

Diagnosis of the vasculitic syndromes is usually based on 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 on 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, eosinophilic granulomatosis with polyangiitis (Churg-Strauss), and 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 eosinophilic granulomatosis with polyangiitis (Churg-Strauss), who have active renal disease. These findings not only distinguish these entities from other causes of glomerulonephritis, but can also confirm the presence of active glomerulonephritis that requires treatment. As a result, 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 IgA vasculitis (Henoch-Schönlein) 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. Because 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. As a result, 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 is used in the assessment of vasculitis including plain radiography, ultrasonography, computed tomography (CT), magnetic resonance imaging (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 provides information on central blood pressure and offers the most precise detail regarding vessel lumen dimensions but carries risks related to dye exposure and the invasive nature of the procedure. Advancements in magnetic resonance (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 in large-vessel vasculitis. However, in patients suspected to have a medium-vessel vasculitis such as polyarteritis nodosa, catheter-directed dye arteriography should still be performed because MR and CT arteriograms do not currently have sufficient resolution to visualize arteries of this size.

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, because 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 and medication-related pneumonitis.



**FIGURE 386e-1** Bilateral nodular infiltrates seen on computed tomography of the chest in a 40-year-old woman with granulomatosis with polyangiitis (Wegener's).