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Imaging assessment and clinical significance of pneumatosis in adult patients

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Abstract Gas detection in the bowel wall and in portomesenteric venous vessels in adults has long been related to intestinal infarction and poor outcome; many case reports have shown that pneumatosis may be associated with a large variety of pathological situations, ranging from absolutely benign and asymptomatic forms to abdominal catastrophes. Several studies have been conducted on this topic with different conclusions, probably due to differences in population so that the clinical value of these signs is still questioned. Intestinal pneumatosis, especially if presenting with a band-like pattern and if associated with portomesenteric vein gas, when observed in an acute abdominal setting should raise the suspicion of mesenteric infarct and prompt a careful search for other signs of intestinal involvement, so as not to miss cases of life-threatening intestinal infarct or allow them to further evolve into extensive necrosis with worse prognosis. In this review we illustrate the most relevant aspects of these debated but significant radiological signs.

Keywords Pneumatosis · Intestinal pneumatosis · Portal pneumatosis · Mesenteric pneumatosis · Mesenteric ischemia · CT · Portal vein gas · Infarct · Intestinal infarction

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Introduction

Intestinal pneumatosis (IP) consists of gas located in the bowel wall, whatever the cause or location (Fig. 1). Portomesenteric pneumatosis (PMP) is the presence of gas in the portal vein and its branches (Fig. 2). PMP was first described by Wolfe and Evans on plain radiography in an infant in [1].

These findings are now more frequently detected with computed tomography (CT), which has a higher accuracy than traditional radiography (Fig. 3). Pneumatosis can occur as a primary (or idiopathic) form in 15 % of cases or as a secondary form in 85 % of cases [2]. In adults, IP and PMP have been usually considered indicative of advanced intestinal infarction [3–5]. Nevertheless, these findings have been reported in a wide range of pathological conditions including intra-abdominal abscess, diverticular disease, pylephlebitis, inflammatory bowel disease, acute gastric or intestinal dilatation, hepatic transplant, barium

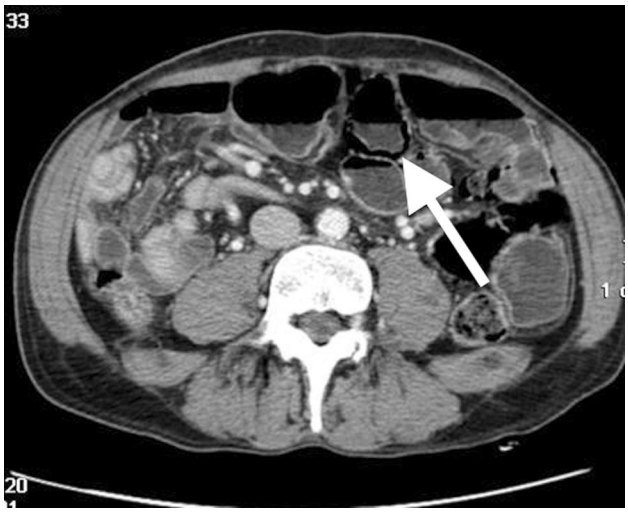

Fig. 1 Intestinal pneumatosis (IP) in small bowel intestinal infarct (*arrows*)

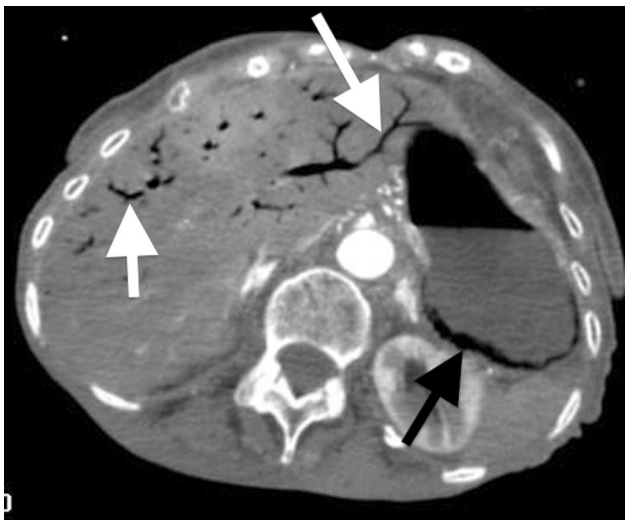

Fig. 2 Gas in portal branches (*white arrows*) in intestinal infarct. Gastric pneumatosis can also be seen (*black arrow*)

enema, umbilical catheterisation, chronic pulmonary disease, corticosteroid therapy, gastric volvulus, intestinal hernias, ingestion of corrosives, carcinoma of the colon, colonoscopy either optical or virtual, drugs, ulcerative disease, and trauma [6–25] (Table 1). Recently, IP and PMP have been described in patients treated with molecular targeted therapy, probably as a result of intestinal toxicity; all patients recovered spontaneously after discontinuation of therapy [26].

The purpose of this paper is to describe the radiological appearances of IP and PMP, and to review the clinical significance and value of these findings.

Pathogenesis

Although IP and PMP are usually discussed separately in the literature, they have always been considered to be related phenomena; in fact, passage of gas into the mesenteric and portal veins starts from the bowel wall collection of gas. In one pathogenetic model gas may pass from the bowel lumen to bowel wall through mucosal defects, leading to IP [27, 28]. Another theory, which is better applied to the congenital form of IP pneumatosis cystoides intestinalis, suggests that increased intestinal permeability would permit penetration of anaerobic, gas-producing bacteria into the intestinal wall, with subsequent IP [28–30]. The subsequent step after IP is migration of gas along the mesenteric veins draining the intestine to the portal vein and its branches into the liver [2, 6, 31]. This mechanism explains the fact that gas is located peripherally in the liver in PMP since it is pushed by blood circulation. This particular peripheral localisation of gas also permits an easy distinction from pneumobilia in which fluid pushes the gas in the opposite direction, towards the hepatic hilum. In this pathogenetic model IP precedes PMP and we can expect that isolated IP is associated with less advanced conditions. There is another theory related to pulmonary conditions (the pulmonary theory) which hypothesise that in patients with chronic obstructive pulmonary disorders or asthma, gas may spread through alveolar rupture with interstitial dissection and spread to the intestinal serosa following a perivascular or perilymphatic route [27, 28].

Radiological appearance

Although both IP and PMP were first described on plain radiography [1], CT is far more effective in detecting pneumatosis (Fig. 3), so that this radiological sign that was previously considered very rare has been observed in many conditions with the growing use of CT [28, 29]. Proper use of window settings allows easy detection of the abnormal presence of intramural or intravascular air and its morphology (Fig. 4).

Three patterns of pneumatosis have been described: a bubble-like or cystoid pattern characterised by separate bubbles of gas with a cystic appearance (Fig. 5), a linear pattern in which the gas has a curvilinear or a crescent shape (Fig. 6) and a circular or circumferential form in the bowel wall (Fig. 7); in some cases all these types may be seen at the same time (Fig. 8). These different patterns appear frequently related to different pathological conditions, with the bubble-like cystic pattern more often observed in primary (or idiopathic) pneumatosis (Fig. 9) [28]. Primitive pneumatosis has a benign course and there

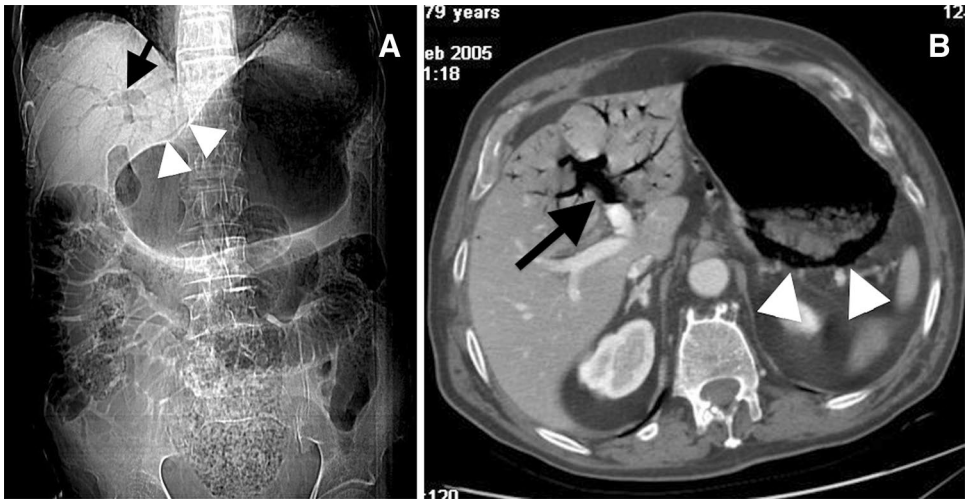

Fig. 3 **a** Plain abdominal X-ray. Portal pneumatosis (*black arrow*) and parietal gastric pneumatosis (*white arrowheads*) in intestinal infarct. **b** Computed tomography (CT). Portal pneumatosis (*black arrow*) and parietal gastric pneumatosis (*white arrowheads*) in intestinal infarct

Table 1 Causes of intestinal pneumatosis (IP) and portomesenteric pneumatosis (PMP)

(a) Increased mucosal permeability
–Pneumatosis cystoides intestinalis
–Drugs (corticosteroids, chemotherapeutic agents)
–Abdominal infections
–Neoplasms
–Solid organ and bone marrow transplantation
–Inflammatory bowel diseases
–Intra-abdominal abscess
–Infectious diseases
(b) Mucosal disruption
–Intestinal ischaemia
–Bowel obstruction
–Ulcerative disease
–Acute gastric or intestinal dilatation
–Blunt trauma
–Diverticular disease
–Ingestion of corrosives
–Intestinal carcinoma
–Iatrogenic causes
–Endoscopy
–Feeding tube (naso-gastric and naso-jejunal)
–Post-surgical anastomoses
–Drugs (corticosteroids, antineoplastic agents)
(c) Pulmonary conditions (alveolar rupture with interstitial dissection by air bubbles)
–Chronic obstructive pulmonary disease (COPD), bronchitis, emphysema
–Asthma
–Cystic fibrosis
–Positive end-expiratory pressure (PEEP)

is clinical and experimental evidence that bacteria are involved in the pathogenesis since the gas was found to be nitrogen on pathological specimens, the experimental

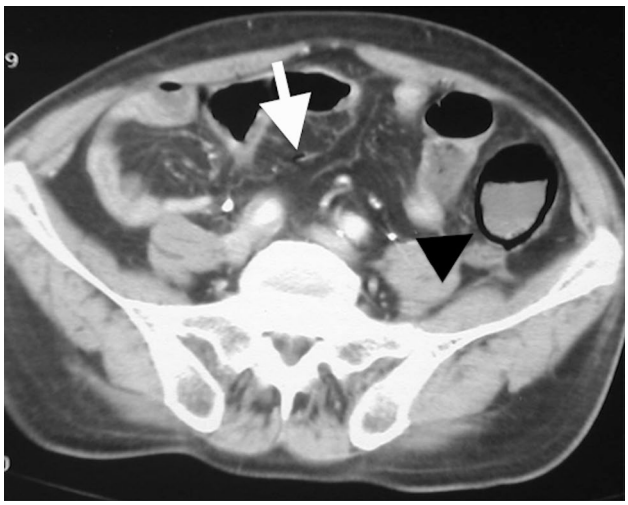

Fig. 4 CT intestinal pneumatosis of a jejunal loop in intestinal infarct (*black arrowhead*). A small amount of gas located in a mesenteric vein (*white arrow*) is clearly visible thanks to the proper window that allows a clear distinction of gas from fat

injection of bacteria in the intestinal submucosa caused pneumatosis, and in some cases pneumatosis resolved after antibiotic or hyperbaric therapy [28–32]. The secondary form of pneumatosis is much more frequent and often has a linear circumferential shape (Fig. 10), even though a bubble-like pattern is also observed (Fig. 11) [21, 33]. Colonic bowel wall enhancement is very difficult to analyse due to its reduced thickness; in this case IP is a capital sign in the recognition of intestinal infarct (Fig. 12). In some cases, and especially in pneumatosis cystoides intestinalis, IP may be associated with pneumoperitoneum [28] (Fig. 9).

PMP is related to presence the of a larger amount of gas in the bowel wall passing into the veins, and often indicates a more advanced stage of pathology (Fig. 13) [21]. Gas is more easily seen in the portal vein and its branches,

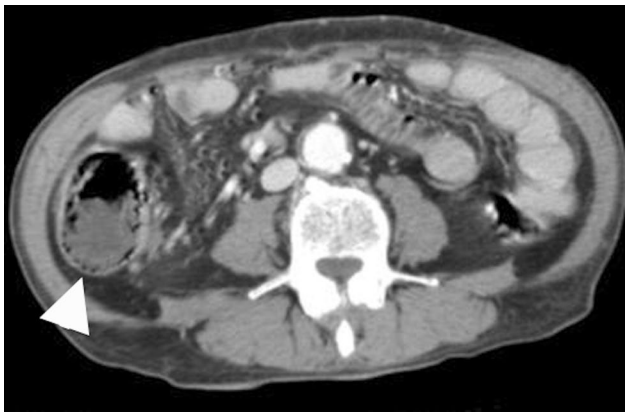

Fig. 5 CT bubble-like intestinal pneumatosis in intestinal infarct (*white arrowhead*)

Fig. 6 Linear pattern of intestinal pneumatosis in intestinal infarct (*white arrowhead*)

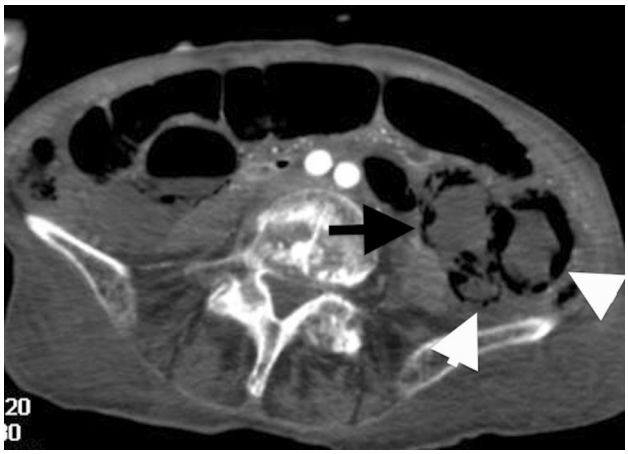

Fig. 8 Bubble-like (*white arrow*), linear (*black arrow*) and circumferential (*white arrowhead*) pattern of intestinal pneumatosis simultaneously present in intestinal infarct

and may sometimes also be seen overimposed on the liver shadow on plain abdominal radiographs (Fig. 3). In some phases, the gas may be seen in the mesenteric veins on CT scans (Fig. 13). Intravascular gas has been rarely observed in systemic veins [34, 35]. In one case it was observed in the systemic veins (gluteal veins, haemorroidal plexus, periprostatic veins, femoral veins) associated with porto-mesenteric pneumatosis and an extensive intestinal infarct [34]. Infrequently, gas may be seen in the spleen; this has been described in an intestinal infarct [36] and in a case of traumatic rupture of the stomach associated with extensive PMP and pneumoperitoneum (Fig. 14) [23].

Clinical significance

IP and PMP have long been considered a capital sign of advanced intestinal infarction and a predictor of poor prognosis. In recent times, however, several papers have

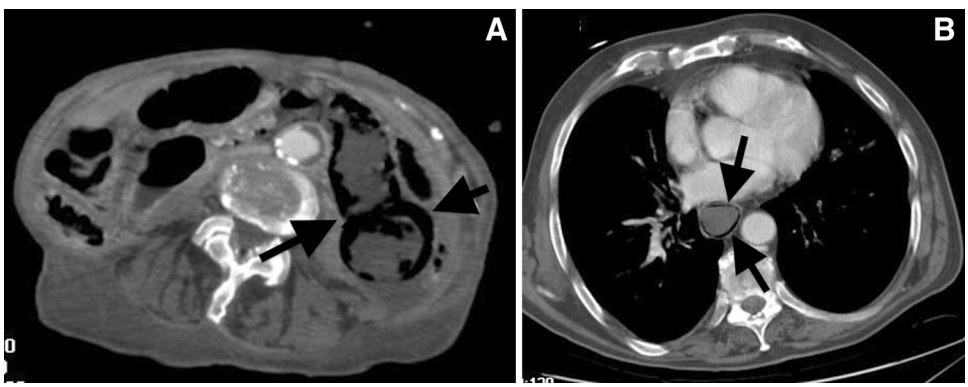

Fig. 7 **a** Circumferential pattern of intestinal pneumatosis in intestinal infarct (*black arrows*). **b** Circumferential pattern of pneumatosis in the oesophagus of a patient with a massive intestinal infarct (*black arrows*)

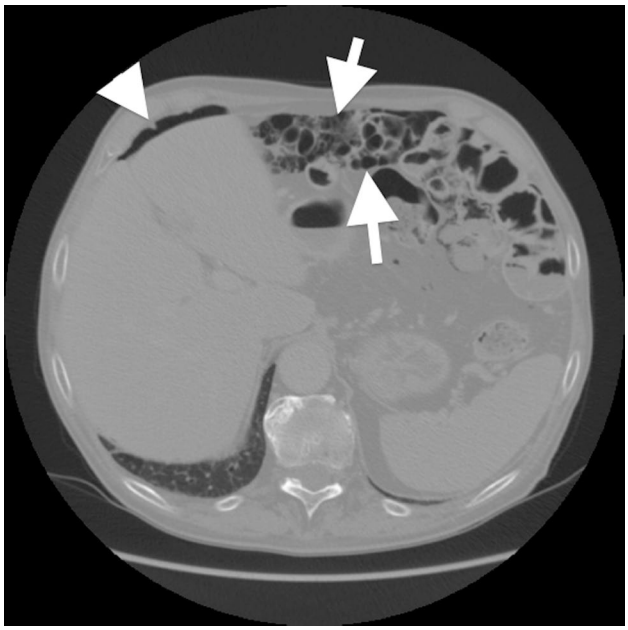

Fig. 9 Bubble-like pattern of intestinal pneumatosis in a patient with pneumatosis cystoides intestinalis (*white arrows*). Rupture of a bubble caused pneumoperitoneum in this patient (*white arrowhead*)

reported these signs in many clinical conditions other than intestinal ischaemia [6–24]. Most of these papers were case reports so that the unusual findings were magnified compared to the more common conditions.

In 1997, Faberman and Mayo-Smith [37] analysed a series of 17 patients with portal venous gas detected by CT and they found an unexpected low mortality in these patients, passing from a previously reported overall mortality of 75 % and of 85 % in non-iatrogenic cases to 29 %. The authors noted that CT has a greater capacity to recognise portal gas compared to plain radiographs; however, they also note the lack of correlation between mortality and the amount of portal gas. Furthermore, in their series there was no association between portal pneumatosis and intestinal pneumatosis. Following this study the significance of portal pneumatosis was questioned and it appeared due to many pathological processes and no longer associated with high mortality rates. In 2001 Wiesner et al. analysed the correlation between IP or PMP, or both, the severity of mural involvement, and the clinical outcome in 23 patients with bowel ischaemia concluding that these findings do not allow prediction of transmural bowel infarction because they may be observed with only partial ischaemic bowel wall damage; furthermore they state that both these signs are unrelated to advanced infarct also in the setting of bowel ischaemia [33].

The clinical significance of IP and portomesenteric venous gas (PVG) changed significantly over time, with the findings appearing more closely related to advanced intestinal infarct, and their prognostic value appearing strongly reduced, with important repercussions on clinical practice.

Wiesner's group also noted a difference between the IP patterns, with the band-like pattern being more closely correlated to transmural infarction than the bubble-like pattern (90 % versus 70 % of patients). They also observed that PMP was associated with transmural bowel infarction in 81 % of patients, and pronounced PMP was slightly more often associated with transmural bowel infarction (86 %) than mild PMP (78 %). Finally if both CT findings of pneumatosis and portomesenteric venous gas were seen, their presence was associated with transmural bowel infarction in 91 % of patients, regardless of their aspect and extent [33].

To better understand the relevance of IP and PMP our group analysed a large number of patients with PMP. Out of 47 patients with PMP, 91.5 % had a surgically proven necrosis along the gastrointestinal tract, the majority had arterial infarcts, perhaps because of fluid congestion preventing air penetration into the bowel wall and mesenteric vessels. Other causes in our series were iatrogenic or due to trauma. Among these patients, there was a high mortality rate (68.1 %), which rose to 79.5 % if only subjects with infarct were considered [22].

In a subsequent study we examined 102 patients with IP. In this study 62 % had gastrointestinal necrosis

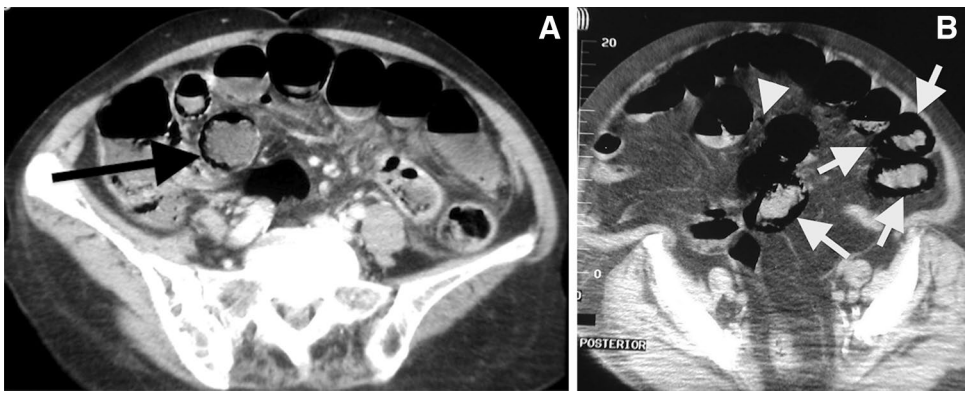

Fig. 10 **a** Intestinal pneumatosis in intestinal infarct. Linear pattern on CT. **b** Intestinal pneumatosis in intestinal infarct. Circumferential pattern on CT (*white arrows*). Gas in mesenteric vein (portomesenteric pneumatosis, PMP) (*white arrowhead*)

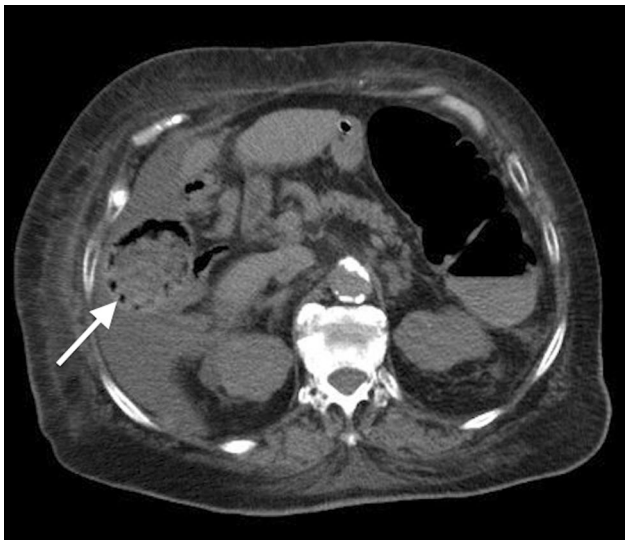

Fig. 11 Intestinal pneumatosis in intestinal infarct. Bubble-like pattern on CT (*white arrow*)

underlying IP, mainly, but not exclusively, arterial infarcts. Other less frequent cases were due to intestinal obstruction, abdominal cancer, peptic ulcer, abdominal trauma, Crohn's disease, diverticulitis,and iatrogenic mucosal injury. A linear pattern was particularly associated with bowel infarction (75.5 %). Among patients with IP and PMP, intestinal infarct was present in 69.2 % cases. IP was associated with death in 30.4 % of patients, a linear pattern was present in 79.2 % of infarcted patients who died, and in advanced cases there was a larger amount of air contributing to IP and PMP. The mortality rate was 50 % when IP was associated with PMP; this rate rose to 72.2 % if only infarcted patients with IP and PMP were considered [21]. These data support once again an association of IP and PMP, and in the appropriate clinical and radiological setting, this finding appears to be an indicator, albeit not pathognomonic, of intestinal infarction, given that intestinal infarction was the single most frequent cause of IP.

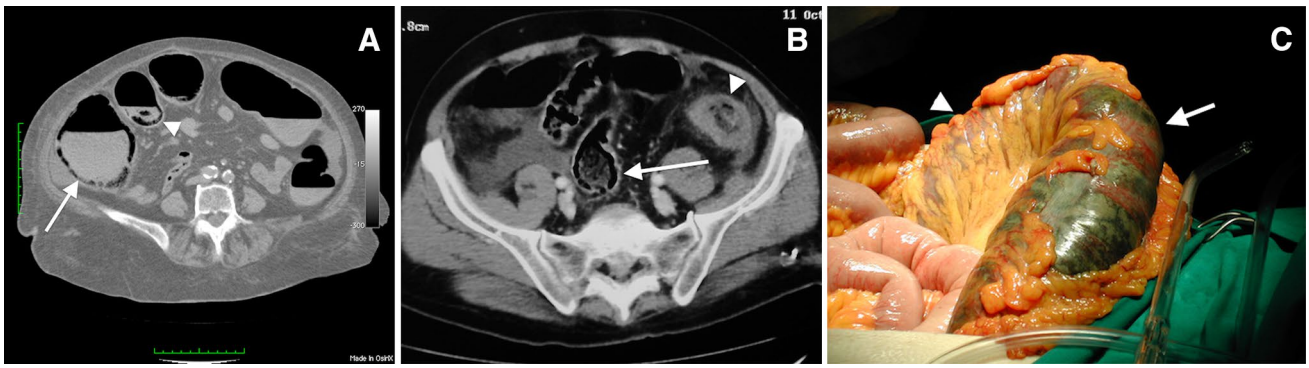

Fig. 12 **a** Right colon infarct due to superior mesenteric artery obstruction. *Bubble*-like (*white arrow*) and linear pattern (*white arrowhead*) on CT. **b** Left colon infarct due to inferior mesenteric artery obstruction. Intestinal pneumatosis with a linear pattern (*white arrow*) of the sigmoid colon. Note thickening of the descending colon (*white arrowhead*) after ischaemic involvement. **c** Surgical specimen of case depicted in Fig. 12b. Infarct of the *left part* of the transverse colon (*white arrow*). Note that *right part* of the transverse colon is still viable and has a normal *pink colour* (*white arrowhead*)

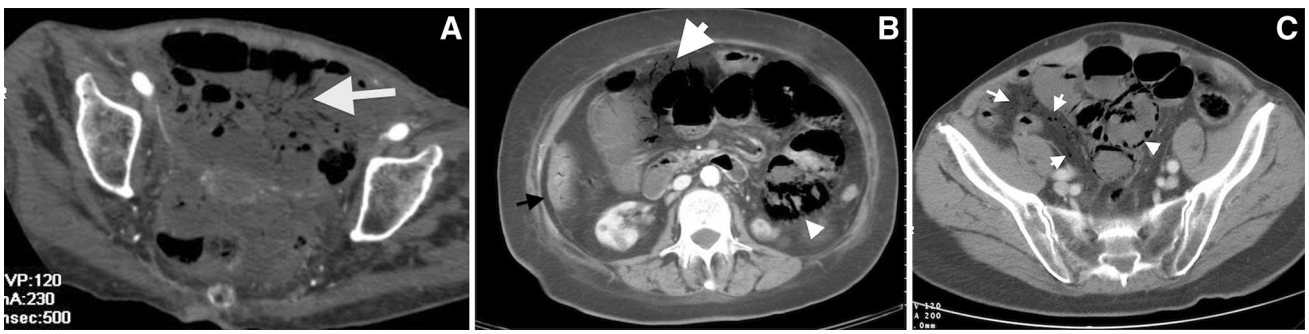

Fig. 13 **a** Gas in mesenteric veins with an arborescent appearance (*white arrow*). **b** Small bowel infarct. Gas in peripheral mesenteric veins (*white arrow*), intestinal pneumatosis (*white arrowhead*) and portal pneumatosis (*black arrow*). **c** Small bowel infarct. Gas in peripheral mesenteric veins (*white arrows*) and intestinal pneumatosis (*white arrowhead*)

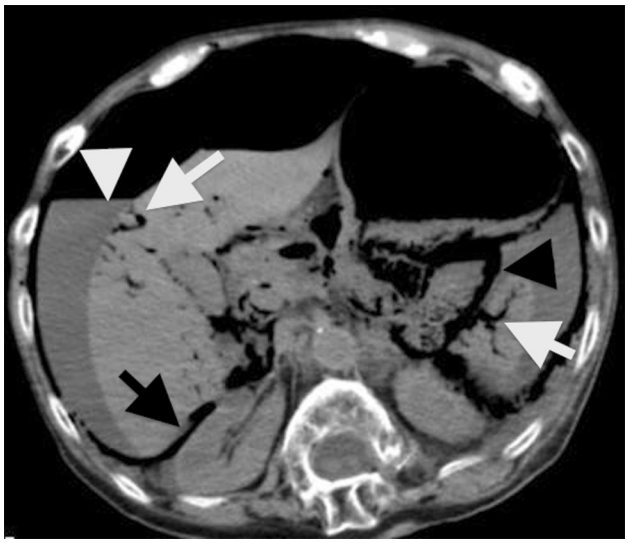

Fig. 14 Traumatic stomach rupture. Pneumoperitoneum with peritoneal fluid and air-fluid level (*white arrowhead*), pneumoretroperitoneum (*black arrow*), portal pneumatosis (*white arrow on the right*), gas in the spleen (*white arrow on the left*), gastric pneumatosis (*black arrowhead*); mesenteric vein gas is also present

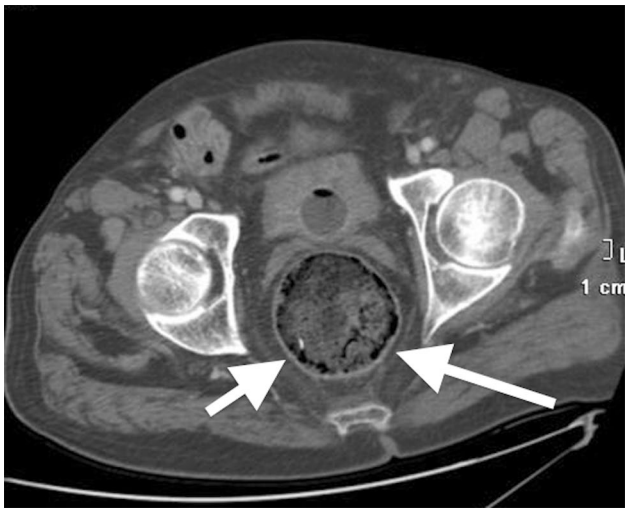

Fig. 15 Air trapped in faeces mimicking intestinal pneumatosis in the rectum

Pitfalls

Some conditions may mimic the radiological appearance of IP or PMP. Bubbles trapped in faeces and between mucosal intestinal folds can appear like intramural bubbles (Fig. 15). The key to distinguish true IP from trapped intraluminal air is the location of bubbles, as in IP these should not be limited to the nondependent part of the bowel but also extend to the posterior side (Figs. 1, 3, 4, 5) [28]. CT examination with a lung window setting helps to differentiate these

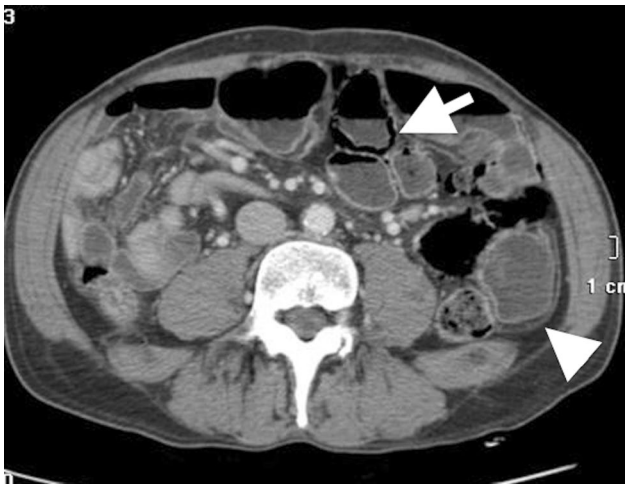

Fig. 16 Careful use of window settings allows for clear distinction between intestinal pneumatosis (*white arrow*) and parietal wall lipomatosis (*white arrowhead*) in a patient with long-standing steroid treatment for Crohn's disease

cases. Proper use of window settings avoids misdiagnosis of IP as lipomatosis. Lipomatosis is observed in particular in patients with long-standing steroid treatment. Occasionally, IP and lipomatosis can coexist in the case of Crohn's disease treated with intense steroid therapy (Fig. 16). Careful examination of images usually permits an easy recognition of IP and PMP from other types of extraluminal air such as abdominal abscesses and diverticula. Pneumobilia also may pose some interpretation doubts; the distinction is based on the fact that gas in the biliary system is usually seen at the hepatic hilum, whereas in PMP intrahepatic gas spreads to the edge of the liver following the blood stream (Fig. 17).

Conclusions

The clinical significance attributed to IP and PMP is continually changing due to recognition of a growing number of pathologies that may display these signs. Their presence was considered a reliable sign of intestinal infarct in adults and was thought to indicate a poor prognosis. Their role has been strongly questioned but, although it is well recognised that they can be observed in a large variety of conditions with very different prognoses, their association with intestinal ischaemia is well documented, and in some studies a high mortality rate has been observed, especially when a linear pattern of IP was found and when IP was associated with PMP.

Recognition of IP and PMP in the appropriate clinical setting should raise a suspicion of mesenteric infarct and prompt a careful search for other signs of intestinal

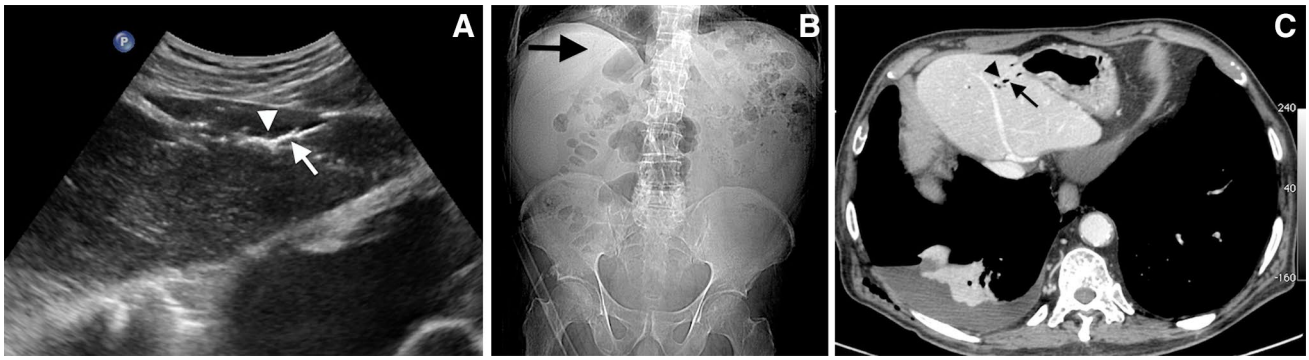

Fig. 17 **a** Ultrasonography. Intrahepatic gas due to pneumobilia (*white arrow*). Although the gas spreads peripherally in this case it is possible to see the associated portal branch (*white arrowhead*). **b** Plain radiograph of the abdomen. Same case as **a**. Intrahepatic gas (*black arrow*). In this case portomesenteric pneumatosis (PMP) cannot be distinguished from pneumobilia. **c** CT intrahepatic gas due to pneumobilia (*black arrow*). Note the associated portal branch (*black arrowhead*)

involvement so as not to miss cases of intestinal infarct or allow them further evolve into wide necrosis, with worsen prognosis.

Conflict of interest The authors declare no conflict of interest.

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