Review Article

A clinical review of spontaneous pneumomediastinum

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Abstract: Primary spontaneous pneumomediastinum is a rare, often benign and self-limited condition defined by air within the mediastinum. However, correctly distinguishing primary spontaneous pneumomediastinum from secondary causes, especially esophageal perforation, remains a diagnostic challenge. There is significant debate regarding the balance of completing a thorough but not overly invasive and costly diagnostic workup. This clinical review aims to gather the limited data regarding spontaneous pneumomediastinum management from case series and retrospective cohort studies, and presents an evaluation algorithm and treatment plan stratified by clinical history. Understanding specifically if the patient presents with coughing versus forceful vomiting is critical to help elucidate the etiology and guide management of pneumomediastinum. Patients who present with forceful vomiting or retching should be considered with higher degree of suspicion for secondary causes of pneumomediastinum, specifically esophageal perforation. However, especially in children, aggressive diagnostic workup is not warranted in every case. After ruling out other etiologies of pneumomediastinum, spontaneous pneumomediastinum can be commonly treated with symptomatic management without the aggressive use of antibiotics or diet restriction. Hospital length of stay may also be minimized on a case-by-case basis. Overall, recurrence of spontaneous pneumomediastinum is rare and outpatient follow up may be safely limited to those at highest risk of recurrence.

Keywords: Pneumomediastinum; spontaneous pneumomediastinum; pneumothorax

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Introduction

Pneumomediastinum is a rare clinical entity (1). It is defined as air within the mediastinum, and is less often termed mediastinal emphysema. The mediastinum is defined as the visceral space bounded laterally by the parietal pleura, superiorly by the thoracic outlet, inferiorly by the diaphragm, and anteriorly by the sternum, and posteriorly by the thoracic vertebral column. Contained within are the heart, tracheobronchial tree, lung, and esophagus. When air escapes from these organs or structures into the mediastinum, this is termed pneumomediastinum. Symptoms from pneumomediastinum most commonly involve chest pain, and may also include dyspnea, neck swelling, cervical pain, dysphagia, odynophagia, and dysphonia (2,3). Pneumomediastinum may be accompanied by pneumothorax, subcutaneous emphysema, or rarely, pneumorrachis, or air within the spinal canal (4).

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Background

Pneumomediastinum is commonly classified into two categories: spontaneous pneumomediastinum and secondary pneumomediastinum. Spontaneous pneumomediastinum occurs in otherwise healthy subjects without obvious causative factor. Spontaneous pneumomediastinum, however, has predisposing factors, such as smoking and recreational drug use (5,6). Secondary pneumomediastinum is due to an identifiable causal factor. These may include iatrogenic causes, such as intubation, thoracic surgery, thoracostomy tube placement, or central vascular access, or may be due to traumatic injuries (7). Other causes of secondary pneumomediastinum include asthma, air trapping, bronchiectasis, child birth, chronic obstructive pulmonary disease (COPD), coronavirus disease 2019 (COVID-19), inhalation of toxic fumes, interstitial lung disease, malignancy, marijuana use, mechanical ventilation, and physical activity (8-15). Exacerbations in respiratory diseases or infections often lead to pneumomediastinum when exacerbations with excessive coughing cause an increase in endopulmonary pressure (7). Similarly, child birth, scuba-diving, retching, and certain types of physical activity may cause increased endopulmonary pressure, leading to pneumomediastinum (7).

The suspected pathophysiology for spontaneous pneumomediastinum is commonly termed the "Macklin effect". Evidence for this arises from Macklin in the 1944 study conducted on cats (16). Increased endopulmonary pressure leads to alveolar rupture; air migrates through the peribronchial and perivascular sheathes towards the mediastinum (16). Increased alveolar pressure may result directly, from coughing or inhalation injury, or indirectly from forceful retching and vomiting leading to increased alveolar pressure, and thus alveolar rupture and pneumomediastinum (17).

Rationale and knowledge gap

Limited evidence exists guiding clinical decision making and treatment for pneumomediastinum. Specifically, no randomized clinical trials exist to guide pneumomediastinum management in either spontaneous or secondary causes of pneumomediastinum. The highest level of evidence guiding pneumomediastinum treatment comes from various case series and reports. However, due to the various etiologies of pneumomediastinum, these may be difficult to apply broadly, and need to be considered on a case-by-case basis (8).

Objective

This review aims to gather the current evidence guiding isolated pneumomediastinum (pneumomediastinum without associated effusion) diagnosis and management, with respect to two clinical scenarios: (I) presentations with low suspicion for esophageal rupture, such as patients with excessive coughing; and (II) presentation with higher concern for esophageal rupture, such as vomiting or retching.

Overview of pneumomediastinum

Pneumomediastinum is a rare clinical entity with incidence of 1/25,000 in age 5–34 years (18). In younger patients, spontaneous pneumomediastinum is more common, with an incidence of 1/14,000 (19). However, some posit that pneumomediastinum is underdiagnosed, due to either patients refraining from seeking medical care, misdiagnosis as musculoskeletal pain, or missed pneumomediastinum on chest X-ray (CXR) (20,21). A male predominance is noted, occurring in a male-to-female ratio of 3.6:1 (22). The prototypical patient with pneumomediastinum exhibits pre-existing lung disease, such as asthma, and is a male of tall and lean habitus (3,23).

Patient presentation: history and physical exam

Patients with pneumomediastinum typically present with the chief complaint of acute retrosternal chest pain radiating to the neck or back (9). This is present in about 60–100% of patients (9). They may also present with dyspnea (75%), neck swelling, cervical pain (36%), dysphagia, odynophagia, and dysphonia (2,3,9). In order to ascertain the causative factor, the patient history should include inciting factors, such as episodes of coughing or episodes of forceful retching or vomiting. For example, an asthma exacerbation may be a precipitating cause of a coughing episode. Alternatively, a patient with hyperemesis gravidarum may present after significant vomiting (7). Predisposing factors, such as respiratory diseases, inhalation injury, smoking, and recreational drug use increase suspicion for pneumomediastinum (7,8). Commonly, the causative factor is unknown, and pneumomediastinum is discovered incidentally (9,10).

Upon exam, patients may present with tachycardia and tachypnea (7). A notable physical exam finding in pneumomediastinum is the Hamman's sign. Hamman's sign, also known as Hamman's crunch, is the presence of

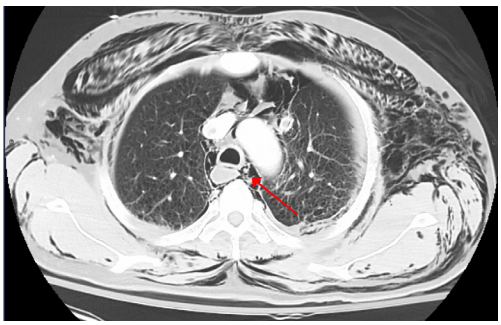

Figure 1 Pneumomediastinum on chest CT as demonstrated by red arrow. This figure is licensed under the Creative Commons Attribution-Share Alike 3.0 Unported License. Initial image is created by James Heilman, MD and is under the public domain because it is a work of medical imaging created in the United States and does not contain additional copyrightable graphics. Informed consent was obtained prior to uploading the image by the initial author. CT, computed tomography.

a "crunch" synchronous with the heart beat on cardiac auscultation (24). Subcutaneous emphysema (70%) may be present as well (10,12).

Diagnostic imaging and other adjuncts

Diagnosis is commonly made on anterior CXR; CXR alone may be diagnostic in 73–90% of patients (3,10,25). Common radiologic signs include the "continuous diaphragm sign" caused by posterior pericardial air, "extra-pleural air sign" caused by air extending between the parietal pleural laterally towards the diaphragm, and the "thymic sail sign" caused by air elevating the thymus. Others include shining bundles surrounding the mediastinal organs and mediastinal pleura separating from the cardiac edge (17,26-28).

Whether CXR alone is sufficient for diagnosis of pneumomediastinum remains contentious. As up to 30% of patients with pneumomediastinum present with normal CXR, it is suggested that inclusion of chest computed tomography (CT) be performed if there is suspicion for pneumomediastinum despite normal CXR, to differentiate between pneumopericardium or subcutaneous emphysema, or to investigate for causes of pneumomediastinum (17,21,23,29). Chest CT can be beneficial in detecting injury to the tracheobronchial system, pneumothorax [present in up to 40% of patients (3)], or esophageal perforation, each of which may affect management (25).

Other minimally invasive adjuncts in diagnosis include bedside thoracic ultrasound, which may aid in more rapid recognition of pneumomediastinum as a cause of acute onset chest pain. Findings on thoracic ultrasound include poor visualization of the heart, diffuse A lines, and normal visualization of the heart from the subxiphoid view (12,30). Laboratory and electrocardiogram (ECG) findings are non-diagnostic and non-specific; laboratory findings may demonstrate leukocytosis or increased C-reactive protein (CRP) while ECG findings may mimic acute pericarditis (31,32). Echocardiography may be used in case of suspicion for pneumopericardium.

Diagnosis and management per etiology

As previously described, the etiology of pneumomediastinum is often challenging to ascertain. The most likely cause of pneumomediastinum is due to alveolar rupture as a result from increased Valsalva pressure from prolonged coughing or retching (33). A more detailed patient history can help narrow the range of possible etiologies of pneumomediastinum, and target diagnostic workup to rule out the most likely associated injury.

Patient with coughing: lower concern for esophageal perforation

Diagnosis

The patient presentation that may raise concern for airway or lung etiology of pneumomediastinum usually demonstrates a significant episode of coughing (8,34). Past medical history that may be associated includes lung-related pathology, i.e., asthma, COPD, bronchiectasis, and interstitial lung disease (7). Clear causes of airway or lung injury, such as mechanical ventilation, intubation, or other non-iatrogenic trauma raise more direct concern for airway or lung etiology of pneumomediastinum.

As standard for pneumomediastinum, one would begin with CXR. If CXR is non-diagnostic, but suspicion remains high for pneumomediastinum, CT chest may be obtained (*Figure 1*) (7,25).

Further diagnostic maneuvers, such as bronchoscopy, laryngoscopy, esophagoscopy, or esophagogram need not be routinely performed, but selectively performed based on clinical suspicion (3,7,25). If clinical history suggests likely airway injury, as in the case of traumatic injury, bronchoscopy should be performed. Clinical instability as defined by unstable vital signs, would also necessitate

more aggressive workup, including repeat CT chest, bronchoscopy to rule out missed air leak and esophagram, and/or esophagoscopy to rule out esophageal injury (7,9,10).

However, if the patient is clinically stable, as determined by normal vital signs and laboratory values, and low suspicion for bronchial or esophageal injury exists, routine bronchoscopy, esophagoscopy, and esophagogram are unlikely to provide clinical benefit. In Song *et al.*, 45 patients with spontaneous pneumomediastinum were studied, 90% underwent esophagogram, 31% bronchoscopy, and 2.2% endoscopy, and no injury was found (25). Similar findings have suggested futility of routine extensive workups, and favor a more selective approach to the workup of pneumomediastinum (10,34).

Management

Management of spontaneous pneumomediastinum largely consists of supportive care, which has historically been defined by oxygen inhalation therapy, pain control, and bed rest (3,9,10,18). The theory behind oxygen inhalational therapy is to increase the diffusion pressure of nitrogen in the interstitium and promote free air absorption in the mediastinum (25,35). Consensus as to admission, length of admission, diet restriction, and antibiotic control has not yet been reached, likely due to limited controlled trials. Though earlier studies suggest hospitalization for approximately 2–5 days, this is based on retrospective data that collects the average length of stay for spontaneous pneumomediastinum. The data lacks granularity to clarify the reason for admission and thus should not be taken as prescriptive (9,10). Hospital admission and length of stay may be minimized in uncomplicated spontaneous pneumomediastinum without compromising patient safety (3,25,31). A clinical judgment for admission versus observation then discharge in each clinical scenario is reasonable. Patients with pain, nausea, or other ongoing symptoms may warrant admission (35,36). During admission, supplemental oxygen may be given as clinically warranted.

Diet restriction and prophylactic antibiotic usage for prevention of mediastinitis is advocated for by some (18,31). However, more recent studies note a lack of evidence for true benefit; in the patient with lower risk features, such as age less than 40 years, presentation with cough, normal white blood cell (WBC) count, and no evidence of pleural effusion, suspicion for esophageal injury is low, and recommendation is instead to advance diet as tolerate and avoid prophylactic antibiotics (37).

If concomitant significant pneumothorax is noted, thoracostomy tube placement may be appropriate. However, this is generally a low percentage of patients (6%), and may be more likely to present in patients who are mechanically ventilated or have associated pulmonary diseases where pneumothoraces are more likely to develop (3,10,23).

In cases that pneumomediastinum progresses to airway compression, video-assisted thoracoscopy (VATS) or thoracotomy may be necessitated for decompression (38). Additionally, tension pneumopericardium may develop, leading to cardiac tamponade, requiring surgical intervention by VATS or pericardial window (9).

Rarely, a tracheal or bronchial injury is found (3,9,10,18,21). In the case of tracheal injury, operative management is usually necessitated. However, in bronchial injuries stenting is increasingly used, particularly in cases of iatrogenic injury (39).

Patient with vomiting/retching: higher suspicion for esophageal perforation

Diagnosis

Pneumomediastinum may also arise in patients with severe and repeated vomiting and retching. This may be due to excessive alcohol ingestion, recreational drug use, hyperemesis gravidarum, and ingestion of caustic substances (7,34). The history of repeated retching could lead to alveolar rupture via Macklin effect (7,16,33).

Given this difference in patient history, the initial CXR should be immediately followed by CT of the chest and abdomen (3,25,34). If high-risk features such as age greater than 40 years, abdominal pain, leukocytosis, CT findings of pleural effusion, or pneumoperitoneum, one should maintain an even higher concern for esophageal perforation and proceed with either contrast esophagram or CT esophagogram with or without esophagoscopy immediately thereafter (34,40,41). For contrast esophagram, it is recommended to start with water-soluble esophagram, and then confirm with barium esophagram to minimize contrast extravasation into the mediastinal or pleural space. A summary of the diagnostic algorithm for both pathways is provided in *Figure 2*.

Management

In the absence of diagnostic evidence of esophageal injury, management of pneumomediastinum in this scenario is similar to that above. The main treatment is supportive care,

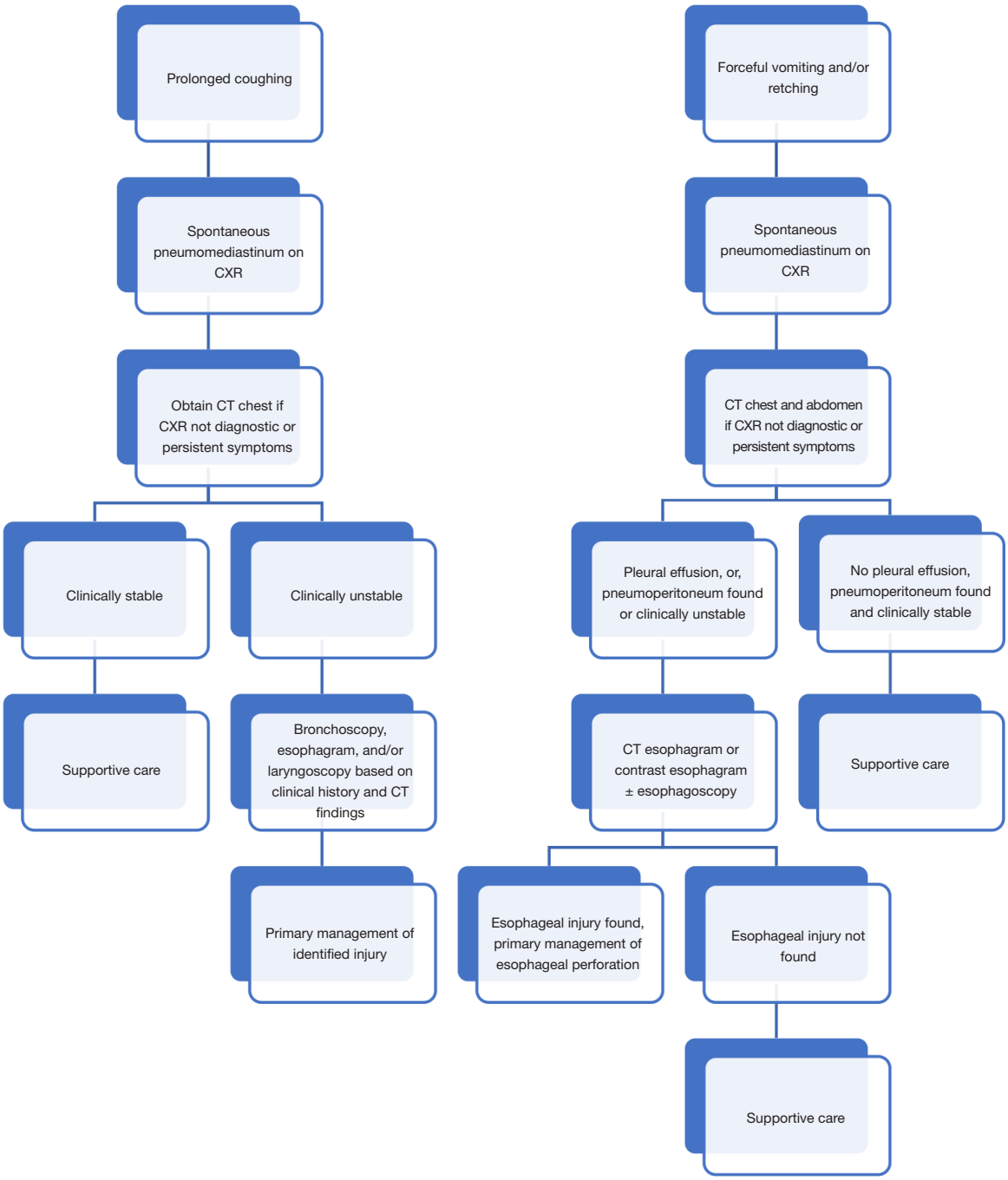

Figure 2 Proposed diagnostic algorithm for spontaneous pneumomediastinum in adults. Clinically unstable defined by any one of fever, tachycardia, hypotension, or leukocytosis. CXR, chest X-ray; CT, computed tomography.

including bowel rest and pain control (3,9,10,25,42). Once symptoms of pneumomediastinum are resolved, patients can be started on a liquid diet and advanced as tolerated. In the absence of evidence for esophageal perforation, prophylactic antibiotics are not required (25,34).

Long-term follow-up of pneumomediastinum

Pneumomediastinum is self-resolving through air resorption in the mediastinal tissues, though in some cases may be notable on imaging for up to 6 months (9,10). Recurrence

of pneumomediastinum is exceedingly rare, approximately 1% (31,43). However, a recent by Kumeda *et al.* in 2023 demonstrates a surprisingly high recurrence rate of 17% (44). Though it is unknown if this particular result is secondary to a small sample size or if the true recurrence rate is higher than previously thought, identifying patients with risk factors for recurrence, such as asthma, is essential (45). Given such low rates of pneumomediastinum recurrence in the majority of studies thus far, long-term follow-up may not be a universal requirement, but evaluated on a case-by-case basis.

Pneumomediastinum in children

Management of spontaneous pneumomediastinum in children is largely similar to that of adult spontaneous pneumomediastinum (19). Spontaneous pneumomediastinum is rarer in children than adults. However, incidence is challenging to ascertain and is likely underdiagnosed (46,47). Asthma remains the most frequent comorbidity in children, with approximately 22% of patients with pneumomediastinum presenting with this comorbidity (47). Secondary spontaneous pneumomediastinum is most common caused by asthma exacerbation, and may also be caused by pneumonia, lower respiratory tract infections, or choking. In a study of 87 pediatric patients at a tertiary children's facility in Taiwan, all patients younger than 6 years had secondary spontaneous pneumomediastinum, versus 60.6% of patients older than 6 years. Diagnosis remains similar, with CXR followed by chest CT only if diagnosis is unclear and suspicion for pneumomediastinum remains high. Further diagnostic testing beyond CXR (after establishing diagnosis) is unlikely to affect management in clinically stable children (48,49).

Routine further invasive testing, such as esophagram and laryngoscopy are unlikely to yield diagnostic information (48). Thus, a difference in diagnostic workup is that for clinically well-appearing children, unless suspicion for associated airway, lung, or esophageal injury is high, extended diagnostic workup including bronchoscopy, laryngoscopy, or esophagram is unwarranted (49). Patients may be managed conservatively with clinical observation, rest, pain control, and oxygen therapy and will likely have spontaneous resolution (47,50).

A key clinical difference is that in the absence of known respiratory disease (i.e., asthma), spontaneous pneumomediastinum in children without known cause should prompt outpatient pulmonary function testing (PFTs) to investigate whether the patient has asthma or other respiratory disease (7,19).

Future spontaneous pneumomediastinum guidelines

Presented in this article is a clinical review of pneumomediastinum and suggested diagnostic and therapeutic pathways based on suspected etiology of pneumomediastinum. The strengths of this article include gathering available evidence arising from a thorough literature review on PubMed for spontaneous pneumomediastinum. Primary evidence regarding spontaneous pneumomediastinum diagnosis and management is a combination of various case reports, case series, and retrospective cohort studies. By combining this literature as well as the author's own diagnostic and management algorithm from a high-volume thoracic surgery regionalized center, a standardized, easy-to-follow algorithm is presented. However, limitations due to the lack of prospective, controlled trial evidence limits the strength of the recommendations for mandated diagnostic and treatment algorithms in this disease process.

Conclusions

Pneumomediastinum is a rare condition with multiple etiologies, and often no known etiology. Spontaneous pneumomediastinum may be diagnosed with limited workup, such as CXR and/or CT. Secondary pneumomediastinum diagnosis and treatment may be streamlined based on suspected etiology. Clues to etiology often come from patient history and presenting symptoms. Although evidence from randomized controlled trials are lacking, case series suggest that pneumomediastinum is generally benign and self-limited if no associated injury is identified. Management is largely supportive care and symptomatic management. Recurrences are rare, and thus limited follow up is necessary. Higher level evidence is needed to solidify treatment algorithms for pneumomediastinum in the future, which may further promote conservative management and limit unnecessary diagnostic procedures.

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Footnote

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Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://med.amegroups.com/article/view/10.21037/med-23-25/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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