CASE REPORT

Pneumatosis intestinalis in an adult patient with antral stenosis and midgut malrotation

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SUMMARY

Pneumatosis intestinalis (PI) is a radiological finding with about 0.03% incidence and incompletely understood pathogenesis. We report a case of PI with a rare presentation of pneumoperitoneum that underwent diagnostic surgery and finally diagnosed by midgut malrotation and antral stenosis. A 40-year-old man with 1-year history of dyspepsia and vomiting which was aggravated by 20 kg underweight since 3 months ago, despite medications was presented. His imaging examinations showed partial antral obstruction besides pneumoperitoneum and gas collection in the small intestine wall, which were in favour of PI. Samplings of the small intestine during laparotomy reported simple serosal cysts. Persistence of his symptoms forced him to recourse to another hospital and their new imaging revealed Ladd's band in addition to mentioned findings in previous studies; he underwent subtotal gastrectomy plus Ladd's band division by Braun gastrojejunostomy. PI could be a presentation of antral stenosis and midgut malrotation. PI with pneumoperitoneum needs surgical interventions. Considering that most of the midgut malrotation cases are diagnosed in the first year of life and it is very rare in adults, it may be that these new findings are due to adhesion band formation after the first surgery in this case.

BACKGROUND

Pneumatosis intestinalis (PI) is a radiological finding, and its pathogenesis is not entirely understood.¹ PI is presented by the presence of free gas in subserosal or submucosal layers of the intestinal wall and cyst formation within the gastrointestinal (GI) tract and tissues around it.^{2,3}

The true PI incidence is currently unknown; however, PI is reported only in 0.03% of the population.³ At present, because of more frequent and improved imaging examinations, it is being reported increasingly,⁴ although several studies reported a higher incidence among male.⁵

Almost 15% of PI cases are primary or idiopathic and 85% are secondary which can be resulted from a wide spectrum of GI or systemic diseases, although primary PI is benign and asymptomatic, but abdominal pain is the most frequent symptom of secondary PI⁶ and some of the secondary PI causes are life threatening.^{1-3,5-8}

CT is the most sensitive imaging modality for diagnosis of PI and might be helpful to distinguish primary PI from secondary as well.^{1,2,5,9}

Whereas PI developed by life-threatening causes that need immediate surgery, it has been estimated that about 27% of patients with primary PI undergone unnecessary operations, although there are no clear and confirmed guidelines for PI management.^{5,10}

We report a case of secondary PI with a rare presentation of pneumoperitoneum and finally diagnosed by midgut malrotation and antral stenosis (atonic stomach). He was undergoing double surgery; the first was emergency diagnostic.

CASE REPORT

A 40-year-old man was brought to the emergency department in Shohadaye Ashayer Hospital, Khorram Abad, Iran, with severe abdominal pain. He had bloating, halitosis, food intolerance, postprandial abdominal distension and recurrent after meal no biliary, no bloody especially self-induced vomiting. He mentioned 20 kg weight loss during the past 3 months. He suffered from mild chronic onset epigastric pain, with no radiation, worsened after feeding, since 6 months ago. His pain was worsened acutely few hours before he came to our hospital. There was normal defecation pattern without any history of constipation or diarrhoea.

One year ago, heartburn and dyspepsia forced him to undergo a GI endoscopic examination. Daily omeprazole 20 mg, sertraline 40 mg and at bedtime ranitidine 40 mg tablet had been prescribed to him with a diagnosis of erosive gastritis. After that, he was partially symptoms free for about 9 months, until 3 months ago when the above-mentioned signs and symptoms developed severely, despite the medications.

In his medical history, there was not any significant internal disease such as hypertension, diabetes mellitus, thyroid disorders and allergic reactions. Smoking or opium addiction was not in his social history and also no internal disease especially GI was in his familial history.

In physical examination, he was awake and alert, not pale and toxic and ill or icteric. Also, he was afebrile (oral temperature: 37.1°C) and other vital signs were stable (blood pressure 125/75 mm Hg, pulse rate 72 beats/min, respiratory rate 16/min).

The abdomen was mildly distended, tympanic in palpitation with generalised tenderness but without rebound tenderness, guarding and hepatosplenomegaly. The bowel sound decreased about two or three times per minute. In review of system examinations, other organs were normal.

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Figure 1 Bilateral free air under diaphragm in chest X-ray.

Laboratory data showed a mild rise in mean corpuscular volume (100.5 fL), mean cell haemoglobin (33.9 pg), blood urea nitrogen (55 mg/dL) and creatinine (1.5 mg/dL), also mild hyponatremia and hypokalemia (Na=130 meq/L, K=2.9 meq/L).

Chest X-ray showed bilateral free air under the diaphragm (figure 1).

Abdominal X-ray revealed pneumoperitoneum, severely distended stomach and air-fluid level in the right side of the abdomen in ascending colon which was in favour of PI (figure 2).

Abdominal CT scan with double (intravenous and oral) contrast also revealed severely distended stomach, partial antral obstruction (stenosis), massive bilateral free air under the diaphragm, free air bubbles throughout the entire length of the small intestine which the latter finding was highly in favour of PI. In this study, there is no sign of gut rotation (figure 3).

DIFFERENTIAL DIAGNOSIS

1. PI.
2. Mesenteric ischaemia.
3. Peptic ulcer disease.
4. Bowel obstruction.
5. Inflammatory bowel disease (IBD).
6. Irritable bowel syndrome.
7. Chronic appendicitis.
8. Meckel's diverticulitis.

Figure 2 Pneumoperitoneum, severely distended stomach and air-fluid level in the right side of abdomen in ascending colon in supine and upright abdominal X-ray.

Figure 3 An axial view of abdominal CT scan showed severely distended stomach, partial antral obstruction, massive bilateral free air under the diaphragm, free air bubbles throughout the entire length of small intestine.

TREATMENT

The patient underwent emergent diagnostic laparotomy in our hospital, Shohadaye Ashayer Khorram Abad. After prep and drape, midline incision was performed, the abdomen cavity was observed and investigated. There were multiple cystic lesions throughout the abdominal cavity which were resected completely and several specimens were sent for pathological study. The liver, spleen and other viscera were normal.

The patient was discharged with the same previous medications for gastritis treatment and referred to GI specialist.

FOLLOW-UP

About 1 month later due to the aggravation of his abdominal pain, dyspepsia, vomiting, nausea and severe anorexia, he decided to visit another abdominal surgeon for further assessment.

The general surgeon in Velayat Hospital in Ghazvin, Iran, re-examined him and re-evaluated his lab tests and imaging which confirmed the previous results of Shohadaye Ashayer Hospital. He performed the new laboratory tests; the results showed white cell count $9500 \times 10^3/L$, haemoglobin 11.4 g/dL, platelets $34700 \times 10^3/L$, alanine transaminase 9 U/L, aspartate aminotransferase 19 U/L, alkaline phosphatase 71 U/L, total bilirubin 0.3 mg/dL, direct bilirubin 0.2 mg/dL, total protein 5.4 mg/dL and albumin 4.02 mg/dL.

He repeated the radiological studies and in the new abdominal double contrast CT scan, the midgut malrotation and partial antral stenosis and atonic stomach were found, which were not reported in the former studies.

Finally, based on physical examinations and new imaging findings, he decided to perform another abdominal surgery, during which subtotal gastrectomy with Ladd's band division and Braun gastrojejunostomy were done.

Then, the patient was discharged with these medications: one tablet of sertraline 50 mg, cefixime 400 mg, folic acid 5 mg, pantoprazole 40 mg, zincplus, calcicare daily, one tablet

of digestive and silax after meals, ursobil 300mg tablet every 12 hours, lorazepam 1 mg tablet at bedtime, B12 and neurobion one ampoule intramuscularly weekly.

Until now, the patient has no GI signs and is symptom free, continuing pantoprazole, sertraline, silax and digestive. He is regularly visited by his GI specialist and surgeon.

DISCUSSION

Du Vernoy, a French pathologist, introduced PI for the first time during cadaveric dissection in 1730 and Mayer named it as PI in 1925.^{3 4 8}

The pathogenesis of PI is still poorly understood but researchers have proposed several theories including: (1) bacterial translocation theory which suggests bacterial invasion to the bowel wall with no mucosal injury, (2) increased mucosal pressure theory which suggests the entering of intraluminal air into the bowel wall secondary to high luminal pressure and (3) mucosal break theory which proposes drive of gas into the bowel wall because of mucosal ischaemia or inflammation.^{3 4}

PI is frequently related to ischaemia but there is no uniform view in its correlation with bowel ischaemia. It is also associated with various non-surgical causes, such as trauma, graft versus host disease, IBD, steroids use, immunosuppressive condition and *Clostridium difficile* infection. Sometimes PI is incidentally found in an asymptomatic patient.³

PI is easily diagnosed by GI endoscopy or abdomen plain radiography, because of radiolucent grape-like clusters or honeycomb-shaped shadows along the wall of the intestine.⁴ However, radiological findings must be considered along with clinical presentations.⁵

In most cases, the primary or benign PI is accidentally found and has no associated symptom,⁵ but secondary or pathological PI is due to various underlying diseases such as chronic obstructive pulmonary disease, systemic sclerosis, obstructive and necrotic GI disease, some of them are life threatening.^{1 2 5}

After the detection of PI, further imaging evaluation should be done. Abdominal CT scan is very useful and important especially in detecting other abdominal pathologies.⁴ CT scan can also help to distinguish these two types of PI.¹

Bindu *et al* reported that elevated lactate level, tachycardia, abdominal tenderness, dilation of small bowel and lack of diabetes mellitus are very useful in distinguishing patients with benign and pathological PI.³

Several studies have reported that leucocytosis, emesis, uraemia and age above 60 can predict the need for surgical treatment; however, various studies found that lactic acidosis is the most frequent consistent predictor of pathological PI.³

Pneumoperitoneum is an uncommon presentation of secondary PI which occurs in less than 3% of PI cases.⁵ Our patient who had pneumoperitoneum in abdominal radiography and CT scan was among this 3% of rare presentations of PI.

The appropriate treatment depends on the underlying cause of PI.⁴ The most fatal causes of PI are due to bowel necrosis secondary to bowel ischaemia, infarction, necrotising enterocolitis, neutropenic colitis, volvulus or sepsis which need emergency surgeries.⁵

Since no standard management guidelines, including indications for surgery, is available for PI treatment, about 27% of patients with benign PI may undergo unnecessary surgery.⁵

Depending on the underlying diseases, most of the cases of PI should receive conservative treatments such as oxygen, intravenous hydration and antibiotics.¹

In general, signs of peritonitis or bowel perforation in radiography and tumour in endoscopy are indications for emergency surgery.⁴

In this case, the patient developed PI with pneumoperitoneum due to antral stenosis and atonic stomach which could not be resolved with conservative therapy and needed surgical interventions. His signs and symptoms and imaging results and exploratory laparotomy confirmed this diagnosis. First evaluations and exploratory laparotomy only showed antral stenosis and PI but second abdominal CT revealed Ladd's band and midgut malrotation as well. So a major surgery with subtotal gastrectomy and Ladd's band division plus Braun gastrojejunostomy was done for him and he discharged with some medications.

Ninety per cent of midgut malrotation cases are diagnosed in the first year of life and it is very rare in adults with an incidence between 0.0001% and 0.19%.¹¹ Ladd's band and midgut malrotation, which are very rare in adults, may be the complications of first surgery due to adhesion band formation in this case.

On the whole, secondary PI could be a presentation of antral stenosis and midgut malrotation (even in adults) in the presence of related clinical signs and symptoms.

Learning points

- ▶ Never forget the differential diagnosis of dyspepsia and epigastric pain.
- ▶ A more accurate evaluation of patients with chronic dyspepsia and history of treatment and normal endoscopy should be done.
- ▶ Pneumatosis intestinalis is easily diagnosed by endoscopy or plain abdomen radiography because of radiolucent grape-like clusters or honeycomb-shaped shadows along the wall of the intestine. Radiological findings must be considered along with clinical presentations.
- ▶ Patients with long-term disease may have uncommon illnesses.
- ▶ Before planning and development of a surgery producer in non-urgent conditions, consider all differential diagnosis specially and collect enough information.

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Contributors ASD is the corresponding author and contributing to conception and design, interpretation of lab findings and other data, drafting the manuscript and revising it, submission and giving the final approval of the revisions, encouraging ISD and PSD to investigate, supervising and agreement for the findings and all aspects of the work, approving the final version of manuscript. MM contributed to the conception of the research, conducting the study, revising the manuscript, approving the final version of the manuscript and agreement for all aspects of the work and acting as corresponding author. ISD and PSD contributed to the designing of the study model and working on almost all the technical details, collecting the acquisition data and sorting them, complying the literature sources and checking the references, writing some of the parts of the manuscript, providing critical feedbacks and helping to shape the research approving the final version of the manuscript and agreement for all aspects of the work.

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