Lung Vasculitis and Alveolar Hemorrhage: Pathology

Gregory A. Fishbein, B.A.1 and Michael C. Fishbein, M.D.2

ABSTRACT

Pulmonary vasculitides are a diverse group of limited and systemic disorders associated with inflammation of pulmonary vessels and parenchyma. These diseases often have distinctive clinical, serological, and histopathological features—extrapulmonary sites of involvement, circulating autoantibodies, predispositions for small or large vessels, and others. Some have characteristic inflammatory lesions; others are characterized by the absence of such lesions. Frequently pathological findings overlap, rendering classification, and diagnosis a challenge.

The anti-neutrophil cytoplasmic antibody (ANCA)-associated small-vessel diseases constitute the major pulmonary vasculitides. These include Wegener granulomatosis (WG), Churg Strauss syndrome (CSS), and microscopic polyangiitis (MPA). Less frequently, diseases such as polyarteritis nodosa, Takayasu arteritis, Behçet syndrome, and connective tissue diseases may involve pulmonary vessels, but these entities are better associated with extrapulmonary disease.

Diffuse alveolar hemorrhage (DAH) is a severe manifestation of pulmonary vasculitis. DAH is most commonly seen in small-vessel vasculitides, specifically MPA and WG. Other syndromes associated with DAH include Goodpasture syndrome, Henoch-Schönlein purpura, and systemic lupus erythematosus. Less commonly, DAH may be secondary to infection or drugs/toxins. Furthermore, in the absence of discernable systemic disease, DAH may be idiopathic—referred to as isolated pulmonary capillaritis (IPC) or idiopathic pulmonary hemosiderosis (IPH), depending on the presence of capillaritis.

KEYWORDS: Vasculitis, Wegener granulomatosis, microscopic polyangiitis, Churg-Strauss syndrome, diffuse alveolar hemorrhage

Literally, vasculitis simply means inflammation of a vessel. However, vasculitis is rarely that simple. In reality, vasculitides are complex and highly variable syndromes. The pathology of a vasculitis is frequently not limited to vessel inflammation. In some cases, vasculitides exhibit phases during which vessel inflammation may not even be present.1,2 Historically, vasculitides have been classified by their histopathology. In practice, they are often diagnosed clinically. Moreover, as laboratory tests become more sophisticated, vasculitides may be classified serologically. Unfortunately, most of these clinical, histopathological, and laboratory features are nonspecific. Therefore, the best attempts at classification have involved all three. The result is an...

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Vasculitides can be categorized based on several features:

1. **Etiology**—primary versus secondary
2. **Vessel type**—arteries, veins, capillaries, lymphatics
3. **Vessel size**—small, medium, large
4. **Inflammatory features**—acute versus chronic, eosinophilic, granulomatous, necrotic, leukocytoclastic, and so forth
5. **Distribution of involvement**—systemic versus limited, organs typically affected
6. **Laboratory findings**—autoantibodies, erythrocyte sedimentation rate, peripheral eosinophilia, and so forth
7. **Clinical history**—rash, asthma, hemoptysis, hematuria, myalgia, claudication, and so forth

Each descriptor is not without its shortcomings. The etiology may be uncertain—perhaps idiopathic, but with risk factors such as family diathesis or history of viral infection. Commonly, multiple types of vessels are affected. Multiple sizes of vessels are involved. Moreover, our definitions of *small*, *medium*, and *large* are arbitrary (some vasculitides have different opinions of what *small* means). Inflammatory features can vary depending on the site of involvement. At times, we resort to accommodating atypical histopathology by inventing variants, for example, bronchiolitis obliterans organizing pneumonia (BOOP)-like variant or eosinophilic variant. Multiple organs may be involved, and there are myriad case reports describing unusual sites of involvement. Laboratory findings may only be markers of disease; loose associations are common. Ultimately, the clinical findings of vasculitis may be overwhelmingly nonspecific (eg, malaise, myalgia, rash). Furthermore, signs and symptoms at presentation represent only a snapshot of the clinical picture, particularly if the disease is chronic or phasic.

Pulmonary vasculitides are as resistant to categorization as any vasculitis. The following text is intended to be descriptive; it is not an attempt at strict classification. Furthermore, though all pulmonary vasculitis is uncommon, there are numerous rare pulmonary vasculitides that will not receive attention. The entities discussed hereafter are enumerated in Table 1.

### Table 1 Vasculitides That Affect the Lung and Diffuse Alveolar Hemorrhage Syndromes

<table>
<thead>
<tr>
<th>Vasculitis commonly involving the lung</th>
<th>Wegener granulomatosis</th>
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<td>Churg-Strauss syndrome</td>
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<td>Microscopic polyangiitis</td>
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<td>Vasculitis uncommonly involving the lung</td>
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<td>Takayasu arteritis</td>
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<td>Diffuse alveolar hemorrhage syndromes</td>
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<td>Wegener granulomatosis</td>
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<td>Goodpasture syndrome</td>
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<td>Systemic lupus erythematosus</td>
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<td>Henoch-Schönlein purpura</td>
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<td>Idiopathic pulmonary hemosiderosis</td>
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<td>Isolated pulmonary capillaritis</td>
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<td>Infection</td>
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<td>Drugs/toxin reaction</td>
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The list above is limited to entities discussed in the text; it is not exhaustive.

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**WEGENER GRANULOMATOSIS**

Wegener granulomatosis (WG) is a necrotizing small vessel vasculitis most commonly affecting the upper and lower respiratory tracts and the kidneys. The pathologist plays an important role in the clinical management of WG because the diagnosis of this disorder requires pathological confirmation.

Grossly, the lungs frequently exhibit bilateral nodular masses, roughly 2 to 3 cm in diameter. The nodules feature irregular borders and areas of central necrosis, appearing dark yellow to red. Areas of consolidations and hemorrhage may be visible, but the surrounding parenchyma is often unremarkable. Some
specimens lack nodules but exhibit features of associated pathology, such as diffuse alveolar hemorrhage, interstitial fibrosis, and lipid pneumonia.

Microscopically, WG is characterized by three major lesions: parenchymal necrosis, vasculitis, and granulomatous inflammation (Fig. 1). The majority of lung biopsies will demonstrate some mixture of these three lesions. The most striking of the three is parenchymal necrosis. Predominant patterns include neutrophilic microabscesses and geographic necrosis. Necrotic areas have characteristic serpentine borders and granular centers. They are often described as blue necrosis, due to the large amounts of basophilic nuclear debris. Peripherally, histiocytes and scattered giant cells surround the necrosis in a manner some describe as a cartwheel pattern.

WG is a vasculitis of small vessels, under 5 mm in diameter, including arteries, venules, and capillaries.

**Figure 1** Wegener granulomatosis with a variety of vascular and parenchymal lesions. (A) Pulmonary capillaritis with alveolar hemorrhage (hematoxylin and eosin stain [H&E], 40×). (B) Venulitis with fibrinoid necrosis of the vein wall (arrow) (H&E, 200×). (C) Arteritis with lymphocytic infiltrate and prominent intimal hyperplasia (H&E, 100×). (D) Arteritis with eosinophils present (H&E, 200×). (E) Granulomatous arteritis with multinucleated giant cell in lumen of artery (H&E, 100×). (F) "Blue necrosis" with adjacent giant-cell in lung parenchyma (arrow) (H&E, 40×).
Vasculitic areas are typically found adjacent to major inflammatory lesions. Arteries and veins exhibit cellular chronic inflammatory infiltrate, neutrophils, fibrinoid necrosis, and poorly formed granulomas. Neutrophilic capillaritis and diffuse alveolar hemorrhage can be present. Cicatricial changes such as intimal proliferation and medial scarring are common.

Extravascular granulomas accompanied by mixed inflammatory cells are characteristic of WG. Lesions composed of lymphocytes, plasma cells, neutrophils, and eosinophils accompany poorly formed collections of palisading histiocytes and scattered giant cells. Often the infiltrate surrounds microabscesses. Sarcoid-like non-necrotizing granulomas are present in rare cases.5

Other minor histological features are frequently found in lung biopsies. These are typically nondiagnostic and are peripheral to major inflammatory lesions. Parenchymal tissue demonstrates changes such as interstitial fibrosis, lymphoid aggregates, alveolar hemorrhage, and organizing pneumonia. Foamy macrophages are frequently present, giving the appearance of endogenous lipid pneumonia. Xanthogranulomatous lesions appear with prominent cholesterol clefts. Tissue eosinophils may be abundant in some cases, and focal areas of eosinophilic pneumonia may even be seen. This eosinophilic variant of WG closely mimics Churg-Strauss disease but typically is not associated with peripheral eosinophilia.6 Additionally, eosinophilic pleuritis is present in roughly half of biopsies. Bronchiolar involvement is not uncommon, though the pathology in WG is typically considered angiocentric. Acute and chronic bronchiolitis and bronchiolitis obliterans can be seen along with prominent organizing pneumonia, representing a cryptogenic organizing pneumonia (COP)-like variant (i.e., BOOP) of WG.7

CHURG-STRAUSS SYNDROME
Churg-Strauss syndrome (CSS), also called Churg-Strauss angiitis or Churg-Strauss granulomatosis, is a disease characterized by asthmatic bronchitis, peripheral eosinophilia, and vasculitis. These features tend to occur in phases, and the disease usually progresses in the order listed earlier.1 Histologically, there are three major pathological findings: vasculitis, tissue eosinophilia, and granulomatous inflammation (Fig. 2). However, given the phasic nature of the disease course, a single biopsy specimen is unlikely to exhibit all three features.5 Therefore, in the context of strong clinical and laboratory evidence, a diagnosis of CSS can be made even in the absence of definitive histology.8

The pulmonary histopathological findings most characteristic of CSS are asthmatic bronchitis, eosinophilic pneumonia, extravascular granulomatous inflammation, and necrotizing vasculitis of small and medium-sized arteries and veins. Typical findings of asthmatic bronchitis include eosinophilic inflammation, subepithelial basement membrane thickening, submucosal gland hyperplasia, and smooth muscle hypertrophy of airways. Additionally, edema and mucous plugging are common. Eosinophilic pneumonia is quite common, featuring numerous intraalveolar eosinophils and mixed degrees of organizing fibrosis. The granulomas of CSS have a unique appearance. Often called allergic granulomas or eosinophilic abscesses, they consist of extravascular collections of palisading histiocytes and multinucleated giant cells around a central core of necrotic eosinophils. Allergic granulomas may involve the vessels but are more often seen extravascularly. As the granulomas heal, they become fibrotic and calcified. Vasculitis in CSS chiefly affects small and medium-sized arteries and veins, but capillaries may be involved as well. Fibrinoid necrosis and transmural chronic inflammation are typical. Numerous eosinophils are present, along with epithelioid cells, multinucleated giant cells, and occasional neutrophils. Alveolar hemorrhage secondary to eosinophilic capillaritis may occur.1

Extrapulmonary pathology is of notable importance in CSS. A major cause of morbidity and mortality in CSS is cardiac involvement, affecting nearly half of patients.9 Eosinophilic and granulomatous myocarditis may be present, which can manifest as heart failure or conduction disturbance. Interstitial fibrosis may be the only finding, depending on the chronicity of the disease. The pericardium commonly demonstrates acute fibrinous pericarditis and pericardial fibrosis.10 Myocardial infarction due to coronary arteritis may also occur.11 Virtually all organs may be affected, but the more common are the gastrointestinal tract,12 skin, and liver. Renal disease is not prominent in CSS but has been reported to occur in 25% of patients.14 Histologically, CSS may be difficult to distinguish from other pulmonary diseases in which eosinophils are a prominent feature. These include the eosinophilic variant of WG,6 parasitic infection, and drug-induced vasculitis—all may demonstrate marked tissue eosinophilia, vasculitis, and granulomatous inflammation. Close attention to history and judicious use of laboratory tests are imperative. Indeed, one must interpret ANCA titers with caution because both CSS and WG may present with p-ANCA positivity, c-ANCA positivity, or neither.14 Cavitating lesions, however, are common in WG and rare in CSS. Furthermore, peripheral eosinophilia greater than 5% is unusual for WG.

MICROSCOPIC POLYANGIITIS
Once inaptly referred to as microscopic polyarteritis, microscopic polyangiitis (MPA) is a necrotizing vasculitis of arterioles, venules, and capillaries. Medium-sized arteries may also be affected. While almost all patients
have glomerulonephritis, the majority of cases of MPA also involve the lung, and, like WG, major extrapulmonary sites of involvement include the kidney and head and neck. Clinically, MPA can present similarly to WG. Histologically, however, MPA differs from WG in that granulomatous inflammation is not seen. Hence, biopsy is often crucial in distinguishing the two diseases. It should be noted that MPA is the most common cause of pulmonary-renal syndrome.

The two major pathological features of MPA are neutrophilic capillaritis and pulmonary hemorrhage (Fig. 3). Alveolar hemorrhage is the predominant feature, with capillaritis present in scattered patchy foci. Accompanying the hemorrhage are nonspecific changes such as interstitial thickening, hemosiderosis, and organizing pneumonia. Hyaline membranes may be present, making MPA difficult to distinguish from hemorrhagic diffuse alveolar damage (DAD).

Figure 2  Churg-Strauss syndrome. (A) Eosinophilic arteritis with marked destruction of the arterial wall (hematoxylin and eosin stain [H&E], 100×). (B) Eosinophilic venulitis (H&E, 200×). (C) Alveolar septal capillaries distended by intraluminal eosinophils (H&E, 400×). (D) Necrotic eosinophils (eosinophilic abscess) (H&E, 200×). (E) “Asthmatic bronchitis” with thickening of the subepithelial basement membrane (asterisk), and eosinophilic bronchitis (H&E, 400×). (F) Eosinophilic/granulomatous myocarditis from a fatal case (H&E, 100×).
Figure 3  Microscopic polyangitis. (A) Neutrophilic capillaritis with acute alveolar hemorrhage (hematoxylin and eosin stain [H&E], 100x). (B) Hemosiderin-laden macrophages (arrow) in a case with chronic hemorrhage (H&E, 200x). (C) Destroyed small artery with arteritis and luminal thrombosis (T) (H&E, 200x).
Necrotizing vasculitis may affect medium and small arteries, venules, and capillaries—in contrast to polyarteritis nodosa, in which veins and small vessels are spared. Granulomatous inflammation and eosinophilic infiltrate are absent, distinguishing MPA from other ANCA-associated vasculitides such as WG and CSS.

POLYARTERITIS NODOSA
Polyarteritis nodosa (PN) is a necrotizing vasculitis of medium-sized arteries. Like MPA, PN lacks both granulomatous and eosinophilic inflammation. However, in PN, arterioles, capillaries, and venules are spared. PN more likely affects bronchial arteries than pulmonary arteries. The fact remains, however, that polyarteritis nodosa rarely affects the lung. It is believed that many
previous reports of pulmonary PN would be considered MPA by current criteria.

**Takayasu Arteritis**

Takayasu arteritis, also called aortic arch syndrome, young female arteritis, and “pulseless disease,” is a large artery vasculitis that primarily affects the aorta and its major branches. Pulmonary and bronchial arteries, however, may be involved, and lung involvement has been linked to the Bw52/Dw12 haplotype. Histologically, Takayasu arteritis appears as chronic inflammation of large elastic arteries with scattered giant cells, predominantly in the outer two thirds of the vessel wall. Typically, there is marked intimal and adventitial scarring. Major complications include stenosis and aneurysm, at a ratio of almost 4:1, respectively. Fistula formation is a rare but deadly complication; pulmonary artery to bronchial artery, and pulmonary artery to coronary artery fistulas have been reported.

**Behçet Syndrome**

Behçet syndrome (pronounced beh ‘chet) is a vasculitis affecting vessels of virtually all sizes. The disease is characterized by the classic triad of oral ulcers, genital ulcers, and uveitis. Other organ systems may be affected, including the central nervous system, skin, gut, and lung. The major histological finding is a necrotizing lymphocytic vasculitis involving small, medium, and large arteries and veins, and capillaries. The most common pulmonary manifestation is pulmonary artery aneurysms, typically presenting as hemoptysis. The aneurysms may be single or multiple, unilateral or bilateral. Thrombi within the aneurysms are common.

**Miscellaneous**

Vasculitis in the lung may be found in virtually all collagen-vascular diseases, even those primarily causing soft tissue injury or those associated with pulmonary interstitial or airways disease. In cryoglobulinemia, pulmonary lesions are not uncommon, with small vessel leukocytoclastic vasculitis or medium-sized artery involvement. In sarcoidosis, classic or necrotizing, granulomatous vasculitis may be seen in systemic and pulmonary arteries. Éven in inflammatory bowel diseases, such as ulcerative colitis, pulmonary vasculitis has been described.

Furthermore, there are several disorders in which the pulmonary vessels are secondarily involved. In lymphoproliferative disorders affecting the lung, pulmonary blood vessels may be infiltrated by the lymphoid cells or may even be necrotic. Certain angioinvasive bacteria and fungi, such as *Aspergillus* and *Phycomyces*, cause vasculitis, vascular necrosis, and vascular occlusion with pulmonary infarcts. Foreign materials injected by intravenous drug abusers can cause a granulomatous vasculitis. Toxic substances, such as L-tryptophan, are associated with eosinophilic vasculitis.

**DIFFUSE ALVEOLAR HEMORRHAGE**

Pulmonary hemorrhage is a serious complication of an extensive variety of diseases and drugs. It can be classified as acute or chronic, localized or diffuse. When the process is diffuse, one can expect a relatively stereotyped histopathology, independent of the etiology (Fig. 5). Diffuse alveolar hemorrhage (DAH) is commonly immunologically mediated, often secondary to vasculitis. Indeed, the presence or absence of immune-complex deposition or capillaritis is an important clue used to distinguish various etiologies of DAH. However, to arrive at a specific diagnosis invariably requires clinical, and often serological, patient information (Table 2).

Biopsy can assist in identifying the specific cause of DAH. It should be emphasized that the diagnosis of DAH does not necessitate an open biopsy. Indeed, open biopsies provide the highest-yield specimens, but transbronchial and transthoracic needle core biopsies have proven sufficient in the majority of cases. Upon microscopic evaluation, one can anticipate the presence of extravascular blood. However, when the presence of intact red cells is the dominant feature, the most likely cause is trauma related to the biopsy procedure. In contrast, hemosiderin-laden macrophages are reliably present in DAH. Macrophages may appear as quickly as within 2 days and may persist for several weeks. Additional signs of “real” pulmonary hemorrhage include fibrin deposition, type II pneumocyte hyperplasia, and acute inflammation. Chronic hemorrhage may result in an organizing pneumonia pattern and vascular encrustation by hemosiderin—so-called endogenous pneumoniosis. Of course, with a clinical history of hemoptysis, one can be more confident that “real” pulmonary hemorrhage is present.

Capillaritis is an important feature of DAH, the presence of which may help narrow the differential diagnosis. Histopathologically, capillaritis consists of varying degrees of neutrophilic interstitial infiltration and fibrinoid necrosis. Nuclear debris in the interstitium may be present, representing leukocytoclasia.

The presence of immune-complex or antibody deposits also helps to distinguish the etiology of DAH. However, deposits cannot be easily assessed using ordinary light microscopy—immunofluorescence (IF) is classically the technique of choice to identify such deposits. Whereas IF is an invaluable diagnostic technique in renal pathology, it is not routinely performed in the context of DAH. Nonetheless, DAH manifests in three patterns identifiable by IF: (1) hemorrhage due to deposition of

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anti–basement membrane antibodies (ABMAs), (2) hemorrhage due to immune-complex deposition, and (3) hemorrhage with little or no deposits. ABMA disease, or Goodpasture syndrome, produces characteristic linear deposits, whereas diseases of immune-complex deposition, such as systemic lupus erythematosus and Henoch-Schönlein purpura (HSP), appear granular. Idiopathic DAH and diseases associated with ANCA yield no findings or unimpressive granular deposits and are therefore referred to as pauci-immune.

Of the pauci-immune DAH syndromes, WG has the most characteristic histopathology. Often there is a profound neutrophilic capillaritis. The presence of microabscesses (blue necrosis) and granulomatous inflammation make the diagnosis of WG a slam dunk. More nonspecific is the histopathology MPA, in which neutrophilic capillaritis may be prominent, but granulomatous inflammation is absent. Extrapulmonary disease is more common in MPA than in WG, and MPA is strongly associated with p-ANCA. The histopathology of DAH in ABMA disease, or Goodpasture syndrome, closely resembles MPA, though capillaritis, if present, tends to be unimpressive. Serology indicating circulating ABMA is highly suggestive but may be absent in extremely rare cases. Furthermore, WG and ABMA disease may coexist.32 Diseases of connective tissue also cause DAH—none more commonly than systemic lupus erythematosus (SLE). The pathophysiology of DAH in SLE involves immune-complex deposition, therefore producing a granular pattern on IF. Capillaritis is also seen.

Table 2 Characteristics of Diffuse Alveolar Hemorrhage Syndromes

<table>
<thead>
<tr>
<th></th>
<th>Goodpasture</th>
<th>WG</th>
<th>MPA</th>
<th>SLE</th>
<th>HSP</th>
<th>IPH</th>
<th>IPC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Capillaritis</td>
<td>Minimal</td>
<td>Abundant</td>
<td>Abundant</td>
<td>Present</td>
<td>Present</td>
<td>Absent</td>
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<tr>
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<td>ANCA</td>
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<td>ANA</td>
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<td>None</td>
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</table>

ABMA, anti–basement membrane antibody; ANA, antinuclear antibody; ANCA, anti-neutrophilic cytoplasmic antibody; HSP, Henoch-Schönlein purpura; IPC, isolated pulmonary capillaritis; IPH, idiopathic pulmonary hemosiderosis; MPA, microscopic polyangiitis; SLE, systemic lupus erythematosus; WG, Wegener granulomatosis.
When extrapulmonary and serological findings are absent, DAH may be idiopathic or drug related. Idiopathic pulmonary hemosiderosis is a chronic illness that tends to affect children more than adults. Clinically, patients will have recurrent episodes of hemoptysis and are frequently anemic. Histopathological findings are completely nonspecific—hemosiderin is abundantly present and there is no capillaritis. In contrast, isolated pulmonary capillaritis is a rare form of DAH—histologically, it resembles any of the other DAH syndromes featuring capillaritis.

REFERENCES