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Pneumatosis Cystoides Intestinalis: A Case Report

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Funds Collection G

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Patient: Female, 84-year-old
Final Diagnosis: Pneumatosis cystoides intestinalis
Symptoms: Abdominal distention
Medication: —
Clinical Procedure: —
Specialty: General and Internal Medicine

Objective: Rare disease

Background: Pneumatosis cystoides intestinalis (PCI) is a rare condition in which cystic gas is found in the submucosal and serosal tissues of the intestinal wall.

Case Report: The patient, an 84-year-old woman, was referred to us because of abdominal distention and diarrhea lasting 2 weeks. On initial physical examination, there was marked abdominal distention without tenderness. Blood tests revealed no abnormalities, but simple abdominal radiographs showed gas in the small intestine. Contrast-enhanced computed tomography showed massive emphysema in the intestinal wall with no signs of portal gas or intestinal ischemia. The patient was diagnosed with PCI, and the prognosis was good. The patient showed improvement when managed with an elimination diet and follow-up.

Conclusions: Herein, we present the characteristics and diagnosis of PCI because the imaging findings of PCI can appear more severe than the actual condition, causing it to be mistaken for other serious diseases, which leads to unnecessary surgical procedures.

Keywords: Diagnosis • Pneumatosis Cystoides Intestinalis

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Background

Pneumatosis intestinal cysticercosis (PCI) is a rare disease wherein cystic gas is observed in the submucosal or serosal tissue of the intestinal wall. It reportedly occurs at all ages, most commonly at 45±15 years of age. Non-surgical treatment results in remission in 70% of patients [1]. However, PCI is often mistaken for other serious diseases due to the associated “severe” imaging findings, which may lead to unnecessary surgical procedures [2]. Here, we present a case of PCI to illustrate the associated key findings and highlight the importance of differentiating PCI from other diseases.

Case report

An 84-year-old woman was referred from a rehabilitation hospital to a tertiary care hospital with abdominal distension and diarrhea that had persisted for 2 weeks. She had an altered state of consciousness and was initially unable to speak; therefore, her subjective symptoms were unknown. Her vital signs were within normal ranges. During the initial physical examination, there was marked abdominal distension without perceptible tenderness. The patient’s medical history included dementia, pneumonia, and cerebral infarction. She was taking multiple medications, including prednisone, lansoprazole, and quetiapine. Blood test results (Table 1) showed that her blood urea nitrogen (10.7 mg/dL, normal: 6 to 24 mg/dL) and creatinine (0.86 mg/dL; normal: 0.7 to 1.3 mg/dL) were within normal limits. Blood gas analysis showed a mildly elevated lactate level (2.1 mmol/L, normal: 0.5-1 mmol/L). Simple abdominal radiography showed the presence of gas in the small bowel (Figure 1). Contrast-enhanced computed tomography (CT) showed free air, copious emphysema in the intestinal wall, and no portal gas or signs of intestinal ischemia (Figures 2, 3). After a multidisciplinary consultation involving surgeons, gastroenterologists, and radiologists, the patient was admitted to the hospital with a diagnosis of PCI with a good prognosis. Surgery was determined to be unnecessary, and the patient was managed with fasting and oral fluids. Oxygen was administered to reduce intestinal emphysema. As there were no signs of infection, no antimicrobials were administered. Three days later, a CT scan revealed that the abdominal distension and intestinal gas had decreased; therefore, oxygen administration was terminated, and food intake was initiated. On day 11 after admission, the patient’s abdominal symptoms improved, and she was discharged.

Discussion

PCI occurs in individuals 50 to 80 years of age, and its incidence is approximately 0.03% [3]. The cause is unknown, but

Table 1. The patient’s laboratory data.

Test item	Unit	Normal range
WBC	8.15 ×10 ³ /μl	3.3-8.6
RBC	466 ×10 ⁴ /μl	386-492
Hb	13.2 g/dl	11.6-14.8
Plt	24.7 ×10 ³ /μl	15.8-34.8
TP	5.3 g/dl	6.6-8.1
Alb	3.2 g/dl	4.1-5.1
T-bil	0.6 mg/dl	0.4-0.5
AST	26 U/L	13-30
ALT	28 U/L	7-23
LDH	248 U/L	124-222
ALP	66 U/L	106-322
γ-GTP	13 U/L	9-32
AMY	64 U/L	44-132
BUN	10.7 mg/dl	8-20
Cre	0.86 mg/dl	0.46-0.79
BS	117 mg/dl	80-112
HbA1c	6.0 %	4.6-6.2
CRP	0.07 mg/L	0.14 or less
Arterial blood gas		
pH	7.452	7.35-7.45
PCO ₂	40.4 Torr	35.0-45.0
PO ₂	85.5 Torr	75.0-100
HCO ₃	27.8 mmol/l	20.0-26.0
BE	4 mmol/l	3.0-3.0
Na	134 mmol/l	138-145
K	3 mmol/l	3.6-4.8
Cl	99 mmol/l	101-108
Ca	1.13 mmol/l	1.16-1.32
Lac	2.1 mmol/l	0.56-1.39

WBC – white blood cell; RBC – red cell concentrate; Hb – hemoglobin; Plt – platelet; TP – total protein; Alb – albumin; T-bil – total bilirubin; AST – aspartate aminotransferase; ALT – alanine aminotransferase; LDH – lactate dehydrogenase; ALP – alkaline phosphatase; GTP – γ-glutamyl transpeptidase; AMY – amylase; BUN – blood urea nitrogen; Cre – creatinine; BS – blood sugar; HbA1c – hemoglobin A1c; CRP – C-reactive protein; pH – potential hydrogen; PCO₂ – arterial partial pressure of carbon dioxide; PO₂ – arterial partial pressure of oxygen; HCO₃ – bicarbonate ion; BE – base excess; Na – sodium; K – potassium; Cl – chlorine; Ca – calcium; Lac – lactate.

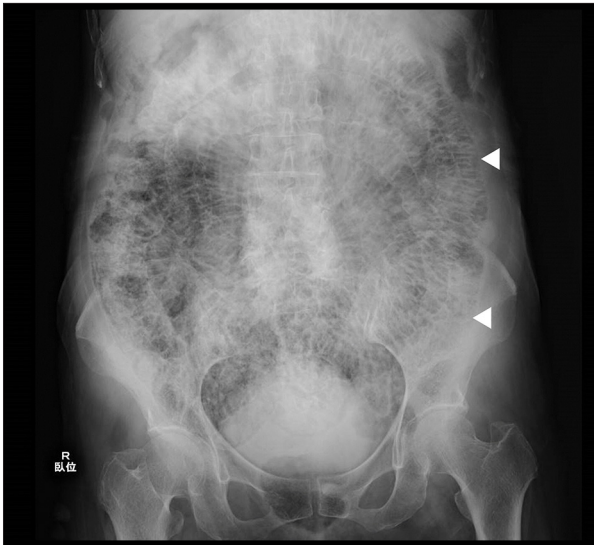


Figure 1. Diffuse dilation of the intestine, a small round translucency, and reticular structures along the intestinal wall suggesting intramural emphysema.

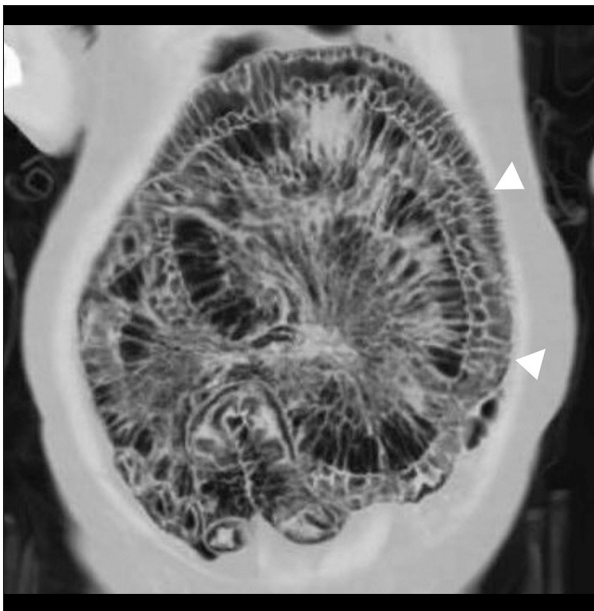


Figure 2. Abdominal computed tomography coronal, showing emphysema in the wall of the intestine over almost the entire length of the ileum.

it may involve multiple factors, including mechanical, biochemical, bacterial, and pulmonary disease-related influences [3]. In such cases, the differential diagnosis of other severe gastrointestinal diseases, such as intestinal ischemia, infarction, necrotizing enterocolitis, intestinal perforation, axial torsion, and sepsis, should be excluded by performing blood tests and imaging studies [4]. **Table 2** shows the poor prognostic factors associated with PCI. Indicators for surgery include age over 60 years, vomiting, and a white blood cell count $>12\,000/\mu\text{L}$ [5].



Figure 3. Abdominal computed tomography axial, showing a small amount of free air in the abdominal cavity and on the surface of the liver.

Table 2. Poor prognostic factors associated with pneumatosis cystoides intestinalis.

- Elderly age
- Unstable vital signs
- Vomiting
- Severe abdominal pain
- Abdominal rigidity
- Increased white blood cell count
- Lactic acid level ≥ 2 mmol/L
- Prolonged international normalized ratio
- Hepatic portal vein gas
- Metabolic acidosis

Although PCI generally has a good prognosis, accurate diagnosis and timely surgery are important. Poor prognostic factors have been proposed, including intrahepatic portal gas, changes in blood test values, and negative predictors such as a pH <7.3 , HCO_3^- level <20 mmol/L, and lactate level >2 mmol/L [6-8]. Despite the patient being elderly, she had normal vital signs and no subjective symptoms except lactate levels; therefore, surgery was not indicated for the patient. There were no findings of wall contrast failure or torsion indicative of intestinal ischemia on CT findings. Based on the diagnostic flowchart for PCI, the vascular risk score was 0 in STEP3 [9] (**Figure 4**). Therefore, the patient was monitored with follow-ups and imaging tests. The sensitivity and negative predictive value in STEP3 were 100%. In addition to abnormal clinical vital signs and severe abdominal pain, it is important to consider these indicators when planning treatment strategies, including surgery, to rapidly identify and manage severe PCI.

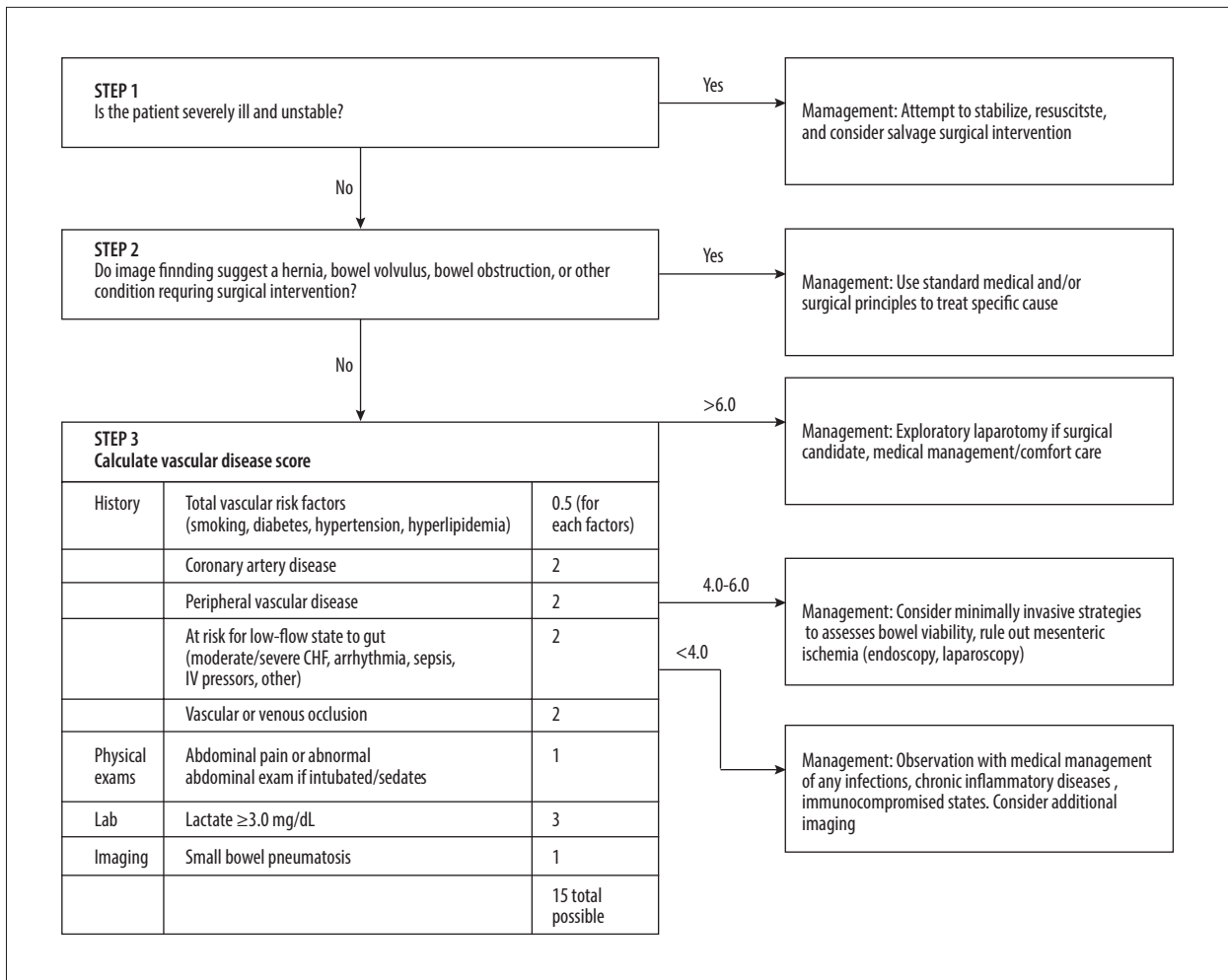


Figure 4. Management algorithm for pneumatosis and/or portal venous gas (PVG) identified on computed tomography scan. CHF – congestive heart failure; IV – intravenous. *Modified from Wayne E, et al. J Gastrointest Surg 2010 Mar; 14(3):437-48.*

Conclusions

PCI is a rare condition with cystic gas in the intestinal wall and is often mistaken for other severe gastrointestinal diseases; therefore, physicians should carefully differentiate PCI from other severe diseases.

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Declaration of Figures' Authenticity

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