A 63-year-old woman with asthma presented with a 1-week history of increasing dyspnea, wheezing, and a productive cough. The patient stated that her grandchild was sick at home with otitis media and sore throat. The patient had a 20-pack-year history of cigarette smoking but had stopped smoking 10 years ago. Her history was significant for systemic lupus erythematosus, diagnosed 10 years ago and controlled with hydroxychloroquine and prednisone (10 mg/d). Twenty-five years ago, the patient had tuberculosis that was treated with isoniazid and a right lower lobe resection. She had a mastectomy and hysterectomy 20 years ago for a benign tumor.

Physical Examination

Vital signs are as follows: temperature, 36.3°C (97.4°F); pulse, 98 beats/min; respiratory rate, 28 breaths/min; BP, 130/70 mm Hg; O₂ saturation, 96% on 2 L/min O₂; and peak flow, 210 L/min increased to 250 L/min after β₂-agonist.

The patient was in no apparent distress. Examination revealed the following: oropharynx, moist mucosa and no lesions; chest, bilateral loud high pitched wheezing diffusely, with prolonged expiratory phase; cardiac, S₁, S₂, diminished, no murmur, regular rate and rhythm, and tachycardia; abdomen, soft, non-tender, normal bowel sounds, and no hepatosplenomegaly; and extremities, no clubbing, cyanosis, or edema. The neurologic examination was unremarkable.

Laboratory Findings

Laboratory findings are as follows: WBC count, 9.8 × 10³/µL with 94% polymorphonuclear neutrophils and 2% band neutrophils; hematocrit, 45%; and platelet count, 276 × 10³/µL. Calcium measurement, liver function test results, and electrolyte levels were normal. No infiltrates were evident on chest radiograph, and ECG revealed no acute changes.

Clinical Course

The patient was admitted to the hospital with a diagnosis of asthma exacerbation. She improved with IV steroids, bronchodilators, and antibiotics. Prednisone was tapered to 60 mg/d. Ten days after admission, she developed neck swelling and hoarseness with pronounced subcutaneous emphysema. Her chest was clear on examination, and the rest of her physical examination was otherwise unremarkable. Her chest radiograph revealed subcutaneous emphysema, pneumomediastinum, and no pneumothorax. On the 11th hospital day, the patient complained of abdominal pain. Examination showed diffuse abdominal tenderness; however, the abdomen was soft with no signs of peritoneal irritation. The patient’s WBC count increased to 23 × 10³/µL, with 4% band neutrophils. CT of the abdomen with iodinated contrast medium (Gastrografin; Schering Diagnostics; Berlin, Germany) was nonrevealing. No intraperitoneal air was identified. A surgeon was consulted on the 12th hospital day; the abdominal pain was attributed to pneumatosis coli. The patient continued to complain of increasing abdominal pain, although her abdominal examination remained unimpressive. The next day, the WBC count increased to 27 × 10³/µL, with 8% band neutrophils. On the 14th hospital day, a second abdominal CT with gastrografin was nonrevealing. A chest radiograph and CT image at the level of mediastinum are shown below.

What would be your next step in managing this patient?

What is the most likely source of mediastinal air?

What is the diagnosis?
Figure 1. Chest radiograph on hospital day 10 showing pneumomediastinum and subcutaneous emphysema.

Figure 2. Chest CT showing air in the mediastinum, most noticeable around the descending aorta.
Answers: Exploratory laparotomy; perforated viscus.

Diagnosis: Sigmoid perforation with retroperitoneal abscess and retroperitoneal air tracking into mediastinum.

DISCUSSION

Pneumomediastinum, or air in the mediastinum, was first described as a complication of trauma in 1819 by Laennec. Spontaneous pneumomediastinum as an entity was initially introduced into the medical literature in 1939 by Hamman, from which “Hamman sign” (air crepitus heard on auscultation with each heart beat) is derived. Pneumomediastinum is usually regarded as a benign, self-limited process that does not require medical intervention, although the etiology must be assessed to exclude rare life-threatening causes. Typical causes of pneumomediastinum are as follows:

1. Secondary to increased intrathoracic pressure, i.e., Valsalva maneuver, strenuous exercise, weight lifting, vaginal delivery, and vomiting.
2. Sniffing cocaine or other drugs.
4. Status asthmaticus.

Unusual causes include arthroscopy, dental extraction, and adenotonsillectomy, scuba diving, trombone playing, and performing a maximal expiratory pressure maneuver.

Pneumomediastinum of a GI origin has been described in the medical literature. It can occur after GI instrumentation, endoscopy, endoscopic retrograde cholangiopancreatography, colonoscopy, or laparoscopic surgery. It may occur with or without evidence of perforation. In most cases, the condition is self-limited and resolves without surgical intervention, although a possible life-threatening etiology should be considered. A known life-threatening cause of pneumomediastinum is spontaneous esophageal rupture (Boerhaave’s syndrome). An esophagogram should, therefore, be included in the routine workup of pneumomediastinum and subcutaneous emphysema in the appropriate clinical setting.

There are four cases of spontaneous bowel perforation presenting as subcutaneous emphysema and pneumomediastinum reported in the literature; each case reported a sigmoid diverticulum perforation into the retroperitoneal space with abscess formation. Each case had a dominant septic picture with a paucity of localizing signs. Abdominal symptoms, if present, were vague without evidence of acute abdomen. Each patient’s septic state did not respond to antibiotic therapy. The diagnostic workup, including imaging studies, was negative for identifying the source of sepsis. In each of these cases, the diagnosis was delayed 8 to 12 days, and exploratory laparotomy was prompted solely by clinical suspicion. Each patient’s septic state did resolve after a Hartmann’s procedure (segmental resection of the sigmoid colon and end-colostomy). One of the patients was receiving chronic oral steroids for temporal arteritis. It has been suggested that when there is a septic state without obvious source that is not responding to broad-spectrum antibiotics, an exploratory laparotomy is justified.

In contrast to the above cases of retroperitoneal perforation, a fifth case described intraperitoneal perforation with acute appendicitis presenting with subcutaneous air and pneumomediastinum. This patient had overt symptoms of acute abdomen, unlike the four previously mentioned cases.

Characteristically, the diagnosis of retroperitoneal perforation is delayed due to the paucity of symptoms or absence of a clinical picture of acute abdomen. Intraperitoneal perforation is diagnosed on average within 1 week of the onset of symptoms, while the diagnosis of retroperitoneal perforation is reported to take as long as 1.7 months after the onset of symptoms.

The location of subcutaneous emphysema of abdominal GI origin depends predominantly on the anatomic location of the perforation. Perforation of the small intestine, appendix, or right colon causes emphysema of the anterior abdominal wall. Perforation of the anorectal area and the left colon is associated with air in the scrotum and perineum.

The symptoms of pneumomediastinum include chest pain or pressure, dysphagia, and hoarseness. Associated abdominal pain should suggest an abdominal cause for the pneumomediastinum. Although rare, bowel perforation should be considered in the differential diagnosis of a patient presenting with pneumomediastinum and/or subcutaneous emphysema because it could be potentially lethal if untreated.

The present patient had pneumomediastinum and extensive subcutaneous emphysema involving the neck and chest that was initially attributed to a peribronchial alveoli rupture from an asthma exacerbation. The abdominal pain in this patient was initially attributed to pneumatisis coli, and the pneumoperitoneum was thought to be caused by pneumomediastinum with air tracking down along the vascular pathways.

However, the patient had a sigmoid retroperitoneal perforation with extensive retroperitoneal abscess with pus, air, and stool around the sigmoid colon, extending to the level of the aorta. Air tracked up the retroperitoneum along the opening for the great vessels and esophagus in the diaphragm and
into the posterior mediastinum and the subcutaneous tissues of the chest and neck. The exact point of the perforation was not apparent, due to dense inflammation. Surgical pathology revealed diverticulitis and peridiverticulitis showing extensive acute and chronic inflammation extending into the pericolic fat with abscess formation.

Because the patient was receiving chronic steroids and because of the retroperitoneal location of the perforation, the clinical picture was vague and not suggestive of an acute abdomen. The diagnostic studies, including an abdominal CT scan with oral Gastrografin contrast, were negative for the presence of intraperitoneal air or other evidence of perforation. The laparotomy was prompted by a high clinical suspicion for acute abdominal process. The pneumomediastinum and subcutaneous emphysema were the unusual initial presentations of a perforated bowel.

The patient underwent a Hartmann’s procedure. On the 17th postoperative day, she was transferred to a rehabilitation facility and eventually discharged to home.

**Pearls**

1. **Pneumomediastinum is generally regarded as a benign self-limited process that does not require medical intervention.**

2. **The symptoms of pneumomediastinum include chest pain or pressure, dysphagia, and hoarseness. When associated with abdominal pain, a perforated abdominal viscus should be suspected as a cause.**

3. **A life-threatening cause of pneumomediastinum is spontaneous esophageal rupture (Boerhaave’s syndrome).**

4. **The diagnosis of retroperitoneal perforation is often delayed due to minimal symptoms or absence of the clinical picture of an acute abdomen.**

**Suggested Readings**

Ciaccia D, Branch MS, Baillie J. Pneumomediastinum after endoscopic sphincterotomy. Am J Gastroenterol 1995; 90:475–477


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