

SPONTANEOUS PNEUMOMEDIASTINUM IN A HEALTHY ADOLESCENT

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SUMMARY – Spontaneous pneumomediastinum is a rare clinical entity defined as the presence of free air in the mediastinal structures without an apparent cause such as trauma. Spontaneous pneumomediastinum is rare in children and most frequently occurs in young male patients. It usually develops after alveolar rupture and air penetration into the pulmonary interstice, followed by air penetration towards the hila and into the mediastinum. Alveolar ruptures may be caused by various pathological and physiological processes, in children most frequently by asthma. Clinical diagnosis is based on the symptom triad including chest pain, dyspnea and subcutaneous emphysema. The diagnosis is confirmed by radiography. On differential diagnosis, esophageal perforation should be considered first, and if suspected, contrast esophagogram should be performed. Spontaneous pneumomediastinum usually resolves spontaneously in several days of treatment, which includes identification of the underlying cause (if possible), rest, analgesics and clinical monitoring. Complications involving spontaneous pneumomediastinum, such as tension pneumomediastinum and tension pneumothorax, are quite rare. A case is presented of pneumomediastinum in a 17-year-old male adolescent with no relevant history but with a clinical picture of intense retrosternal pain and subcutaneous emphysema of the neck and supraclavicular region. Thorough examinations including chest x-ray, chest computed tomography, bronchoscopy and esophagoscopy failed to identify the cause of pneumomediastinum. After eight days of conservative treatment, the pneumomediastinum symptoms completely disappeared and x-ray showed resolution of pneumomediastinum.

Key words: *Mediastinal emphysema – diagnosis; Mediastinal emphysema – therapy; Adolescence; Case report*

Introduction

The term pneumomediastinum is derived from the Greek word *pneuma*, meaning air, and actually means mediastinal emphysema, i.e. a condition involving the presence of air within the mediastinum¹. It was first described by Laennec back in 1819 as a result of traumatic injury². In 1939, Hamman was the first to describe a series of patients with spontaneous pneumomediastinum (SPM) and precordial crunching noise synchronous with the heart beat, known as Hamman's sign³. SPM

refers to pneumomediastinum cases that do not occur after a thoracic trauma, endotracheal or endoesophageal procedure, mechanical ventilation, cardiac catheterization or thoracic surgeries¹. The most common clinical picture of SPM involves chest pain, dyspnea and subcutaneous emphysema; however, dysphagia, neck pain, neck swelling, hoarse voice and rhinolalia may also be quite frequently present⁴⁻⁶. Subcutaneous emphysema mostly affects the neck, often spreading over the thorax to cause Hamman's sign⁶.

Various factors may be involved in the development of SPM. These are mostly conditions that lead to raised intra-alveolar pressure, such as forcible cough, screaming, crying, vomiting, defecation, strenuous physical activities and Valsalva maneuver^{4,5,7,8}.

Chest radiography is an option in diagnosing SPM in patients having no history of potential perforation

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of intrathoracic organs or the need of further treatment⁹. If pneumomediastinum is not confirmed by x-ray, it is recommended to do contrast-enhanced chest computed tomography (CT)⁸⁻¹¹.

Therapy for SPM is largely conservative and includes rest, strict avoidance of any procedures requiring forcible expiration, observation and cardiopulmonary monitoring of children due to an extremely rare risk of developing complications such as tension pneumomediastinum or tension pneumothorax^{9,12-14}.

A case is reported of a 17-year-old adolescent admitted for medical check-up due to intense chest and neck pains, aggravated on inhalation and swallowing, manifest subcutaneous emphysema of the neck and both supraclavicular regions, with normal vital functions and without a history of predisposition for the disease development. Diagnostic work-up failed to identify the cause of SPM and the disease resolved after eight days of conservative therapy.

Case Report

A 17-year-old male adolescent was admitted to our emergency service in evening hours for intense chest



Fig. 1. Anteroposterior chest x-ray showing pneumomediastinum (vertical lucent streak along the aortic arch) and subcutaneous emphysema in the cervical region.

and neck pains that had begun early that morning. The pain was localized centrally in the thorax and was also present at rest, but intensified on inhalation and swallowing. He presented with no relevant medical history or asthma history.

Physical examination revealed 97% O₂ saturation in room air, blood pressure 120/80 mm Hg and pulse 60/min. Thoracic expansion was symmetric with clear pulmonary sounds. Subcutaneous emphysema was present on the neck and in the supraclavicular regions with palpable crepitations. Complete blood count and serum biochemistry findings were free from any significant deviation. The urine toxicology test was negative.

X-ray of the lung and cervical region pointed to bilateral subcutaneous emphysema in the area of the neck soft tissues, with normal lung expansion. At the aortic arch on the left, a sharply delineated linear shadow of trapped air was seen in the form of transparent band, raising suspicion of a small amount of air in the mediastinum (Fig. 1). That is why chest CT was done. In the lower half of the neck, a series of air bubbles appeared between the muscles and the vessels in the cervical region and on both sides supraclavicularly, a finding corresponding to subcutaneous emphysema (Fig. 2). A relatively copious evenly distributed air quantity was visible around the ana-

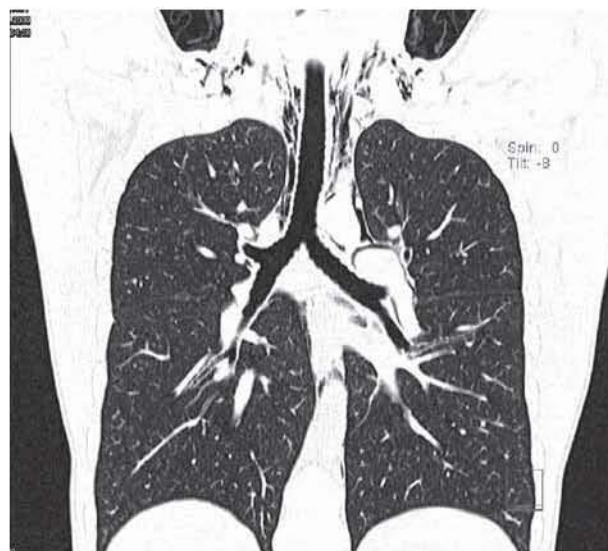


Fig. 2. Spontaneous pneumomediastinum with mediastinal and subcutaneous emphysema in the cervical and supraclavicular region, detected by computed tomography scan of the chest.

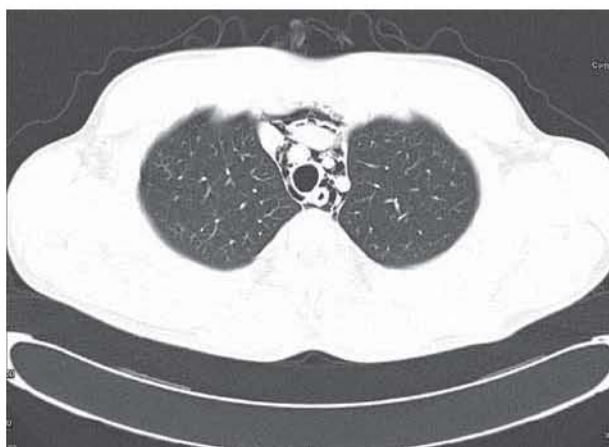


Fig. 3. Computed tomography confirmed massive emphysema (air) of the superior mediastinum and around the tracheal bifurcation and vessels.

tomic structures of superior mediastinum (Fig. 3). In all sections, the esophagus showed normal width and regular contours. The trachea and the main bronchial branches were normally shaped and ramified, and there were no signs suggesting a lesion of bronchial branches. The pneumomediastinum was largely seen in the upper part of the chest, with minimum air quantities being visible in the inferior and posterior mediastinum around the esophagus (Fig. 4). Pulmonary parenchyma was properly ventilated without any signs of pneumothorax.

On fiberoptic endoscopy of the trachea and esophagus, laryngeal structures and the entire trachea showed normal appearance, with visualization of all segmental and subsegmental bronchi on the left and right, free from any pathologic changes. The esophagus showed normal appearance all the way to the gastroesophageal junction.

Based on these examinations, SPM was diagnosed. Conservative treatment consisting of rest and analgesics if required resulted in complete recovery. The patient was discharged after eight days of supportive therapy. Follow up chest x-ray finding was normal.

Discussion

Pneumomediastinum is not common in pediatric practice. It is defined as free air in the mediastinum that leaks as a rule from the alveolar space or, rarely, from the airways. The etiology of pneumomediastinum is multifactorial^{4,15}. Pneumomediastinum may be the

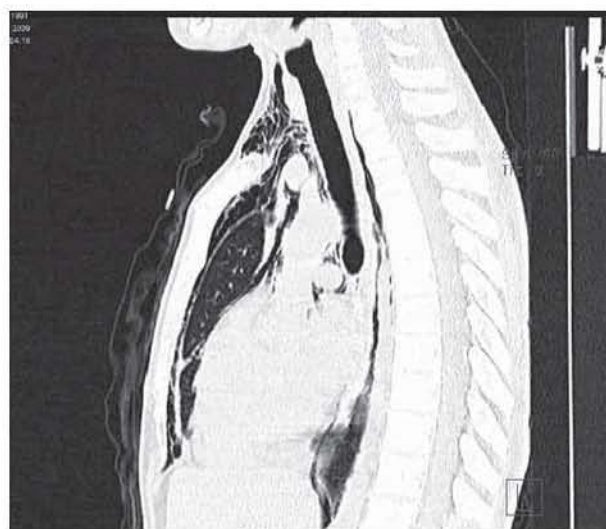


Fig. 4. Free air expanding in the middle and inferior mediastinum.

result of an open or penetrating injury or leakage of air produced by microorganisms, or after esophageal rupture accompanied by vomiting¹⁶. SPM is not common and occurs in patients without a history of the above mentioned troubles. Many authors distinguish SPM as a form of pneumomediastinum not associated with an open trauma, endobronchial or esophageal procedure, mechanical ventilation, surgical operation of the thorax, or some other invasive procedure¹⁵. It is the result of conditions that lead to raised pressure in the alveoli causing their rupture and thereby air penetration in the peribronchial and perivascular spaces into the mediastinum¹⁷. According to the review of the available literature on the issue reported by Chalumeau *et al.* in 2001, the prevalence of SPM ranges from 1 *per* 800 to 1 *per* 42,000 pediatric patients hospitalized at emergency service⁹. SPM often occurs in young, lanky persons^{10,11}. All patients are usually of good health and without a serious pulmonary disease. According to the latest reports, the incidence of SPM is on a rise, which is surely a result of improved examinations, but also of using inhaled drugs by young people¹⁸. As early as 1939, Macklin described the pathophysiology of SPM as alveolar rupture, air dissection along the bronchovascular tree and penetration of free air into the mediastinum¹⁹. Air volume determines the severity of SPM, and the air spreads around the trachea and esophagus into the neck and subcutaneous spaces. Free air is present in superior mediastinum, spread-

ing by progression to the middle and inferior mediastinum¹¹. The most common clinical picture of SPM includes sudden retrosternal chest pain potentially spreading to the shoulders, the back and the arms (in 50%–90% of SPM cases), subcutaneous emphysema and dyspnea (that may be associated with asthma or tension pneumomediastinum)^{1,5,8}. Neck pain, dysphagia, and hoarseness may also be present^{4,8,10,12}. The most common finding in SPM patients is subcutaneous neck emphysema, which, however, is not pathognomonic for pneumomediastinum. According to some authors, Hamman's sign is pathognomonic for pneumomediastinum; in other words, precordial systolic crepitations synchronous with cardiac action, intensified during inspiration, and weakened cardiac sounds are described in 50% of SPM patients^{3,4,6,8,13}.

The causes of SPM can be a variety of factors leading to an increase in the intra-alveolar (intrathoracic) pressure, such as forcible cough, crying, screaming, vomiting, defecation, strenuous athletic activities, scuba diving, playing of wooden wind instruments, etc.^{7,8,10,12–14}. In the literature, SPM is often described in association with conditions leading to Valsalva maneuver, such as childbirth, strenuous exercises, stool strains, coughing, sneezing, attempted vomiting or vomiting^{7,8,12,14}. Another risk factor for SPM development are obstructive pulmonary diseases (asthma, bronchiolitis, foreign body aspiration), especially in intubated and mechanically ventilated patients^{20,21}. SPM often occurs during the administration of inhaled drugs, however, SPM cases following the administration of methylenedioxymethamphetamine (ecstasy tablets) have also been recently reported^{22–24}. Along with the administration of inhaled drugs, marijuana and cocaine, many persons perform Valsalva maneuver, forcibly suspended breathing aimed to reinforce the effect of drugs, which may lead to alveolar rupture and pneumomediastinum²³. While the mechanism of pneumomediastinum development associated with ecstasy consumption is not exactly known, it is thought that a high level of physical strain and reduced interstitial pressure may lead to a pressure gradient that causes alveolar rupture²⁴.

Based on data collected so far, the trigger can be found in 70%–90% of SPM cases^{9,10,12}. The most common of them in pediatric population is asthma, followed by vomiting, plus situations reproducing Val-

salva maneuver, e.g., screaming, coughing, intensive sports activity, etc.^{7–10,20,21}. Spontaneous esophageal perforation (Boerhaave syndrome) or esophageal perforation due to esophageal candidiasis may lead to pneumomediastinum development with frequent serious complications, particularly mediastinitis and a dismal clinical picture^{25,26}.

The laboratory work-up we use in the diagnosis of pneumomediastinum includes blood gas analysis, cardiac enzymes (rare cases of infarction in children are possible in those with coronary vasculitis, Kawasaki syndrome, or congenital anomalies of coronary arteries), and urine and blood toxicology for illicit drugs, as indicated by the history or physical examination^{4,8–10,22,23}. Chest x-ray is an option in the diagnosis of SPM in patients having no history of potential perforation of intrathoracic organs or the need for further treatment. X-ray of the lung and cervical region may reveal air in the mediastinum area in the form of transparent bands around the heart, mostly on the left, and subcutaneous neck emphysema; associated diseases such as pneumothorax or pneumopericardium may also be visualized^{9,11–16}. If pneumomediastinum is not confirmed by chest x-ray, it is recommended to perform chest CT, which has two main diagnostic roles, i.e. to display pneumomediastinum not seen on x-ray and to anatomically locate the air in the mediastinum. CT often reveals additional diseases such as perforated esophagus, and it is as sensitive method in diagnosing mild pneumomediastinum, especially when the clinical picture is atypical^{11–16}. Bronchoscopy is used to evaluate the tracheobronchial tree and it is performed when perforation of the tracheobronchial tree following a chest trauma is suspected, in order to localize and remove the foreign body^{4,9,10,18}. Fiberoptic esophagoscopy (without insufflation) and esophagogram are used in the diagnosis of esophageal perforation and play a part in excluding a transparent foreign body^{14,25,26}. If the Boerhaave syndrome (spontaneous esophageal perforation with vomiting) is suspected, contrast examination of the esophagus with swallowing is obligatory^{25,26}.

The management of SPM is largely conservative because most SPM cases resolve spontaneously. The management includes patient observation along with strict rest, clinical monitoring and cardiopulmonary monitoring of children to avoid the risk of tension

pneumomediastinum and pneumothorax, and the administration of 100% O₂ if the air in the mediastinum compromises cardiac function^{8,10,12,15}. Procedures that require forcible expiration, such as peak expiratory flow (PEF) measurement or radiography with forcible expiration should be avoided^{5,12-14}. Mediastinoscopy is very rarely employed; it is reserved for life-threatening cases of tension pneumomediastinum, thoracotomy and drainage in case of pneumothorax^{4-6,12,14,27}. It is recommended to avoid physical activities by the time of pneumomediastinum resolution. Air reabsorption follows after seven to ten days^{9,12,13}. Subcutaneous and mediastinal emphysema are resorbed into the tissues by diffusion in accordance with the partial pressure gradient.

It should be noted that potentially threatening complications may arise from SPM, although very seldom in practice. They have been defined by Macklin and Macklin in animal model experiments and include tension pneumomediastinum, unilateral or bilateral pneumothorax or tension pneumothorax, and pressure increase in pulmonary interstice which causes respiratory troubles¹⁷. Tension pneumomediastinum with cardiac compression and tamponade effect is indeed a rare complication of SPM^{9,27}. These complications are extremely rare owing to the fact that the visceral layer of the deep cervical fascia has continuity with the mediastinum, resulting in air decompression in the neck and thus preventing the tamponade and pneumothorax. Mediastinitis as a complication develops especially in association with the Boerhaave syndrome, i.e. spontaneous esophageal rupture accompanied by vomiting. The Boerhaave syndrome should always be kept in mind in individuals with a history of vomiting followed by chest pain, dyspnea and potential cardiovascular collapse²⁵.

The young patient admitted to our emergency service had a history of intense chest pain that worsened on inhalation and swallowing. The presence of marked subcutaneous emphysema of the neck and supraclavicular regions raised suspicion of pneumomediastinum. No auricular, nasal or pharyngeal lesions were detected on physical examination. Chest x-ray indicated subcutaneous neck emphysema, confirming the suspicion of pneumomediastinum by visualizing a transparent band at the aortic arch. Thoracic CT finding confirmed the diagnosis of pneumomediasti-

num with the presence of air mainly in the superior mediastinum, but with a tendency to spread into the inferior and posterior mediastinum. Subcutaneous emphysema of the neck and supraclavicular regions was also present, however, without signs of tracheal, bronchial and esophageal lesions. Fiberoptic bronchoscopy and esophagoscopy showed no lesions of the tracheobronchial tree and esophagus, so SPM was diagnosed. Thorough history revealed no trigger factor as a possible cause of increased intra-alveolar pressure and SPM development.

Once the diagnosis was made, conservative therapy was initiated, consisting of strict rest, cardiopulmonary monitoring and analgesics as required. The patient was discharged after eight days of normal clinical status and normal follow up x-ray.

Conclusion

Pneumomediastinum should be taken in consideration in a patient presenting with severe chest pain, subcutaneous emphysema, Hamman's sign and dyspnea. Chest x-ray can help make the diagnosis. CT is used in the diagnosis of pneumomediastinum when clear clinical and x-ray findings are absent. Also, esophageal and tracheobronchial perforations as the possible cause of pneumomediastinum should not be overlooked in cases involving thoracic traumas or diseases that may lead to pneumomediastinum, such as asthma, foreign body aspiration or Boerhaave syndrome. SPM caused by various factors that lead to an increased intra-alveolar pressure and alveolar rupture should be suspected when trauma, i.e. perforation of thoracic organs, has been excluded as the cause of pneumomediastinum. SPM patients must be on strict bed rest regimen and subjected to cardiopulmonary monitoring due to the risk of the possible albeit rare complications in the form of tension pneumomediastinum, pneumothorax or pneumopericardium. SPM is a rare disease with a typically benign course that usually resolves spontaneously on conservative treatment.

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Sažetak

SPONTANI PNEUMOMEDIJASTINUM KOD ZDRAVOG ADOLESCENTA

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Spontani pneumomediastinum (SPM) je rijedak klinički entitet obilježen prisutnošću slobodnog zraka oko medijastinalnih struktura bez jasnog uzroka kao što je trauma. SPM je rijedak u djece i javlja se uglavnom u muških adolescenata. Obično se pojavljuje sekundarno nakon rupture alveola i prodora zraka u plućni intersticij, a potom slijedi prodor zraka prema hilusima i u medijastinum. Različiti patološki i fiziološki događaji mogu dovesti do rupture alveola, a u djece je najčešći uzrok astma. Klinička dijagnoza se temelji na trijasi simptoma, a to su bol u prsištu, dispneja i supkutani emfizem. Dijagnoza se potvrđuje rentgenskom snimkom. U diferencijalnoj dijagnozi treba prvenstveno misliti na perforaciju jednjaka, a postoji li sumnja na perforaciju treba učiniti ezofagogram s kontrastom. SPM se obično spontano povlači nakon nekoliko dana liječenja koje uključuje pronalaženje osnovnog uzroka (ako je moguće), odmor, analgetike i kliničko praćenje. Rijetke su komplikacije spontanog pneumomediastinuma kao što je tenzijski pneumomediastinum i tenzijski pneumotoraks. Ovdje se prikazuje slučaj pneumomediastinuma u 17-godišnjeg mladića bez značajne medicinske anamneze, s kliničkom slikom intenzivnih retrosternalnih bolova te supkutanim emfizemom vrata i supraklavikularnih područja. Intenzivnim pretragama uključujući rentgensku snimku pluća, kompjutoriziranu tomografiju prsnog koša, bronhoskopiju i ezofagoskopiju nije se pronašao uzrok pneumomediastinuma. Nakon osam dana konzervativnog liječenja došlo je do potpunog nestanka simptoma i povlačenja pneumomediastinuma na rentgenskoj snimci.

Ključne riječi: Emfizem medijastinuma – dijagnostika; Emfizem medijastinuma – terapija; Adolescencija; Prikaz slučaja