CASE REPORT

Benign pneumatosis intestinalis after bilateral lung transplantation

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SUMMARY

This series details two cases of benign pneumatosis intestinalis (PI) in patients post-lung transplant, which were discovered incidentally on routine surveillance chest radiographs during ambulatory clinic visits. Both patients had uneventful post-transplant recovery and were asymptomatic at presentation. The patients were admitted for observation. Contrast-enhanced abdominal CT scans confirmed the plain film findings. Both cases were managed conservatively with bowel rest, intravenous hydration and serial abdominal examinations. The patients had unremarkable hospital courses and were both discharged in good condition. Our current understanding of benign PI in patients post-transplant is limited to a few case series and case reports. Greater awareness of this entity may decrease unnecessary invasive procedures and improve management of these patients.

BACKGROUND

Pneumatosis intestinalis (PI) is a radiological and pathological finding characterised by gas within the bowel wall.^{1–4} PI may have a benign aetiology or harbinger a life-threatening cause such as intestinal ischaemia, mesenteric vascular disease or intestinal obstruction.⁵ Benign PI is an uncommon and little understood phenomenon, which has been reported in conjunction with associated conditions such as steroid therapy, chronic obstructive pulmonary disease (COPD), asthma, scleroderma, endoscopy and organ transplantation.⁵ Benign PI in patients post-lung transplant is even less recognised,⁶ with the literature limited to case reports and case series. We report incidental discovery and follow-up of two cases of PI in patients post-lung transplant.

CASE PRESENTATION**Case 1**

A 68-year-old man, patient A, status post-bilateral lung transplant for COPD, presented for a routine clinic visit on postoperative day (POD) 49. He was asymptomatic and was recovering well from the transplantation. His immunosuppressive regimen comprised of tacrolimus (satisfactory trough level of >7 ng/mL), mycophenolate mofetil and prednisolone. Vital signs and physical examination were unremarkable. Serum blood work was within normal limits except for a mild lactic acidosis of 2.6 mmol/L. Serological testing for cytomegalovirus (CMV) infection was negative. On routine surveillance chest radiograph, a large amount of pneumoperitoneum was seen, and the bowel had a mottled appearance, which raised the possibility of PI

(figure 1). Subsequent abdominal radiographs demonstrated pneumoperitoneum and PI involving the right hemicolon. The patient was admitted for observation. A contrast-enhanced abdominal CT scan confirmed extensive colonic PI, most pronounced around the hepatic flexure, and pneumoperitoneum with otherwise normal bowel wall architecture. No pericolic stranding, ascites, or portal venous gas was seen. The patient was managed conservatively with bowel rest, intravenous hydration and serial abdominal examinations. He remained asymptomatic throughout the inpatient observation period and was discharged the next day in good condition.

Case 2

A 52-year-old woman, patient B, status post-bilateral lung transplant for scleroderma, presented for a routine follow-up clinic visit on POD 60. She was asymptomatic and recovering well from her procedure. Her medications included tacrolimus (trough level of 16.7 ng/mL), mycophenolate mofetil and prednisone. Vital signs and physical examination were unremarkable. Serum laboratory work showed a mild uraemia and alkalosis but was otherwise within normal limits, including complete blood count, basic metabolic panel and lactic acid of 1.6 mmol/L. Serological testing for CMV was negative. The patient's routine chest radiograph was suspicious for PI and pneumoperitoneum.

Figure 1 Posteroanterior chest radiograph showing large amount of pneumoperitoneum subjacent to the diaphragm (thick arrows) and bowel with mottled appearance (thin arrows).

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Figure 2 Upright abdominal radiograph showing extensive pneumatosis intestinalis along entire ascending colon (thick arrows) and also pneumoperitoneum under the diaphragm and along the right paravertebral region, likely extending into the retroperitoneum (thin arrows).

Abdominal radiographs demonstrated ascending colonic PI and free air in the peritoneum as well as retroperitoneum (figure 2). The patient was admitted for further work up. On CT imaging with contrast, extensive PI was found in the ascending colon, with right-sided intraperitoneal and retroperitoneal free air (figure 3). There was no inflammatory stranding evident on imaging. The patient was managed conservatively and remained asymptomatic throughout the hospitalisation.

Figure 3 Coronal CT image showing extensive pneumatosis intestinalis along entire ascending colon, with right-sided intraperitoneal and retroperitoneal free air (arrows).

OUTCOME AND FOLLOW-UP

Patient A returned to the clinic for a routine visit 35 days after discharge (POD 84), and imaging demonstrated resolution of PI and pneumoperitoneum. However, the follow-up chest radiograph of patient B, on POD 74, showed persistent PI as well as intraperitoneal and retroperitoneal free air. However, she remained asymptomatic.

DISCUSSION

PI is a rare clinical finding with aetiologies varying from innocuous to life-threatening. PI has been associated with various conditions such as transplantation, AIDS, cancer, COPD and scleroderma.^{2–12} The pathophysiology of benign PI remains unclear. There are two popular postulated mechanisms of benign PI: mechanical and bacterial. The mechanical theory posits increased positive pressure and consequent leakage of air through microscopic breaks in the bowel wall.^{1 2 11 13} The bacterial theory puts forth translocation of gas-producing bacteria among the normal gut flora into the bowel wall and gas production within the mucosal layers.^{2 11}

One apparent commonality between our patients and earlier reported cases of benign PI is the use of steroids and (medical or pathological) immunosuppression.^{4 10 11} Atrophy of the intestinal lymphoid tissues has been postulated to compromise mucosal integrity and to lead to PI.⁶ CMV infection has also been put forth as a potential cause of PI in patients post-transplant,⁷ but our observations do not corroborate this hypothesis.

Although clinical parameters guide management of PI, clinico-radiological correlation has been attempted to delineate benign from more sinister aetiologies. PI limited to the large bowel is associated with benignity whereas isolated PI of the small bowel is associated with a more grave aetiology.^{9 11} Other worrisome imaging features include decreased or absent mural enhancement, bowel wall thickening, mesenteric stranding, ascites, bowel dilation and portomesenteric venous gas.^{11 14} We had similar observations where the PI in both asymptomatic patients was limited to the colon and other worrisome features were not noted.

The presence of pneumoperitoneum has not been associated with life-threatening causes of PI.¹¹ Pneumoperitoneum in the absence of concerning clinical findings may not warrant invasive surgical procedures. Similar to two earlier case reports of benign spontaneous pneumoperitoneum in the presence of PI,^{12 15} our patients had a benign course with conservative management despite having pneumoperitoneum. Patient A's pneumoperitoneum resolved spontaneously whereas patient B's pneumoperitoneum persisted, though without symptoms or complications.

Serum lactic acid >2 mmol/L has been associated with a mortality rate of $>80\%$ in patients with PI.⁵ Nonetheless, Patient A whose serum lactic acid was 2.6 mmol/L, had a benign course. In a recent study, Lee *et al*¹⁴ proposed a score to predict the mortality in patients with PI instead of arbitrary assignment as either benign or life-threatening. The score accounts for the radiological findings (ie, bowel wall enhancement) as well as the clinical presentation (ie, signs of peritoneal irritation). According to Lee *et al*'s scoring system, our patients' estimated mortality risk was 0.0185 and 0.0408 on the first and seventh day of PI, respectively.

In conclusion, benign PI is an uncommon incidental finding in asymptomatic patients post-lung transplant. Although the threshold for intervention is low in patients post-transplant,

limited literature shows that conservative management may be sufficient in appropriate cases. Greater understanding of this entity may lead to improved patient care.

Patient's perspective

Patient A

It was a routine visit. There was nothing wrong. I felt no pain. I felt great. As we were leaving the clinic, they called us back in. What they were telling us was so frightening at the time. The gastroenterologists and the surgeons came to look at me, and my doctor ended up admitting me to the hospital. After they were done with performing the tests, they said I could go. I felt good that I did not have to have another operation. I've had enough operations.

Patient B

During the transplant follow-up, an X-ray was ordered. The finding was that some air was found. I did not have any symptoms. There was no nausea or pain. They scared me to death when they told me I might have to have surgery again. They said things like, 'the tear could get bigger and burst,' and, 'sepsis,' etc. That really scared me. After the admission, the team decided to wait it out and put me on liquid diet for a week. I felt relieved when they told me I didn't have to have emergency surgery. I was anxious throughout all of this but was so glad that I didn't have to have another surgery. The transplant was stressful enough! I am grateful that it was something treatable with monitoring. I am really happy that I am still sitting here breathing. (By the patient's sister, for the patient.)

Learning points

- ▶ Pneumatosis intestinalis (PI) is associated with various conditions such as transplantation, AIDS, cancer, chronic obstructive pulmonary disease and scleroderma.
- ▶ Benign PI is a rare incidental finding in asymptomatic patients post-lung transplant and may not require invasive management.
- ▶ In patients with PI, worrisome imaging features on CT include small bowel involvement, bowel wall thickening, mesenteric stranding, ascites, bowel dilation and portomesenteric venous gas.

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