CASE PRESENTATION

An 87-year-old woman presented to the emergency department with a one-day history of haematuria and fever. Significant past medical history included chronic left hydroureteronephrosis secondary to a left vesicoureteric junction stricture, complicated by multiple episodes of urinary tract infection.

Clinical examination revealed a mildly tender left-sided abdominal mass with no other localising signs, peritonism or cardiorespiratory compromise. Serological markers showed signs of acute kidney injury (creatinine 148 μmol/L from a baseline of 129 μmol/L) and raised inflammatory markers (C-reactive protein 54 mg/dL and procalcitonin 0.44 ng/mL). Preliminary urinalysis confirmed gross haematuria with pyuria (red blood cell count 149 cells/μL and white blood cell count > 2,000 cells/μL).

Initial radiographs did not reveal any definitive source of infection. Unenhanced computed tomography (CT) of the abdomen and pelvis (Fig. 1) was subsequently performed. What do the images show? What is the diagnosis?

Fig. 1 Unenhanced (a) axial and (b) reconstructed coronal CT images of the abdomen and pelvis. (c) US image of the left kidney.
Medical Education

Fig. 2 Bowel ischaemia. An 84-year-old woman presented with abdominal distension. (a) Contrast-enhanced axial abdominal CT image shows diffuse portal venous gas in the liver with air-fluid levels within the portal vein (black arrow) and intrahepatic branches (black arrowhead). (b) CT image shows diffusely reduced small and large bowel wall enhancement with pneumatosis with multiple air-fluid levels in the bowel loops. No features of bowel perforation, free fluid or drainable abscess collection are identified. CT image shows critical stenosis of (c) the proximal coeliac artery (white arrow) and (d) superior mesenteric artery (white arrow). The imaging features are diagnostic of extensive bowel ischaemia involving the ascending and descending colon with associated portal venous gas secondary to critical arterial vascular compromise.

IMAGE INTERPRETATION

The unenhanced axial CT image of the abdomen and pelvis (Fig. 1a) demonstrates an atrophic left kidney with severe hydronephrosis containing a large-volume gas within the collecting system (arrows) and ureteric intramural gas (arrowheads). There is small-volume left retroperitoneal perinephric fat stranding but no focal perinephric collection or parenchymal abscess. No ureteric calculus is present.

The unenhanced coronal CT image of the abdomen and pelvis (Fig. 1b) shows multiple small-calibre branching peripheral lucencies within the liver, compatible with portal venous gas (arrow). There are numerous small gallstones within the gallbladder (not shown) but no evidence of biliary tree dilatation or other hepatobiliary abnormality.

Ultrasonographic evaluation of the kidney (Fig. 1c) confirms gross hydroureteronephrosis with renal cortical thinning.

DIAGNOSIS

Severe left emphysematous pyelonephritis complicated by hepatic portal venous gas.

CLINICAL COURSE

The patient was admitted to an acute medical ward and successfully treated with intravenous antibiotics (initially piperacillin/tazobactam and amikacin, which was subsequently de-escalated to ampicillin). Urinary decompression of the infected obstructed system was offered from the outset but declined by the patient. Urine cultures on admission grew Klebsiella pneumoniae, and culture clearance was achieved prior to the patient’s discharge two weeks later.

DISCUSSION

The presence of portal venous gas is commonly thought to be a radiological indicator of abdominal catastrophe and a harbinger of poor outcomes, especially when associated with intestinal ischaemia. Portal venous gas is most commonly attributed to gastrointestinal tract diseases, approximately 60% due to bowel ischaemia and mesenteric pathology, 15% from gastrointestinal tract inflammation and 10% secondary to bowel obstruction. The pathophysiological mechanism is attributed to damage of the gastrointestinal mucosal barrier, coupled with gaseous bowel over-distension and gas-forming bacterial proliferation, allowing the transition of gas into the mesenteric and subsequent portal venous system. No identifiable causes have been reported in up to 15% of cases.

To the best of our knowledge, there have only been four published cases of portal venous gas secondary to emphysematous pyelonephritis. Given the absence of primary bowel involvement, the pathophysiology of gas in the hepatic venous system remains unclear. Other seemingly unrelated pathologies such as chronic obstructive pulmonary disease and diabetes mellitus can also result in portal venous gas, although the exact mechanism remains unknown.

While portal venous gas, when associated with intestinal ischaemia, is a harbinger of poor outcomes, reported cases of portal venous gas-related emphysematous pyelonephritis have had better outcomes, similar to our presented case, showing clinical improvement after medical treatment without the need for surgical intervention.
It is important to distinguish portal venous gas from pneumobilia, a separate entity that has similar imaging appearances (described later in this article), as pneumobilia has a separate host of aetiological factors ranging from pathological processes such as cholangitis to iatrogenic, benign conditions such as post-sphincterotomy endoscopic retrograde cholangiopancreatography.

As part of the initial imaging workup, abdominal radiographs may reveal linear, branching lucencies overlying the hepatic parenchyma. However, the sensitivity of abdominal radiography in detecting portal venous gas is lower than that of CT. Additionally, portal venous gas may be less appreciated than pneumobilia, which typically has a greater volume of gas, rendering it more readily perceptible.

Ultrasonography (including Doppler flow imaging) has been proposed as an effective modality in the diagnosis and follow-up of portal venous gas. Ultrasonographic features include direct visualisation of mobile intraluminal hyperechogenic foci in the portal vein, with marked shadowing or reverberation and sharp bidirectional spikes superimposed on the normal portal vein flow waveform. Previous attempts to pinpoint prognostic, predictive findings on ultrasonography have yielded mixed results. Furthermore, limitations such as operator technique and patient habitus significantly impact diagnostic efficacy.

Cross-sectional imaging with CT is considered the most sensitive and informative modality in identifying portal venous gas and exploring potential aetiologies such as pneumatosis intestinalis in bowel ischaemia or generalised bowel dilatation in mechanical bowel obstruction, the former with a reported specificity of > 95%. The classical appearance of portal venous gas on cross-sectional imaging involves linear, small-calibre, branching hypodensities of gaseous density in the portal vein and its tributaries, extending to within 2 cm of the liver capsule, its location within the liver primarily explained by the centrifugal flow of portal venous blood. This phenomenon requires distinction from pneumobilia, which has similarly branching, tubular lucencies that are conversely more centrally placed, close to the porta hepatis, owing to centripetal biliary flow.

In conclusion, portal venous gas should always raise concerns for a potentially life-threatening intra-abdominal pathology. Portal venous gas is not a disease entity but rather a consequence of intra-abdominal organs drained by one of the portal vein’s tributaries. Initial differentiation from pneumobilia, coupled with evaluation of the constellation of clinical and radiological
findings, is paramount in order to arrive at the correct diagnosis and appropriately direct management.

REFERENCES
SINGAPORE MEDICAL COUNCIL CATEGORY 3B CME PROGRAMME
(Code SMJ 202205B)

Question 1. Regarding portal venous gas:
(a) It refers to accumulation of gas in the intrahepatic biliary system.
(b) Up to 15% of cases have no identifiable causes.
(c) The majority of cases are due to gastrointestinal tract diseases.
(d) Roughly 10% of portal venous gas cases are secondary to bowel ischaemia.

Question 2. The aetiology of portal venous gas are:
(a) Pregnancy
(b) Chronic obstructive pulmonary disease
(c) Bowel ischaemia
(d) Emphysematous pyelonephritis

Question 3. Regarding imaging evaluation of portal venous gas:
(a) Ultrasonography is the best imaging tool for the diagnosis of portal venous gas.
(b) Portal venous gas is often initially diagnosed on plain radiography.
(c) Computed tomography (CT) is often performed to confirm portal venous gas and investigate its causative factors.
(d) Doppler ultrasonography is ineffective in the diagnosis and follow-up of portal venous gas.

Question 4. Possible imaging features of portal venous gas on plain radiography are:
(a) Linear branching lucencies leading to hepatic parenchyma
(b) Pneumatosis intestinalis
(c) Common bile duct stent
(d) Bowel dilation

Question 5. Regarding the CT features of portal venous gas:
(a) It appears as centrally located gaseous lucencies in the liver.
(b) It appears as linear branching hypodensities extending to within 2 cm of the hepatic edge.
(c) It appears as gaseous lucencies in the portal vein and tributaries.
(d) Centripetal biliary flow is responsible for the CT features.

Doctor’s particulars:
Name in full: ____________________________ MCR no.: ____________________________
Specialty: ____________________________ Email: ____________________________

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Visit the SMJ website: http://www.smj.org.sg/current-issue and select the appropriate quiz. You will be redirected to the SMA login page.
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RESULTS:
(1) Answers will be published online in the SMJ July 2022 issue. (2) The MCR numbers of successful candidates will be posted online at the SMJ website by 29 July 2022. (3) Passing mark is 60%. No mark will be deducted for incorrect answers. (4) The SMJ editorial office will submit the list of successful candidates to the Singapore Medical Council. (5) One CME point is awarded for successful candidates. (6) SMC credits CME points according to the month of publication of the CME article (i.e. points awarded for a quiz published in the December 2021 issue will be credited for the month of December 2021, even if the deadline is in February 2022).