Vasculitis (Polyarteritis Nodosa, Microscopic Polyangiitis, Wegener’s Granulomatosis, Henoch-Schönlein Purpura)

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Ronald J. Falk

The kidneys are affected by a variety of systemic vasculitides [1,2]. This is not surprising given the numerous and varied types of vessels in the kidneys. The clinical manifestations and even the pathologic expressions of vasculitis often are not specific for a particular diagnostic category of vasculitis. An accurate precise diagnosis usually requires the integration of many different types of data, including clinical signs and symptoms, associated diseases (eg, asthma, systemic lupus erythematosus, rheumatoid arthritis, hepatitis virus, polymyalgia rheumatica), vascular distribution (ie, types and locations of involved vessels), histologic pattern of inflammation (eg, granulomatous versus necrotizing), immunopathologic features (eg, presence and composition of vascular immunoglobulin deposits), and serologic findings (eg, cryoglobulins, hypocomplementemia, hepatitis B antibodies, hepatitis C antibodies, antineutrophil cytoplasmic autoantibodies, anti-glomerular basement membrane [GBM] antibodies, antinuclear antibodies). Specific diagnosis of a vasculitis is very important because the prognosis and appropriate therapy vary substantially among different types of vasculitis.

A general overview of the major categories of vasculitis that affect the kidneys is presented. The focus is primarily on polyarteritis nodosa, Henoch-Schönlein purpura, Wegener’s granulomatosis, and microscopic polyangiitis.
SELECTED CATEGORIES OF VASCULITIS

Large vessel vasculitis
  Giant cell arteritis
  Takayasu arteritis
  Medium-sized vessel vasculitis
  Polyarteritis nodosa
  Kawasaki disease
Small vessel vasculitis
  ANCA small vessel vasculitis
  Microscopic polyangiitis
  Wegener’s granulomatosis
  Churg-Strauss syndrome
  Immune complex small vessel vasculitis
  Henoch-Schönlein purpura
  Cryoglobulinemic vasculitis
  Lupus vasculitis
  Serum sickness vasculitis
  Infection-induced immune complex vasculitis
  Anti–GBM small vessel vasculitis
  Goodpasture’s syndrome

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M any different approaches to categorizing vasculitis exist. We use the approach adopted by the Chapel Hill International Consensus Conference on the Nomenclature of Systemic Vasculitis [3]. The Chapel Hill System divides vasculitides into those that have a predilection for large arteries (i.e., the aorta and its major branches), medium-sized vessels (i.e., main visceral arteries), and small vessels (predominantly capillaries, venules, and arterioles, and occasionally, small arteries). However, there is so much overlap in the size of the vessel involved by different vasculitides that other criteria are very important for precise diagnosis, especially when distinguishing among the different types of small vessel vasculitis. ANCA — anti-neutrophil cytoplasmic antibody.

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FIGURE 2-1

Predominant distributions of renal vascular involvement. This diagram depicts the predominant distributions of renal vascular involvement by large, medium-sized, and small vessel vasculitides [2]. Note that all three categories may affect arteries, although arteries are least often affected by the small vessel vasculitides and often are not involved at all by this category of vasculitis. By the Chapel Hill definitions, glomerular involvement (i.e., glomerulonephritis) is confined to the small vessel vasculitides, which provides a concrete criterion for separating the diseases in this category from those in the other two categories [3].
RENAL INJURY CAUSED BY DIFFERENT CATEGORIES OF VASCULITIS

Large vessel vasculitis
- Ischemia causing renovascular hypertension (uncommon)

Medium-sized vessel vasculitis
- Renal infarcts (frequent)
- Hemorrhage (uncommon)
- ANCA small vessel vasculitis
  - Pauci-immune crescentic glomerulonephritis (common)
  - Arcuate and interlobular arteritis (occasional)
  - Medullary angiitis (uncommon)
  - Interstitial granulomatous inflammation (rare)

Immune complex small vessel vasculitis
- Immune complex proliferative or membranoproliferative glomerulonephritis with or without crescents (common)
- Arteriolitis and interlobular arteritis (rare)

Anti-GBM small vessel vasculitis
- Crescentic glomerulonephritis (common)
- Extraglomerular vasculitis (only with concurrent ANCA disease)

FIGURE 2-3
The type of renal vessel involved by a vasculitis determines the resultant renal dysfunction. Large vessel vasculitides cause renal dysfunction by injuring the renal arteries and the aorta adjacent to the renal artery ostia. These injuries result in reduced renal blood flow and resultant renovascular hypertension. Medium-sized vessel vasculitides most often affects lobar, arcuate, and interlobular arteries, resulting in infarction and hemorrhage. Small vessel vasculitides most often affect the glomerular capillaries (ie, cause glomerulonephritis), but some types (especially the antineutrophil cytoplasmic antibody vasculitides) may also affect extraglomerular parenchymal arterioles, venules, and capillaries. Anti-GBM disease is a form of vasculitis that involves only capillaries in glomeruli or pulmonary alveoli, or both. This category of vasculitis is considered in detail separately in this Atlas.

Large Vessel Vasculitis

NAMES AND DEFINITIONS FOR LARGE VESSEL VASCULITIS

Giant cell arteritis
- Granulomatous arteritis of the aorta and its major branches, with a predilection for the extracranial branches of the carotid artery. Often involves the temporal artery. Usually occurs in patients older than aged 50 years and often is associated with polymyalgia rheumatica.

Takayasu arteritis
- Granulomatous inflammation of the aorta and its major branches. Usually occurs in patients younger than aged 50 years.

FIGURE 2-4
The two major categories of large vessel vasculitis, giant cell (temporal) arteritis and Takayasu arteritis, are both characterized pathologically by granulomatous inflammation of the aorta, its major branches, or both. The most reliable criterion for distinguishing between these two disease is the younger age of patients with Takayasu arteritis compared with giant cell arteritis [3]. The presence of polymyalgia rheumatica supports a diagnosis of giant cell arteritis. Clinically significant renal disease is more commonly associated with Takayasu arteritis than giant cell arteritis, although pathologic involvement of the kidneys is a frequent finding with both conditions [4,5].
Medium-sized Vessel Vasculitis

### NAMES AND DEFINITIONS FOR MEDIUM VESSEL VASCULITIS

<table>
<thead>
<tr>
<th>Disease</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Polyarteritis nodosa</td>
<td>Necrotizing inflammation of medium-sized or small arteries without glomerulonephritis or vasculitis in arterioles, capillaries, or venules.</td>
</tr>
<tr>
<td>Kawasaki disease</td>
<td>Arteritis involving large, medium-sized, and small arteries, and associated with mucocutaneous lymph node syndrome. Coronary arteries are often involved. Aorta and veins may be involved. Usually occurs in children.</td>
</tr>
</tbody>
</table>

The medium-sized vasculitides are confined to arteries by the definitions of the Chapel Hill Nomenclature System [3,6]. By this approach the presence of evidence for involvement of vessels smaller than arteries (i.e., capillaries, venules, arterioles), such as glomerulonephritis, purpura, or pulmonary hemorrhage, would point away from these diseases and toward one of the small vessel vasculitides. Both polyarteritis nodosa and Kawasaki disease cause acute necrotizing arteritis that may be complicated by thrombosis and hemorrhage. The presence of mucocutaneous lymph node syndrome distinguishes Kawasaki disease from polyarteritis nodosa.

Photograph of kidneys showing gross features of polyarteritis nodosa. The patient died from uncontrollable hemorrhage of a ruptured aneurysm that bled into the retroperitoneum and peritoneum. The cut surface of the left kidney and external surface of the right kidney are shown. The upper pole of the left kidney has three large aneurysms filled with dark thrombus. These aneurysms are actually pseudoaneurysms because they are not true dilations of the artery wall but rather are foci of necrotizing erosion through the artery wall into the perivascular tissue. These necrotic foci predispose to thrombosis with distal infarction, and if they erode to the surface of a viscus they can rupture and cause massive hemorrhage. The kidneys also have multiple pale areas of infarction with hemorrhagic rims, which are seen best on the surface of the right kidney.

Antemortem abdominal CAT scans showing polyarteritis nodosa (A–E). These are the same kidneys shown in Figure 2-6. Demonstrated are echogenic oval defects in both kidneys corresponding to the aneurysms (pseudoaneurysms), and a perirenal hematoma adjacent to the right kidney (left sides of panels) that resulted from rupture of one of the aneurysms.

(Continued on next page)
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Vasculitis (Polyarteritis Nodosa, Microscopic Polyangiitis, Wegener’s Granulomatosis, Henoch-Schönlein Purpura)

FIGURE 2-7 (Continued)
Antemortem abdominal CAT scans showing polyarteritis nodosa.

FIGURE 2-8 (see Color Plate)
Micrograph of transmural fibrinoid necrosis of an arcuate artery in acute polyarteritis nodosa. The fibrinoid necrosis results from lytic destruction of vascular and perivascular tissue with spillage of plasma constituents, including the coagulation proteins, into the zone of destruction. The coagulation system, as well as other mediator systems, is activated and fibrin forms in the zone of necrosis, thus producing the deeply acidophilic (bright red) fibrinoid material. Marked perivascular inflammation is seen, which is the basis for the archaic term for this disease, i.e., periarteritis nodosa. Note that the glomerulus is not inflamed. (Hematoxylin and eosin stain, ×200.)

FIGURE 2-9
Micrograph of extensive destruction and sclerosis of an arcuate artery in the chronic phase of polyarteritis nodosa. Severe necrotizing injury, probably with thrombosis as well, has been almost completely replaced by fibrosis. A few small residual irregular foci of fibrinoid material can be seen. Extensive destruction to the muscularis can be discerned. Infarction in the distal vascular distribution of this artery was present in the specimen. (Hematoxylin and eosin stain, ×150.)
2.6 Systemic Diseases and the Kidney

Small Vessel Vasculitis

NAMES AND DEFINITIONS FOR SMALL VESSEL VASCULITIS

<table>
<thead>
<tr>
<th>Name</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Henoch-Schönlein purpura</td>
<td>Vasculitis with IgA-dominant immune deposits affecting small vessels, ie. capillaries, venules, or arterioles. Typically involves skin, gut and glomeruli, and is associated with arthralgias or arthritis.</td>
</tr>
<tr>
<td>Cryoglobulinemic vasculitis</td>
<td>Vasculitis with cryoglobulin immune deposits affecting small vessels, ie. capillaries, venules, or arterioles, and associated with cryoglobulins in serum. Skin and glomeruli are often involved.</td>
</tr>
<tr>
<td>Wegener's granulomatosis</td>
<td>Granulomatous inflammation involving the respiratory tract, and necrotizing vasculitis affecting small to medium-sized vessels, eg. capillaries, venules, arterioles, and arteries. Necrotizing glomerulonephritis is common.</td>
</tr>
<tr>
<td>Churg-Strauss syndrome</td>
<td>Eosinophil-rich and granulomatous inflammation involving the respiratory tract and necrotizing vasculitis affecting small to medium-sized vessels, and associated with asthma and blood eosinophilia.</td>
</tr>
<tr>
<td>Microscopic polyangiitis</td>
<td>Necrotizing vasculitis with few or no immune deposits affecting small vessels, ie. capillaries, venules, or arterioles. Necrotizing arthritis involving small and medium-sized arteries may be present. Necrotizing glomerulonephritis is very common. Pulmonary capillaritis often occurs.</td>
</tr>
</tbody>
</table>

FIGURE 2-10
The small vessel vasculitides have the highest frequency of clinically significant renal involvement of any category of vasculitis. This is not surprising given the numerous small vessels in the kidneys and their critical roles in renal function. The renal vessels most often involved by all small vessel vasculitides are the glomerular capillaries, resulting in glomerulonephritis. Glomerular involvement in immune complex vasculitis typically results in proliferative or membranoproliferative glomerulonephritis, whereas ANCA disease usually causes necrotizing glomerulonephritis with extensive crescent formation. Involvement of renal vessels other than glomerular capillaries is rare in immune complex vasculitis but common in ANCA vasculitis.

FIGURE 2-11
Algorithm for differentiating among the major categories of small vessel vasculitis that affect the kidneys. In a patient with signs and symptoms of small vessel vasculitis, the type of glomerulonephritis is useful for categorization. Identification of IgA nephropathy is indicative of Henoch-Schönlein purpura. Type I membranoproliferative glomerulonephritis (MPGN) suggests cryoglobulinemia and/or hepatitis C infection, and pauci-immune necrotizing and crescentic glomerulonephritis suggest some form of ANCA-associated vasculitis [1,2]. The different forms of ANCA vasculitis are distinguished by the presence or absence of certain features in addition to the necrotizing vasculitis, ie, granulomatous inflammation in Wegener's granulomatosis, asthma and blood eosinophilia in Churg-Strauss syndrome, and neither granulomatous inflammation nor asthma in microscopic polyangiitis. Approximately 80% of patients with active untreated Wegener's granulomatosis or microscopic polyangiitis have ANCA, but it is important to realize that a small proportion of patients with typical clinical and pathologic features of these diseases do not have detectable ANCA.
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Vasculitis (Polyarteritis Nodosa, Microscopic Polyangiitis, Wegener’s Granulomatosis, Henoch-Schönlein Purpura)

**APPROXIMATE FREQUENCY OF ORGAN SYSTEM INVOLVEMENT IN SMALL VESSEL VASCULITIS**

<table>
<thead>
<tr>
<th>Organ system</th>
<th>Henoch-Schönlein purpura, %</th>
<th>Cryoglobulinemic vasculitis, %</th>
<th>Microscopic polyangiitis, %</th>
<th>Wegener’s granulomatosis, %</th>
<th>Churg-Strauss syndrome, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Renal</td>
<td>50</td>
<td>55</td>
<td>90</td>
<td>80</td>
<td>45</td>
</tr>
<tr>
<td>Cutaneous</td>
<td>90</td>
<td>90</td>
<td>40</td>
<td>40</td>
<td>60</td>
</tr>
<tr>
<td>Pulmonary</td>
<td>&lt;5</td>
<td>&lt;5</td>
<td>50</td>
<td>90</td>
<td>70</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>60</td>
<td>30</td>
<td>50</td>
<td>50</td>
<td>50</td>
</tr>
<tr>
<td>Ear, nose, and throat</td>
<td>60</td>
<td>30</td>
<td>50</td>
<td>50</td>
<td>50</td>
</tr>
<tr>
<td>Musculoskeletal</td>
<td>75</td>
<td>70</td>
<td>60</td>
<td>60</td>
<td>50</td>
</tr>
<tr>
<td>Neurologic</td>
<td>10</td>
<td>40</td>
<td>30</td>
<td>50</td>
<td>70</td>
</tr>
</tbody>
</table>

**FIGURE 2-12**

All of the small vessel vasculitides share signs and symptoms of small vessel injury in multiple different tissues; however, the frequency of involvement varies among the different diseases [1]. Combined renal and pulmonary involvement (pulmonary-renal syndrome) is most common in ANCA vasculitis, whereas combined renal and dermal involvement (dermal-renal syndrome) is most common in immune complex vasculitis. The cutaneous involvement in small vessel vasculitides usually manifests as purpura caused by venulitis, but occasionally is more nodular or necrotizing secondary to arteritis or granulomatous inflammation. Nodular cutaneous lesions, as well as neuropathies, abdominal pain, and musculoskeletal symptoms also can be caused by medium sized vessel vasculitis (eg, polyarteritis nodosa), and thus these clinical manifestations are not specific for a small vessel vasculitis; whereas glomerulonephritis, purpura, or alveolar capillaritis are.

**Henoch-Schönlein Purpura**

**FIGURE 2-13**

Cutaneous purpura in a patient with Henoch-Schönlein purpura. This clinical appearance could be caused by any of the small vessel vasculitides, and thus is not specific for Henoch-Schönlein purpura. Henoch-Schönlein purpura is the most common small vessel vasculitis in children [7]. In a young child with purpura, nephritis and abdominal pain, the likelihood of Henoch-Schönlein purpura is approximately 80%; however, in an older adult with the same clinical presentation, the likelihood of Henoch-Schönlein purpura is very low and the patient has an approximately 80% chance of having an ANCA-associated vasculitis.

**FIGURE 2-14**

Skin biopsy from a patient with small vessel vasculitis demonstrating the typical dermal leukocytoclastic angiitis pattern of venulitis that results in vasculitic purpura. This histologic lesion is nonspecific and can be a component of any of the small vessel vasculitides. Additional immunohistologic, serologic, and clinical observations are required to determine what is causing the leukocytoclastic angiitis (Figs. 2-9 and 2-10). (Hematoxylin and eosin stain.)
2.8 Systemic Diseases and the Kidney

**FIGURE 2-15**
Direct immunofluorescence microscopy demonstrating granular IgA-dominant immune complex deposits in dermal vessels, which is indicative of Henoch-Schönlein purpura. This procedure typically would show vascular IgM, IgG, and C3 cryoglobulinemic vasculitis, and little or no staining for immunoglobulins in a specimen from a patient with an ANCA vasculitis (a paucity of staining for immunoglobulins in vessel walls indicates pauci-immune vasculitis).

**FIGURE 2-16**
Direct immunofluorescence microscopy demonstrating granular, predominantly mesangial IgA-dominant immune complex deposits in a glomerulus. This is indicative of some form of IgA nephropathy, including the form that occurs as a component of Henoch-Schönlein purpura.

**FIGURE 2-17**
Electron micrograph showing mesangial dense deposits representative of the pattern of deposition seen in patients with Henoch-Schönlein purpura glomerulonephritis. The dense deposits are immediately beneath the paramesangial basement membrane.

**FIGURE 2-18**
Severe crescentic proliferative glomerulonephritis in a patient with Henoch-Schönlein purpura and rapidly progressive glomerulonephritis (Masson trichrome stain). Approximately half of patients with Henoch-Schönlein purpura have mild nephritis with hematuria and proteinuria, but less than a quarter develop renal insufficiency, and rapidly progressive glomerulonephritis is rare. Less than 10% of patients have persistent renal disease that progresses to end-stage renal disease.
Vasculitis (Polyarteritis Nodosa, Microscopic Polyangiitis, Wegener’s Granulomatosis, Henoch-Schönlein Purpura)

FIGURE 2-19
Fibrinoid necrosis obliterating the wall of an arteriole in a renal biopsy specimen from a patient with Henoch-Schönlein purpura (hematoxylin and eosin). Involvement of renal vessels other than glomeruli is rare in Henoch-Schönlein purpura.

ANCA Small Vessel Vasculitis

FIGURE 2-20 (see Color Plate)
C-ANCA staining pattern of ethanol-fixed normal human neutrophils in an indirect immunofluorescence assay of serum. Approximately 90% of C-ANCA are specific for proteinase 3 (PR3-ANCA) in specific immunochemical assays, such as enzyme immunoassay (EIA) [8-10].

FIGURE 2-21 (see Color Plate)
P-ANCA staining pattern of ethanol-fixed normal human neutrophils in an indirect immunofluorescence assay of serum. Approximately 90% of P-ANCA in patients with nephritis or vasculitis are specific for myeloperoxidase (MPO-ANCA) in specific immunochemical assays, such as EIA. P-ANCA in patients with other types of inflammatory disease, such as inflammatory bowel disease are typically not specific for MPO. Using ethanol-fixed neutrophils as substrate, nuclear staining caused by anti-nuclear antibodies (ANA) cannot be distinguished confidently from nuclear staining caused by P-ANCA. Using formalin-fixed neutrophils as substrate, P-ANCA stain the cytoplasm but ANA do not. The difference in staining pattern between ethanol and formalin fixed cells is due to the artifactual diffusion of solubilized cationic ANCA-antigens to the nucleus during substrate preparation of the ethanol-fixed cells, as opposed to immobilization of the antigens in the cytoplasm by covalent crosslinking during formalin fixation.
2.10 Systemic Diseases and the Kidney

**FIGURE 2-22**
Approximate relative frequency of P-ANCA/M PO-ANCA versus C- ANCA/PR 3-ANCA in patients with pauci-immune necrotizing and crescent glomerulonephritis without systemic vasculitis ("renal-limited vasculitis"), microscopic polyangiitis, and Wegener’s granulomatosis. Note that most patients with renal-limited disease have P-ANCA/M PO-ANCA, most patients with Wegener’s granulomatosis have C-ANCA/PR 3-ANCA, and patients with microscopic polyangiitis do not have a major preponderance of either ANCA specificity.

**FIGURE 2-23** *(see Color Plate)*
Early segmental fibrinoid necrosis and infiltration by neutrophils in an ANCA-positive patient with Wegener’s granulomatosis (Masson trichrome stain). There also is fibrin (red/fuchsinophilic material) in Bowman’s space, which is a precursor event to crescent formation.

**FIGURE 2-24**
Glomerulus from a patient with ANCA and a pauci-immune necrotizing and crescentic glomerulonephritis showing a large circumferential crescent and segmental lysis of glomerular basement membranes (combined Jones silver and hematoxylin and eosin stain). Also note the adjacent tubulointerstitial inflammation, which often is pronounced in ANCA disease. This pattern of glomerular injury can be seen with any of the ANCA-small vessel vasculitides.

**FIGURE 2-25** *(see Color Plate)*
Direct immunofluorescence microscopy demonstrating intense staining of a crescent and adjacent segmental glomerular fibrinoid necrosis with an antiserum specific for fibrin in a renal biopsy from a patient with ANCA small vessel vasculitis. There was no staining of glomeruli in this specimen with antisera specific for IgG, IgA, or IgM.
Vasculitis (Polyarteritis Nodosa, Microscopic Polyangiitis, Wegener’s Granulomatosis, Henoch-Schönlein Purpura)

2.11

**FIGURE 2-26**
Chronic ANCA-associate glomerulonephritis with effacement of the architecture of a glomerulus by extensive sclerosis. Bowman’s capsule has been destroyed and there is periglomerular fibrosis and chronic inflammation.

**FIGURE 2-27**
Necrotizing arteritis involving an interlobular artery in a renal biopsy specimen from a patient with ANCA-positive microscopic polyangiitis (hematoxylin and eosin). There is focal transmural fibrinoid necrosis with intense perivascular inflammation. This pattern of arteritis is nonspecific, and could be seen, for example, in a patient with polyarteritis nodosa, microscopic polyangiitis, or Wegener’s granulomatosis. The presence of ANCA or glomerulonephritis in the patient would exclude polyarteritis nodosa.

**FIGURE 2-28**
Direct immunofluorescence microscopy demonstrating intense staining of the fibrinoid necrosis in the wall of an interlobular artery with an antiserum specific for fibrin in a renal biopsy from a patient with microscopic polyangiitis.

**FIGURE 2-29**
Medullary leukocytoclastic angiitis involving vasa recta in a patient with Wegener’s granulomatosis (hematoxylin and eosin). When this process is severe, papillary necrosis may result. The frequency of this process is unknown because the medulla often is not sampled in renal biopsy specimens.
2.12 Systemic Diseases and the Kidney

**FIGURE 2-30**
Poorly defined focus of necrotizing granulomatous inflammation in the cortex in a renal biopsy obtained from a patient with ANCA-positive Wegener's granulomatosis (hematoxylin and eosin). Granulomatous inflammation is only very rarely observed in renal biopsy specimens.

**FIGURE 2-31**
Necrotizing granulomatous inflammation in a wedge biopsy of lung from a patient with Wegener's granulomatosis (hematoxylin and eosin). Note the scattered large multinucleated giant cells on the left side and the extensive necrosis and neutrophilic infiltration on the right side. The granulomatous inflammation of acute Wegener's granulomatosis has much more neutrophilic infiltration and liquefactive necrosis than most other forms of granulomatous inflammation, which is why the lesions in the lung tend to cavitate, and why the lesions in the nose and sinuses tend to destroy bone.

**FIGURE 2-32**
Hemorrhagic alveolar capillaritis in a wedge biopsy from the lung of a patient with microscopic polyangiitis (hematoxylin and eosin). Note the neutrophils within alveolar capillaries and the massive hemorrhage into the air spaces. This pattern of injury can be seen in both microscopic polyangiitis and Wegener's granulomatosis. The pulmonary hemorrhage of anti-GBM disease usually does not have conspicuous neutrophils in alveolar capillaries.

**FIGURE 2-33**
Categorization of patients with crescentic glomerulonephritis with respect to both the immunopathologic category of disease (immune complex versus anti-GBM versus ANCA) and the clinicopathologic expression (glomerulonephritis alone versus Wegener's granulomatosis versus Goodpasture's syndrome versus other small vessel vasculitis) [11]. Note that most patients with ANCA have some expression of systemic vasculitis rather than glomerulonephritis alone. Most patients with Wegener's granulomatosis have C-ANCA/PR3-ANCA but some have P-ANCA/MPO-ANCA. Also note that some patients with anti-GBM and some patients with immune complex disease also are ANCA positive. (Adapted from Jennette [11]).
Vasculitis (Polyarteritis Nodosa, Microscopic Polyangiitis, Wegener’s Granulomatosis, Henoch-Schönlein Purpura)

References