Pulmonary Vasculitis

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KEYWORDS

- Diffuse alveolar hemorrhage • ANCA-associated vasculitis
- Granulomatosis with polyangiitis (Wegener) • Microscopic polyangiitis
- Antiphospholipid syndrome • Antiglomerular basement membrane disease
- Takayasu arteritis • Behçet syndrome

KEY POINTS

- Pulmonary vasculitis most frequently occurs in the context of antineutrophil cytoplasmic antibody (ANCA)–associated vasculitis.
- In these disorders, pulmonary capillaritis with diffuse alveolar hemorrhage is the most common manifestation of small-vessel pulmonary vasculitis.
- Treatment of ANCA-associated vasculitis should be tailored to disease severity; diffuse alveolar hemorrhage represents a severe disease manifestation and warrants aggressive induction therapy.
- Pulmonary artery involvement in large vessel vasculitis such as Behçet syndrome and Takayasu arteritis may present as aneurysmal, thrombotic, or stenotic disease.

INTRODUCTION

Systemic vasculitis refers to a clinicopathologically heterogeneous group of diseases classified most commonly by the size of the inflamed vessels and the organ systems affected. Pulmonary vasculitis encompasses inflammation in the pulmonary vasculature, with involved vessels varying in caliber from large elastic arteries to capillaries. Small pulmonary capillaries are the vessels most commonly involved in vasculitis affecting the lung.[1] The antineutrophil cytoplasmic antibody (ANCA)–associated vasculitides (AAVs), which include granulomatosis with polyangiitis (GPA, formerly Wegener granulomatosis), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome), are the small-vessel vasculitides in which pulmonary vasculitis is most frequently observed and

Disclosure Statement: The authors have nothing to disclose.
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Rheum Dis Clin N Am 41 (2015) 315–331
http://dx.doi.org/10.1016/j.rdc.2015.01.004 **rheumatic.theclinics.com**
0889-857X/15/$ – see front matter

are the major focus of this article. Vasculitic involvement of the large pulmonary vessels, as may occur in Behçet syndrome and Takayasu arteritis (TA), is also discussed.

SMALL-VESSEL VASCULITIS

Diagnosis

Clinical presentation and physical examination

Although vasculitis of small pulmonary capillaries is the most common pathologic manifestation of pulmonary vasculitis, the clinical presentation is highly variable. Capillaritis and resultant destruction of the capillary-alveolar basement membrane leads to diffuse alveolar hemorrhage (DAH), characterized by extravasation of red blood cells into the alveolar spaces. A wide-spectrum of clinical signs and symptoms are associated with DAH; patients may be asymptomatic at presentation or present with acute respiratory failure. Symptoms usually arise over the course of several days, although more subacute presentations can occur. Most patients experience hemoptysis, although approximately one-third of patients with DAH do not report this condition at presentation.[2] Cough, fever, and dyspnea are other frequently occurring presenting manifestations. Similar to the presenting symptoms, physical examination findings in patients with DAH are typically nonspecific. Pulmonary auscultation may reveal decreased breath sounds or inspiratory crackles. Because DAH is the common denominator of many disease states injuring the pulmonary capillaries, specific clinical manifestations or examination findings suggestive of an underlying systemic disorder are discussed.

Laboratory findings

Anemia or a serially decreasing hemoglobin measurement is the most common laboratory finding in DAH, reflecting the accumulation of red blood cells in the alveolar spaces.[2] Leukocytosis or elevated inflammatory markers may be present, especially if the patient has an underlying systemic vasculitis. A retrospective analysis of almost 100 patients hospitalized for an initial episode of DAH suggested that a plasma lactate dehydrogenase (LDH) level greater than 2 times the upper limit of normal was an independent risk factor for in-hospital mortality.[3] In many cases, DAH is present as part of a pulmonary-renal syndrome with concurrent glomerular disease; thus, urinalysis may reveal elevated serum creatinine or active urine sediment levels. Specific serologic tests and detectable autoantibodies, which can be helpful in diagnosis, are highlighted in the discussion of the individual disease entities.

Radiology

Demonstration of bilateral air-space consolidation or opacities is the radiographic hallmark of DAH. Patchy, bilateral pulmonary infiltrates may be present on chest radiograph. Because the chest radiograph may be normal in DAH, a high-resolution computed tomography (CT) scan of the chest is recommended in patients with suspected DAH.[4] Chest CT often demonstrates ground glass opacities (GGOs) and can simultaneously rule out other causes of pulmonary hemorrhage, such as bronchiectasis or endobronchial tumor, in a patient with hemoptysis. In DAH, GGOs are often diffuse and bilateral; however, in approximately one-quarter of patients, the opacification is restricted to dependent areas of the lower lobes.[5] Imaging may lag behind clinical improvement and may take days to weeks to resolve after cessation of acute bleeding into the alveolar space. Following acute DAH, a radiographic pattern of septal thickening, known as crazy-paving, can occur, although, like GGO, this pattern is not specific for capillaritis or DAH.[6] Recurrent DAH can result in the development of pulmonary fibrosis, which is also readily apparent on high-resolution chest CT.