

Recurrent spontaneous pneumomediastinum and pneumorrhachis accompanied by Raynaud's phenomenon

Raynaud fenomeninin eşlik ettiği tekrarlayıcı spontan pnömomediastinum ve pnömoraşi

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Pneumorrhachis is characterized by the air in the spinal epidural space. In this article, we report a 21-year-old male case with Raynaud's phenomenon and recurrent spontaneous pneumomediastinum episodes accompanied by subcutaneous emphysema who was radiographically diagnosed with pneumomediastinum accompanied by pneumorrhachis during his follow-up visits.

Key words: Pneumorrhachis; Raynaud's phenomenon; recurrent spontaneous pneumomediastinum.

Pneumomediastinum (PM) is characterized by the presence of air in the mediastinal area while pneumorrhachis (PR) is a radiological condition with air in the spinal epidural space. The coexistence of these two conditions along with recurrent spontaneous PM have rarely been reported in the literature.^[1-3]

Raynaud's phenomenon is a vasospastic disorder featuring attacks that usually affect the upper limb arteries, but occasionally the lower limb arteries are involved. To our knowledge, the coexistence of PM, PR, and Raynaud's phenomenon has not been previously reported in the literature.

Herein, we present a male patient who was first diagnosed with Raynaud's phenomenon. We then observed the recurrent spontaneous PM attacks accompanied by subcutaneous emphysema, and he was diagnosed with PR radiologically at his follow-up visits.

Pnömoraşi, spinal epidural alanda hava bulunması ile karakterizedir. Bu yazıda, Raynaud fenomeni ve subkutan amfizemin eşlik ettiği tekrarlayıcı spontan pnömomediastinum atakları olan takip vizitleri sırasında radyolojik olarak pnömomediastinuma eşlik eden pnömoraşi tanısı konulan 21 yaşında bir erkek olgu sunuldu.

Anahtar sözcükler: Pnömoraşi; Raynaud fenomeni; tekrarlayıcı spontan pnömomediastinum.

CASE REPORT

A 21-year-old male patient was admitted to our cardiovascular surgery outpatient clinic with complaints of pain and bruising in both hands because of exposure to cold. Intensive smoking (two packs of cigarettes a day) was present in his medical history. Hyperemia was observed in both hands during a physical examination, and a triphasic changes in the colour of his hands triggered by cold exposure were also seen. The patient was diagnosed with Raynaud's phenomenon, but serological tests for the etiology of secondary Raynaud's phenomenon were normal. Treatment with vasodilators and calcium channel blockers was then initiated. Since smoking is one of the major causes of Raynaud's phenomenon and because the patient was an intensive smoker, he was educated on how to break his habit. At the follow-up visit 14 days later, subcutaneous emphysema in both dorsal forearms that was spreading to the thorax was



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Figure 1. The pneumomediastinum as revealed on chest X-ray.

observed. Therefore, the patient was given a chest X-ray due to the suspicion of pneumothorax, and PM was detected (Figure 1). He was then hospitalized in the thoracic surgery clinic where it was determined that his hemogram and routine biochemical tests were normal. Soon afterwards, the patient was discharged because of spontaneous regression. At the one-month follow-up visit after his discharge, the patient stated that he had not stopped smoking. Subcutaneous emphysema was observed, and PM and PR were

detected on chest radiography and chest computed tomography (CT) (Figure 2); thus, the patient was rehospitalized in the thoracic surgery clinic. The subcutaneous emphysema disappeared while he was being followed up at our facility, and spontaneous regression of the PM and PR was observed in his control CT scan (Figure 3).

DISCUSSION

Pneumomediastinum is classified as spontaneous PM when there is no clear primary source, whereas secondary PM develops due to a specific pathological event such as trauma or intrathoracic infection.

Spontaneous PM was first described by Louis Hamman in 1939.^[4] It is a benign condition that is usually seen in young adults that is caused by exposure to a sudden pressure change in the intrathoracic space.^[5] As a result of the sudden increase in intra-alveolar pressure caused by an acute cough or vomiting, spontaneous PM may arise. When alveolar rupture occurs, it moves along the lung interstitium and reaches the mediastinum through the perivascular spaces.^[3] Predisposing factors for spontaneous PM are smoking in 30% of patients and bronchial asthma in 15%, with coughing and heavy physical activity being less frequent causes. However, it can occur without any triggering event or abnormal finding on a chest radiograph. Patients who present with this clinical picture usually regress because of a lack of treatment.^[5] Common clinical symptoms are chest pain, dyspnea, coughing, and subcutaneous emphysema. Although mediastinal air is usually associated with subcutaneous emphysema,



Figure 2. The pneumomediastinum and pneumorrhachis as seen on chest computed tomography at the one-month follow-up visit after discharge.



Figure 3. Spontaneous regression of the pneumomediastinum and pneumorrhachis as shown on control computed tomography.

it may rarely be found in conjunction with a pneumopericardium or pneumoperitoneum.

In a comparative study by Caceres et al.^[5] that focused on secondary PM and spontaneous PM, they determined that spontaneous PM occurs more often in younger patients and that it has a lower mortality rate along with a shorter hospital stay. However, in contrast to spontaneous PM, secondary PM was associated more with the presence of subcutaneous air, as observed on CT, pneumothorax, pleural effusion, and chest tube insertion.^[5]

Pneumorrhachis, or epidural pneumatosis, is a rare radiological condition characterized by air in the spinal epidural area that is usually detected accidentally. The causes of PR can be classified as traumatic, non-traumatic, or iatrogenic. Pneumorrhachis is usually iatrogenic and occurs following the administration of epidural analgesia. However, there was no history of trauma or epidural analgesia in our case. Furthermore, PR may very rarely occur when mediastinal air passes through to the epidural space, which was the case with our patient. This is necessitated by the lack of real fascia barriers between the posterior mediastinum and epidural space.^[1] The coexistence of PM and PR has been reported in only a few case reports in the literature. In a review by Goh and Yeo,^[6] it was observed that seven out of 13 traumatic PR cases had occurred secondary to traumatic pneumothorax and PM. In addition, previous studies suggested that traumatic PR is primarily associated with subcutaneous emphysema, pneumothorax, and PM following minor trauma. However, to our knowledge, the coexistence of PM, PR, and Raynaud's phenomenon has not been previously reported.

Recurrent spontaneous PM has also been infrequently reported in the literature, with only six previous cases.^[7-12] In the Caceres study^[5] which compared secondary PM with spontaneous PM, no recurrence was observed in the patients who were followed up for between one and 10 years.^[5] We believe that our patient represents the seventh reported case of recurrent spontaneous PM and the first case accompanied by PR in the literature.

The presence of Raynaud's phenomenon in our patient was another unique feature of our patient. Although no cases involving the coexistence of Raynaud's phenomenon and spontaneous PM have been reported in the literature, there was a case report that described the association of spontaneous pneumoperitoneum with Raynaud's phenomenon.^[13] Their patient was similar to ours in that they were both

young, but contrary to our case, their case involved a follow-up patient with Raynaud's phenomenon. Our case was admitted to our hospital with only symptoms of Raynaud's phenomenon. Furthermore, no increase in white blood cells was observed in the patient in the literature, whereas this was true in our case. That case was also similar to ours in terms of the absence of any etiological cause and the presence of spontaneous recovery.

Whether or not a relationship exists between spontaneous PM and Raynaud's phenomenon remains controversial. Since smoking is one of the most important factors in the etiology of both Raynaud's phenomenon and recurrent spontaneous PM, we believe that intensive smoking might have been responsible for the coexistence of these two conditions in our patient.

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