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CASE REPORT

Pneumatosis intestinalis, to treat or not to treat?

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Abstract

We present the case of an 81-year-old patient who was admitted to the intensive care unit of our hospital with severe septic shock due to acute mesenteric ischaemia and pulmonary artery embolism. This case has several diagnostic, therapeutic and prognostic dilemmas. The diagnostic and prognostic accuracy of pneumatosis intestinalis and portal venous gas are discussed.

Background

Acute mesenteric ischaemia (AMI) is a severe medical condition with a mortality rate up to 50-69%.[1] Untreated, it will cause intestinal necrosis and an excessive inflammatory response leading to multi-organ failure and death. Early intervention is pivotal for a chance of survival. However, diagnosis is difficult, especially in the early stages when intervention is most beneficial.

Case presentation

An 81-year-old man, with a history of type II diabetes mellitus, hypertension, dyslipidaemia and mild proteinuria, presented to the emergency department with progressive abdominal pain existing for several hours. He reported fever and vomiting. The patient was pale, disorientated, tachycardic (97/min), tachypnoeic (24/min) and hypoxic (SaO2 90% in room air). Further physical examination revealed signs of peritonitis.

Arterial blood gas analysis showed hypoxaemia and a respiratory compensated metabolic acidosis with increased lactate level (pH 7.38; PaCO2 4.0 kPa; PaO2 7.4 kPa; HCO3 16.2 mmol/l; lactate 3.9 mmol/l). Furthermore, he had renal insufficiency (creatinine 206 µmol/l, previously 150 µmol/l), leukocytosis (20.7 x 10^9/l) and an increased CRP (41 mg/l). A chest X-ray showed perihilar oedema, an electrocardiogram showed sinus rhythm and right bundle branch block.

During the diagnostic work-up, the patient's condition deteriorated. A computed tomography (CT) with intravenous contrast of the thorax and abdomen detected two embolisms: a longitudinal embolism reaching from the superior to the inferior mesenteric artery and a second embolism located in the pulmonary truncus and right pulmonary artery *(figure 1)*. Severe pneumatosis intestinalis of both the colonic and intestinal wall and additionally portal venous gas were present. The patient was taken to the operating room. A thrombectomy of the longitudinal thrombus was performed and simultaneously 350 cm of necrotic small bowel was resected.

Postoperatively, the patient was treated in the intensive care unit for septic shock with antibiotics, fluids, vasopressors, inotropes and corticosteroids. Low-molecular-weight heparin was started because of the pulmonary embolism. The next day, a second-look laparotomy was performed, resulting in the removal of the necrotic caecum and gallbladder. After the second operation the patient deteriorated rapidly with combined septic and cardiogenic shock. Cardiac ultrasound showed right atrial overload, an open foramen ovale was not observed. Thrombolysis for the pulmonary embolism was contraindicated because of recent surgery. The patient was considered too unstable for thrombectomy. Further medical treatment was regarded as futile and palliative care was started. The patient died in the presence of his family. Autopsy was performed and revealed a recent pulmonary embolism, severe atherosclerosis of the aorta and the coronary arteries, cardiac hypertrophy, and a shock aspect of the liver, spleen and pancreas. Neither macroscopic nor microscopic examination revealed malignancy.

Discussion

In Europe and the USA, AMI accounts for about 1:1000 hospital admissions.[1] It is an abdominal emergency requiring rapid diagnosis and treatment. Diagnosis, however, is difficult, due to non-specific clinical signs and lack of non-invasive diagnostic tests. Despite a decline in mortality rates over

the past 50 years, they remain high at 50-69%[1]. AMI may be provoked by various mechanisms and has different risk factors. Consequently, there are different diagnostic methods and therapeutic options. Obstructive AMI can be caused by bowel strangulation or volvulus.[1-3] Four aetiological forms of vascular AMI have been identified: arterial embolism (EAMI), arterial thrombosis (TAMI), venous thrombosis (VAMI) and non-occlusive mesenteric ischaemia (NOMI).[4] EAMI accounts for 45% of causes of AMI and is associated with comorbidities that predispose to formation of arterial thrombi, such as atrial fibrillation.[5] A sudden onset of pain, with prompt emptying of the bowel, without specific physical symptoms is a classical sign of EAMI. In approximately 25% of the cases, arterial thrombosis is the underlying mechanism of AMI. Risk factors include atherosclerotic disease and dyslipidaemia. Patients present with prodromal abdominal angina.[6] Patients with hypercoagulable states are prone to venous thrombosis. Venous occlusion is usually peripheral, involving different ischaemic bowel segments. The presentation is subacute and can take up to two weeks of abdominal pain, nausea and vomiting. NOMI may be caused by profound and disproportionate splanchnic vasoconstriction during low flow states in critically ill patients. Alternatively, it can occur perioperatively during major aortic surgery when splanchnic blood flow is disrupted or mesenteric arteries are sacrificed.[7,8]

Diagnosis of acute mesenteric ischaemia

Early diagnosis is crucial for reversal of ischaemic damage. Delayed intervention may result in intestinal necrosis, multiple organ dysfunction syndrome, and death. However, diagnosis is difficult, particularly in the early stages when treatment is most beneficial. The accuracy of the currently available laboratory tests (sensitivity and specificity of l-lactate is 86% and 44%, white blood count 80% and 50%, and d-dimer 89% and 40%, respectively) is suboptimal for routine clinical use.[9,10] The best diagnostic test apart from diagnostic laparotomy remains contrast CT (angiography) (sensitivity 94%, specificity 95%).[9] This is, however, time-consuming, costly, invasive and requires expertise. Several new serological markers may facilitate diagnostic accuracy. Intestinal fatty acid-binding protein, ischaemia modified albumin and α- glutathione S-transferase report a sensitivity of 79%, 95% and 68%, respectively, and a specificity of 91%, 86% and 84%, respectively.[10] Citrulline is a promising marker as well with a high reported specificity (100%).[10] Further research is required to identify optimal diagnostic accuracy and clinical utility.

Prognosis at presentation

Patients with significant comorbidities and poor performance status are unlikely to benefit from intervention. When the patient is seriously ill but not moribund it is more difficult to predict prognosis at presentation based only on clinical findings. A number of factors are associated with increased mortality, including advanced age, admission from a nursing home, partial dependence, coma, artificial ventilation, chronic obstructive pulmonary disease and recent (<6 months) myocardial infarction. Also, presentation >24 hours after the onset of symptoms and signs of peritonitis, organ failure, and sepsis, are independent risk factors for mortality.[4] So, according to the European Society for Trauma and Emergency Surgery guidelines on AMI, the decision to either proceed to curative treatment or palliative care should be based on the full history and pre-hospital performance status, clinical and lab findings. However, the guidelines are unclear about the value of radiological findings in this decision-making process. With this in mind we endeavoured to assess the prognostic accuracy of CT findings.

Prognostic predictive value of pneumatosis intestinalis

Pneumatosis intestinalis is defined as the presence of gas in the submucosa or subserosa of the intestinal wall. In some cases, pneumatosis intestinalis is an incidental finding associated with a benign aetiology, and patients can be observed without intervention. In others, it is a symptom of a life-threatening disease and urgent surgical resection of ischaemic bowel is required. The pathophysiology of this clinically challenging phenomenon is still poorly understood. Theoretically, it can be caused by intraluminal pressure and gas producing bacteria. Raised intraluminal pressure, for example due to obstruction, causes mechanical injury to the intestinal wall, which allows gas migration into the submucosal or subserosal layer.[11] In case of increased intrathoracic pressure, air leakage from alveolar rupture in pulmonary diseases migrates to the retroperitoneum through the mediastinum and locates within the bowel mesentery.[12] Also, pneumatosis intestinalis has been reported in patients taking α-glucosidase inhibitors. The increased production of intraluminal air in patients taking these drugs is attributed to the fermentation of undigested carbohydrates by intestinal bacteria.[13] When intramural gas enters the mesenteric veins, air can migrate to the portal venous system. This is defined as portal venous gas.

Clinically, the presence of pneumatosis intestinalis has always been considered an alarming radiological feature, due to its association with advanced mesenteric ischaemia and poor outcome.[14,15] Since the widespread use of the CT, the detection of pneumatosis intestinalis and portal venous gas has significantly improved in the last decade. However, these findings are not always predictive of poor outcome. DuBose et al. evaluated 500 patients with pneumatosis intestinalis and/or portal venous gas in a retrospective multicentre study, and reported an incidence of ischaemic pneumatosis intestinalis of only 40%.[16] In a separate retrospective review of 149 emergency patients with pneumatosis intestinalis, intestinal ischaemia was diagnosed in 54% of the cases. Ischaemia was significantly related to mortality.

The overall mortality was 42% and the mortality of patients with ischaemic pneumatosis intestinalis was 53%.[17] Higashizono et al., reported an incidence of bowel necrosis of 33% in a case series of 52 patients with pneumatosis intestinalis and/or portal venous gas. Overall in-hospital mortality was 23%, whereas 59% of patients with bowel necrosis died.[18]

It has been questioned whether anatomical location and extension of intraluminal and portal venous gas predicts incidence, severity, and mortality of AMI. On CT scans of 23 patients with AMI, mortality increased gradually with the number of bowel segments involved: 14% for a single segment, up to 80% for two or three infarcted bowel segments.[19]

Bowel necrosis is suggested to be related to the distribution of pneumatosis intestinalis, which can present as particles or air bubbles, referred to as a 'bubble-like' pattern. When the amount of air increases, it seems likely that air bubbles could merge together to form a 'band-like' pattern. Reports on the distribution of pneumatosis intestinalis have been conflicting. Higashizono et al. found no significant difference in distribution between patients with and without bowel necrosis.[18] Wiesner and colleagues reported an incidence of bowel necrosis of 70% for bubble-like pneumatosis and 88% for band-like pneumatosis. However, the association was nonsignificant.[19]

As the amount of gas increases, portal venous gas generally expands from the left lobe of the liver to the right anterior and right posterior lobe. Treyaud et al. did not find a significant correlation for portal gas distribution with underlying ischaemia. Similarly, neither the anatomical location of pneumatosis intestinalis (small bowel, large bowel or both), nor the length of intestinal involvement was correlated to mesenteric ischaemia. However, the combination of pneumatosis intestinalis and portal venous gas was significantly associated with underlying ischaemia.[17] As for Wiesner's population, 91% of the patients with both pneumatosis intestinalis and portal venous gas had transmural bowel infarction, compared with 81% of those with portal venous gas only. Mortality for patients with transmural bowel infarction was 53%.[19]

Further considerations

One could argue that our 81-year-old patient with a history of diabetes, hypertension and dyslipidaemia should not have undergone surgery. However, the patient was a non-smoker, mobile, self-supporting and the abdominal pain had existed for several hours. During diagnostic work-up, the pulmonary embolism seemed of no significant haemodynamic importance, therefore the decision for surgical intervention was made.

Conclusion

We present a case with severe AMI with pneumatosis intestinalis and presence of portal venous gas on CT scan and have reviewed the prognostic accuracy of pneumatosis intestinalis on CT for AMI.

Pneumatosis intestinalis is associated with AMI in 33-54% of the cases and has a mortality of 53 - 59%. Some distinctions can be made regarding mortality, namely the number of affected bowel segments, the pattern of pneumatosis intestinalis and the presence of portal venous gas in combination with pneumatosis intestinalis. When pneumatosis intestinalis spreads to two or more bowel segments, is distributed in a band-like pattern or is combined with portal venous gas, incidence of bowel infarction is increased to up to 80%, 88% or 91%, respectively.

Disclosures

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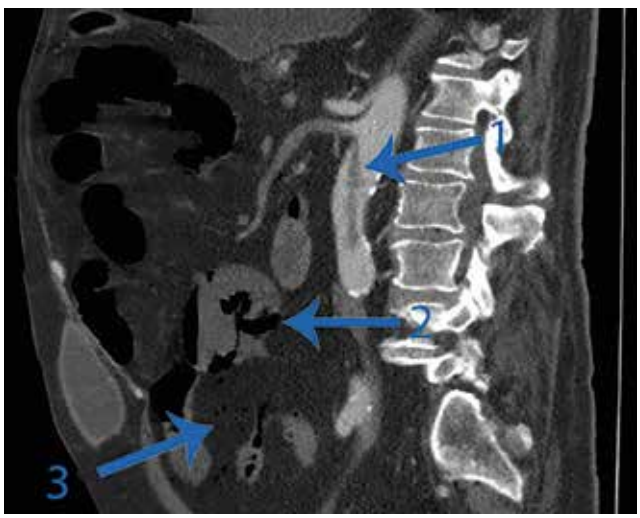

Figure 1. CT angiography of our patient showing: 1) a longitudinal thrombus from the superior to the inferior mesenteric artery, 2) pneumatosis intestinalis and 3) gas in the mesenteric veins

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