Diagnostic Challenge

Answer

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The correct answer in this case is "d," appendicitis. The patient was taken to the operating room. On exploration, he had mild diffuse distension of his small bowel and no evidence of bowel ischemia. At the base of his cecum, he was found to have a focally perforated appendix with a small associated phlegmon but no abscess. The specimen was sent to pathology where it was diagnosed as acute necrotizing appendicitis with perforation and marked periappendicitis.

Portal venous gas (PVG) is frequently identified in patients seen in the emergency department. It has classically been associated with ischemic bowel disease and a poor prognosis.[1] In fact, 9 years separated the original reported case of PVG in 1955 in infants with necrotizing enterocolitis[2] from the first reported survivor in 1964, a patient with segmental necrosis of the small and large bowel.[3] Over the next decade, as more cases of PVG were reported, the outcomes still appeared poor. In the seminal review from 1978, a case series of 64 patients with PVG was presented. Of this group, 72% were found to have necrotic bowel, most underwent surgery and 75% ultimately died.[1] As a result of these historical reports, many physicians have interpreted PVG as a frequently fatal disease best treated with a surgical approach.

However, with the onset of widespread use of CT, PVG has been reported in cases involving colonoscopy, diverticulitis, massive digestive tract distension, intra-abdominal abscess, inflammatory bowel disease, trauma, gastric ulcer, upper endoscopy, intraperitoneal tumours, cholangitis, enteritis, barium enema, endoscopic retrograde cholangiopancreatography, hypertrophic pyloric stenosis, liver transplantation, fulminant hepatitis and pancreatitis, as well as in a number of other miscellaneous cases.[4–12]

Kinoshita and colleagues[5] better characterized the etiologies of PVG in a literature review of 182 cases. The authors found that PVG was still most frequently seen with bowel ischemia (43%). However, of all instances of PVG, nonischemic cases made up the majority (57%), with digestive tract dilatation (12%), intraperitoneal abscess (11%), ulcerative colitis (4%), gastric ulcer (4%), Crohn disease (4%), complications of endoscopic procedures (4%) and intraperitoneal tumour (3%) leading the way. As a result of the detection of more cases of PVG secondary to etiologies other than bowel ischemia and better modalities of treatment, patients in the review by Kinoshita and colleagues[5] had a mortality rate of only 39%, despite the (unchanged) 75% mortality rate of bowel ischemia. Furthermore, there were no deaths reported when PVG was associated with ulcerative colitis, intraperitoneal tumour, Crohn disease, cholangitis, pancreatitis or complications of endoscopic procedures.[5]

Although the number of reported cases of PVG has increased, there has been little consensus in the literature on its management. In their report on PVG, Hong and colleagues[11] present guidelines with respect to conservative and surgical approaches. In their opinion, PVG associated with inflammatory bowel disease, barium enemas, endoscopic retrograde cholangiopancreatography, colonoscopy or liver transplantation should be managed nonoperatively. All other patients should be treated surgically.[11] Hou and colleagues[13] created an algorithm with similar conclusions; the authors broadly divided PVG patients into ischemic and nonischemic groups and recommended management with urgent laparotomy or conservative therapy, respectively.[13]

Both of these decision-making trees appear to have shortcomings. How does one approach treatment of

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Submitted Dec. 18, 2009; Accepted Jan. 8, 2010

This article has been peer reviewed.

CJEM 2010;12(6):538-9

pancreatitis? Should a patient with inflammatory bowel disease and peritonitis be observed with intravenous antibiotics? In addition, our patient with appendicitis is not included in any algorithm. For this reason, we agree with the assessment of others that PVG is a radiographic finding. The clinical presentation of the patient should be the determining factor that dictates management.[5] In our review of the literature, isolated PVG associated with appendicitis has been reported only once. In this case, the patient had findings of PVG seen on ultrasonography but not on CT.[14] In the case of our patient, we had no algorithm to follow and relied on the clinical history and physical examination to develop a treatment plan. Given the numerous possible etiologies of PVG, it is important that physicians view PVG as a radiological finding and not as an indicator of prognosis or treatment modality.

Portal venous gas has traditionally been associated with ischemic bowel disease and a poor prognosis. As more cases have been reported, the differential diagnosis has broadened and includes many nonfatal conditions. As our case demonstrates and the literature confirms,[5] PVG alone (and the amount of PVG present) should not dictate management. Rather it highlights the importance of treating the patient's condition and not the PVG.

Competing interests: None declared.

Keywords: abdominal pain, portal venous gas, appendicitis

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