

Pneumatosis intestinalis with obstructing intussusception: A case report and literature review

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Abstract

Pneumatosis intestinalis (PI) often represents a benign condition that should not be considered as an argument for surgery. We report a patient with PI and obstructing intussusception who underwent urgent colectomy and review the literatures regarding PI with intussusception. A 20-year-old man presented at our hospital with a 3-d intermittent lower abdominal pain history. He underwent steroid therapy for membranoproliferative glomerulonephritis for 4 years. Computed tomography revealed ascending colon intussusception with air within the wall. Intraoperative colonoscopy revealed numerous soft polypoid masses with normal overlying mucosa and right hemicolectomy was performed. Histological examination of colonic wall sections revealed large cysts in the submucosal layer. The pathological diagnosis was PI. Nine cases of intussusception associated with primary PI have been reported. Although primary PI often represents a benign condition that should not be considered as an argument for surgery, if the case involves intussusception and obstruction, emergent laparotomy should be considered.

Key words: Pneumatosis intestinalis; Intussusception; Urgent surgery; Immunosuppressive drug; Ischemia of the intestine

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Core tip: We report a patient with pneumatosis

intestinalis (PI) and obstructing intussusception who underwent urgent colectomy and review the literatures regarding PI with intussusception. A 20-year-old man presented at our hospital with abdominal pain, and has undergone steroid therapy for 4 years. Computed tomography revealed ascending colon intussusception with air within the wall, and colectomy was performed. Histological examination of colonic wall sections revealed large cysts in the submucosal layer. Nine cases of intussusception associated with primary PI have been reported. Although primary PI often represents a benign condition, if the case involves intussusception and obstruction, emergent laparotomy should be considered.

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INTRODUCTION

Pneumatosis intestinalis (PI) is a rare condition characterized by the presence of gas within the wall of the gastrointestinal tract. This condition can result from a wide variety of pathologies, including chronic obstructive lung disease, collagen diseases, necrotizing enterocolitis in premature infants, intestinal infections, ischemic bowel disorders, and immunosuppressive drug therapy^[1]. PI often represents a benign condition that should not be considered as an argument for surgery^[2]; however, immediate surgery may be required in some life-threatening circumstances such as the presence of bowel obstruction, perforation, or ischemia^[3].

Here we describe a case of PI in the ascending colon with obstructing intussusception for which urgent surgery was performed, and review the available published literature on PI with intussusception. Written informed consent was obtained from the patient.

Search strategy

The literature search strategy for this study was based on published systematic review guidelines^[4]. Literature databases such as PubMed MEDLINE (National Library of Medicine) were searched from 1980 to 2015 using the following medical subject headings: "PI (or Pneumatosis cystoides intestinalis)" and "intussusception" or "PI (or Pneumatosis cystoides intestinalis)" and "invagination". In addition, references within the retrieved articles were reviewed. We identified 24 manuscripts using this search strategy and selected 8 case reports for this review. Nineteen articles were excluded because their content was not applicable to this review and 7 articles were excluded because they were not written in English.

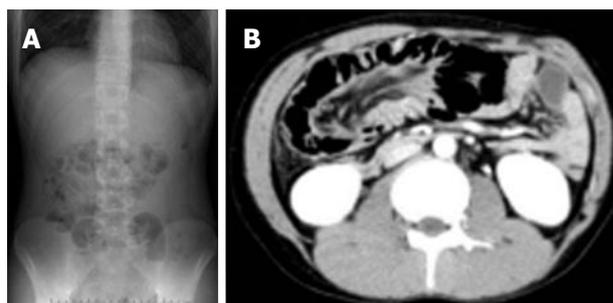


Figure 1 Abdominal radiograph. It showing multiple distended loops of small bowel with fluids and multiple air pockets (A) and computed tomography showing multiple gas-filled cysts, a streaky collection of air in the bowel wall, and an intussusception of the colon (B).



Figure 2 Intraoperative findings showed intussusception of the ascending colon with palpable soft polypoid masses.

CASE REPORT

A 20-year-old man presented our hospital with a 3-d history of intermittent lower abdominal pain. He had been on steroid therapy (methylprednisolone 25 mg/d) for membranoproliferative glomerulonephritis for 4 years. A physical examination revealed tenderness in the lower right quadrant of the abdomen. His body temperature was 37.7 °C and pulse was 81 beats per minute. All serum levels tested were within the normal range, with the exception of serum total bilirubin (1.5 mg/dL; normal range, 0.3-1.2 mg/dL). White blood cells (WBCs) (21000/μL), hemoglobin concentration (17.1 g/dL), and the C-reactive protein concentration (0.5 mg/dL) were also elevated, indicating acute inflammation and dehydration, with a level of base excess of 2.4 mmol/L. Abdominal X-ray showed multiple air-filled lucencies in the right upper quadrant and multiple distended loops of small bowel with fluid (Figure 1A). Computed tomography (CT) revealed intussusception of the ascending colon with air within the wall (Figure 1B). We performed an urgent laparotomy under the diagnosis of acute abdomen with obstructing intussusception. The colo-colic intussusception that was caudal to the polypoid lesion easily resolved using Hutchinson's maneuver and soft polypoid masses were palpable from the cecum to the ascending colon (Figure 2). Intraoperative colonoscopy revealed the presence of numerous soft polypoid

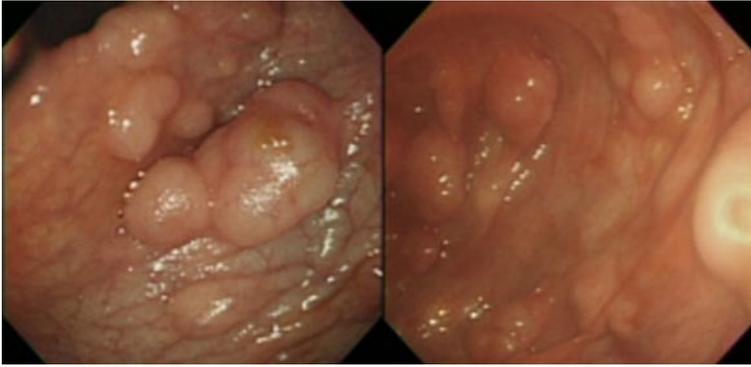


Figure 3 Intraoperative colonoscopy showed numerous soft polypoid masses with normal overlying mucosa located between the ascending colon and middle part of transverse colon.

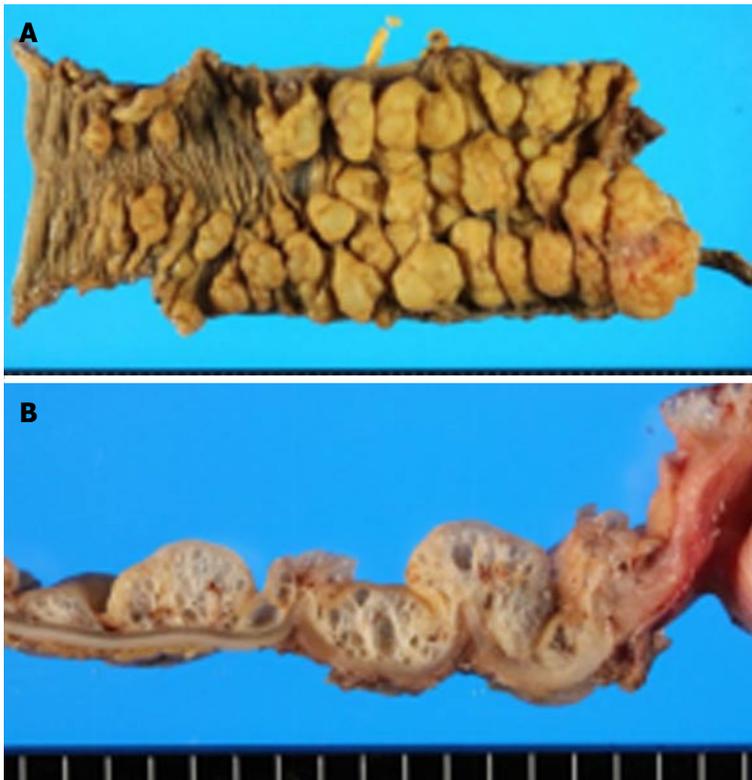


Figure 4 The resected specimen revealed polypoid lesions with normal mucosa and cystic structures (A), submucosal cysts had a spongy consistency (B).

masses with normal overlying mucosa (Figure 3). A right hemicolectomy was performed because other polyposis diseases or intussusception relapse could not be ruled out, and to help make a final pathological diagnosis. A gross examination showed that the mucosa of the resected colon appeared normal with no evidence of ulceration or ulcer-related lesions, but instead a number of soft, yellowish cystic masses (Figures 4). A histological examination of sections of the colonic wall revealed large cysts in the submucosal layer (Figure 5A). The cysts were empty, but were surrounded by a distinct fibrous wall and were lined by macrophages that frequently coalesced to form multinucleated giant cells (Figure 5B). Based on these findings, the patient was diagnosed with pneumatosis cystoides intestinalis. The postoperative course was uneventful and the patient was discharged on postoperative day 9. No recurrence was noted on radiographic imaging performed on postoperative 14 mo.

DISCUSSION

Classically, PI can be subdivided into 2 distinct groups: primary PI, representing 15% of cases, and secondary PI, representing 85% of cases^[5]. Secondary PI, where the gas accumulates as linear collections and reflects a pathological condition, has been attributed to endoscopic procedures, immunological disturbances, bowel mucosal disruptions, and intra-abdominal pathologies. In contrast to secondary PI, primary PI is characterized by intramural gas that is cystic and benign in nature and does not always require urgent laparotomy^[6]. Although PI may occur in association with acquired immunodeficiency^[7], transplant status^[8], cancer treatment^[9,10], scleroderma^[11], cystic fibrosis^[12], systemic lupus^[13], inflammatory bowel disease^[2], intestinal ischemia^[14], colitis^[15], or trauma^[16], the exact etiology of both primary and secondary PI remains unknown.

There is currently no consensus on the appropriate

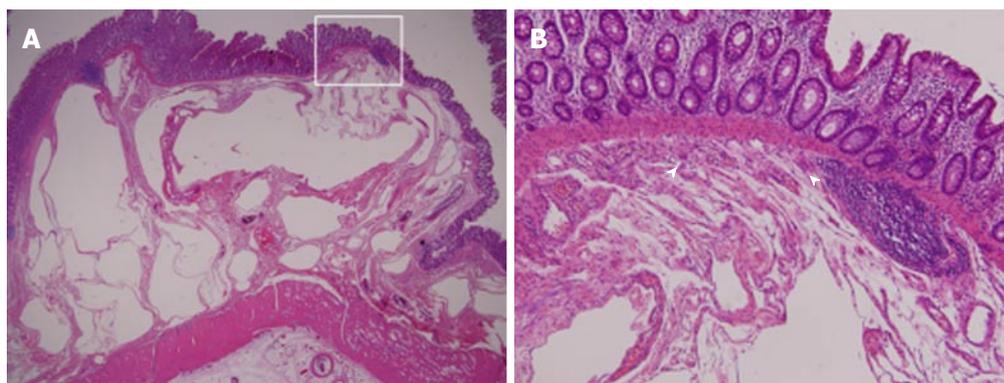


Figure 5 Histopathological examination revealed cystic air-filled spaces within the submucosa, which were partially lined by clusters of foreign-body macrophages (arrow heads) (hematoxylin-eosin stain; A: × 40, B: × 400).

Table 1 Reported cases that had intussusception associated with primary pneumatosis intestinalis										
Ref.	Year	Sex	Immunosuppressive drug	Ischemia	PVG	Site	Treatment	Indication of surgery	Co-morbidity	
Nagata <i>et al</i> ^[20]	23 yr	Male	No	No	No	A/C	CS→Surgery	Abdominal pain	None	
Emanuel <i>et al</i> ^[21]	48 yr	Male	NR	NR	NR	D/C	Surgery	Obstruction	Hybrid perineurioma-schwannoma	
Sugita <i>et al</i> ^[22]	5 yr	Female	Yes	No	Yes	A/C	BE	-	CML	
Stern <i>et al</i> ^[23]	32 yr	Male	No	No	No	A/C	BE	-	None	
Morrison <i>et al</i> ^[24]	3 mo	NR	Yes	Yes	NR	T/C	BE→Surgery	could not resolved intussusception	Peter's anomaly	
Dubinsky <i>et al</i> ^[25]	1 yr	Male	Yes	NR	NR	A/C	Surgery	Obstruction	Crohn's disease	
Navarro <i>et al</i> ^[26]	13 yr	Male	No	No	NR	T/C	BE→Surgery	Obstruction	None	
Ahrar <i>et al</i> ^[27]	29 yr	Male	No	No	NR	A/C	BE→Surgery	NR	None	
Our case	20 yr	Male	Yes	No	No	A/C	Surgery	Obstruction	MPGN	

NR: Not referred; PVG: Portal venous gas; A/C: Ascending colon; D/C: Descending colon; T/C: Transverse colon; CS: Colonoscopy; BE: Barium enema; CML: Chronic myelogenous leukemia; MPGN: Membranoproliferative glomerulonephritis.

management of PI, although many mechanical, bacterial, and pulmonary hypotheses have been proposed regarding PI etiopathogenesis, and its management can be challenging for surgeons^[17]. Many studies have investigated the use of risk factors as predictors of a compromised bowel and the probable need for surgery, such as patient age and the presence of hypotension, peritonitis, renal failure, or serum lactate levels^[18]. Other studies have attempted to create algorithms for PI management that, while helpful, are also tedious and may be difficult to apply clinically in circumstances where the patient requires rapid evaluation^[19]. In this case, we performed urgent laparotomy because he had intestinal obstruction due to intussusception and several inflammatory symptoms.

Although the course of primary PI may be benign or may not frequently result in a need for urgent surgery, laparotomy should be considered in cases with intestinal obstruction due to intussusception. To our knowledge, 9 reported cases, including the present case, had intussusception associated with primary PI (Table 1)^[20-27]. The mean patient age was 19.0 ± 16.0 years (range 0-48 years), which is younger than has been reported previously^[19]. It is notable that more than

44% of these patients received an immunosuppressive drug, and few patients appeared to have intestinal ischemia. All patients had the intussusception in the colon, particularly on its right side. Five out of 9 patients had co-morbidities, most of which required an immunosuppressive drug. With the exception of the article placed as reference number 22, no other articles referred to the presence of portal venous gas, which often indicates ischemic bowel disease. Only 2 cases experienced successful reduction of the intussusception with barium enema^[22,23], although long-term outcomes were not reported. Among the 4 cases with initial successful reduction of the intussusception by colonoscopy or barium enema, all of these patients eventually needed surgery: One case had persistent abdominal pain after solution of intussusception; in one case, the intussusception could not be completely resolved; one case had bowel obstruction; and for one case the indication of surgery was not clearly stated. For the present case, we performed an urgent laparotomy because of bowel obstruction with intussusception of the ascending colon. Right hemicolectomy for this case was performed because of the possibility of there being another polyposis disease present and because relapse

of the intussusception could not be ruled out.

In conclusion, although primary PI often represents a benign condition that should not be considered as an argument for surgery, emergent laparotomy should be considered for cases with intussusception, obstruction, and unsuccessful resolution of intussusception by colonoscopy or barium enema.

COMMENTS

Case characteristics

A 20-year-old man presented our hospital with a 3-d history of intermittent lower abdominal pain.

Clinical diagnosis

The authors performed an urgent laparotomy under the diagnosis of acute abdomen with obstructing intussusception.

Differential diagnosis

A right hemicolectomy was performed because other polyposis diseases or intussusception relapse could not be ruled out.

Laboratory diagnosis

All serum levels tested were within the normal range, with the exception of serum total bilirubin (1.5 mg/dL; normal range, 0.3-1.2 mg/dL). White blood cells (WBCs) (21000/ μ L), hemoglobin concentration (17.1 g/dL), and the C-reactive protein concentration (0.5 mg/dL) were also elevated.

Imaging diagnosis

Computed tomography revealed intussusception of the ascending colon with air within the wall.

Pathological diagnosis

A gross examination showed that the mucosa of the resected colon appeared normal with no evidence of ulceration or ulcer-related lesions, but instead a number of soft, yellowish cystic masses, suggesting that to be pneumatosis cystoides intestinalis.

Treatment

A right hemicolectomy was performed because other polyposis diseases or intussusception relapse could not be ruled out, and to help make a final pathological diagnosis.

Related reports

To our knowledge, 9 reported cases, including the present case, had intussusception associated with primary pneumatosis cystoides intestinalis.

Experiences and lessons

Although primary pneumatosis intestinalis (PI) often represents a benign condition that should not be considered as an argument for surgery, emergent laparotomy should be considered for cases with intussusception, obstruction, and unsuccessful resolution of intussusception by colonoscopy or barium enema.

Peer-review

This is an interesting article summarising a case of PI and intussusception with a review of the cases in the literature.

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