Pneumatosis Intestinalis Associated with Pulmonary Disorders

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Purpose To determine the clinical features, imaging findings and possible causes of pneumatosis intestinalis (PI) in thoracic disorder patients.

Materials and Methods From 2005 to 2017, Among 62 PI patients, four of PI related with thoracic disease (6%) were identified. Medical records were reviewed to determine the clinical presentation, laboratory findings and treatment at the time of presentation of PI. Two experienced chest radiologists reviewed all imaging studies and recorded specific findings for each patient.

Results The causative thoracic diseases for each four patient were severe asthma, emphysema and airway obstruction. The imaging appearance of PI, including the involved bowel segment and pattern of the air, were divided into two mesenteric vascular territories; three of our cases showed linear pattern of PI located in the ascending & proximal transverse colon and the fourth case (lung cancer) had bubbly and cystic PI in the distal transverse and descending colon. All of the remaining 3 patients, except one patient who had not been followed up, improved within 1 month by conservative treatment.

Conclusion Thoracic disorder with obstructive lung disease may result in the development of benign PI. Such PI in thoracic disease patients has a similar linear and cystic appearance with ischemic bowel disease, but can nevertheless be managed by conservative treatment.

Index terms Pneumatosis Cystoides Intestinalis; Asthma; Emphysema; Airway Obstruction

INTRODUCTION

Pneumatosis intestinalis (PI) is a radiologic finding characterized by formation of gas bubbles within the bowel wall and is a rare disorder which has recently come to increased clinical attention due to improved radiographic identification (1, 2). PI has been reported in association with many clinical conditions ranging from benign to the life-
threatening, and it is important to promptly diagnose this disorder, because the overall morta-
ity is estimated to be between 20% and 47% (3, 4). Since secondary PI suggesting underly-
ing pathology accounts for about 85% of cases (5) and can represent a wide range of pathologies, delineating the certain etiology may be difficult. The reported cause of the PI is 
not exclusively limited to gastrointestinal tract pathology and pulmonary causes, while de-
batable, have been suggested such as asthma, cystic fibrosis and other obstructive broncho-
pathologies e.g., chronic obstructive pulmonary disease (COPD) (6).

To our knowledge, there has not been a series of secondary PIs associated with pulmonary 
disease in the radiologic literature. The purpose of our study was to describe the clinical and 
imaging features and outcomes of benign PI from a pulmonary disorder in four patients 
treated at our institute. We emphasize the importance of early recognition of the overall clin-
ic picture for decision-making in a patient with dyspnea and a radiological finding of PI or 
pneumoperitoneum to efficiently distinguish between benign and life-threatening PI.

MATERIALS AND METHODS

PATIENTS

From March 2005 to October 2017, consecutive four (6%) of 62 PI patients who developed PI related with thoracic disease were identified in our institute. This study included four pa-
tients (two females and two males; age range, 43–79 years; mean age, 63 years) with CT scan 
findings suggestive of PI. Abdomen CT (n = 3) and chest CT (n = 4) scans were available. Initially, a chest CT (n = 4, three enhanced chest CT and one low-dose chest CT) and brain CT (n = 1) were performed because of major patient complaints (dyspnea and mental change), and 
abdomen CT in three patients were subsequently performed. The time interval between the 
chest CT and the abdomen CT was within 24 hours. Patient 2 was first diagnosed with lung 
cancer by chest CT, and he refused further evaluation, including abdomen CT and followed 
up the loss. Our Institutional Review Board approved this retrospective study, and the re-
quirement for informed consent was waived (EUMC 2018-08-001).

CT PROTOCOL

Abdominal CT examinations were performed by using a 16-channel multidetector-row CT 
(MDCT) scanner (SOMATOM Sensation, Siemens Medical Solutions, Forchheim, Germany) 
for two patients and a 64-channel MDCT scanner (SOMATOM Sensation 64, Siemens Medical 
Solutions) for one patient. MDCT images were obtained from the diaphragm to the symphy-
sis pubis during a single breath-hold. A detector configuration of 1.5 mm × 16 and a table 
speed of 24 mm per gantry rotation were used for 16-channel MDCT scans. A detector con-
figuration of 0.6 mm × 32 with a Z-flying spot system and a table speed of 38.4 mm per gan-
try rotation were used for 64-channel MDCT scans. A pitch of 1, a reconstruction thickness of 
5 mm, 120 kVp and variable tube current (90–140 mAs) for both MDCT scanners were used. 
Single-phase contrast-enhanced images were obtained at 90 or 100 seconds after the injec-
tion of 120 mL of the contrast agent (iohexol, Omnipaque 300; Nycomed, Zurich, Switzerland 
or iopromide, Ultravist 300; Schering, Berlin, Germany) at a rate of 3 mL/s.

Chest CT scans were performed using a 16-channel MDCT scanner (SOMATOM Sensation)
for all patients. The parameters for helical chest CT imaging were 120 kVp, 80–100 mAs, 5 mm collimation and a 10 mm/sec table feed. Contrast-enhanced chest CT scans were obtained after injection of 30 g of iodinated contrast agent (100 mL iopromide, Ultravist 300) at a rate of 2.3 mL/s with the use of a power injector (OP100, Medrad, Pittsburgh, PA, USA). The scan data were displayed directly on monitors (two monitors, 512 × 512 image matrices, 12-bit viewable gray scale) coupled to a picture archiving and communication system (Starpacs, Infinitt, Seoul, Korea).

CLINICAL ANALYSIS
A total of 62 patients had PI at our institution between March 2005 and May 2016. Among these patients, the four patients (6%) who had PI with thoracic disorder without abdominal lesion were included in this study. The medical records of the four patients were recorded and included the following criteria: age, sex, clinical presentations, past medical histories, physical examinations, laboratory data, treatments and outcome from PI or pneumoperitoneum.

IMAGING ANALYSIS
The patient’s abdominal radiographs, chest radiographs, abdomen CT and chest CT scans were reviewed by two radiologists with 15 years and 3 years of experience, respectively. On abdomen CT, the extent of PI involved bowel segment, combined mesenteric and/or retroperitoneal gas and other abdominal lesions were recorded. On chest CT, an underlying pulmonary finding was obtained, indicating the presence of an additional mediastinal or bony lesion. All CT imaging data were retrospectively evaluated by 2 experienced radiologists, and a final correlation of imaging findings were taken.

RESULTS

CLINICAL
The clinical findings for the four patients are summarized in Table 1. The underlying known thoracic diseases were asthma, pulmonary emphysema and lung graft-versus-host disease (GVHD) after peripheral blood stem cell transplantations for acute myeloid leukemia in three patients (Patient 1, 2, 3). One patient (Patient 2) was diagnosed with lung cancer at this hospital visit. The chief complaint of all patients was dyspnea, and one patient (Patient 4) had dyspnea with a mental change. Abdominal symptoms were indigestion, poor oral intake, and abdominal distension in three cases and one had no specific symptoms. All four patients had soft abdomens and no tender points; only one patient had an abdominal distention. There is no known history of a connective tissue disorder, inflammatory bowel disease, acquired immune deficiency syndrome (AIDS), recent trauma, operative or endoscopic procedure history in all the patients. Three of the patients had leukocytosis based on laboratory data.

All three patients (except follow up loss patient) were treated conservatively by medication and parenteral nutrition, kept at nothing by mouth. Three patients showed improved PIs or pneumoperitoneum within 1 month (range 7 days to 1 month). The guidance of the patient 2 (his son) came to the hospital after 1 month and told that patient complained the systemic pain.
Table 1. Clinical Findings in Four Patients with Pneumatosis

<table>
<thead>
<tr>
<th>No.</th>
<th>Sex/Age</th>
<th>Initial Chief Complaint</th>
<th>Duration</th>
<th>Abdomen Symptom</th>
<th>Underlying Disease</th>
<th>Initial Department</th>
<th>Initial Diagnostic Study</th>
<th>PaO₂</th>
<th>Final Diagnosis</th>
<th>Medication</th>
<th>Outcome of Pneumatosis Intestinalis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F/43</td>
<td>Dyspnea</td>
<td>4 days</td>
<td>Indigestion</td>
<td>Asthma</td>
<td>Emergency room</td>
<td>Chest PA Chest CT</td>
<td>81.7</td>
<td>Asthma exacerbation</td>
<td>Methylon</td>
<td>Resolved in 1 month</td>
</tr>
<tr>
<td>2</td>
<td>M/79</td>
<td>Dyspnea</td>
<td>7 days</td>
<td>Poor oral intake</td>
<td>Pulmonary emphysema</td>
<td>Respiratory medicine</td>
<td>Chest PA Chest CT</td>
<td>-</td>
<td>Lung cancer (left main bronchus obstruction)</td>
<td>-</td>
<td>Follow up loss</td>
</tr>
<tr>
<td>3</td>
<td>F/53</td>
<td>Dyspnea</td>
<td>3 days</td>
<td>-</td>
<td>AML</td>
<td>Hematologic medicine</td>
<td>Abdomen CT</td>
<td>51.2</td>
<td>GVHD and pneumonia</td>
<td>Methylon</td>
<td>Resolved in 17 days</td>
</tr>
<tr>
<td>4</td>
<td>M/71</td>
<td>Dyspnea &amp; mental change</td>
<td>Abrupt</td>
<td>Abdomen distension</td>
<td>-</td>
<td>Emergency room</td>
<td>Chest PA Brain CT Chest CT</td>
<td>33.1</td>
<td>Airway obstruction by food impaction and aspiration pneumonia</td>
<td>Endoscopic removal of food material</td>
<td>Resolved in 7 days</td>
</tr>
</tbody>
</table>

AML = acute myeloid leukemia, GVHD = graft-versus-host disease, PA = posteroanterior, PaO₂ = partial pressure of oxygen

Table 2. Radiologic Findings in Four Patients with PI

<table>
<thead>
<tr>
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</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Pneumomediastinum</td>
<td>Pneumomediastinum</td>
<td>Asthma</td>
<td>N</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
</tr>
<tr>
<td>2</td>
<td>Lung mass PI</td>
<td>Lung cancer in left lung Pulmonary emphysema</td>
<td>N</td>
<td>Y</td>
<td>N*</td>
<td>Y*</td>
<td>Distal transverse colon to descending colon*</td>
</tr>
<tr>
<td>3</td>
<td>- PI</td>
<td>Stable TB Pulmonary emphysema</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>Cecum to hepatic flexure of colon</td>
</tr>
<tr>
<td>4</td>
<td>Atelectasis in BLLZ PI</td>
<td>Atelectasis in LLL Partial atelectasis in RLL</td>
<td>N</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>Cecum to hepatic flexure of colon</td>
</tr>
</tbody>
</table>

*Scanned abdominal images on chest CT.
PI = pneumatosis intestinalis, BLLZ = both lower lung zones, IVC = inferior vena cava, LLL = left lower lobe, RLL = right lower lobe, TB = tuberculosis
RADIOLOGIC FINDINGS

The radiologic findings are summarized in Table 2. PI was initially detected on chest radiographs of three of four patients who had PI and the diagnosis was subsequently confirmed by CT scans. In one patient (Patient 3) who did not have a chest radiograph when visiting the outpatient clinic, PI (cecum to proximal ascending colon) was detected for the first time on a follow-up abdomen CT. The amount of PI ranged mostly from moderate to extensive gas collections in all patients. The cecum and ascending colon were involved with PI in three patients. In one patient (Patient 2), the PI extended from the distal transverse colon down to the descending colon. None of the patients had PI involving the small bowel.

The underlying chest CT and chest X-ray showed findings compatible with asthma (n = 1) (Fig. 1A, B) and pulmonary emphysema (n = 2). Two patients showed abruptly developed complete airway stenosis due to an obstructive mass or aspirated material; initial detection of presumptive lung cancer (Patient 2) (Fig. 2A, B) and abruptly developed total atelectasis in left lower lobe with food impaction (Patient 4) (Fig. 3A). A patient with severe asthma exacerbations had a pneumomediastinum visible on a chest X-ray and a CT scan (Patient 1). Evidence of bronchiolitis with small airway disease was seen in Patient 3.

In three patients with substantial PI, the intramural gas had very similar appearances. The PI was dominated by a linear and tubular appearance that involved mainly the ascending colon, with adjacent focal or extensive mesenteric or peritoneal gas collection (Figs. 1C, D, 3B, C). The one patient with lung cancer (Patient 2) involving the distal transverse and the descending colon showed an intramural cystic and bubbly bowel-wall appearance (Fig. 2C). There was no case of bowel wall thickening or congestion of mesentery, which suggests the presence of bowel-wall ischemia. There was no evidence of intestinal obstruction or enterocolitis on the CT scan.

All three patients with a cecum and an ascending colon involving PI also had a pneumoperitoneum detected on a CT scan. The extraluminal gas ranged from focal gas collection to a large area extending to the retroperitoneum and small bowel mesentery without portal venous gas. Radiological resolution of PI occurred within 1 month in this study, without significant association between the extent of involvement and the duration.

Fig. 1. A 43-year-old woman with severe asthma.
A. Chest X-ray posterior-anterior views shows pneumomediastinum (arrows) with PI (arrowheads) on initial chest radiography.
B. These lesions are completely resolved after 1 month with conservative treatment.
C, D. Transverse and coronal abdomen CT scans (2.0-mm slice thickness) obtained with lung and wide window show linear patterns of PI involving from the cecum to proximal transverse colon.
PI = pneumatosis intestinalis
DISCUSSION

This is the first reported series, to our knowledge, of PI in adult patients with thoracic diseases without history of lung transplantations. There have been several case reports of PI with a thoracic disorder, including asthma, chronic bronchiectasis, GVHD after chemotherapy for lung cancer and a series of PI after lung transplantation (2, 5, 7-11). Among these, COPD have been a well-known pulmonary cause and all our cases have been consistent with obstructive lung disease such as asthma, emphysema, and airway obstruction. This study, which excludes cases with other possible abdominal or systemic causes, demonstrated relatively short-term and severe events of dyspnea as initial symptoms of abrupt airway obstruction or acute exacerbations of pre-existing obstructive lung disease in all cases.

In the reported cases of PI with a thoracic disorder, all the initial presentation is acute developed dyspnea consistent with this study, except for a case report of a 66-year-old women who subsequently developed PI due to chronic bronchiectasis and presented with chronic
Pneumatosis Intestinalis with Pulmonary Disorder

Choi et al. (7) described PI with asthma (similar to our patient 1) occurring in a 74-year-old woman who complained of dyspnea during management of acute asthma exacerbations; the patient had no abdominal complaint, and PI resolved spontaneously.

Although PI can occur in the form of a primary PI without an apparently associated disease, most of the PI is secondary, due to various causes, to the point that the definition of secondary PI is in fact inadequate. The disease is caused by a variety of acute and chronic diseases, including intestinal, pulmonary, systemic, iatrogenic, medication, and transplantation in which there is a life-threatening disease that requires urgent treatment. Therefore, radiologists and clinicians need to be familiar with various underlying diseases of PI and their radiologic findings. However, among these conditions, the incidence of PI due to pulmonary disease is low, and most of them are not life-threatening; extensive PI seen on CT scans in patients with respiratory symptoms can cause unnecessary evaluations, including invasive procedure due to unfamiliar imaging findings, especially to the chest-imaging subspecialist.

Efforts have been made to understand the pathogenesis of PI and the pathophysiological assessment of PI by other lesions, other than the gastrointestinal tract disorder, remains unclear. According to some theories, PI due to chest disease is presumed to be caused by the ruptured alveoli entering the vasculature and migrating caudally to the retroperitoneum and mesenteric vascular channels. This explanation is more convincing than transmural infiltration by subserosal location of air, however, has met with skepticism due to lack of pulmonary and mesenteric interstitial air of patients (5, 12, 13). It has been also proposed that the association between pulmonary disease and PI may simply be due to fluctuations in intra-abdominal pressure caused by pulmonary obstruction. Patients with chronic cough have increased intra-abdominal pressure (14).

Three of our cases showed a linear pattern of PI located in the ascending and the proximal transverse colon, and the one case had a different pattern and location of the air; bubbly and cystic PI in the distal transverse and the descending colon. All previously reported cases of asthma demonstrated a linear pattern of PI in the ascending and the proximal transverse colon which was consistent with our asthma case (Patient 1) (7, 15). The association between the air-collection pattern and distributed colon segment can be evaluated more in a further study with a larger number of cases. Nevertheless, the imaging appearance of the involved bowel segment divided by two vascular territories can support a presumed spectrum of PI; dissection of air along vascular channels to the mesentery of the bowel. CT is the best imaging modality for establishing the diagnosis of PI, as denoted by findings such as intramural gas parallel to the bowel wall.

Our study has some limitations. The major limitation is the sample size, which was small because of the low incidence of PI related with the thoracic disease. Second, the biopsy was not performed for each intestinal lesion. However, all the clinical symptoms, laboratory and radiologic findings did not suggest the possibility of malignant or ischemic bowel disease and completely improved without any treatment for these illnesses on follow-up radiologic assessments.

Patients in two cases of our study with airway obstruction (Patient 2 and 4) showed only lung lesion and focal PI without combined pneumoperitoneum at initial chest X-ray. And the major complaint in patients that presented dyspnea without related abdominal manifes-
tation in all our cases might cause a clinician to overlook or misdiagnose the PI. Radiologists and clinicians not only need to be aware of these imaging findings and but also correlate them with clinical and laboratory data, before assuming that the patient has ischemia or a malignant lesion of the colon and performing unnecessary further evaluation.

Our results have emphasized that PI detected in the abdominal field of chest CT or chest PA in a patient with severe dyspnea or a cough might not be managed as an emergent or life-threatening condition, since these lesions are almost benign PI and can be managed by conservative treatment. In conclusion, thoracic disorder with obstructive lung disease with abrupt onset may result in the development of benign PI. Such PI in thoracic disease patients has a similar linear and cystic appearance with ischemic bowel disease, but can nevertheless be managed by conservative treatment.

Conflicts of Interest
The authors have no potential conflicts of interest to disclose.

REFERENCES


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폐병변과 연관된 장벽 공기를
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목적 흉부 질환과 연관하여 장벽 공기증을 가지는 환자의 임상 특징 및 영상 소견을 검토하여 장벽 공기증의 가능한 원인을 알아보고자 한다.

대상과 방법 2005년부터 2017년까지, 장벽 공기증을 가진 62명의 환자 중 흉부 질환과 연관하여 발생한 4명(6%)의 증례가 확인되었다. 장벽 공기증 발생 시 임상 기록, 검사 소견 및 치료를 검토하였으며 2명의 흉부영상의학과 의사가 이들의 영상 검사를 검토하고 각 환자의 특정 소견을 기록했다.

결과 4명 환자의 원인이 되는 흉부 질환은 심한 천식, 폐기종 및 기도 폐색이었다. 장벽 공기증이 발생한 장관 범위 및 공기의 패턴을 포함하는 장벽 공기증의 영상 소견은 두 개의 장간막 혈관의 해당 영역별로 분리되어 나타나는 결과를 보였다. 우리의 증례 중 3명의 환자에서 흉부 질환과 연관하여 발생한 장벽 공기증의 발생 소견을 보였으며, 제 4의 증례는 흉부 질환과 연관하여 발생한 장벽 공기증의 발생 소견을 보였다. 결과적으로, 추적검사가 되지 않은 1명의 환자를 제외하고 나머지 3명의 환자는 모두 보존적인 치료로 1개월 내에 호전되었다.

결론 폐쇄성 폐질환을 동반하는 흉부 질환은 양성 장벽 공기증의 발생을 초래할 수 있다. 흉부 질환 환자에서에서는 이러한 장벽 공기증은 단순 흉부 촬영이나 컴퓨터단층촬영 검사에서 허혈성 장 질환과 같은 형태의 선형 및 낭종 형태의 장벽 공기증 모양을 보이지만 보존적 관리에 의하여 치료되어야 한다.

이화여자대학교 이대목동병원 ¹영상의학과, ²호흡기 및 중환자의학과