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A Case of Pneumatosis Intestinalis in Ph+ Acute Lymphoblastic Leukemia

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Abstract

Pneumatosis intestinalis (PI) is an uncommon condition and it is characterized by the presence of gas within bowel wall. The cause of PI is diverse, although specific etiologic factors remain unknown. PI can be seen infrequently in leukemia and be associated with several medical and surgical conditions. Sometimes, PI can be presented from a benign to a fatal situation. Plain radiographs can diagnose all most all cases. Now a day, using CT scan is increased. And that makes easy to diagnose of PI. Here, we report a case of PI that developed in recovery phase after induction chemotherapy in a 46 year-old woman, with Ph+ ALL.

Key Words : Acute lymphoblastic leukemia, Pneumatosis intestinalis, Pneumoperitoneum

Introduction

Pneumatosis intestinalis represents air in the intestinal wall [1]. The cause of PI is variable, but specific etiologic factors remain unknown [2]. Common causes of PI are lung disease like asthma and bronchiectasis, inflammatory bowel disease (e.g. Crohn's disease), connective tissue disease, infection, cytotoxic agents, steroid, laxative, iatrogenic cause like stab wound and endoscopic procedure [3]. There are some etiology for explain PI [4]. The one, increasing intramediastinal or intraabdominal pressure makes space of intestinal wall and air leak to that space. And the other theory is the cause of increasing permeability of mucosal layer, gas forming bacteria infiltrated in submucosa of intestine [5-7]. And that bacteria makes gas and air trap in the bowel wall. The process of PI is almost mild and needs no specific treatment. Just bowel rest and supportive care are needed. But in

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rare cases, complications like bowel obstruction or perforation are occurred, and that conditions are needed surgical procedure [3,7,8]. For early detection of that complication, CT scan is useful.

In leukemia patients, both of children or adults, PI is occurred associated with using steroid, chemotherapy, bone marrow transplantation and infection [2,5,9,10]. In rare cases, PI resulted from the leukemic process [11]. We experienced a case of PI in Ph+ ALL. The time of PI onset was recovery phase after chemotherapy. And that time, the patient already got remission state.

Case Report

A fourty-six years old woman visited emergency room due to recent onset general weakness. Initial white cell counts were 137 × 10^6/L, hemoglobin was 11.9 g/dL, platelet counts were 47 × 10^6/L. Bone marrow aspiration revealed 95% blast out of nucleated cells and CD 10, CD 19, CD 34, TdT and HLA-DR positive. BCR/ABL fusion transcription genes were detected in RT-PCR. She was diagnosed Ph+ALL. The patient was treated with daunorubicin, vincristine, prednisone and imatinib. After cell counts were recovered, follow up bone marrow biopsy was done and the result was complete remission. Two weeks after confirmed remission, she complained abdominal distension and whole abdominal pain. At that time, cell counts were completely recovered from cytopenia following induction chemotherapy. Laboratory results revealed a leukocyte count of 6.45 x 10^6/L, 77.5% neutrophils, a platelet count of 160 x 10^6/L, a hemoglobin level of 9.9 g/dL. Other laboratory findings, including electrolytes, liver function tests, amylase, and CRP were within normal ranges. No acidosis was shown by arterial blood gas analysis. She had no fever and checked normal range vital sign. There were no evidence of infection. Plain radiographs which were checked 26 days after induction chemotherapy, revealed gas-filled small bowel loops and large bowel loops (Fig. 1A, B). CT scan showed extensive mural air in bowel loop and free air in subphrenic space, but there were no evidence of bowel necrosis or other complication to need surgical procedure (Fig. 2A, B, C). Patient received conservative treatment with parenteral nutrition and antibiotics. She fasted, and were provided IV nutrition daily 1,500 Kcal for 28 days. To prevent infection, we used levofloxacin 750 mg IV infusion once a day and vancomycin 1 g twice a day for 12 days. Eventually the bowel gases were completely absorbed 3 weeks later (Fig. 3A, B). At that time, there were no fever and blood culture was reported no growth. And she did not complain abdominal pain. She completely recovered from PI. She discharged safely, and got consolidation chemotherapy after 2 months later. During the nadir period of 2nd consolidation chemotherapy, *E. coli* and *E. faecium* were isolated from the blood culture. She died because of Sepsis.

Discussion

PI is a rare complication after chemotherapy in leukemia [1,6]. That condition needs treatment from just supportive care to surgical procedure, disease severity of PI is variable. If diagnosed early, almost all PI does not require surgical intervention and can be managed successfully by parenteral nutrition and broad-spectrum antibiotics [7,12,13]. Correlation with clinical history, physical examination and laboratory test results are the best indicator of whether PI is due to a benign or life-threatening causes [7]. But in rare cases, complications like bowel obstruction or perforation are occurred, and that conditions are needed surgical procedure

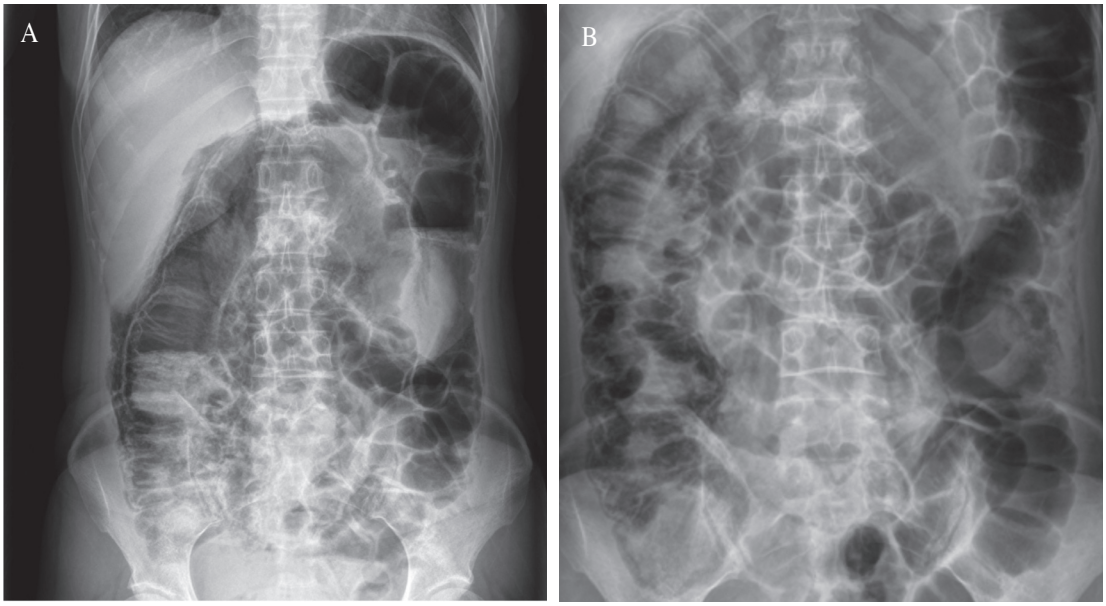

Fig. 1. Initial simple abdomen X-ray. Erect (A) and supine (B) view, when she complained abdominal pain. It shows intramural air in small bowel wall.

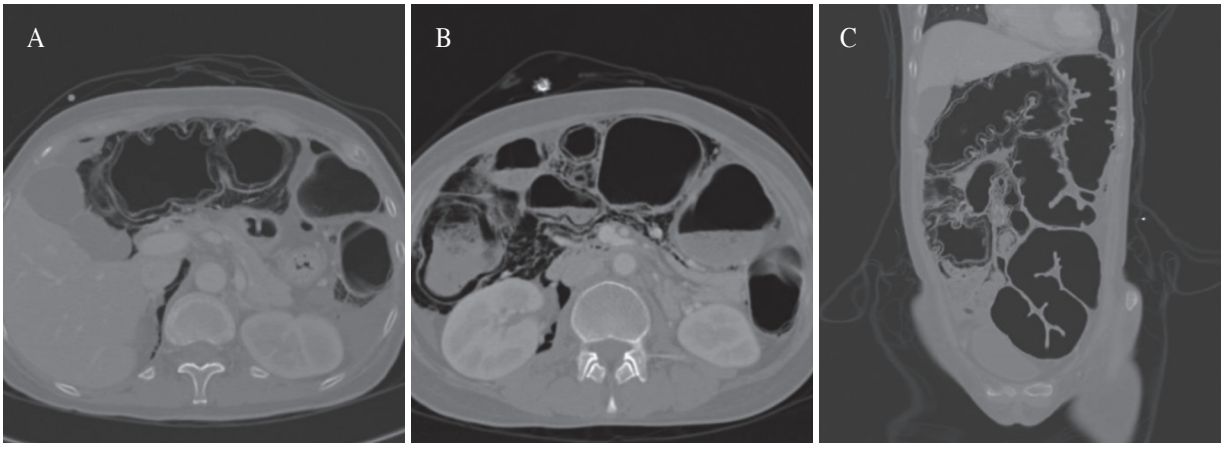

Fig. 2. CT scan shows intramural air (arrow) in the transverse colon (A), ascending colon and small bowel (arrow) (B). Extensive mural air in small bowel loops, ascending colon and transverse colon but, normal bowel enhancement and no visible air in mesenteric vessels and portal vein. No evidence of bowel necrosis. (C) is coronal view of CT scan.

[3,7,8]. Now a day, imaging technique was advanced. And CT scan can detect complications of PI, which required surgical procedure. Most of the leukemia related PI developed in neutropenic period. And almost all cases of PI were following by chemotherapy and immunosuppressive therapy. Because, at that time, patients are particularly vulnerable to infection. Bae *et. al*. [14], reported a case of PI, that was developed with leukemic cell infiltration during leukemia progression. Jaffe N *et.*

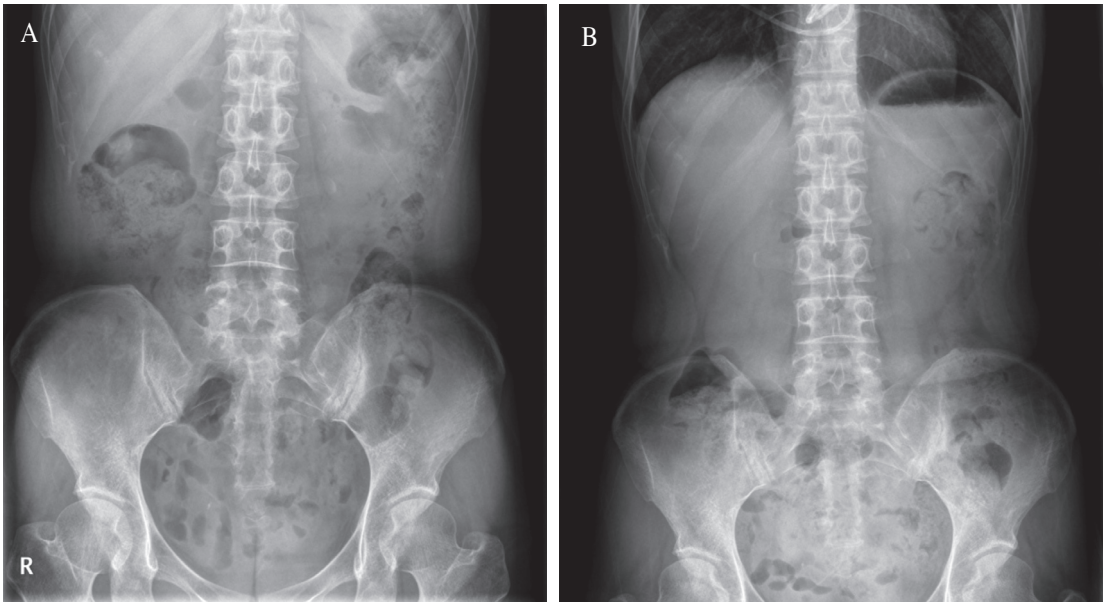

Fig. 3. After treatment, mural air was absorbed and simple x-ray became normalization (A, B).

al. [11], published some cases of pneumatosis intestinalis in leukemia patients. Two of all that cases were caused by leukemic cell infiltration revealed by autopsy. But our case developed in complete recovery state from myelosuppresive phase. Medication side effects can be an overlooked cause of unexplained PI. Corticosteroid administration is the most common cause of medication-induced PI [15]. Steroids have been shown to cause atrophy of lymphoid aggregates (Peyer's patches) in the gastrointestinal tract, which can in turn lead to loss of submucosal structural integrity and allow dissection of intraluminal air into the intestinal wall. The timing of our PI case was recovery period of induction chemotherapy. Patient received high dose corticosteroid over 2 weeks and cytotoxic chemotherapy. Both agents could be the cause of this PI. For these reasons, we excluded leukemia progression as the cause of PI. We briefly summarize in recovery period PI in Ph+ ALL. Base on all that reasons, we assess that long term use of steroid and cytotoxic chemotherapy caused PI in this patient.

In conclusion, PI should be considered as a complication of recovery phase of hematologic malignancies.

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