Diffuse Alveolar Hemorrhage in Primary Antiphospholipid Syndrome

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Introduction Diffuse alveolar hemorrhage (DAH) is a rare manifestation of antiphospholipid syndrome (APS). APS is primarily associated with thrombotic complications, and most patients are systemically anticoagulated. This presents a challenge in the management of pulmonary hemostasis both in the acute phase and maintenance of remission. Case Presentation A 55 year-old gentleman with a 10-year history of primary APS with recurrent deep vein thromboses on rivaroxaban, and proteinuric chronic kidney disease stage III, who presented with acute-on-chronic progressive hypoxic respiratory failure and acute tubular necrosis. Outpatient rheumatologic workup for dyspnea on exertion prior to admission was notable only for moderately elevated antinuclear antibody. Computed tomography of the chest revealed bilateral ground-glass opacities (GGOs). Bronchoscopy performed in the Intensive Care Unit revealed diffuse alveolar hemorrhage with 95% iron-positive macrophages, and was negative for infection or malignancy. He received pulse-dose steroids, improved, and was eventually discharged with supplemental oxygen and a steroid taper. Efforts to transition to steroid-sparing interventions included a brief course of rituximab, hydroxychloroquine and Intravenous Immune Globulin. He has been intermittently admitted with respiratory complaints since initial presentation and worsening renal disease prompted a renal biopsy, previously deferred due to bleeding risk. Biopsy results were consistent with thrombotic microangiopathy (TMA) and eculizumab was initiated. Serial thoracic imaging demonstrates waxing and waning diffuse GGOs that correlated with his symptoms, which are most recently resolved. His last pulmonary function testing reveals mild restriction. At this time, he remains steroid-dependent but no longer requires supplemental oxygen. Discussion In the absence of connective tissue disease, APS-associated DAH is less common than the diffuse alveolar damage that occurs with catastrophic APS. A recent review identified 91 case reports of primary DAH in APS since the first published account in 1991. The review characterizes a male-predominant cohort of patients, presenting in middle-age years with hypoxia, cough, hemoptysis, and bilateral GGOs on cross-sectional imaging. Antiphospholipid antibody-mediated capillaritis is the hypothesized mechanism of DAH in APS. Although complement deposition is rarely noted in APS-associated DAH, complement activation is an attractive therapeutic target for management of other manifestations of the disease. Eculizumab, an anti-C5 antibody, is being used as a salvage therapy for renal manifestations of APS-associated TMA. Additional investigation is required to further elucidate the mechanism of eculizumab in the lungs and its role in treating pulmonary manifestations of APS.
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