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Case Report

Pneumorrhachis in children: A report of two cases and review of the literature

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ABSTRACT

Pneumorrhachis refers to the clinical presentation of air within the spinal canal, and it is rarely associated with pneumomediastinum, particularly in young children. Pneumorrhachis associated with pneumomediastinum is generally asymptomatic. Here we report 2 unusual cases involving very young children with pneumorrhachis secondary to pneumomediastinum and present a review of the relevant literature. Case 1 involved a 4-year-old girl who presented with wheezing, violent coughing, and dyspnea associated with bronchiolitis. Case 2 involved a 3-year-old boy who presented with wheezing, violent coughing, and dyspnea associated with interstitial pneumonia possibly caused by graft-versus-host disease with human herpesvirus 6 infection after allogeneic hematopoietic stem cell transplantation. In both cases, pneumorrhachis improved with oxygen inhalation therapy and treatment of the underlying disease. Pneumorrhachis is rarely associated with neurological problems; however, decompressive laminectomy may be indicated to relieve the air block. Because pneumorrhachis is rare in children and neurological sequelae may be difficult to identify, close clinical, and radiographic observations are necessary. Plain radiography is not sufficient, and computed tomography should be performed to rule out intraspinal air.

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Introduction

Pneumorrhachis is characterized by the presence of air in the epidural space. It is most commonly associated with iatrogenic or traumatic etiologies, including epidural anesthesia, lumbar puncture, spinal surgery, spinal injury, and traumatic pneumothorax [1]. Although pneumorrhachis is rarely associated with pneumomediastinum complicated by bronchial asthma and violent cough [2,3], Tsuji et al. first reported cases of pneumorrhachis associated with asthma and/or violent coughing [4]. In such cases, the peripheral pulmonary alveoli may rupture because of a sudden increase in the intraalveolar pressure. Subsequently, the air present in the pulmonary perivascular interstitium may migrate along the fascial planes and move from the posterior mediastinum or retropharyngeal space to the epidural space through the neural foramina. There are no fascial barriers to block connections between the posterior mediastinum or retropharyngeal space and the epidural space [4-6].

There are few reports of young children with pneumorrhachis secondary to pneumomediastinum. Here we describe 2 unusual cases involving very young children and present a review of the relevant literature.

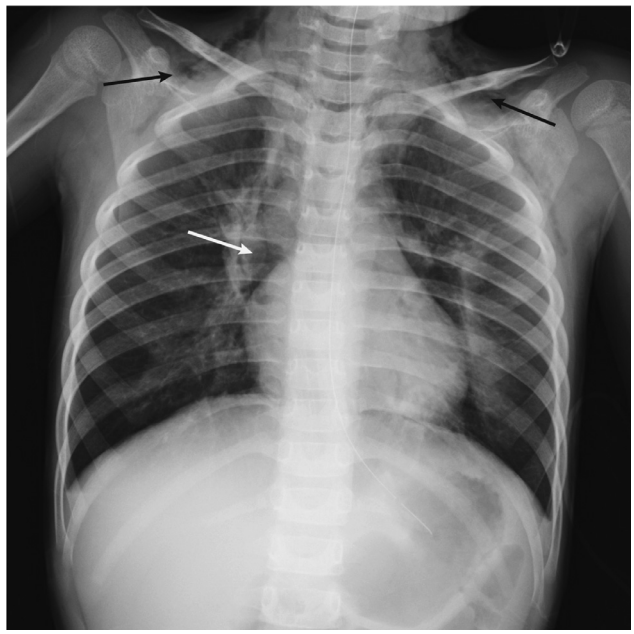

Fig. 1 – Chest radiography findings at admission of a 4-year-old girl with pneumorrhachis. Subcutaneous emphysema (black arrows) and pneumomediastinum (white arrow) can be observed.

Case Presentation

Case 1

A 4-year-old girl presented with wheezing, violent coughing, and dyspnea since 3 days. She had initially consulted the community hospital for her symptoms. She had a history of viral-induced wheezing, which was treated with bronchodilators, montelukast sodium, and dexamethasone. However, her symptoms did not improve, and physical examination indicated subcutaneous emphysema in the chest and neck. Chest radiography showed subcutaneous emphysema and pneumomediastinum (Fig. 1), while computed tomography (CT) further indicated pneumorrhachis (Fig. 2). She was transferred to our institute and admitted to the pediatric intensive care unit. Her respiratory rate was 40/min, while the oxygen saturation (SO_2) was 100% in pulse oximetry with oxygen delivery at 4 L/min through a mask. Physical examination revealed subcutaneous emphysema in the chest and neck with retractive breathing. Auscultation revealed decreased inspiratory sounds and bilateral wheezing. Her general and neurological conditions were normal. Arterial blood gas analysis revealed the following: pH, 7.355; partial pressure of carbon dioxide, 30.5 mmHg; partial pressure of oxygen, 148 mmHg; SO_2, 98.6%; and bicarbonate (HCO_3^-), 17 mEq/L. A blood test revealed a white blood cell count of 14600/μL (neutrophils: 91%) and C-reactive protein level of 4.3 mg/dL. Other biochemical parameters showed no abnormalities. She was treated with broad-spectrum antibiotics, inhalation bronchodilators, systemic corticosteroids, and oxygen inhalation therapy. We also performed polymerase chain reaction for a throat swab specimen, whole blood, and serum. The throat swab specimen was positive for adenovirus (Ct value: 37.371). Her condition improved and she was shifted from pediatric intensive care unit on the second day of admission. A respiratory function test was not performed because of her age. She was discharged from the hospital on day 10 of hospitalization and prescribed inhaled corticosteroid treatment because of the history of recurrent bronchiolitis.

Case 2

A 3-year-old boy with refractory juvenile myelomonocytic leukemia received chemotherapy and 2 rounds of allogeneic stem cell transplantation. However, his condition relapsed, and he received HLA-haploidentical bone marrow transplantation. At 3 months after the final transplant procedure, he presented with tachypnea, violent coughing, and wheezing. Chest radiography revealed bilateral ground-glass opacities, subcutaneous emphysema, and pneumomediastinum (Fig. 3). These findings were confirmed on chest CT, which also demonstrated cervical epidural air (Fig. 4). There were no neurological abnormalities. A blood test revealed a white blood cell count of 8800/μL (neutrophils: 60%) and C-reactive protein level of 0.15 mg/dL. Aspartate aminotransferase (normal range: 24–43 IU/L) and alanine aminotransferase (9–30 IU/L) levels were 45 and 63 IU/L, respectively. The serum level of KL-6, a marker for interstitial pneumonia, was elevated to 1470 U/mL (normal: <500 U/mL). *Candida* and *Aspergillus* antigens were absent. Polymerase chain reaction for a throat swab specimen, whole blood, and serum revealed human herpesvirus 6 (HHV6) infection (1.2×10^7 copies/mL, 2.6×10^9 copies/mL, and 4.0×10^6 copies/mL, respectively). The patient was diagnosed with interstitial pneumonia possibly caused

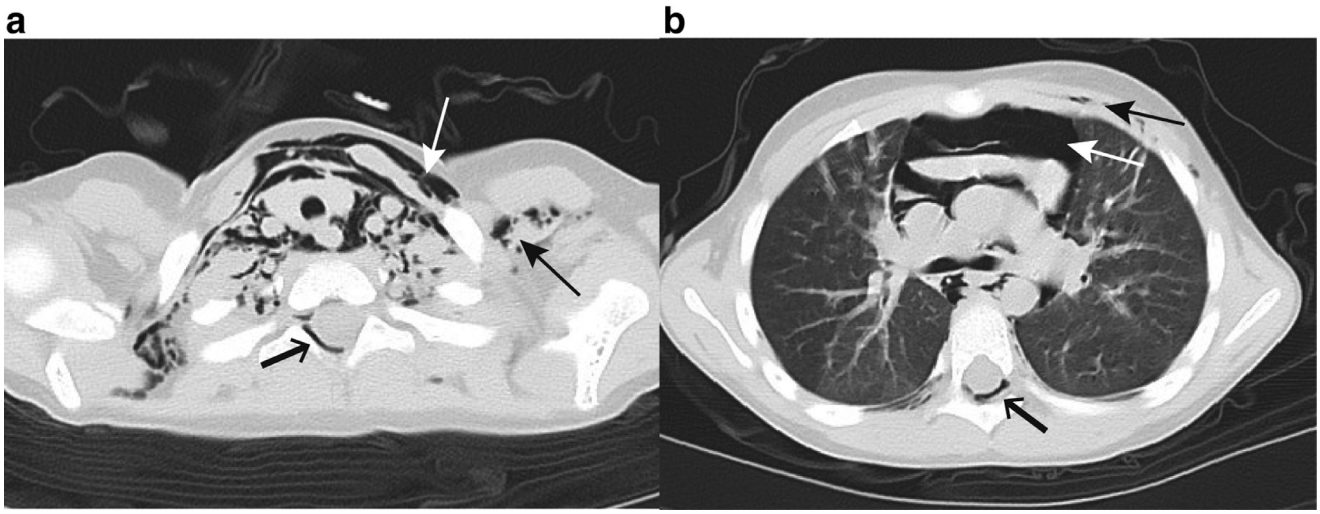

Fig. 2 – Computed tomography findings at admission of a 4-year-old girl with pneumorrhachis.
(A) Axial reconstruction of the cervical spine shows subcutaneous emphysema (black arrow), pneumomediastinum (white arrow), and epidural emphysema (short arrow).
(B) Subcutaneous emphysema (black arrow), pneumomediastinum (white arrow), and epidural emphysema (short arrow) can be observed at the level of the tracheal bifurcation.

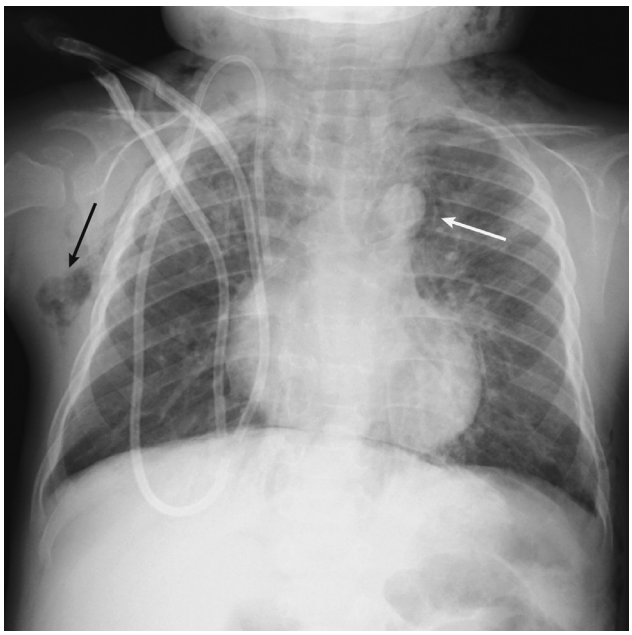

Fig. 3 – Chest radiography findings at admission of a 3-year-old boy with pneumorrhachis. Bilateral ground-glass opacities, subcutaneous emphysema (long arrow), and pneumomediastinum (white arrow) can be observed.

by graft-versus-host-disease with HHV6 reactivation. He received prednisolone, antiherpesvirus agents, and oxygen inhalation therapy, and the subcutaneous emphysema resolved in 7 days.

Discussion

We presented 2 important cases of pneumorrhachis associated with pneumomediastinum in very young patients. The findings are clinically significant because most previous reports involved young adults. We reviewed the relevant literature regarding cases involving children aged <15 years and retrieved 25 articles reporting 32 cases [3,4,7–29]. In total, 22 boys and 10 girls aged between 1 and 15 years (mean age: 9 years) were identified. Pneumorrhachis occurred concomitantly with pneumomediastinum in all cases. Underlying conditions included bronchial asthma ($n = 15$); lower respiratory tract infection or bronchiolitis ($n = 3$); foreign body aspiration, graft-versus-host disease, and upper respiratory tract infection ($n = 2$); and anorexia nervosa and vomiting ($n = 1$). Two patients developed neurological symptoms that did not require surgical intervention.

Chaichana et al. described neurological complications in patients with pneumorrhachis associated with both trauma and pneumomediastinum [2]. In addition, Song reported a case of pneumorrhachis secondary to pneumomediastinum that presented with progressive motor weakness and sensory deficits in the lower extremities. These neurological symptoms resolved after C7 laminectomy [30]. Our patients did not present with any neurological abnormalities, and their condition resolved after treatment for the underlying disease and high flow oxygen therapy.

In summary, pneumorrhachis presents with subtle clinical findings and is difficult to diagnose on chest radiographs. Noncontrast-enhanced CT is additionally required for further neurological evaluations and treatment planning. Once abnormal neurological findings are recognized or ruled out, further CT studies are necessary for confirming the etiology [1].

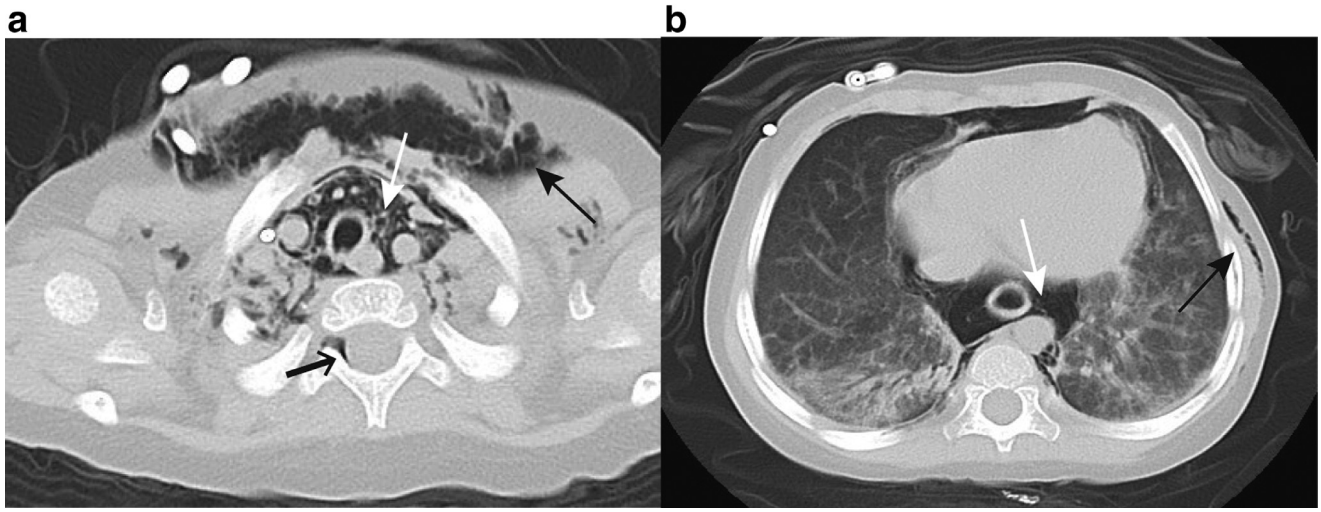

Fig. 4 – Computed tomography findings at admission of a 3-year-old boy with pneumorrhachis.
(A) Axial reconstruction of the cervical spine shows subcutaneous emphysema (black arrow), pneumomediastinum (white arrow), and epidural emphysema (short arrow).
(B) Bilateral ground-glass opacities, predominantly dorsolateral lobes, subcutaneous emphysema (black arrow), and pneumomediastinum (white arrow) can be observed at the T10 level.

Pneumomediastinum is also difficult to identify in young children, necessitating close clinical examinations and noncontrast-enhanced CT for neurological evaluations.

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