CASE REPORT

Pneumorrachis Secondary to Spontaneous Pneumomediastinum in an Adolescent

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Abstract

Pneumorrhachis is the accumulation of free air around the dura mater and is rare in pediatric patients. In this report, the development of pneumorrhachis secondary to spontaneous pneumomediastinum is described in a 15-year-old adolescent. Spontaneous pneumomediastinum was most likely related with the patient's underlying asthma, which became exacerbated following a respiratory infection, probably of viral etiology. The patient was asymptomatic regarding pneumorrhachis, and its diagnosis was made through computed tomography, performed following the diagnosis of pneumomediastinum. The patient was hospitalized and pneumorrhachis was treated with oxygen supplementation and analgesia. After clinical improvement, the patient was discharged at day six of hospitalization. One month later, a new computed tomography revealed the complete resolution of the pneumorrhachis. The present case report is in agreement with what has been described in the literature, and its presentation will increase the awareness of pediatricians to this infrequent condition.

Keywords: Adolescent; Asthma/complications; Mediastinal Emphysema; Pneumorrhachis/diagnostic imaging; Pneumorrhachis/therapy; Computed Tomography

Introduction

Pneumorrhachis is the accumulation of air around the dura mater,[1] and its causes may be iatrogenic, traumatic, and non-traumatic.[1-3] Non-traumatic pneumorrhachis is less frequent and has several etiologies, such as asthma, viral infections, conditions associated with a sudden increase of intrathoracic pressure and barotrauma, anorexia nervosa, foreign body inhalation, physical exercise, and drugs.[3-5] Pneumorrhachis can occur spontaneously, when it is normally associated to pneumomediastinum and subcutaneous emphysema.[2,3] Pneumomediastinum is defined as the presence of interstitial air in the mediastinum.[6] Pneumomediastinum is normally associated with a good prognosis and has a low prevalence in patients seen in the emergency department.[6,7]

The first description of non-traumatic pneumorrhachis was in 1989, in two pediatric patients with pneumomediastinum, with one of them being asthmatic.[8] This was also the first description of pneumorrhachis in pediatrics. Since then, more cases have been reported, although pneumorrhachis occurrence in this age group is uncommon.[3-5,9]

The authors report the diagnostic and therapeutic approach to pneumorrhachis associated with spontaneous pneumomediastinum in a 15-year-old adolescent.

Case Report

A 15-year-old male with a personal history of allergic asthma, diagnosed at 5 years old, controlled with fluticasone 250 µg twice/day, and montelukast 5 mg/day was admitted in the emergency department due to a sudden onset of right cervical pain. This occurred when his neck was jerked abruptly during a car trip. Besides cervical pain, the patient denied other accompanying symptoms. He also reported having an upper respiratory infection with a duration of five days, which occurred two weeks before admission. This was treated with acetaminophen 1500 mg/day and levodropropizine 180 mg/day. At the initial physical examination at the emergency department, the patient was eupneic, with an arterial oxyhemoglobin saturation (SpO_2) measured by pulse oximetry of 99% in room air, tympanic temperature of 37.2 °C, heart rate of 92 bpm, and blood pressure of 117/63 mmHg. The patient had right cervical pain, which worsened with cervical movement. Palpation

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of the right anterior inferior and supraclavicular cervical regions elicited pain. In the same areas, at palpation the presence of subcutaneous emphysema was also felt. There was a positive Hamman sign at cardiac auscultation. Pulmonary auscultation and the remaining physical examination, including neurological examination, were unremarkable.

Cervical and thoracic radiographs and blood analysis were conducted. The former showed subcutaneous emphysema with no other changes. Thoracic radiography demonstrated pneumomediastinum in the superior mediastinum. Blood analysis revealed the presence of mild leukocytosis (14 000 cells/µL), neutrophilia (11 500 cells/µL), and a slight increase in urea (47.2 mg/dL) and creatinine (1.1 mg/dL) levels, with all the remaining values within the normal range. Arterial blood gases revealed pH 7,223, partial pressure of carbon dioxide (pCO_2) 10.9 mmHg and partial pressure of oxygen (pO_2) 153 mmHg, without abnormalities in the remaining parameters.

The patient was admitted to the pediatric department for further investigations. In addition, conservative treatment was started with oxygen supplementation through a high-flow oxygen non-rebreather mask (10-15 L/min) and analgesia with paracetamol as required.

In the first day of hospitalization, the patient underwent thoracic computed tomography (CT), which confirmed pneumomediastinum involving the heart and the great vessels, extending to the lateral cervical vessels. It also showed right anterior and lateral cervical subcutaneous emphysema and the presence of air bubbles within the spinal canal, which is a finding compatible with the diagnosis of pneumorrhachis at the epidural level. Pneumorrhachis was visible in the cervical and dorsal spinal regions, from C7 to D6 (Figs. 1 and 2). Testing for several infectious agents was also performed. The results of these were a profile suggestive of a previous but currently non-active Epstein Barr viral infection, negative for cytomegalovirus, parvovirus 19, enterovirus, adenovirus, echovirus, respiratory syncytial virus, influenza A and B, and *Mycoplasma pneumoniae.* The patient was also observed by neurology and pulmonology which concluded that this was a case of pneumorrhachis secondary to spontaneous pneumomediastinum.

During the hospitalization, the patient always remained eupneic and hemodynamically stable without fever or neurological signs. There was a progressive improvement of pain complaints, with the patient becoming asymptomatic and with the complete resolution of subcutaneous emphysema by the sixth day of hospitalization. A new thoracic radiograph at the fourth day of the hospitalization revealed an almost complete resolution of the pneumomediastinum. Treatment with supplemental oxygen with a high-flow non-rebreather mask continued until the fifth day. On the sixth day, the patient tolerated the transition to room air, maintaining normal SpO_2 levels.

The patient was discharged on the sixth day, asymptomatic, with SpO_2 of 99% in room air, and without any changes at the physical examination (including neurological examination). He received the advice to continue his regular asthma medication and rest.

A month later, he was reevaluated in a pediatric consultation. He remained asymptomatic and the clinical examination (including neurologic examination) was unremarkable. A thoracic CT was repeated and was normal, showing the resolution of the pneumomediastinum and pneumorrhachis. He also performed respiratory function tests (spirometry and plethysmography) whose results were a forced vital capacity (CVF) of 111.8%, forced expiratory volume in one second (FEV1) of 129, 6% in relation to the respective reference values (both within normal values), FEV1 with partial response to bronchodilator of 7.9%/90 mL, FEV1/FVC ratio of 114.7%, which was also normal. Plethysmography yielded a residual volume of 214% of the reference value, a reduction of -6% with the bronchodilator test and a specific resistance of 249.7% of the reference value, with a significant reduction to

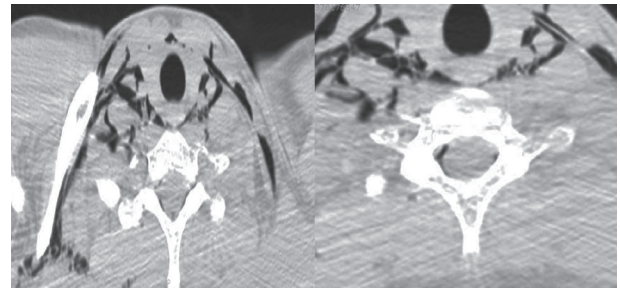

Figure 1. Pneumorrhachis in the cervical region. Computed tomography of the cervical region showing epidural pneumorrhachis in the spinal canal and subcutaneous emphysema in the anterior and right lateral cervical regions (left image), cervical epidural pneumorrhachis in a magnified view (right image).

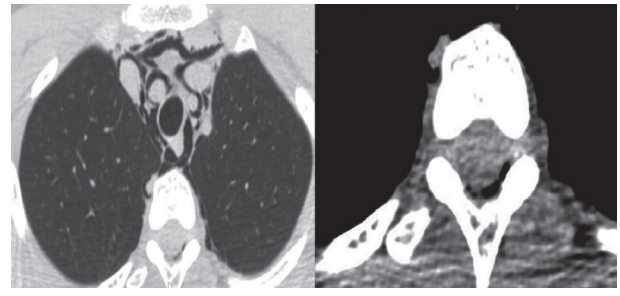

Figure 2. Pneumorrhachis in the thoracic region. Thoracic computed tomography showing the presence of pneumomediastinum in the cranial mediastinum and epidural pneumorrhachis (left image) and epidural pneumorrhachis in a magnified view (right image).

153.9% (-95.8%) after the bronchodilator test. These studies were consistent with pulmonary insufflation, with a partial response to inhaled bronchodilator (< 12% increase in forced expiratory volume in one second), increased residual volume, and airway resistance. Based on these results, it was decided that the patient should continue his current medication.

Discussion

Pneumorrhachis can be classified into three types: epidural, intradural/extra-axial, and intradural/intra-axial.[1-3] The latter two are often associated with traumatic injuries, where the occurrence of pneumorrhachis is considered a marker of greater severity.[1] Epidural pneumorrhachis is the only type that is found in non-traumatic patients and may occur following spontaneous pneumomediastinum, such as the case described here.[2-5]

The pathway through which air reaches the epidural space in pneumomediastinum patients remains incompletely understood. It is known that, when the pressure gradient between the intra-alveolar and interstitial spaces increases, air reaches the interstitium through small alveolar openings or through alveolar or bronchiolar rupture. From the interstitium, air can reach the mediastinum, passing through the pulmonary bronchovascular sheath and hilum, which are pathways that offer less resistance. From there, air passes to the subcutaneous cervical tissue (contiguous to the mediastinum) and to the posterior paraspinal and/or retropharyngeal spaces.[3,6] Then, it can reach the epidural space through the *foramina* that serve as conduit for the spinal nerves and vessels.[3,6,10] Once in the epidural space, air will concentrate mainly in the posterior epidural space, as this offers less resistance than the anterior epidural space. The reason is because the former is composed mainly of loose connective tissue, whereas the latter is filled with an important and rich vascular network.[11]

Spontaneous pneumomediastinum is a rare condition in pediatrics, with an estimated incidence of 1/800 to 1/42,000 of all presentations in the emergency department.[12-14] It affects mainly tall, thin male adolescents with average age of 15 years.[14-16] In 70%-90% of cases, it is possible to identify an underlying etiology, with the most common being asthma, respiratory infections, vomiting, and other conditions associated with Valsalva maneuver (**e.**g. cough, laughter, shouting, intense physical exercise).[14-17] In adolescents with mild spontaneous pneumomediastinum, asthma is the most likely etiology, accounting for 50% of cases.[15-17] As our patient had a personal history of asthma and maintained abnormal findings in respiratory function tests one month after admission, compatible with asthma, we believe that asthma was probably the main factor underlying pneumomediastinum and consequently pneumorrhachis in this case.

The patient also had a history of upper respiratory infection prior to coming to the emergency department. The development of spontaneous pneumomediastinum and pneumorrhachis following viral infections has been described previously in the literature.[4,18-20] In the present clinical case, none of the several infectious agents that were tested was identified. However, other viruses that have been associated with the development of pneumomediastinum and pneumorrhachis, including rhinovirus or bocavirus, were not tested.[4,18] Therefore, it cannot be ruled out that these or other agents could have contributed to the development of pneumomediastinum and pneumorrhachis, either directly or indirectly, by aggravating the patient underlying asthma.

None of the clinical signs presented by the patient unequivocally pointed out to the diagnosis of pneumorrhachis. This is in agreement with the literature that describes the majority of pneumorrhachis patients either as asymptomatic, or with nonspecific clinical signs, with a minority having neurological signs or symptoms.[1-3,21] In the current case, pneumorrhachis diagnosis was only established after performing thoracic CT, as it was not apparent on thoracic radiographs. Thoracic radiographs are still indicated in the initial diagnostic evaluation of pneumorrhachis, although it can only be identified by this method when it has a significant size.[1] Thoracic radiographs can also detect the underlying lesions that are associated to pneumorrhachis development.[1] However, in the cases where pneumorrhachis is not significantly large, as in the current case, CT is considered the diagnostic method of choice.[1-3]

In the absence of specific neurological signs, it is unlikely that CT is performed in the emergency department to exclude the presence of pneumorrhachis. Moreover, the role of thoracic CT for the diagnosis of spontaneous pneumomediastinum in pediatric patients is not defined and consequently, is not performed in most cases. Therefore, it is possible that the actual incidence of pneumorrhachis in this context is higher than the one described, which is of 9.5% of all the pediatric patients observed in the emergency department with spontaneous pneumomediastinum having concomitant pneumorrhachis.[3] Because of this, some authors suggest that, to exclude pneumorrhachis associated with spontaneous pneumomediastinum, the diagnostic

approach should always include a complete neurological examination and a thoracic and cervical CT.[22]

Regarding treatment, given the rarity of pneumorrhachis, no published guidelines are available. Consequently, it should be individualized and according to the clinical condition.[1,2] Pneumorrhachis is normally a benign and self-limiting condition, with most patients showing the reabsorption of spinal air in the first days after the initial clinical presentation.[2,21,22] Because a minority of patients can develop neurological signs, the most consensual approach suggested by the literature is to admit the patient to the hospital and to start conservative treatment.[1,22] This consists in rest and high flow oxygen supplementation to promote nitrogen excretion and consequently air reabsorption from the epidural space. Other treatments described in the literature include intravenous dexamethasone and hyperbaric oxygen administration and epidural air aspiration through a percutaneously inserted Tuohy needle.[1] In some cases with neurologic signs and symptoms, surgical decompression may be necessary. In the current case, the patient was admitted and responded favorably to conservative treatment, confirming the good prognosis of this clinical condition.

In conclusion, this case is a good example of the clinical presentation and diagnostic and therapeutic approach to pneumorrhachis associated to spontaneous pneumomediastinum in an adolescent. Because this is a rare situation, its presentation will contribute to raising the awareness of pneumorrhachis among pediatricians and pointing out the best approach to it.

WHAT THIS CASE REPORT ADDS

- Accumulation of air around the dura mater is called pneumorrhachis and it is a rare condition in pediatrics.
- It can be caused by iatrogenic, traumatic, and non-traumatic etiologies.
- When it is spontaneous, it is often associated with pneumomediastinum.
- Computed tomography is the diagnostic method of choice.
- In most cases, pneumorrhachis is asymptomatic and responds favorably to conservative treatment consisting of rest, high flow oxygen supplementation, and analgesia.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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Pneumoráquis Secundário a Pneumomediastino Espontâneo num Adolescente

Resumo:
O pneumoráquis define-se como a acumulação de ar em redor da dura-máter e é raro em pediatria. Descreve-se um caso de pneumoráquis associado a pneumomediastino espontâneo num adolescente de 15 anos. O pneumomediastino terá estado provavelmente relacionado com a condição asmática do doente, agravada após infecção respiratória, de possível origem viral. O doente permaneceu sempre assintomático em relação ao pneumoráquis. O diagnóstico foi estabelecido através de tomografia computorizada, realizada para esclarecimento diagnóstico do pneumomediastino. A abordagem terapêutica passou por internamento e tratamento conservador, com analgesia, repouso e administração de oxigénio por alto débito. A resposta foi favorável, tendo o doente tido alta assintomático, seis dias após a admissão hospitalar. A tomografia computorizada realizada um mês após o internamento não revelou pneumoráquis. O caso clínico relatado corrobora o que está descrito na literatura. A sua apresentação contribui para uma maior sensibilização dos pediatras acerca desta condição clínica rara.

Palavras-Chave: Adolescente; Asma/complicações; Enfisema Mediastínico; Pneumoráquis/diagnóstico por imagem; Pneumoráquis/terapia; Tomografia Computorizada