A Case Report of Spontaneous Pneumoperitoneum Related to Scleroderma without Evidence of Pneumatosis Cystoides Intestinalis

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We report a case of a 72-year-old male with a history of scleroderma who presented to the emergency room with an asymptomatic pneumoperitoneum after undergoing a CT thorax for evaluation of interstitial lung disease. Abdominal imaging was performed with oral contrast and did not reveal extravasation of the contrast into the peritoneum suggesting there was no perforated viscus. The patient was managed conservatively with serial abdominal exams, bowel rest, empiric antibiotics, supplemental oxygen, and total parenteral nutrition. The patient was subsequently diagnosed with a spontaneous pneumoperitoneum related to scleroderma without evidence of pneumatosis cystoides intestinalis. Over a course of 3 months the pneumoperitoneum resolved and the patient remained well.

INTRODUCTION

In 1915, Hugo Popper first described the technique of utilizing radiographic methods to detect pneumoperitoneum. Radiological techniques that are available for the diagnosis include ultrasound, plain X-ray and computed tomography (CT). Pneumoperitoneum appears as a radiolucency below the diaphragms on chest radiography or in a superiorly dependent location on an abdominal radiograph. Miller and Nelson described a process that can detect as little as one cubic centimeter of gas in the abdominal cavity. This includes placing the patient in the left lateral decubitus position for 10 to 20 minutes then exposing a left lateral decubitus film. The patient is then moved into an erect position where an anteroposterior chest film is taken as well as an erect abdominal film. The patient is next tilted to a horizontal position where a supine abdominal film is taken. If there is concern that the pneumoperitoneum is from an area above the diaphragm then abdominal x-rays should demonstrate air in the paraspinal area of the retroperitoneum. Computed tomography is more sensitive for detecting intraperitoneal air than upright chest radiographs. Visceral perforation may not always be detected by contrast examination with computed tomography. The causes of most pneumoperitoneum have been linked to intra-abdominal disease. Approximately 90% of the cases of pneumoperitoneum are due to visceral perforation, usually associated with a gastric or duodenal ulcer. Pneumoperitoneum due to perforation is a surgical emergency. Generally these patients will present with fever, leukocytosis and peritoneal signs. There are, however, cases of an occult leakage due to visceral perforation where the patient may only demonstrate mild abdominal pain without fever, leukocytosis, or peritoneal signs. These occult leakages may be due to sigmoid diverticulitis, fecal impaction, or peptic ulcer perforations. It has been noted that patients with peptic ulcer perforations may present less symptomatic than other sites of perforation or even asymptomatic with subsequent healing without intervention.

The remaining 10% of cases of pneumoperitoneum not associated with visceral perforation fall into the classification of spontaneous pneumoperitoneum. The term spontaneous pneumoperitoneum has been used synonymously with aseptic spontaneous, idiopathic, spontaneous asymptomatic and nonsurgical pneumoperitoneum. Causes of spontaneous pneumoperitoneum can be divided into abdominal, thoracic, gynecologic, pseudopneumoperitoneum, miscellaneous and idiopathic etiologies. Abdominal causes of spontaneous pneumoperitoneum...
are the most common. The etiologies include abdominal surgery, peritoneal dialysis, and most endoscopic gastrointestinal procedures. By far the most common abdominal cause is due to open abdominal surgery. It generally resolves within 2 days in two thirds of the cases and within 5 days in 97% cases when followed by abdominal radiographs. If followed by CT, which is more sensitive, intraperitoneal air at 3 days will be detected in 85% of the cases and 50% of cases at day six. Older, leaner adults more commonly retain a greater volume of free air over a longer period of time.

Pneumatosis cystoides intestinalis, also known as cystic lymphomatosis or enteromesenteric emphysema, is the most common abdominal cause of nonsurgical pneumoperitoneum. It was first described in 1730 by DuVernois during a cadaver dissection. The development of pneumatosis cystoides intestinalis has been associated with idiopathic development, scleroderma, bone marrow transplant, AIDS, diverticular disease, small bowel resection, intestinal pseudo obstruction, dermatomyositis, nontropical sprue, jejunal-ileal bypass, gastric outlet obstruction, sclerotherapy, and heart transplantation. It is characterized by intramural gas filled cysts in any portion of the gastrointestinal tract; however, it is most commonly found in the terminal ileum. Even though the small bowel is the most common site for these cysts, they can be found anywhere in the gastrointestinal tract including the stomach (gastric pneumatosis cystoides) and the colon (pneumatosis coli). It is the rupture of these cysts that lead to pneumoperitoneum. The cysts are constantly filling, rupturing, sealing, and then filling again providing a constant spontaneous pneumoperitoneum. This condition generally resolves spontaneously, but has been known to reoccur. There have been five possible explanations for the development of these cysts. The first two theories suggest that an increase in acidic byproducts result in increased levels of lactic acid leading to a decrease in carbon dioxide and oxygen resorption leading to the cyst formation. The mechanical theory states that gas is forced into the bowel by obstructions, anastomotic sites, the pulmonary system, trauma, mucosal breaks, increased pressure, or increased peristalsis. The bacterial theory suggests that bacteria gain entrance to the bowel wall and produce the cysts. The last theory is mentioned due to historical interest, which is a neoplastic theory described by Bangs.

Outside of the abdomen, other locations that may cause pneumoperitoneum include thoracic, gynecologic, and other locations. Thoracic causes include mechanical ventilation, cardiopulmonary resuscitation, and pneumothorax. Thoracic causes are the second most common cause of spontaneous pneumoperitoneum and barotraumas represents the most frequent of the intrathoracic causes. Gyneologic etiologies are rare compared to the other locations. These generally occur due to air traveling through the genital tract into the peritoneum via the uterus and fallopian tubes. Examples include vaginal insufflation by orogenital sex, vaginal douching, postpartum knee-chest exercises, use of bulb aspiration in pelvic exam, tubal insufflation in hysterosalpingogram, pelvic inflammatory disease, and coitus. In the differential of pneumoperitoneum is pseudoneuropneumoperitoneum. This was first described in the 1930s and represents a simulated appearance of free intraperitoneal air. This is frequently due to adventitial air shadows, over distension of hollow viscer, the basal lung appearing to lie in the diaphragm, gas trapped...
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in wounds, basal pulmonary atelectasis resembling subphrenic air, subdiaphragmatic extraperitoneal fat, and positioning of colonic hepatic flexure between the right lobe of the liver and diaphragm, also known as Chilaiditi sign.6 Failure of the air to shift during different positioning or to localize to the most superior aspect of the radiographic imaging should lead to the suspicion of pseudopneumoperitoneum. Other causes of pneumoperitoneum include scleroderma without pneumatosis cystoides intestinalis, idiopathic, cocaine use, diving and decompression, and dental extraction.

There are many reports of pneumoperitoneum in patients who have mild abdominal pain but no leukocytosis, fever, or peritoneal signs that were treated successfully with conservative management.2 When spontaneous pneumoperitoneum is suspected, emergency surgery is not required. A difficult situation arises when the physical examination is unreliable such as in an unresponsive patient on a ventilator or a patient with a depressed immune system causing a delay in the signs and symptoms of peritonitis. In these cases, a diagnostic peritoneal lavage may aid in the decision where to perform surgery or to treat conservatively.8 Methylene blue may be instilled via a nasogastric tube to help increase the accuracy of detecting an upper gastrointestinal perforation.8 If the lavage is negative and the patient is unremarkable, then continued observation is warranted. Also aiding in the decision for surgery would be to instill water-soluble contrast through a nasogastric tube with a subsequent abdominal film.8

CASE REPORT

Here we report a 72 year-old white male who was having thoracic CT surveillance without contrast for evaluation of interstitial lung disease due to his history of scleroderma. Thoracic CT incidentally revealed free air in his abdomen and he was subsequently instructed to seek further evaluation in the emergency department. The patient’s past medical history included rheumatoid arthritis, scleroderma and a previous gastrointestinal bleed with an unclear etiology five years prior to admission. The patient did not have any surgeries or procedures in the last six months.

Upon admission to the emergency department, his blood pressure was 153/76 mm Hg, heart rate 78 bpm, temperature 97.7 °F and an oxygen saturation of 99% on room air. The patient’s physical exam was benign. The initial complete blood count (CBC) and complete metabolic panel (CMP) were within normal limits. While in emergency department, the patient had a chest X-ray demonstrating free air within the abdomen. A CT of the abdomen without contrast confirmed a pneumoperitoneum (Figure 1). The patient was evaluated by both gastroenterology and surgery. Conservative measures of bowel rest along with serial abdominal and x-ray examinations were undertaken which remained stable throughout the admission. Abdominal imaging did not show evidence of pneumatosis cystoides intestinalis. An upper gastrointestinal series was completed and did not demonstrate extravasation of contrast, shown in Figure 2. The patient’s diet was advanced over six days; he tolerated this well and was discharged.

Approximately six weeks later, the patient had a follow up abdominal CT with oral contrast. It demonstrated a mild increase in the pneumoperitoneum along with right pleural calcification, which was noted on a previous thoracic CT. He was again instructed to return to the emergency department. Upon arrival to the emergency department his physical exam, vital signs, CBC and CMP were all normal. The patient remained asymptomatic. Another upper gastrointestinal series was done, again demonstrating no obvious extravasation of contrast. He was treated with bowel rest, supplemental oxygen and placed on total parenteral nutrition for several weeks. Cefazolin and metronidazole were started as empiric treatment for a bacterial source of the gas and antibiotics were continued for one week. An abdominal x-ray one week later demonstrated a resolving pneumoperitoneum. A follow up abdominal CT scan two months after being discharged from his second hospital admission demonstrated no free air in the abdomen.

DISCUSSION

Pneumoperitoneum generally occurs in the presence of a visceral perforation with peritoneal signs; however, in some cases where the patient is asymptomatic, the physician should consider causes of spontaneous pneumoperitoneum. In the case above, we reported a spontaneous pneumoperitoneum thought to be due to scleroderma without relation to pneumatosis cystoides intestinalis. After a thorough search of the medical literature only five published case reports are available for review, making this an extremely rare entity.

Once our patient was in the emergency department, a CT of the abdomen was completed which did not demonstrate any evidence of pneumatosis cystoides intestinalis. When attempting to identify pneumatosis...
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intestinalis with computed tomography, it is best to view the bowel wall in the lung window for evidence of air within the gut wall. However, there was always a chance that the pneumatosis cystoides intestinalis was below the resolution of the computed tomography in our patient, although this is unlikely. It is also possible that there was a microperforation in the stomach that had subsequently healed. As noted above, patients can be minimally symptomatic, if not asymptomatic, with a microperforation of the stomach. It is interesting to note that the patient had a gastrointestinal bleed in the preceding five years with an unclear etiology. The patient had a colonoscopy in the past, but was never evaluated with an esophagogastroduodenoscopy, leaving room for gastrointestinal pathology. The patient did have two upper gastrointestinal series and a CT with oral contrast that did not demonstrate extravasation of contrast. However, that does not rule out a perforation but does make it less likely. Since our patient had a history of scleroderma and demonstrated no obvious source of visceral perforation, was asymptomatic and radiographic imagining did not show evidence of pneumatosis cystoides intestinalis, this led us to believe that the pneumoperitoneum was related to scleroderma.

As stated by Rowe et al., determining which patient with a pneumoperitoneum needs to undergo exploratory laparotomy versus those that can be managed conservatively can be challenging. However, Rowe et al. suggests that the laparotomy rate can be decreased to near zero if a thorough history and physical examination is performed and does not suggest perforation. Since our patient was stable a conservative approach was taken with antibiotics, increased fractional inspired oxygen content to 27% and total parenteral nutrition and he responded well. In the other five known cases, one case report performed a laparotomy, another case report performed a peritoneal lavage, and in the remaining case reports, conservative treatment without invasive intervention was the preferred approach. The conservative approach, rather than an unnecessary exploratory laparotomy, saved our patient significant morbidity and he responded well.

Our patient’s pneumoperitoneum resolved in three months. The other cases reported resolution ranging from six months to two years. This is in drastic contrast to those patients who undergo abdominal surgery or have endoscopic procedures, which generally have resolution of the pneumoperitoneum in five days in approximately 97% of cases followed by abdominal radiographs.

In patients with scleroderma associated with spontaneous pneumoperitoneum without evidence of pneumatosis cystoides intestinalis, the pathophysiology of the pneumoperitoneum is still unclear. It could be that pneumatosis cystoides intestinalis in many of these cases are at an undetectable level from radiographic imaging and could be causing a pneumoperitoneum. The cysts have been postulated to form in scleroderma due to gut hypomotility with subsequent bacterial overgrowth. It was for that reason that empiric antibiotics were used in the conservative approach in our patient to reduced bacterial overgrowth and possible cyst formation. It has also been postulated that there are recurrent microperforations in colonic diverticula causing a pneumoperitoneum in scleroderma without evidence to pneumatosis cystoides intestinalis. The microperforations may arise due to intestinal ischemia due to the vasculitic nature of scleroderma. It may also be related to the atrophy of the muscularis propria and collagen tissue replacing it, which occurs throughout the gastrointestinal tract in patients with systemic sclerosis.

CONCLUSION

Spontaneous pneumoperitoneum associated with scleroderma without evidence of pneumatosis cystoides intestinalis is an extremely rare presentation. Surgical intervention is not generally needed unless the history and physical suggests perforation. Patients can be treated conservatively with bowel rest and total parenteral nutrition. Patients should also be educated about the prolonged time of resolution of the pneumoperitoneum as well as the likelihood of the recurrence.

References