

Spontaneous Pneumomediastinum Due to Achalasia: An Unusual but Benign Cause

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ABSTRACT

Pneumomediastinum is usually first identified radiographically in the emergency department. Distinguishing benign from more ominous causes, such as esophageal rupture, is imperative, particularly in the setting of associated esophageal disease. We describe a case, with correlative imaging, of spontaneous pneumomediastinum as the initial presentation of achalasia. A general discussion of spontaneous pneumomediastinum is also provided, including the pathophysiology, precipitating and predisposing factors, clinical manifestations, role of radiology in the diagnosis as well as the radiographic signs.

CASE REPORT

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A 21-year-old previously healthy Caucasian female presented to our emergency department with chest pain and vomiting. She described a chronic cough and progressive gagging, which had recently been treated presumptively with oral antibiotics as an upper respiratory tract infection. She additionally reported a 20 pound weight loss over several weeks, which she attributed to food aversion from progressive solid food dysphagia. She denied fever or chills, and reported no foreign travel, nor personal or family history of esophageal disease.

On examination, she appeared ill from dehydration, and biochemistry revealed pre-renal azotemia, with creatinine of 2.3 mg/dL (normal 0.5 - 1.4). She was mildly anemic with hemoglobin of 11.2 g/dL (normal 12.0 - 16.0) with a moderate leukocytosis of 17.1 K/uL (normal 5.0 - 10.0).

Chest radiography (figure 1) demonstrated pneumomediastinum and subcutaneous emphysema, without pneumothorax or evidence of mediastinal fluid. Chest CT (figures 2 and 3) confirmed extension of pneumomediastinum into the neck, axillae, and epidural space, along with marked esophageal dilatation, which terminated abruptly at the level of the diaphragm. No associated mediastinal or pleural fluid was present.

Subsequent water soluble esophagography (figure 4) revealed typical findings of achalasia, with marked esophageal dilatation and a beak-like transition at the gastro-esophageal junction, without evidence of contrast extravasation.

After several days of aggressive intravenous hydration and prophylactic antibiotics, endoscopic distal esophageal dilatation was performed. Her pain, swallowing difficulty, abnormal laboratory parameters, and pneumomediastinum all completely resolved. She was discharged home with appropriate follow-up arrangements.

DISCUSSION

Spontaneous pneumomediastinum manifests as mediastinal air in the absence of trauma or injury. Usually first identified with chest radiography or thoracic computed tomography (CT), spontaneous pneumomediastinum has been described in a wide variety of clinical scenarios. It was first described by Louis Hamman in 1939 [1].

The most commonly reported clinical manifestation is chest pain followed by dyspnea, cough, and subcutaneous emphysema or neck swelling [2]. Precipitating factors are many (table 1), and include violent coughing, sneezing, vomiting, asthma attacks, inhalational drug use (particularly marijuana, cocaine, and methamphetamine), choking, heavy exertion and even defecation, sex, and childbirth [2, 3, 4]. Predisposing factors (table 1) include smoking and underlying lung disease [2, 5, 6].

To our knowledge, achalasia has been described only once as an etiology of spontaneous pneumomediastinum [7], but that report included no correlative imaging. The pathophysiology, as in other cases of spontaneous pneumomediastinum, is presumably alveolar rupture from repeated retching and vomiting. Such migration of air from the alveoli back along the bronchovascular bundle was first described by Macklin and Macklin [8] by overinflating a cat's lung to induce alveolar rupture, and subsequently injecting hot gelatin containing minute carmine granules to permit subsequent tissue evaluation.

Sources of pneumomediastinum are numerous and have been divided into intrathoracic and extrathoracic causes [9]. Intrathoracic etiologies include air trapping in the setting of asthma, straining against a closed glottis during vomiting or weight-lifting, esophageal perforation in Boerhaave's syndrome, infection from gas-producing microorganisms, and airway and alveolar damage from blunt chest trauma [3, 9]. Extrathoracic etiologies include dissection of air from the head and neck (such as from sinus fracture or dental extraction), or from intra- or retroperitoneal sources (such as from intestinal perforation) [9]. Spontaneous pneumomediastinum presents most in healthy young male adults. While most authors have suggested a male predominance [3], this has been debated [2].

Pneumomediastinum has classically been described frequently in the setting of esophageal rupture [10], referred to as Boerhaave's syndrome, and is particularly important because of its high associated morbidity and mortality [10]. Unlike most causes of spontaneous (and typically clinically benign) pneumomediastinum, this more ominous etiology, as with achalasia, may occur in the setting of underlying esophageal disease. Rapid differentiation of the two is thus imperative. Contrast esophagography should provide definitive characterization, since spontaneous pneumomediastinum from achalasia is associated with classic esophagram findings. When extravasation, however, is present, a diagnosis of esophageal rupture can be established, which usually results in emergency thoracotomy [10]. In contrast, achalasia is usually treated less urgently, with endoscopic balloon dilatation [11].

Several previously reported radiographic signs of pneumomediastinum are summarized in table 2 [9]. While some studies have indicated that more than 30% of cases may be occult on plain radiography [2, 12], others have suggested a sensitivity of greater than 90% [5]. Lateral radiography can be particularly helpful, since up to 50% of all cases are not identifiable with a posteroanterior view alone [3]. Given these limitations, chest CT is considered the gold standard for the diagnosis of pneumomediastinum [2].

To facilitate the early diagnosis of esophageal perforation, it is recommended that contrast esophagography be performed in cases of pneumomediastinum associated with emesis, dysphagia, gastrointestinal disease, trauma, fever, leukocytosis, pleural effusion, and pneumoperitoneum [11]. In other cases, a contrast swallowing study is of little, if any, benefit, particularly when patients are not ill-appearing [11]. It is worth mentioning, however, that false negative results in contrast esophagography have been reported in some patients with esophageal perforation [10], presumably because extravasation is either intermittent or radiographically obscured. When clinical suspicion is sufficiently high, CT with oral contrast may be beneficial.

Given the importance of imaging in the diagnosis and evaluation of pneumomediastinum, radiologists play extremely instrumental roles in the emergency setting. A thorough understanding of various modalities and their roles will facilitate the diagnosis of spontaneous pneumomediastinum, and differentiate it from entities which require more aggressive intervention.

TEACHING POINT

Spontaneous pneumomediastinum typically presents with chest discomfort in young healthy adults as a benign self-limiting condition and is generally diagnosed radiographically. When pneumomediastinum is associated with esophageal disease, contrast esophagography may be helpful in distinguishing the benign variety associated with achalasia from esophageal perforation.

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FIGURES

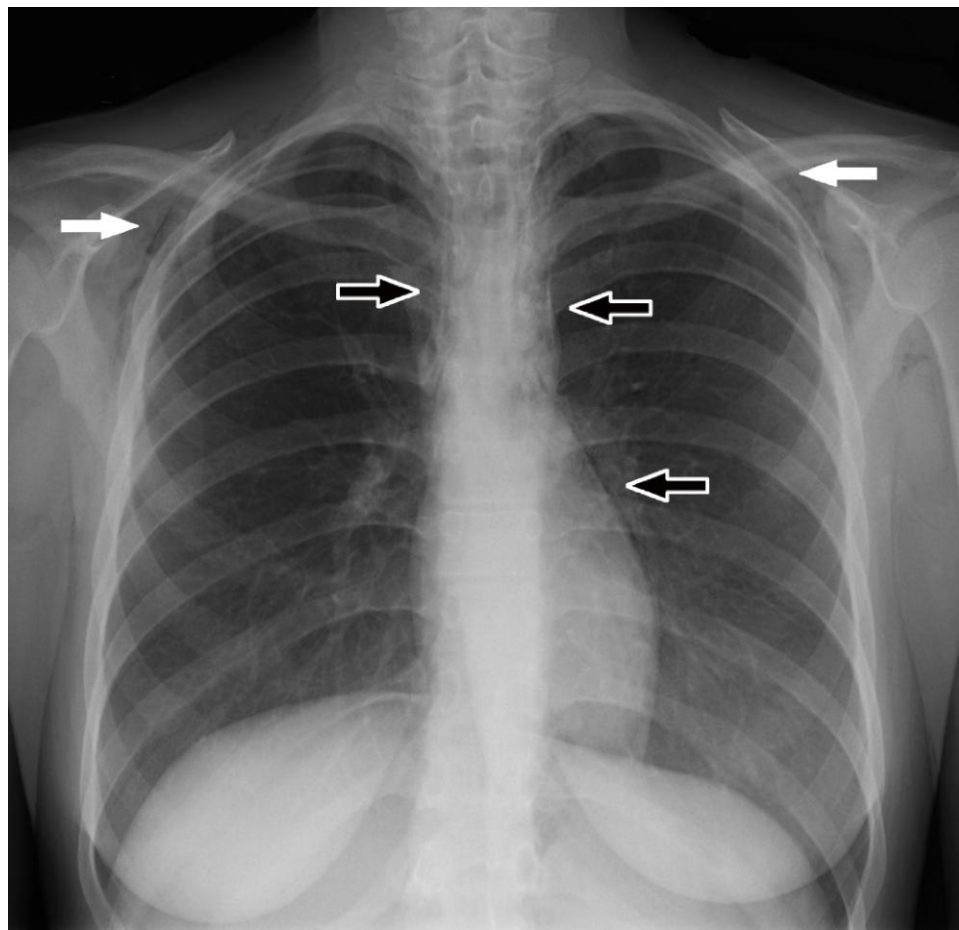


Figure 1: 21-year-old female with spontaneous pneumomediastinum. Frontal chest radiography at the time of emergency department presentation demonstrates pneumomediastinum (open arrows) and associated subcutaneous emphysema (white arrows).

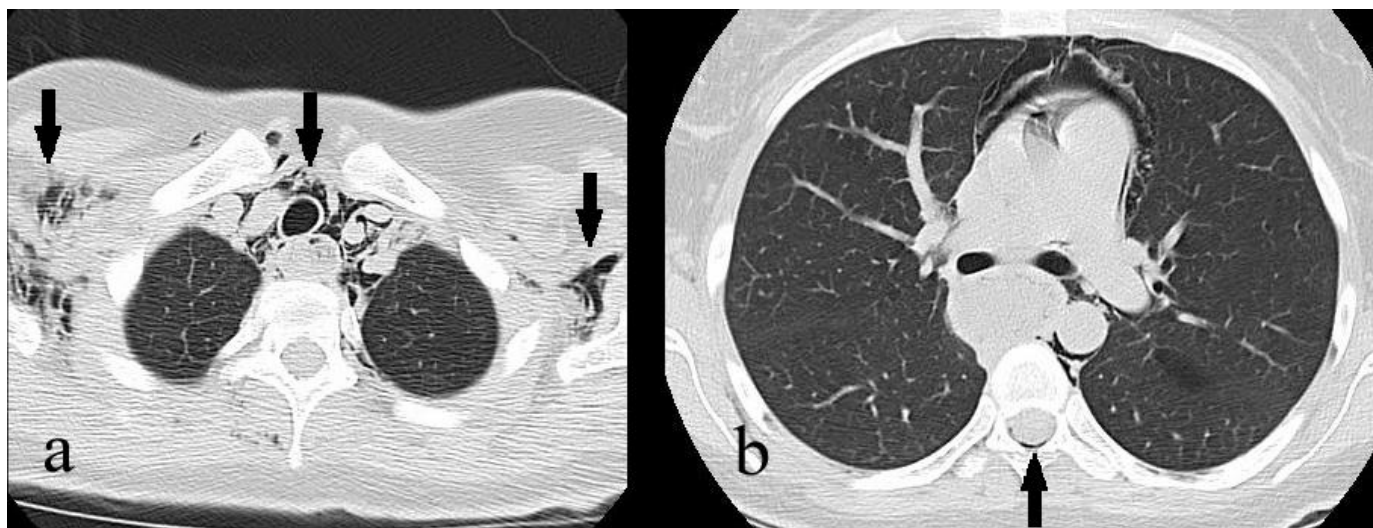


Figure 2: 21-year-old female with spontaneous pneumomediastinum and achalasia. Axial CT with lung windows demonstrate extensive pneumomediastinum, with extension into soft tissues of the neck and axillae (a) and the epidural space (b). (Protocol: standard algorithm, 300 mAs maximum (automated), 100 kV, 2.5 mm slice thickness, axial reconstruction with lung windows, no contrast material)



Figure 3: 21-year-old female with achalasia. Coronal CT imaging with soft tissue window demonstrates marked diffuse esophageal dilatation (open arrows), with associated pneumomediastinum (closed arrow). (Protocol: standard algorithm, 300 mAs maximum (automated), 100 kV, 2.5 mm slice thickness, coronal reconstruction with mediastinal windows, no contrast material)

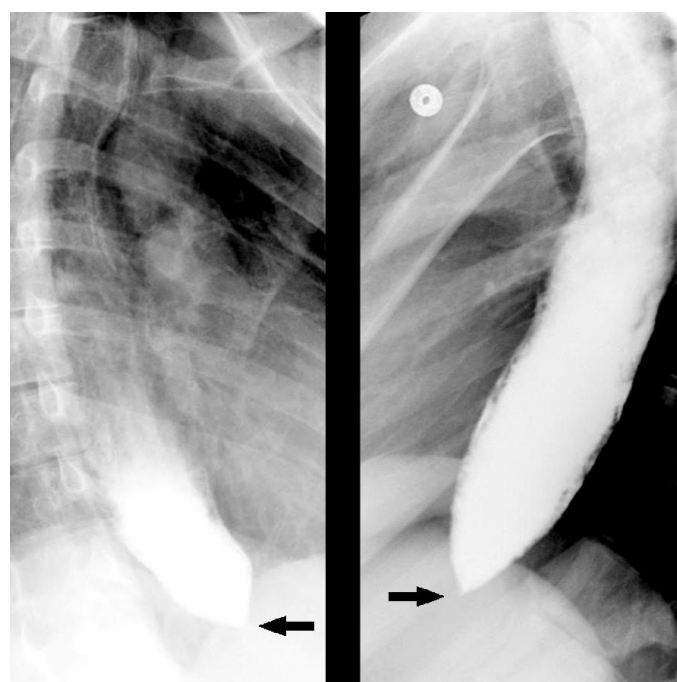


Figure 4: 21-year-old female with achalasia. Bilateral oblique images from a water soluble esophagram reveal marked esophageal dilatation, with abrupt smooth narrowing at the expected level of the gastro-esophageal junction (arrows), characteristic of achalasia. No contrast extravasation is present. Pneumomediastinum is difficult to discern on these fluoroscopic spot images.

Risk Factors	
Precipitating Factors	Emesis, asthma exacerbation episode, sneezing, cough, physical activity, choking episode, defecation, childbirth, bronchospasm, inflating balloons, playing wind instruments, mountaineering, spirometry
Predisposing Factors	Interstitial lung disease, asthma, bronchiolitis obliterans syndrome, bronchiectasis, chronic obstructive pulmonary disease, bronchogenic or metastatic disease, cystic lung lesions, idiopathic pulmonary fibrosis, smoking history, upper respiratory tract infection, illicit drug use, inhalation of irritants, diabetic ketoacidosis

Table 1. Precipitating and predisposing factors for spontaneous pneumomediastinum, as identified at literature review. Precipitating factors refer to recent activities or events leading to the development of pneumomediastinum [3, 5, 13]. Predisposing factors refer to pre-existing conditions which make patients more prone to pneumomediastinum [3, 5, 14].

Radiographic Signs and Location of Air	
Subcutaneous emphysema	Beneath the skin and subcutaneous tissue
Thymic sail sign	Around the thymus gland, outlining its borders
Pneumoprecardium	Anterior to the pericardium
Ring around the artery sign	Around the pulmonary artery or either of its main branches
Tubular artery sign	Medial to a major branch of aorta outlined laterally by lung
Double bronchial wall sign	Next to a major bronchus outlining the walls
Extrapleural sign	Between parietal pleura and the diaphragm
Continuous diaphragm sign	Trapped posterior to the pericardium (seen in the anteroposterior view as a continuous collection of air)

Table 2. Radiographic signs of pneumomediastinum [adapted from Zylak], identifiable at chest radiography and CT.

Spontaneous Pneumomediastinum	
Etiology	Numerous, detailed in table 1
Incidence	1 in 30,000 emergency department visits
Gender ratio	Male predominance with ratios of slightly above 1 to more than 3
Age predilection	Younger population with mean age of 17-30, all ages possible
Risk factors	Numerous, detailed in table 1
Treatment	Conservative medical management (bed rest, oxygen therapy, reassurance, and analgesics)
Prognosis	Good
Findings on imaging	Discussed in table 2

Table 3. Summary table of spontaneous Pneumomediastinum [5, 15].

Achalasia	
Etiology	Failure of lower esophageal sphincter to relax
Incidence	1 per 100,000 per year
Gender ratio	No sex predilection
Age predilection	Increased incidence between ages 20-40, any age possible
Risk factors	Chagas' disease, malignancy, amyloidosis, sarcoidosis, neurofibromatosis, eosinophilic gastroenteritis
Treatment	Heller myotomy, botulinum toxin injection, pneumatic dilatation, calcium channel blockers or nitrates
Prognosis	Generally good, however repeated treatments are necessary. Long-standing achalasia likely has increased risk of esophageal cancer
Findings on imaging	Dilated distal esophagus with "bird's beak" or "rat's tail" appearance on esophagography, distal esophageal dilatation on CT

Table 4. Summary table of Achalasia [16].

Differential Diagnosis	
Intrathoracic causes	Spontaneous pneumomediastinum Esophageal perforation due to ingested material Boerhaave's syndrome Blunt chest trauma with injury to airways Tracheal perforation Infection from gas-producing bacteria
Extrathoracic causes	Intestinal or gastric perforation Sinus fracture Dental extraction

Table 5. Table of differential diagnosis of spontaneous pneumomediastinum.

ABBREVIATIONS

CT = Computed Tomography

KEYWORDS

Spontaneous Pneumomediastinum; Boerhaave's Syndrome; Achalasia; Pneumomediastinum

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