CASE REPORT

Pneumatosis intestinalis presenting as pneumoperitoneum in a teenage girl with pyloric stenosis

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SUMMARY

A 16-year-old girl presented with free gas under the diaphragm after endoscopic balloon dilation for pyloric stenosis. There was no perforation site identified on laparotomy. However, there was massive pneumatosis intestinalis involving the entire small bowel.

BACKGROUND

Pneumatosis intestinalis (PI) is defined as the presence of air within the bowel wall.¹ It may be a radiological or intraoperative finding. In the paediatric population, the most common pathological cause is necrotising enterocolitis in premature neonates, in which there is a risk of bowel ischaemia and infarction.² However, it has been reported in the literature that there are non-ischaemic and relatively more benign causes of PI presenting beyond infancy. These patients may have associated underlying conditions that constitute an immunosuppressed state. Examples include steroid use, chemotherapy, AIDS and mucosal disruption (due to inflammatory bowel disease, intestinal obstruction and indwelling catheters, etc).²⁻⁴ In this report, we describe a 16-year-old patient with pyloric stenosis and extensive PI, presenting with abdominal pain and pneumoperitoneum. This patient is, to the best of our knowledge, the first reported case of such association in the Asian paediatric population.

CASE PRESENTATION

A 16-year-old girl presented to us with episodic epigastric pain since childhood. Before attending our hospital, she had previously sought medical advice in China, where an upper endoscopy was performed, showing chronic gastritis and gastric outlet obstruction. There was a pinhole opening at the pylorus and the endoscope was unable to pass through. Balloon dilation of the pylorus was performed. *Campylobacter*-like organism (CLO) test confirmed *Helicobacter pylori* infection. There was no radiological imaging performed in China. The patient subsequently returned to Hong Kong for further treatment. On admission, she was noted to have epigastric tenderness and a succussion splash. An erect chest X-ray showed free gas under the diaphragm (figure 1). The complete blood count, electrolytes and blood gas were normal. In view of the X-ray finding and the recent history of endoscopic dilation, emergency operation was performed to rule out perforated viscus. Diagnostic laparoscopy was first performed to look for the site of

pathology. On laparoscopic assessment, the peritoneal cavity was clean but there were multiple gas pockets on the bowel wall, obscuring the view for a thorough examination of the internal organs (figure 2). Thus the operation was converted to laparotomy. There was no perforation site identified in the stomach and bowels after thorough examination. However, there were extensive pneumatosis and multiple gas pockets along the antimesenteric side of the bowel wall from proximal to mid small bowel (figure 3). Since the peritoneal cavity was not contaminated and there was no perforation site identified, peritoneal lavage was performed and a biopsy was taken from the ileum, which showed fibrovascular connective tissue with rounded pseudocystic changes, consistent with pneumatosis. We hypothesised that the free intraperitoneal gas was due to the spontaneous rupture of an intramural gas pocket. The patient was treated for her *H. pylori* infection with triple therapy and had an uneventful recovery. She tolerated her diet well after the surgery.

One month later, an elective gastroscopy was performed on the patient. The endoscopic findings were pyloric stenosis with no peptic ulcers. Balloon dilation to 10 mm diameter was performed under

Figure 1 The erect chest X-ray of the 16-year-old girl on admission.

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Figure 2 Numerous gas pockets hindering proper examination of organs on laparoscopy.

fluoroscopy. Her gastric outlet obstruction initially improved with balloon dilation. However, the condition progressively worsened again and she eventually underwent pyloroplasty as a definitive treatment. Intraoperative findings showed a thickened pylorus and a dilated stomach. Diffuse intestinal pneumatosis of the small bowel was still present but was less extensive when compared with that in the first operation. After the pyloroplasty, the patient was able to tolerate a normal diet well and is currently free of symptoms.

OUTCOME AND FOLLOW-UP

Since the gastric outlet obstruction of this teenage girl has been settled with pyloroplasty, we are very interested in repeating a set of CT scans of her abdomen and pelvis in order to reassess

Figure 3 Laparotomy findings of extensive pneumatosis intestinalis involving the small bowel.

the degree of pneumatosis. However, we do not want to expose her to too much radiation within such a short period of time. Therefore, our plan is to keep following her up in the future and see if she incidentally needs a CT scan for some other reasons, during which radiological reassessment of her condition would be possible.

DISCUSSION

PI has been described to be associated with various comorbidities in adult⁵ and in paediatric patients. Fenton and Buonomo reviewed 37 paediatric patients with PI and found that 32 children had identifiable risk factors such as immunosuppression, post-transplant state, short gut syndrome, chronic pulmonary disease, malignancies, congenital heart disease and collagen vascular disease. Immunosuppression was the most common association and was found in 25 patients.⁴ Our patient did not appear to have any of the risk factors mentioned by Fenton and Buonomo. However, the only significant medical history she had was pyloric stenosis, leading to gastric outlet obstruction.

Literature on the association of PI with pyloric stenosis in children is lacking. There are only a few isolated case reports. It has been reported that pyloric stenosis may contribute to massive gastric pneumatosis.^{6,7} However, the described patients were of infantile age and the pneumatosis was localised in the stomach wall. Our teenage patient presented with the alarming feature of free gas under the diaphragm and extensive pneumatosis involving most of the small bowel. Gastric outlet obstruction (mostly due to peptic ulcer disease) has been reported as a cause of PI in adult patients,^{8,9} but is rarely the cause in children. A similar scenario has been described in a 37-year-old adult man who had peptic ulcer-induced pyloric stenosis and PI of the terminal ileum.¹⁰ He also presented with free gas under the diaphragm, thus requiring an emergency laparotomy for suspected perforated peptic ulcer; but there was no perforation site identified on laparotomy. In this adult patient, the pneumatosis only affected a short segment of his terminal ileum and was not as extensive as that in our patient.

There are a number of proposed theories as to the pathogenesis of PI.³ Among these, we believe the mechanical theory best explains the situation in our patient. This theory suggests that increased intraluminal pressure and bowel trauma give rise to a break in mucosal integrity, allowing intraluminal gas to enter the bowel wall.¹¹ Moreover, the inflation of air during upper endoscopy in our patient might have produced a high-pressure jet of gas, which entered the duodenum via the stenotic pylorus. The guidewire used during balloon dilation could have further caused breaks in the mucosal integrity. The gas would then be distributed to more distal areas by peristalsis. This may explain why the pneumatosis was in the small bowel while the obstruction was at the pylorus. We also postulate that the stomach is less easily involved by pneumatosis because the stomach wall is thicker and much more muscular. In order for gas to form sub-serosal cysts, it must break through the muscularis layer.¹⁰ The mechanical theory is frequently used to explain pneumatosis seen in gastric outlet or intestinal obstruction, Hirschsprung's disease, inflammatory bowel disease and gastroenteritis.

The bacterial theory proposes that overgrowth of anaerobic bacteria in the gut leads to overproduction of intraluminal gas, in particular hydrogen,¹² and concurrent loss of mucosal integrity. Gas can thus escape into the bowel wall.

Other theories involving pulmonary conditions,^{13,14} such as increased mucosal permeability, chemical and nutritional factors, neoplastic conditions and many others, have been suggested, implying that the aetiology of PI is likely multifactorial.

Learning points

- ▶ Pneumatosis intestinalis is never an isolated condition.
- ▶ It may not always require surgical intervention.
- ▶ However, the underlying associated disease must be identified and treated accordingly.

Contributors CWYW wrote the case report under the supervision of PHYC, KKYW and PKT. PHYC provided the intraoperative photos.

Competing interests None.

Patient consent Obtained.

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