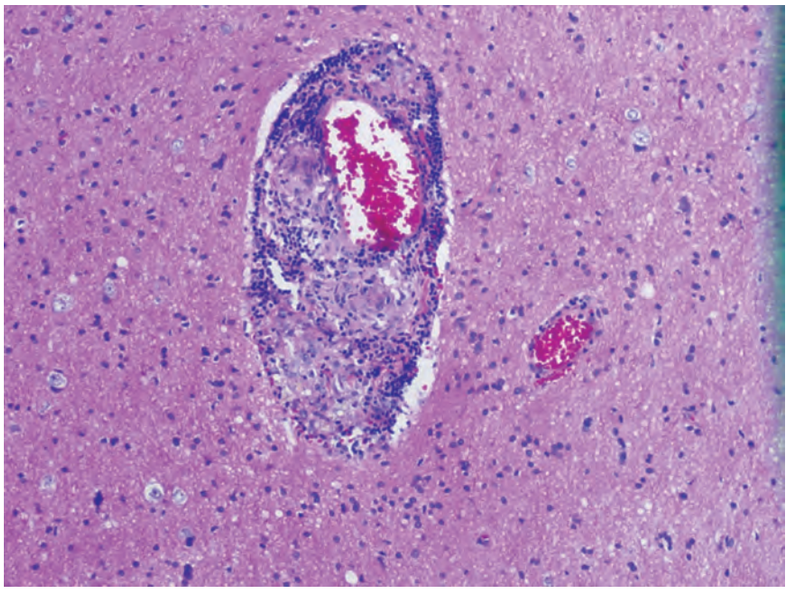

Figure e40-21 Granulomatous primary angiitis of the central nervous system. This brain biopsy demonstrates a medium-sized artery with granulomatous inflammation present within the vessel wall indicative of a granulomatous vasculitis. This patient presented with progressive headache, clinical and radiographic features of a stroke, and had arteriographic features consistent with vasculitis. As no evidence of vasculitis could be found outside of the brain, this was consistent with granulomatous primary angiitis of the central nervous system (PACNS).