Catastrophic Antiphospholipid Syndrome

Gabriel Hocum  
*Providence St. Vincent, Internal Medicine Residency, Portland, Oregon*, Gabriel.Hocum@providence.org

Jeff Youker  
*Providence St. Vincent, Portland, Oregon*, jeff.youker@providence.org

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Catastrophic Antiphospholipid Syndrome (CAPS) is a rare and extreme manifestation of Antiphospholipid Syndrome (APS) that features widespread thrombotic disease affecting multiple small vessels in a short time frame. CAPS affects only 0.8% of APS patients, but when it occurs it is has a mortality rate of approximately 50%.

Most of our understanding of this condition comes from retrospective analyses of patients in the “CAPS Registry”, which in 2016 included about 500 patients. It is more common in women, and the average age of onset is 38. 60% of these patients had an underlying primary diagnosis of APS; 30% had an associated SLE diagnosis; 65% of cases had an identifiable antecedent, most commonly, infection.

Diagnostic Criteria:
1. Involvement of three or more organs, systems, and/or tissues
2. Development of manifestations simultaneously or in less than a week
3. Presence of antiphospholipid antibodies
4. Histopathologic evidence of small vessel occlusion in at least one organ or tissue

Treatment:
1. Treatment of the inciting factor if one is identifiable (e.g. antibiotics for bacterial infection)
2. Anticoagulation, usually with heparin
3. High dose intravenous steroids
4. Plasma exchange and/or IVIG
5. Other considerations: cyclophosphamide, rituximab, eculizumab

This case highlights the severity of the CAPS disease process, and the importance of early recognition and aggressive management. The primary prognostic factor for this patient, however, was not prompt diagnosis or treatment, but the extent of irreversible damage at the time of presentation.