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Case report
Prikaz slučaja
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SPONTANEOUS PNEUMOMEDIASTINUM WITH PNEUMOTHORAX, PNEUMORRHACHIS AND PNEUMOPERITONEUM IN A CHILD – A CASE REPORT

SPONTANI PNEUMOMEDIJASTINUM SA PNEUMOTORAKSOM, PNEUMORAHISOM I PNEUMOPERITONEUMOM KOD DETETA – PRIKAZ SLUČAJA

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Summary

Introduction. Pneumomediastinum is defined as the presence of free air in the mediastinum. Primary, idiopathic, spontaneous pneumomediastinum is very rare and it affects healthy children with no identifiable cause. Secondary pneumomediastinum may be caused by underlying respiratory disorders, iatrogenic causes or trauma. The most common clinical sign of pneumomediastinum is subcutaneous emphysema, and the most common symptoms are acute chest pain and dyspnea. The diagnosis is confirmed by a chest X-ray or chest computed tomography. Pneumomediastinum is rarely associated with pneumothorax, pneumoperitoneum, and pneumorrhachis. **Case Report.** In this report, we present a case of a spontaneous pneumomediastinum in a child aged 2 years and 6 months. A child was admitted to our hospital due to massive subcutaneous emphysema. On admission, the patient was without a history of chest trauma or any chronic respiratory tract diseases. He had a mild upper respiratory tract infection 6 days before admission. The diagnosis of pneumomediastinum was confirmed by chest X-ray and computed tomography. After conservative treatment, on the eighth day of hospitalization, there was a complete regression of the pneumomediastinum with normalization of the clinical and radiological findings. **Conclusion.** Spontaneous pneumomediastinum is the most common benign condition that spontaneously regresses after conservative treatment. Life-threatening complications require surgical decompression. The use of antibiotic therapy in the prophylaxis of mediastinitis has not been proven to be useful. Opinions on the routine use of chest computed tomography in patients with spontaneous pneumomediastinum are still not uniform.

Key words: Mediastinal Emphysema; Pneumothorax; Pneumoperitoneum; Pneumorrhachis; Child; Signs and Symptoms; Radiology

Sažetak

Uvod. Pneumomedijastinum se definiše kao prisustvo slobodnog vazduha u medijastinumu. Primarni, idiopatski, spontani pneumomedijastinum se javlja jako retko kod zdrave dece, bez poznatog uzroka. Sekundarni pneumomedijastinum može biti uzrokovan bolestima respiratornog trakta, jatrogenim faktorima ili traumom. Najčešći klinički znak pneumomedijastinuma je supkutani emfizem, a najčešći simptomi su akutni bol u grudima i dispnea. Dijagnoza se potvrđuje na osnovu rendgenskog snimka grudnog koša ili kompjuterizovane tomografije grudnog koša. Pneumomedijastinum je retko udružen sa pneumotoraksom, pneumoperitoneumom i pneumorahisom. **Prikaz slučaja.** Opisan je slučaj pneumomedijastinuma kod deteta uzrasta dve godine i šest meseci. Dete je primljeno u našu bolnicu zbog masivnog supkutanog emfizema. Nije imalo traumu grudnog koša niti hronične bolesti respiratornog trakta. Dete je imalo blagu infekciju gornjih respiratornih puteva šest dana pre prijema u bolnicu. Dijagnoza pneumomedijastinuma potvrđena je radiografijom i kompjuterizovanom tomografijom grudnog koša. Nakon primene konzervativnog tretmana osmog dana hospitalizacije došlo je do potpune regresije pneumomedijastinuma uz normalizaciju kliničkog i radiološkog nalaza. **Zaključak.** Spontani pneumomedijastinum je najčešće benigno stanje koje spontano regredira nakon konzervativnog tretmana. Životno ugrožavajuće komplikacije zahtevaju hiruršku dekompresiju. Nije dokazana korist od primene antibiotske terapije u profilaksi medijastinitisa. Stavovi o rutinskoj primeni kompjuterizovane tomografije grudnog koša kod pacijenata sa spontanim pneumomedijastinumom još uvek nisu unifromni.

Ključne reči: pneumomedijastinum; pneumotoraks; pneumoperitoneum; pneumorahis; dete; znaci i simptomi; radiologija

Introduction

Pneumomediastinum, defined as the presence of free air in the mediastinum, is a rare condition in children [1]. Primary, idiopathic, spontaneous pneumomediastinum occurs most often in healthy children with no identifiable cause. On the other hand, secondary pneumomediastinum may be caused by disorders of the respiratory tract (such as asthma, infections of the respiratory tract, interstitial lung disease, foreign body aspiration, etc.), iatrogenic causes (such as mechanical ventilation, endoscopy, intubation, central venous access procedures, thoracostomy, and chest or abdominal surgery) or trauma (esophageal rupture, bronchial injury) [1–3].

Rarely, pneumomediastinum may be associated with pneumothorax, pneumorrhachis, and pneumoperitoneum. Pneumothorax is defined as free air in the pleural space and can be spontaneous or traumatic. Pneumorrhachis is the presence of free air in the spinal

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Abbreviations
CRP – C-reactive protein
CT – computed tomography

canal and can be primary (spontaneous) or secondary. Spontaneous pneumorrhachis is usually asymptomatic and extra-dural, while secondary pneumorrhachis is most often intra-dural and symptomatic [4, 5].

The most common clinical manifestations of spontaneous pneumomediastinum are subcutaneous emphysema, acute chest pain, and dyspnea [6, 7]. The medical history, clinical findings, and radiological imaging, chest X-ray and chest computed tomography (CT) are sufficient to confirm the diagnosis of the pneumomediastinum. In rare cases, such as suspected foreign body aspiration or esophageal rupture, endoscopic evaluation should also be considered [6–8].

In most patients, spontaneous pneumomediastinum is a benign condition with a high rate of complete regression after conservative treatment. Rarely, pneumomediastinum may be a life-threatening condition that requires immediate diagnosis and surgical treatment [8, 9].

Case Report

A male child aged 2 years and 6 months presented with nasal secretion and dry cough six days before admission to our hospital. The child had a fever for two days. On admission, facial and neck swelling was observed, without dyspnea or any other symptom. Medical history ruled out the possibility of chest trauma. Apart from the delay in speech development, he was a healthy child without any respiratory diseases.

On admission, the boy was afebrile, his respiratory rate was normal and oxygen saturation was 97% on room air. His cheeks, neck, and upper part of the thoracic wall were extremely swollen, palpation of these body parts revealed crepitations. Chest auscultation was normal, as well as the rest of physical findings. The neurological status was also normal. The C-reactive protein (CRP) was 9.33 mg/L, with normal blood count, normal blood gas exchange, and regular biochemical findings. The alpha-1 antitrypsin level was in the reference range. Blood cultures and serological tests for viruses were negative. A chest X-ray was done and revealed subcutaneous emphysema and pneumomediastinum **(Figures 1 and 2)**. Chest CT confirmed pneumomediastinum associated with massive diffuse subcutaneous emphysema affecting the head, neck and chest walls **(Figures 3, 4 and 5).** There was a small amount of free air in the upper retroperitoneum, upper anterior abdomen (pneumoperitoneum), and spinal canal (pneumorrhachis) **(Figure 6).** A smaller partial pneumothorax in the apex of the left lung was also confirmed **(Figure 4).** Signs of interstitial emphysema have been described on both sides of the lungs, mostly in the left apical and lingular segment (described as the Macklin effect), without any other lung parenchymal disorders. The electrocardiogram and echocardiographic findings were normal.

The boy was examined by a pulmonologist and a pediatric thoracic surgeon. He was treated conservatively with monitoring of his vital signs during the

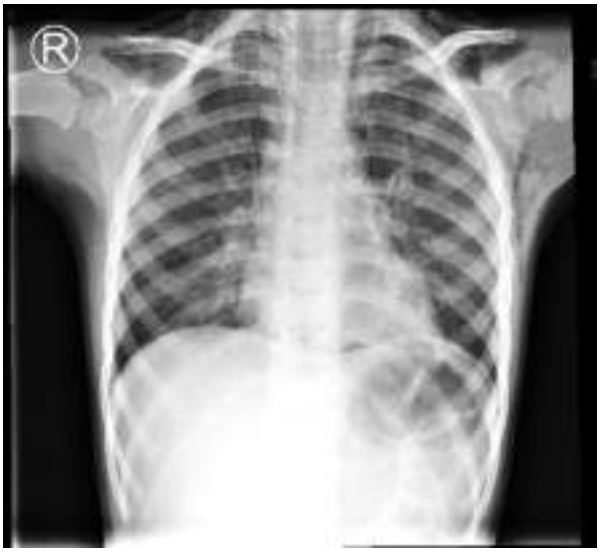

Figure 1. Anteroposterior chest X-ray showing pneumomediastinum and subcutaneous emphysema
Slika 1. *Rendgenografija grudnog koša u anteroposteriornoj projekciji: pneumomedijastinum i supkutani emfizem*

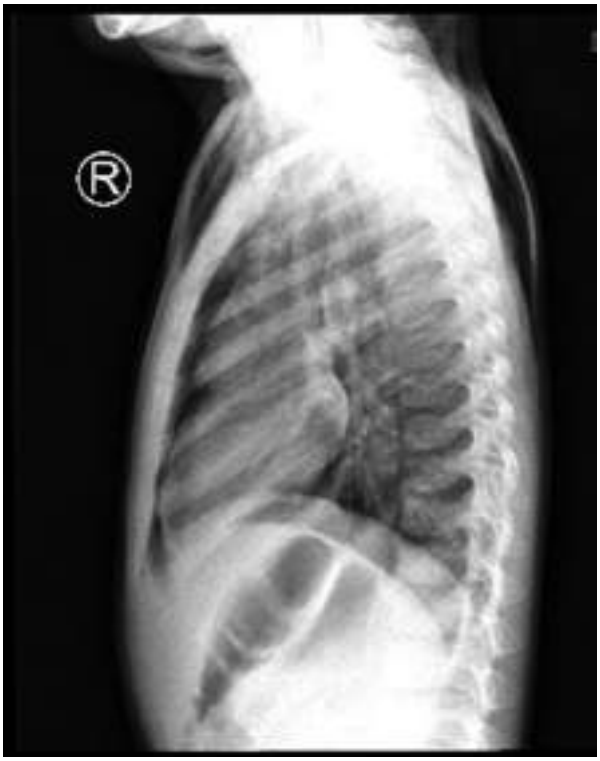

Figure 2. Lateral neck X-ray shows subcutaneous emphysema of the neck soft tissues. Lateral chest X-ray confirmed pneumomediastinum and chest subcutaneous emphysema
Slika 2. *Rendgenografija vrata u lateralnoj projekciji: subkutani emfizem mekih tkiva vrata. Rendgenografija grudnog koša u lateralnoj projekciji: pneumomedijastinum i supkutani emfizem grudnog koša*

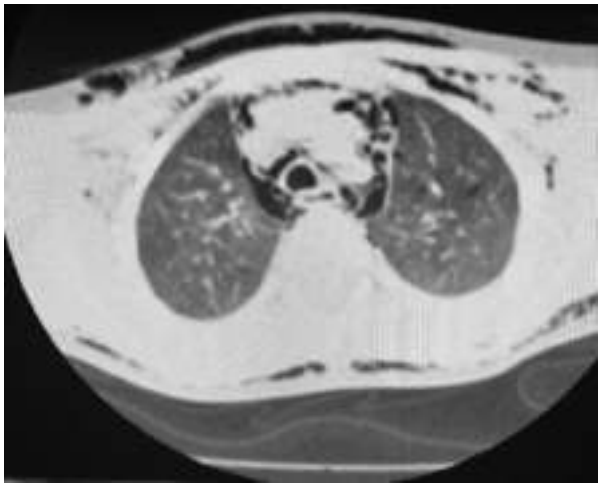

Figure 3. Chest CT shows pneumomediastinum and subcutaneous emphysema
Slika 3. *Kompjuterizovana tomografija grudnog koša: pneumomedijastinum i supkutani emfizem*

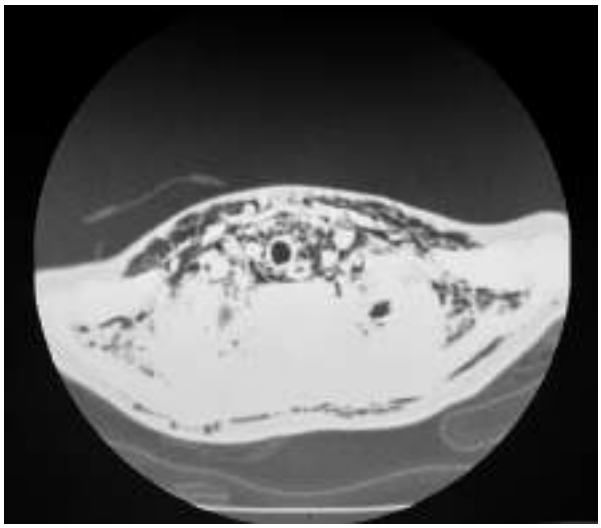

Figure 4. CT of the upper part of the chest shows chest wall emphysema and a smaller partial pneumothorax in the apex of the left lung
Slika 4. *Kompjuterizovana tomografija gornjih partija grudnog koša: emfizem struktura torakalnog zida i mali parcijalni pneumotoraks u predelu apeksa levog plućnog krila*

follow-up. Empirical intravenous antibiotic therapy was introduced, but it was discontinued when negative blood cultures were obtained, the child was afebrile without any signs of infection and the CRP values decreased. On the eighth day of hospitalization a complete spontaneous regression of the subcutaneous emphysema and pneumomediastinum was observed, with normalization of the clinical and radiological findings.

Discussion

Spontaneous pneumomediastinum in children is a rare and mostly benign clinical entity. It is defined as

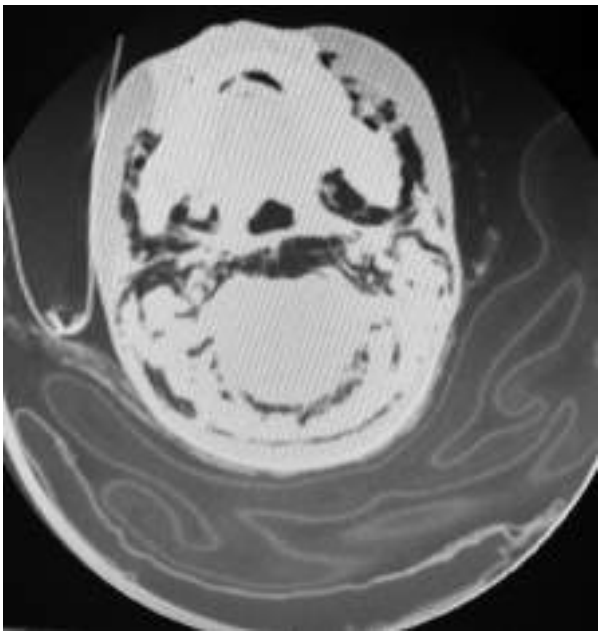

Figure 5. CT of the head (suprahyoid neck region) shows subcutaneous and deep neck emphysema
Slika 5. *Kompjuterizovana tomografija glave (suprahioidni region vrata): supkutani emfizem i emfizem dubokih struktura suprahioidnog regiona vrata*

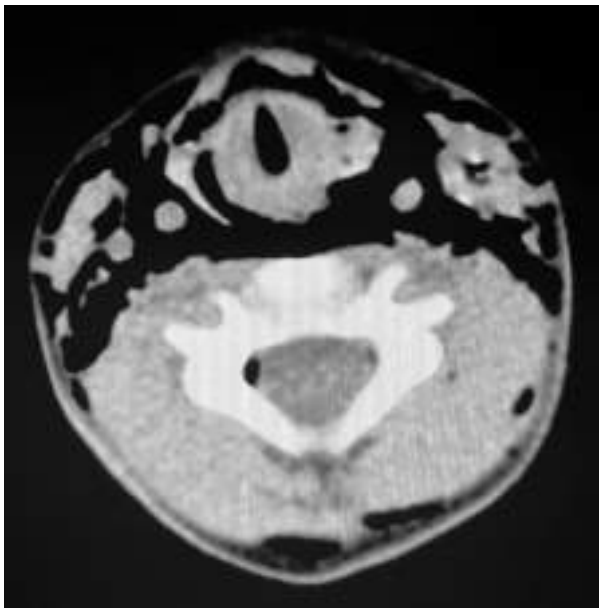

Figure 6. CT of the infrahyoid neck shows pneumorrhachis
Slika 6. *Kompjuterizovana tomografija infrahioidnog dela vrata: pneumorahis*

the presence of free air in the mediastinal space in the absence of previous respiratory disease, chest trauma, or iatrogenic causes [1–3]. Primary spontaneous pneumomediastinum affects healthy children. Voluntary alterations in the breathing pattern (playing a wind instrument, pulmonary function tests, smoking of cannabis) or involuntary breathing pattern alterations (in-

tense physical activity, coughing and vomiting) are described as possible triggers. Secondary pneumomediastinum develops in children with certain respiratory diseases, iatrogenic causes or trauma. In the case of pneumomediastinum, it is very important to rule out predisposing conditions that would require immediate specific treatment [3, 6–9]. Our patient was without a history of trauma and without any respiratory disease at the time of admission, so the diagnosis of primary spontaneous pneumomediastinum was established. Pneumomediastinum presented 6 days after a mild acute upper respiratory tract infection.

Pneumomediastinum occurs as a result of increased intra-alveolar pressure, which causes alveolar rupture. The alveolar free air then enters the mediastinum through the peribronchial and perivascular lung spaces [9, 10]. Since there are no fascial barriers surrounding the posterior mediastinum, mediastinal air may spread into the spinal canal (pneumorrhachis), soft tissues (subcutaneous emphysema), pericardium (pneumopericardium), and peritoneum (pneumoperitoneum) [4]. The neck and mediastinum communicate with the peritoneal cavity and retroperitoneum through a common visceral space that inverts the esophagus and trachea and follows the esophagus through the diaphragmatic hiatus. This can explain the airflow from the mediastinum into the peritoneum [5]. Our patient had pneumomediastinum associated with a smaller partial pneumothorax, pneumorrhachis and pneumoperitoneum.

Epidemiological data on pneumomediastinum in children are not precisely determined, but it is evident that pneumomediastinum is an extremely rare condition in pediatric population [3]. The incidence of spontaneous pneumomediastinum in children under the age of 18 is 1/8000 to 1/15000, with the first peak at the age of 6 months to 4 years, and the second peak between the age of 15 and 18 years. The incidence of neonatal pneumomediastinum is 1.7 - 2.5/1000 [1, 11]. Our patient was 2 years and 6 months old, being classified in the first peak age group. There is no available data in the literature about the incidence of the association of pneumomediastinum, pneumothorax, pneumoperitoneum, and pneumorrhachis. To the best of our knowledge, this is the first case report that describes this association in children.

The clinical manifestations of pneumomediastinum are determined by the amount of free air in the mediastinum and by the underlying condition. Although symptoms of spontaneous pneumomediastinum may be mild, secondary pneumomediastinum may be potentially lethal, depending on the underlying disease [3]. The most common clinical presentations include facial and neck swelling as a sign of subcutaneous emphysema, acute chest pain, and dyspnea. The chest pain is usually retrosternal radiating to the back, shoulders, and arms. It is more common in patients with primary spontaneous pneumomediastinum than in patients with secondary pneumomediastinum. Other symptoms and signs include cough, shortness of breath, speech difficulties, cyanosis, dysphagia, and wheezing [9–11]. At the time of admission, our patient had facial and neck swelling, without any other symptoms. Hamman's sign is characteristic in pneumomediastinum with subcutaneous emphysema, but it is rare (11.6%). It represents the crepitations detected by cardiac auscultation [1, 8]. In our patient, palpation of the cheeks, neck, and chest revealed crepitation, while the auscultatory finding on the lungs was normal. Hamman's sign was not recorded.

Radiological evaluation is required to confirm the diagnosis of pneumomediastinum. Chest X-ray is the basic diagnostic tool, while lateral neck radiography is more sensitive for free air detection in the soft neck tissues [12–14]. On chest X-ray, pneumomediastinum presents with lucent, vertical streaks or bubbles of gas within the mediastinum, which also can extend into the neck or chest wall. The characteristic radiological signs are the ring sign (free air surrounds the pulmonary artery), thymic sail sign (free air causes thymus elevation), continuous diaphragm sign (the superior surface of the diaphragm is surrounded by mediastinal air), and double bronchial wall sign (both sides of the bronchial wall are visible due to presence of air in the mediastinum) [3, 10]. In our patient, the frontal and lateral chest X-ray and lateral neck radiography confirmed the diagnosis of pneumomediastinum with subcutaneous emphysema of the neck and chest walls.

There are still different attitudes towards the routine use of chest CT in patients with pneumomediastinum. Some recent studies support the use of the chest CT to determine the possible underlying cause of pneumomediastinum and to assess the amount of free air in the mediastinum and surrounding spaces [10, 13, 15]. Furthermore, recent data show that about 30% of children with pneumomediastinum have a normal chest X-ray [3]. Other studies do not recommend routine use of additional radiological tools due to radiation exposure [2]. In our patient, chest CT was performed due to prominent clinical signs of subcutaneous emphysema. The pneumomediastinum with diffuse massive subcutaneous emphysema on the head, neck and chest walls was described.

In 1944, Macklin and Macklin first observed that released air from an alveolar rupture centripetally dissects through the pulmonary interstitium along the bronchovascular sheaths. Then the released alveolar air is directed to the pulmonary hila and into the mediastinum. The Macklin effect appears on chest CT as linear collection of free air in the mediastinum contiguous to the bronchovascular sheaths [16], as it was the case in our patient.

Pneumomediastinum is very rarely associated with pneumothorax, pneumorrhachis, pneumopericardium, and pneumoperitoneum [13–15]. In our patient, a smaller partial pneumothorax in the apex of the left lung was detected on CT, as well as interstitial emphysema. Also, a small amount of free air in the spinal canal and upper part of the retroperitoneum was described. Pneumopericardium was not apparent. Pneumomediastinum was probably caused by a small partial pneumothorax.

Severe complications, such as respiratory distress and cardiac disorders, are caused by airway and heart compression by a large amount of free air in the me-

diastinum. Massive pneumothorax or pneumopericardium are life-threatening conditions and require surgical decompression (thoracic drainage or video-assisted thoracoscopy). Gas embolism and laryngeal compression are also described [13, 14, 17].

The majority of patients with primary spontaneous pneumomediastinum should be treated conservatively (analgesics, monitoring, bed rest, avoiding triggering factors such as Valsalva maneuver, physical activity, and vomiting). Complete spontaneous regression is expected during 3 to 15 days [3, 12–15], which was the case in our patient. Physical activity should not be strictly limited after recovery [3]. The treatment of the secondary pneumomediastinum is determined by the underlying disease [12–15].

The use of antibiotics in the treatment of pneumomediastinum is controversial. Although antibiotics may be used to prevent mediastinitis and soft tissue infections of the neck, recent research has not proven the benefit of prophylactic antibiotic use [8, 12, 13]. However, antibiotics are indicated in patients with confirmed infection of the respiratory tract, bronchial tree trauma, or esophageal trauma [3]. In our patient, antibiotic therapy was initiated empirically, but it was discontinued in the further course.

Patients with spontaneous pneumomediastinum without severe complications have a good prognosis, high survival rate, and minimal risk of recurrence [11–15].

Conclusion

Spontaneous pneumomediastinum, pneumothorax, pneumorrhachis, and pneumoperitoneum are usually benign but potentially life-threatening conditions in children. In the majority of the patients, conservative treatment results in complete spontaneous regression. Life-threatening complications require immediate surgical decompression. Antibiotic therapy in mediastinitis prophylaxis has not been proven to be useful in recent studies. The opinions on the routine use of chest computed tomography in patients with spontaneous pneumomediastinum are still not uniform. Recent studies suggest that patients without complications of spontaneous pneumomediastinum should not be exposed to radiation, but more evidence is needed to confirm this assumption.

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