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Title: Diffuse alveolar hemorrhage caused by primary antiphospholipid syndrome

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Body: Introduction: Diffuse alveolar hemorrhage (DAH) is a severe complication of primary antiphospholipid syndrome (PAPS). We describe treatment and outcomes of DAH due to PAPS. Methods: Retrospective review of all adults evaluated at Mayo Clinic with DAH due to PAPS between 01/01/97 and 12/31/11. PAPS diagnosis met the revised Sapporo criteria. DAH was defined as bilateral pulmonary infiltrates with BAL documenting bloody return and/or >20% hemosiderin laden macrophages (HLM). Results: Seventeen patients (men=12) were identified. Median age (IQR) was 43 years (36-47). Three patients underwent lung biopsy showing capillaritis. Median % of HLM was 87% (81-98), BAL differential was predominantly neutrophilic, median 30% (18-60). All patients were treated with high doses of glucocorticoids, 6 of whom did not respond. Sixteen patients were on anticoagulation at DAH diagnosis. Number of patients treated with immunosuppressants/number that achieved remission are described as follows: Mycophenolate mofetil 7/0, azathioprine 6/0, cyclophosphamide 7/3, plasma exchange 2/0, intravenous gamma-globulin 4/1, rituximab 6/3. Only 2 patients are off glucocorticoids(all treated with rituximab). Five patients died, 4 from complications of DAH and one from complications of autologous stem cell transplant conditioning regimen for treatment of refractory DAH/ APS. Conclusions: To the best of our knowledge, we present the largest series of DAH secondary to PAPS. This disease carries a poor prognosis with limited successful therapeutic options. Glucocorticoids are first line therapy and B-cell targeted immunosuppression with cyclophosphamide or rituximab may have the highest likelihood to induce remission and should be considered early.