Araştırma Makalesi / Research Article

Hamman's Syndrome (Spontaneous Pneumomediastinum)

Hamman Sendromu (Spontan Pnömomediastinum)

Mehmet Akif TEZCAN [1], İbrahim Ethem ÖZSOY [1]

1 Department of Thoracic Surgery, Kayseri Health Practice and Research Center, University of Health Sciences, Kayseri, Turkey

Abstract

Background: In this study, we wanted to draw attention to the spontaneous pneumomediastinum which rarely seen and to share our experiences.
Materials and Methods: Between April 2016 - April 2018, we retrospectively evaluated 12 patients who were hospitalized and treated with SPM diagnosis at Health Practice and Research Center.
Results: Of the 12 patients with spontaneous pneumomediastinum, 9 (75%) were male and 3 (25%) were female. Patients had a mean age of 30.5 years (range 15-42 years), 2 patients admitted to the emergency service with swelling and dyspnea complaints, 1 patient with cough and vomiting, 1 patient with cough and chest pain, 3 patients with dyspnea and chest pain, 3 patients with sore throat and chest pain, 2 patients with swelling in the neck after the tooth extraction. 6 (50%) of the patients had a smoking history.
Conclusions: Spontaneous pneumomediastinum is a disease that is seen rarely in emergencies and should be treated. Sometimes life-threatening complications may develop. Other disorders in differential diagnosis should be eliminated in spontaneous pneumomediastinum.
Keywords: Spontaneous pneumomediastinum, Dyspnea, Chest pain, Subcutaneous emphysema

Öz

Amaç: Bu çalışmada nadir görülen spontan pnömomediastinuma dikkat çekmek ve klinik tecrübelerimizi aktarmak istedik.
Materyal ve Metod: Nisan 2016- Nisan 2018 tarihleri arasında Sağlık Uygulama ve Araştırma Merkezinde yatırılıp tedavi edilen spontan pnömomediastinum'lu 12 hastayı retrospektif olarak değerlendirdik.
Bulgular: Spontan pnömomediastinumlu 12 hastanın 9'u erkek (%75) ve 3'ü (%25) kadındı. Hastaların yaş ortalaması 30,5 (15-42 yaş) idi. İki hasta şişme ve dispne, 1 hasta öksürük ve kusma, 1 hasta öksürük ve göğüs ağrısı, 3 hasta dispne ve göğüs ağrısı, 3 hasta boğaz ve göğüs ağrısı ve 2 hasta diş çekimi sonrası boyunda şişme şikâyeti ile acil servise kabul edildi. Hastaların 6'sında (%50) sigara hikayesi mevcuttu.
Sonuç: Spontan pnömomediastinum, acilde nadir görülen, hayatı tehdit eden komplikasyonlar gelişebilecek tedavi edilmesi gerekli bir hastalıktır. Spontan pnömomediastinum'lu hastalarda diğer hastalıklar elimine edilmelidir.
Anahtar kelimeler: Spontan pnömomediastinum, Dispne, Göğüs ağrısı, Cilt altı amfizem

Sorumlu Yazar / Corresponding Author

Mehmet Akif Tezcan
Assistant Professor
Department of ThoracicSurgery,
Kayseri Health Practice and Research Center,
University of Health Sciences, Kayseri, Turkey

Kayseri Şehir Hastanesi, Molu, Kocasinan, Kayseri, Turkey

Tel: +90 532 7279366

e mail:mehmetakiftercan@gmail.com

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Introduction

Spontaneous pneumomediastinum (SPM) is a rare disease characterized by the presence of free air in the mediastinum without any underlying cause. The pathophysiology of this condition is based on the existence of a pressure gradient between the alveoli and the lung interstitium; this pressure difference may lead to alveolar rupture and the consequent escape of air into the interstitium. In the development of SPM, there are some predisposing and triggering factors such as asthma, Marfan syndrome, ethanol intoxication, vomiting, defecation, exercise, inhaler drug use, screaming and smoking. Chest pain, dyspnea, and subcutaneous emphysema are the most common clinical manifestations.

The objective of this study was to discuss our experience in the diagnosis and management of spontaneous pneumomediastinum

Materials and Methods

This study was approved by the local ethics committee. The study was conducted in accordance with the principles of the Declaration of Helsinki.

Between April 2016 - April 2018, we retrospectively evaluated 12 patients who were hospitalized and treated with SPM diagnosis at Health Practice and Research Center. We evaluated the patients with respect to age, gender, referral symptoms, white blood cell-leukocytes (WBC), neutrophil / lymphocyte ratio (N/L), intrathoracic pathologies such as accompanying pneumothorax, pleural effusion, predisposing factors and mortality.

Results

Of the 12 patients with SPM, 9 (75%) were male and 3 (25%) were female. Patients had a mean age of 30.5 years (range, 15-42 years), 2 patients admitted to the emergency service with swelling and dyspnea complaints, 1 patient with cough and vomiting, 1 patient with cough and chest pain, 3 patients with dyspnea and chest pain, 3 patients with sore throat and chest pain, 2 patients with swelling in the neck after the tooth extraction. 6 (50%) of the patients had a smoking history. There were not drug history 10 patients except two of them. One of these patients was using opioids, other one was using drugs for asthma. All of the patients were evaluated by on chest radiography and computerized thorax tomography (CT) (Figure 1). All of the patients had mediastinal emphysema on their tomography. Chest radiography was normal in three patients, while subcutaneous emphysema was present in on chest radiography of other patients. Pneumothorax was present in one patient, but tube thoracostomy was not performed. None of the patients had pleural effusion. Fiberoptic bronchoscopy and esophagoscopy were performed on two patients and no pathology was detected. At the first admission to the hospital, the leukocyte and N/L ratios of all patients were analyzed. Counts of leukocytes were 5810 – 18500 (mean 10203). Ratios of N/L were mean 3.45 (range 1.83-7.04). The patients were treated with bedrest, oxygen support, bronchodilators, analgesic and antibiotics. Antibiotics were used to reduce the risk of mediastinitis.

The patients were followed up with daily chest radiographs for mediastinal emphysema and pneumothorax. There was no mortality. The duration of hospitalization in the patients was mean 5.16 days (range 3-10 days) (Table 1).

Discussion

SPM is a benign disease, which is very rare and often self-limiting, usually seen in young adults with an increase in intrathoracic pressure (1-7). It was defined by Louis Hamman in 1939 (8). However, pneumomediastinum has been known since 1819 when it was described by Rene´ Laennec. SPM is defined as the accumulation of air in the interstitial space in the mediastinum without any defined reason. The mediastinum can occur in three different ways;

1. Entering of the air, by the penetration of the tracheabronchial tree or esophagus from the traumatic or nontraumatic rupture, through the cutaneous or mucosal barier into the mediastinum
2. Mediastinum or neighboring structure infections with gas producing organism
3. Alveolar rupture may develop due to a decrease in pressure gradient between the lung interstitium and the alveolar. Pressure gradient; It occurs due to the increase of intraalveolar pressure in Valsalva maneuver, a decrease of pleural pressure in Mueller maneuver and interstitial pressure drop due to frequent breathing and vasoconstriction in asthma. Alveolocapillary membrane anomalies and interstitial lung diseases can lead to the development of alveolar rupture. Asthma was detected as a predisposing factor in 8- 39% of patients (5,9).

This latest mechanism is known as SPM. SPM pathogenesis was first revealed by Macklin (10). The air leak, which is caused by the rupture, reaches hilum and the mediastinum via the interstitium and bronchovascular tissue (Macklin effect) (11). The air can reach thorough submandibuler and retropharengeal gaps between tissue planes, also it can reach diaphragm and retroperitoneum with periaortic and periesophageal planes.

Although the incidence is not completely known, different ratios have been reported in different series. Newcomb and Clarke (12) reported this ratio as 1/29670 in their series. It was 1/32896 in the study conducted by Igor et al (13) and 1/42000 in the study conducted by Bodey (14). Recurrence is rare in SPM. The first recurrence was reported by Yellin et al. (15).

Table 1. Details of 12 patients with SPM

Patients	Gender	Age	Compliant	Radiology (emphysema) X-Ray	CT	Background	Endoscopy (FOB,Esophgoscopy)	WBC	N/L	Hospitalization Time	Mortality
1	F	37	D,CP	+	+	Asthma	-	14130	1.83	5	-
2	M	28	C,CP	-	+	Cigarette	-	12100	2.1	3	-
3	M	33	D,S	+	+	Cigarette	+	9610	2.57	6	-
4	M	21	D,CP	-	+	Cigarette	+	18500	4.67	4	-
5	M	27	S	+	+	-	-	10960	2.9	10	-
6	*M*	*32*	C,V	-	+	Cigarette	-	7310	2.35	3	-
7	F	15	D,CP	-	+	-	-	8480	4.83	6	-
8	F	42	TP,CP	-	+	Exodontia	-	11710	2.36	7	-
9	M	38	TP,CP	-	+	Cigarette	-	5810	3.13	8	-
10	M	41	TP,CP	-	+	Cigarette	-	7260	3.83	4	-
11	M	21	D,S	-	+	Opioid	-	10230	7.04	4	-
12	M	20	S	-	+	Exodontia	-	6345	3.8	2	-

D: Dyspnea, CP: Chest Pain, S: Swelling, C: Cough, V: Vomit, TP: Throat Pain, Cigarette: Smoking

Several factors (vomiting, coughing, defecation, birth, medication use, Marfan syndrome, ethanol intoxication, bonding, woodwind and bronchospasm) have been reported in the literature that have triggered and predisposing SPM (Table 2). In most of the publications, there is consensus that most patients with SPM do not have a triggering factor. When the sources used were listed, the ratio of predisposing factors was found to be 39-100%. When all publications were evaluated, predisposing factor was defined in 32% of patients with SPM (16). In our series, there were predisposing and triggering factors in the ratio of 83.3% (smoking in 6, 1 opioid drug using in 1, asthma in 1 and exodontia in 2 patients).

In SPM, chest pain, sore throat, vomiting, dyspnea, abdominal pain, subcutaneous emphysema and hypotension are significant clinical symptoms. Chest pain, dyspnea, neck pain and discomfort are the most frequently reported symptoms. Chest pain is the most commonly reported symtom in pleuritic type, retrosternal expanding through the back and shoulders (3,5,10,17). Chest pain (58.3%) and dyspnea (41.6%) were the most common symptoms in our patients.

Patients with SPM are usually hemodynamically stable and have not respiratory distress.

One of the most frequent findings are subcutaneous emphysema reported 40-100% (18,19). Some authorities consider subcutaneous emphysema as a good clinical symptom, because the escape of air to the subcutaneous site prevents pressure on the large vessels by preventing the increase in pressure in the mediastinum. In ausculta tion, the sounds of crackles or bubbles heard in each shot of the heart are called the Hamman sign. Hamman sign is seen less frequently than subcutaneous emphysema in studies. Also in Boerhaave's syndrome, because of similar predisposing factors (coughing, straining or vomiting) can be seen, it should be considered in the diagnosis. If accompanied by Boerhaave's syndrome, tachypnea, tachycardia, hydrothorax and fever can be seen.

In the differential diagnosis, acute coronary syndrome, pulmonary embolism, pericarditis, pneumothorax and tracheobronchial rupture should be considered.

SPM is an acute disease and the patient's admission to the hospital is between 45 minutes and 17 hours with the begining of symptoms. The diagnosis is confirmed by posteroanterior and lateral chest radiography. Lateral radiography is necessary because in 50% of the undiagnosed cases a small hyperlucent air is seen in the retrosternal region (20). In the SPM, there may be many radiologic findings such as hyperlucent band parallel to the left cardiac silhouette, elevation of mediastinal pleura, hyperlucent line extending from mediasten to neck, especially air in mediastinal structures such as aorta, trekea, esophagus and thymus gland.

Three patients had chest radiography subcutaneous emphysema, while all patients had mediastinal emphysema in thorax CT. Pneumomediastinum subcutaneous emphysema with pneumothorax and pleural effusion may be seen on chest radiography when accompanied by Boerhaave's Syndrome. Kaneki and his friends (7) reported that 30% of chest radiographs were normal in patients

with SPM. Chest radiography revealed pathology in 16.6% of our patients. Chest tomography is considered as the gold standard in the diagnosis of SPM and small amount of air or Macklin effect can be detected. Eliminate to Boerhaave Syndrome, contrast thoracic CT or contrast esophagography may be performed. Water-soluble substances should be used in esophagography. As barium is more opaque, it can cause inflammatory changes, leading to mediastinal fibrosis. Some authorities find endoscopic examinations risky. Bronchoscopy can be performed if there is a suspicion of tracheobronchial rupture (9). We performed endoscopy in 2 patients with subcutaneous emphysema, and could not detect any pathology. WBC and C-Reactive Protein (CRP) can be checked if an inflammatory cause is considered. In our series, a WBC 5800-18500 (mean 10203), neutrophil / lymphocyte (N / L) ratios were in the range of 1.83-7.02 (mean 3.45).

Table 2. Triggering and Predisposing factors for SPM.

Triggering Factors	Predisposing Factors
Emesis, Asthma	Smoking
Cough, Physical activity	Asthma
Choking, Defecation	Idiopathic pulmonary disease
Vomiting, Inhaled drugs	Chronic obstructive pulmonary
Crying, Unknown maneuvers	disease

Treatment, except in severe cases, is usually rest, analgesic, oxygen and bronchodilators. We have applied rest, analgesic, oxygen and bronchodilators treatment to our patients. The pure oxygen treatment increases the diffusion pressure of nitrogen in the interstitium and promotes rapid absorption of the free air.

Antibiotics are used if mediastinitis is suspected (21,22). Some previous studies recommend prophylactic antibiotics for preventing mediastinitis (5). We used prophylactic antibiotics to prevent mediastnitis Patients respond well to this treatment: clinical manifestations resolve and radiographic signs of the condition diminish. Patients should be hospitalized for a minimum of 24 hours to avoid potential complications (23). Different hospitalization times were reported between 24 hours and 6.3 ± 3.9 days in different reports (13,24). In our series, the patients' duration of hospital stay was 5.16 (range 3-10). Reported the low incidence of recurrence, outpatient follow-up is not necessary.

If there is an excessive subcutaneous emphysema, small skin incisions and subcutaneous drainage catheters can be placed (25). Malignant pneumomediastinum aims to reduce the high pressure in the alveoli by evacuating the air in the mediastinum with the use of multiple subcutaneous aspirations or incisions. Cervical mediastinotomy rarely may be necessary. We did not perform any drainage procedure for subcutaneous emphysema.

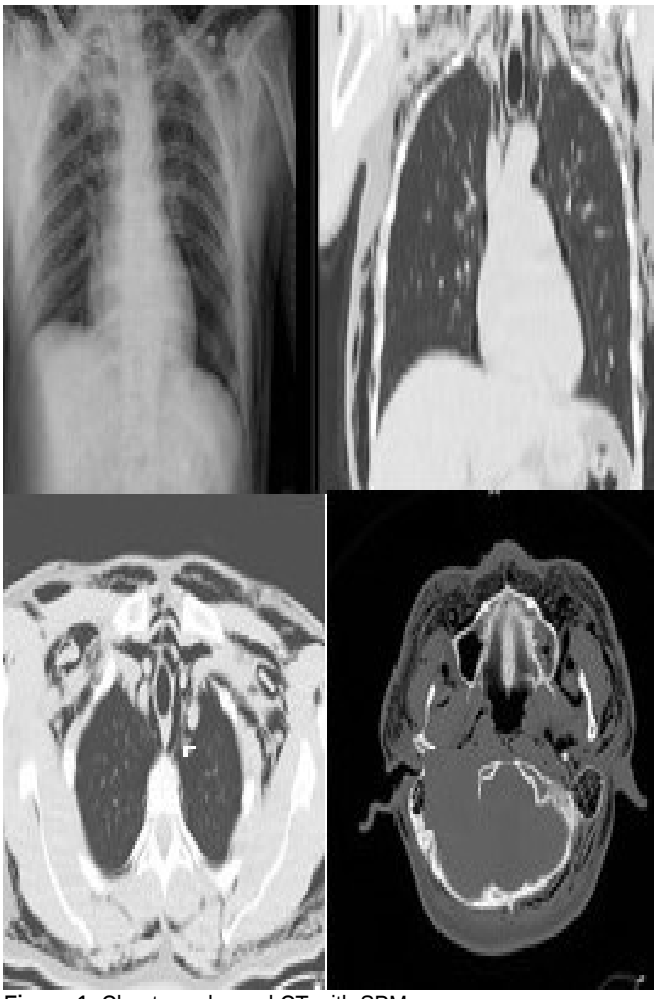

Figure 1. Chest graphy and CT with SPM

Should a large pneumothorax produce respiratory embarrassment or the lung be collapsed by more than 50% aspiration and underwater drainage or suction are necessary.

In our series, 1 patient had a pneumothorax, however, the treatment was not at the level of treatment, regressed with medical treatment.

For the evaluation of the large vessels, esophagus, lung and trachea occasionally video-assisted thoracoscopic surgery, even thoracotomy may be required. In our series, none of the patients underwent major surgical intervention.

Conclusion

SPM is a disease that is seen rarely in emergencies, and should be treated. Conservative treatment and follow-up are usually sufficient. But sometimes life-threatening complications may develop. Other disorders in differential diagnosis should be eliminated in SPM. Unlike the Boerhaave's Syndrome, esophageal rupture should be kept in mind. FOB, upper gastrointestinal CT with oral contrast and esophagoscopy have a very important role in the diagnosis in order to detect the tracheobronchial and esophageal injuries.

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