INTRODUCTION
Pneumatosis intestinalis (PI) refers to the radiographic finding of gas within the small or large bowel wall. It can be seen in association with gas in the portal venous system. The clinical significance of these findings is variable and depends on the patient’s etiology and initial presentation. PI can be seen anywhere in the gastrointestinal tract distal to the stomach and is caused by a myriad of conditions. We present a case of a patient admitted to the intensive care unit with PI and portal vein gas.

CASE DESCRIPTION
A 46 year-old woman with a past medical history of lupus and recent stroke presented to the emergency department (ED) with chief complaint of syncope. She was boarding a flight when she became suddenly unresponsive. She was awoken by her family and taken to the nearest ED. On arrival, she reported intermittent hematochezia and diffuse abdominal pain for a week prior to the presentation. Her past medical history was pertinent for a stroke two weeks prior to presentation for which she was started on anti-platelet agents (aspirin and clopidogrel). She was taking prednisone and had started belimumab therapy for her lupus, with the most recent treatment one month earlier.

She was afebrile with an initial blood pressure of 65/30 mm Hg, heart rate of 74 beats per minute, and respiratory rate of 30 breaths per minute. On exam, she was distressed and had mottled skin on her extremities. She was alert and oriented to person, place, and time. Cardiac and pulmonary exam was normal. Her abdomen was soft and distended with mild, diffuse tenderness to soft palpation. Guaiac stool testing was positive. Admission laboratory values revealed a hemoglobin of 6.4 g/dL, white blood cell count of 28,200 per cubic millimeter, lactate of 15.0 mmol/L, and an arterial blood pH of 7.07. Other inflammatory markers were not checked. She was given three liters of intravenous fluid in the ED and had an emergent computed tomography (CT) scan of the abdomen and pelvis. These scans showed air within the colonic wall and the portal venous system (Figures 1 and 2).

Figure 1. Coronal image of non-contrast CT scan showing air within the colonic wall.

Figure 2. Axial image of non-contrast CT scan showing air within the portal venous system.
DIFFERENTIAL DIAGNOSIS

Imaging allowed the medical team to focus the differential diagnosis on likely causes of PI in this patient. Stool, mucosal lesions, or submucosal lesions may be mistaken for PI, so making the diagnosis should involve clinical correlation and consultation with a radiologist. Once the diagnosis is confirmed, an etiology should be determined. This finding has a broad differential, which included ischemic, infectious, inflammatory, immunologic, endocrine, and iatrogenic causes. Lupus is occasionally associated with mesenteric ischemia or ischemic colitis, a known cause of pneumatosis intestinalis. Infectious colitis and intra-abdominal sepsis may present similarly and have explained her hypotension and acidosis. Inflammatory conditions of the bowel such as celiac disease have been associated with the finding as well. The use of immunosuppressive monoclonal antibody therapy may have contributed to the development of her PI and affected healing of the bowel wall. Cases have been described with bevacizumab and sunitinib. This patient was on belimumab, which inhibits B-cell activating factor. Iatrogenic PI can be seen after bowel surgery or endoscopy and typically has a benign course; this did not seem likely in this patient.

OUTCOME

Treatment with fluid resuscitation, vasopressors, packed red cell transfusion, and broad-spectrum antibiotics was continued on admission to the intensive care unit. General surgery was consulted for urgent laparotomy, but it was felt that the patient was too unstable and would not survive the surgery. She remained hypotensive despite maximum dosages of four vasopressors. She developed worsening encephalopathy and urine output ceased. Five hours after presentation, the patient had a cardiac arrest and died after receiving advanced cardiovascular life support.

DISCUSSION

PI can be found incidentally or in patients who have intra-abdominal catastrophe. This case demonstrates the rapid clinical decline that can be associated with this finding. The portal vein gas shown in Figure 2 is subtle and can be mistaken for air in the biliary tree. It is seen when there is significant bowel ischemia and portends an even worse prognosis. In critically ill patients, urgent surgical intervention is indicated as was initially sought here. In non-critically ill patients, culprit medications should be discontinued and antibiotics given until resolution of PI. Data also supports the use of an elemental diet.

Although this patient warranted surgical intervention, her rapid deterioration prevented her from undergoing surgery. This patient’s lupus was presumed to be severe, as it had recently caused a stroke despite medical therapy. A diagnosis of antiphospholipid antibody was possible, but diagnostic studies were not sent. The chronic colonic inflammation and vasculitis associated with lupus had been contributing to the ongoing bloody diarrhea and eventual ischemic colitis. The ongoing abdominal insult eventually led to her shock and death.

KEY POINTS

- Pneumatosis intestinalis seen on imaging is associated with significant mortality and concomitant portal venous gas worsens the prognosis.
- Syncope and gastrointestinal bleeding in a patient with underlying autoimmune disease may represent intra-abdominal catastrophe and merits expedited workup.

REFERENCES