Crohn’s Disease Causes a Catastrophe

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Abstract

Patients with Crohn’s disease are at higher risk of developing antiphospholipid antibody syndrome (APS), of which 1% develop life-threatening, rapidly progressive clotting known as catastrophic APS (CAPS). A 17-year-old male presented with intermittent bloody diarrhea and abdominal pain. He developed myopericarditis, alveolar hemorrhage, left frontal and parieto-occipital infarct, superior sagittal sinus venous thrombosis, disseminated intravascular coagulation, and a pulmonary embolus. He was treated with high-dose pulse steroids, anticoagulation, and plasma exchange. Colonoscopy revealed a flare of Crohn’s disease, and azathioprine was initiated.

Introduction

Patients with inflammatory bowel disease (IBD) are at 2-fold higher risk of developing venous thromboembolism (VTE) than are individuals without IBD.1 IBD has been established as an independent risk factor for developing VTE, especially in a hospitalized setting. Isolated VTE or VTE as part of a syndrome such as antiphospholipid antibody syndrome (APS) has been demonstrated in patients with Crohn’s disease. Catastrophic APS (CAPS) is a rare variant of APS that occurs in less than 1% of APS patients, and 50–60% of cases of CAPS occur after a precipitant such as trauma, surgery, infections, or a lupus flare.2 CAPS associated with Crohn’s disease has not been previously described.

Case Report

A 17-year-old male presented with abdominal pain, bloody diarrhea, and weight loss for 2 months. He quickly developed worsening shortness of breath and hemoptysis. Troponin was elevated (17 ng/mL), and electrocardiogram was consistent with pericarditis and suggestive of myopericarditis with pericardial effusion. Serial imaging demonstrated ground glass opacities in the lungs (Figure 1); bronchoscopy was performed, which showed alveolar hemorrhage. Two days later, the patient had an acute hemorrhagic stroke involving the left fronto-parietal region with symptoms of right-sided neglect and homonymous hemianopia (Figure 2). Cranial CT showed a cerebral venous thrombosis. The patient then developed marked thrombocytopenia with coagulopathy (PT 19.1, PTT 48.5, LDH 446 U/L, D-dimer >20 μg/mL, and fibrinogen 129 mg/dL).

Lupus anticoagulant and anti-ß2 glycoprotein antibodies were present in high titers (105 GPI). On hospital day 6, the patient developed a pulmonary embolism. Treatment for CAPS with systemic anticoagulation and intravenous steroids was initiated. However, he continued to deteriorate, and plasma exchange was initiated.

During his hospitalization, the patient continued to have intermittent rectal bleeding. Stool studies were negative for infectious etiology, but revealed an elevated calprotectin level (>2100 μg/g). Abdominal CT revealed abnormal wall enhancement and diffuse colonic wall thickening. Colonoscopy revealed chronic active ileitis in terminal ileum. A diagnosis of Crohn’s disease was made and intravenous steroids were restarted. Azathioprine
was chosen for maintenance therapy due to its dual role in treating both Crohn’s disease and CAPS. His shortness of breath, neurologic deficits, and bloody diarrhea gradually resolved over the next 2 weeks and he was discharged in good health and with recommendation of lifelong anticoagulation. He continues to do well at 1-year follow-up.

Discussion
The diagnostic criteria of CAPS consists of 4 clinical features: involvement of 3 organ systems, development of manifestations within 1 week, histopathology of small vessel occlusion, and presence of anti-ß2 glycoprotein antibodies in high titers. Early diagnosis is important for improved outcomes; however, the diagnosis can be challenging because antiphospholipid antibody test results may not be available at the early stage of the disease, and organ biopsy to demonstrate small vessel involvement is often not feasible. In addition to recognizing this rare syndrome, underlying precipitants must be identified and corrected to prevent morbidity and mortality.

Disclosures
Author contributions: A. Khan wrote the case report and is the article guarantor. Y. Natarajan performed the literature search and wrote the discussion. J. Sellin wrote the case report.

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