CASE REPORT
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Epidural Pneumorrhachis Accompanying to Spontaneous Pneumomediastinum in a Boy: A Rare Association

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Abstract
Epidural pneumorrhachis, or emphysema, refers to the presence of air within the spinal epidural space. It is a well-described sequela of trauma and of instrumentation. It is more rarely seen, as in our case, in association with a spontaneous pneumomediastinum. Isolated case reports describe in association spontaneous pneumomediastinum and epidural pneumorrhachis. There are no fascial barriers between the posterior mediastinum and the retropharyngeal and epidural spaces; thus, air can diffuse freely to the epidural space and produce an epidural pneumatosis. We herein report a case in a 16-year-old boy with spontaneous pneumomediastinum and epidural pneumorrachis and discuss this rare condition.

Keywords: Spontaneous pneumomediastinum, epidural pneumorrachis, plain radiography, computed tomography

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INTRODUCTION

Pneumomediastinum is due to air in the mediastinal space especially in the antero-superior mediastinum. This may be spontaneous or secondary to a primary event [1].

Spontaneous pneumomediastinum (SPM), also known as mediastinal emphysema, is the result of a sudden increase in intra-alveolar pressure resulting from acute bout of cough or vomiting. When the alveolar rupture occurs, the air dissects along lung interstitium and reaches the mediastinum along perivascular spaces. If the air extends towards the visceral pleura, a pneumothorax is a likely event [2]. It is an uncommon condition that is reported to occur in 1 in 33,000 in the general population [3]. SPM occurs more often in young men, with a mean age group of 18 years for first presentation [4]. Epidural pneumorrhachis, or pneumatosis accounts for the visualization of air in the spinal epidural space. Epidural pneumorrhachis with SPM is rare [5]. There are only sporadic cases reported in the literature on this condition. In this report, we present a boy patient with epidural pneumorrhachis associated with SPM which was diagnosed with chest computed tomography (CT).

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CASE PRESENTATION

A 16-year-old male patient was admitted to our hospital who complained of a sudden onset of a sharp chest pain and dyspnea at rest. The patient was asymptomatic until the present admission and he had no marked respiratory disease. There was no dysphagia or history of foreign body ingestion, trauma or surgery. He denied rhinitis, cough, aspiration of vomitus, difficulty with defecation, and illicit drug use. The patient was afebrile and vital signs were normal. Cervical and thoracic subcutaneous emphysema were noted on initial physical examination. Electrocardiography, blood cell count, and serum biochemistry, including serum protein electrophoresis and alpha1-antitrypsin level, were normal. Tests for common autoantibodies and proteinuria was negative. On auscultation of his chest, a systolic crunching sound was heard at the apex and left sternal border that was more pronounced on inspiration consistent with a Hamman's crunch. The lungs were clear in auscultation and percussion. Neurological examination and other systemic examination were normal. Results of the oesophagogram, otorhinolaryngologic examination, and bronchoscopy performed in the emergency department were normal. The initial chest radiograph showed subcutaneous emphysema in the cervical, thoracic, and axillary regions with no evidence of rib fracture (**Figure 1**).

To better define the extent of air dissection into the neck and mediastinum, the patient underwent computed tomography (CT) (Picker PQS scanner, OH, USA). CT scan confirmed the extent of the stripping, which was demonstrated air in the subcutaneous, visceral and carotid spaces of the neck, extending along the anterior mediastinal space down to the aortic arch (**Figure 2**). High resolution CT (HRCT) sections of the lung showed no evidence of pulmonary interstitial emphysema or any bullae. No pneumothorax was recognized. In addition, the CT scan showed a small amount of air in the spinal epidural space on multiple cervical and thoracic levels (**Figures 3a, b**). The patient was admitted to the cardiothoracic surgery service for observation, and antiobiotic therapy was administered to prevent mediastinitis. His hospital course was uneventful and further chest X-ray studies showed no progression

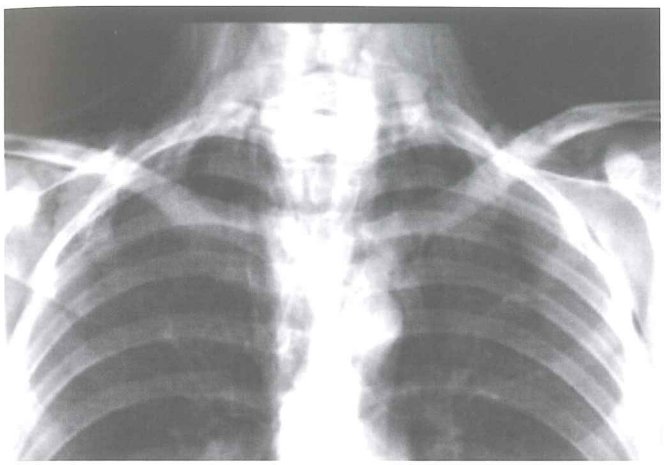

Figure 1. Posteroanterior chest radiograph showing subcutaneous emphysema and pneumomediastinum.

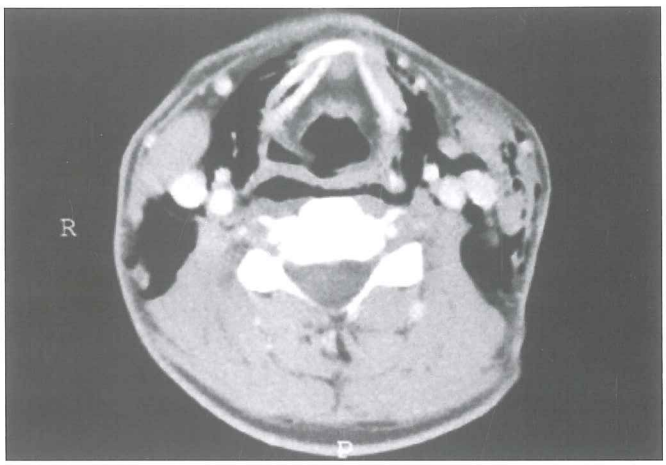

Figure 2. Axial CT image of the neck demonstrating subcutaneous emphysema and air in multiple tissue planes.

of the pneumomediastinum. On follow-up evaluation 1 month later the patient was clinically well.

DISCUSSION

SPM is an uncommon clinical condition and should be a diagnosis of exclusion. SPM was first described in 1617 by Louise Bourgeois whilst Hamman in 1939 named Hamman's sign which is the finding of audible crepitis occurring with the heart beat due to the presence of mediastinal air [6].

SPM is usually seen in healthy young men (in particular, male adolescents) or parturient women resulting from the rupture of peripheral pulmonary alveoli due to sudden increase of intra-alveolar pressure after exaggerated Valsalva maneuvers [weight lifting, Heimlich maneuver, defecation, parturition, inhalation of nitrous oxide, marijuana, cocaine, methylenedioxymethamphetamine (mdma) or 'Ecstasy'] [7–9]. Childbirth, barotrauma during mechanical ventilation, ascent phase of dive, seizures, inhalational drug use or hyperbaric treatment are other possible causes. The most common cause in children is asthma [2]. SPM may also complicate obstructive airway processes such as asthma or foreign bodies [10]. Inhalational drug use has been reported as the most common associated activity, accounting for approximately 76% of cases [8].

SPM is usually self-limited due to the transient nature of the clinical course (e.g. cough, asthma, and bronchiolitis). However, in the presence of a foreign body, persistence of expiratory outflow resistance and associated coughing leads to an uninterrupted air leak and progressive emphysema [11].

A possible explanatory mechanism for pneumomediastinum is high bronchoalveolar pressure during cough resulting in air leakage into pulmonary perivascular interstitium. The air dissects the paths of least resistance into the mediastinum to the fascial planes of the neck. There are no fascial barriers to prevent communication of the posterior mediastinum or retropharyngeal space with the epidural space. Air can thus communicate freely via the neural foramina [12].

Epidural pneumorrhachis, also called pneumatosis, emphysema or aerorachia, accounts for the visualization of air in the spinal epidural space. The causes of pneumorrhachis have broadly been classified into iatrogenic, non-traumatic and traumatic. Iatrogenic causes are the most common, and follow the administration of epidural analgesia. Non-traumatic causes have been described following spontaneous or non-trauma related pneumomediastinum or pneumothorax [12]. In a recent review of the English literature, Goh and Yeo [13] found 31 cases of traumatic pneumorrhachis, of which only 13 were epidural. Most of these were secondary to pneumothorax or pneumomediastinum (7 of 13 cases), as in our case. The combination of pneumomediastinum with epidural pneumorrachis without thoracic trauma has rarely been reported in the medical literature. In extensive literature search on association of non-traumatic epidural pneumorrachis associated with spontaneous pneumomediastinum, we revealed only thirteen cases reports on this subject [2–10, 12, 14–16].

Laman and McCleskey [17] propose that air introduced into the epidural space during epidural anesthesia may escape from the epidural space via the neural foramina to enter the mediastinum. Radicular pain and paraplegia due to extradural air have been reported [11]. However, in most patients this is only an incidental finding and does not require specific treatment, and the air gets absorbed over 2–3 weeks. Because this condition usually accompanies subcutaneous or mediastinal emphysema, treatment is primarily directed towards these complications. Further air transfer occurs, by dissection through the deep fascia of the mediastinum and neck, to ultimately reach the subcutaneous tissue. The theory of air transfer that explains the occurrence of intraspinal air with pneumothorax and pneumomediastinum correlates well with this hypothesis of air transfer between the epidural space and subcutaneous tissues. Because of the absence of real fascial barriers between

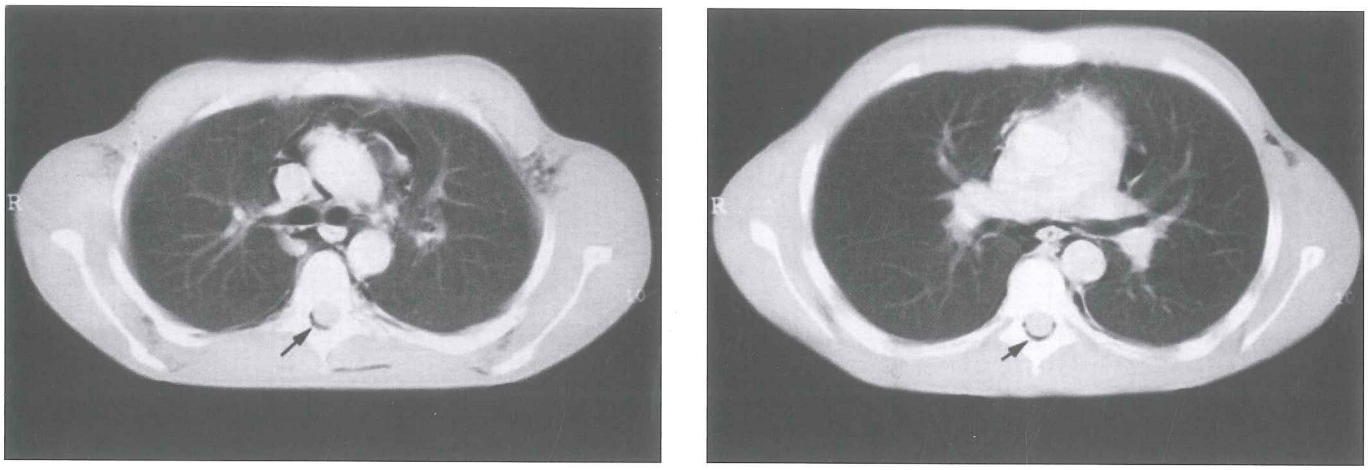

Figure 3. CT images of the upper thorax (a) and mid-thorax (b) levels on lung windows demonstrating extensive pneumomediastinum and epidural pneumorrhachis (black arrows).

the posterior mediastinum or the retropharyngeal space and the intrarachidian epidural space, pneumorachis seems to be the consequence of the pneumomediastinum. The air typically collects in the posterior epidural space because of less resistance from the loose connective tissue, as compared with the rich vascular network present anteriorly [10]. This posterior location was also seen in our patient. The epidural pneumorrhachis associated with spontaneous pneumomediastinum in our patient did not affect his clinical outcome nor did it require specific treatment [6, 7, 10, 13].

Although recognized in the adult population and documented in the literature, pneumorrhachis is very rare in children, and thus may be clinically alarming, as in the case we present. Detailed imaging allows the distinction between air within the epidural space and subarachnoid air which, due to breach of the dura, has further clinical implications. Awareness of the mechanism by which air reaches the epidural space secondary to pneumothorax and pneumomediastinum is reassuring, and allows a conservative approach to be taken, as treatment of the underlying cause will lead to spontaneous resorption of the pneumorrhachis [5, 8, 12, 13, 17].

The treatment is conservative and consists of bed rest, analgesics, and avoidance of the Valsalva maneuver [7].

In conclusion, epidural pneumorrhachis associated with SPM usually has a benign character, therefore, does not need a special treatment. However, follow-up of this rarely seen entity still remains significant, because in some cases serious neurologic symptoms may occur. If a traumatic or infectious cause of epidural pneumorrachis is not present, the epidural air on CT scan may represent a benign finding secondary to pneumomediastinum that does not require further work up or treatment. To our opinion, while the chest X-ray is able to define the presence of a pneumomediastinum and subcutaneous emphysema, the diagnosis of epidural pneumorrhachis can only be made by CT.

REFERENCES

1. Chalumeau M, Le Clainche L, Sayeg N et al. Spontaneous pneumomediastinum in children. Pediatr Pulmonol 2001;31:67-75.
2. Dosios T, Fytas A, Zarifis G. Spontaneous epidural emphysema and pneumomediastinum. Eur J Cardiothorac Surg 2000;18:123.
3. Defouilloy C, Galy C, Lobjoie E et al. Epidural pneumatosis: a benign complication of benign pneumomediastinum. Eur Respir J 1995;8:1806-7.
4. Aribas OK, Gormus N, Aydogdu Kiresi D. Epidural emphysema associated with primary spontaneous pneumothorax. Eur J Cardiothorac Surg 2001;20:645-6.
5. Caramella D, Bulleri A, Battolla L et al. Spontaneous epidural emphysema and pneumomediastinum during an asthmatic attack in a child. Pediatr Radiol 1997;27:929-31.
6. Balachandran S, Guinto FC, Goodman P et al. Epidural pneumatosis associated with spontaneous pneumomediastinum. Am J Neuroradiol 1993;14:271-2.
7. Koelliker PD, Brannam LA. Epidural pneumatosis associated with spontaneous pneumomediastinum: case report and review of the literature. J Emerg Med 1999;17:247-50.
8. Hazouard E, Koninck JC, Attucci S et al. Pneumorachis and pneumomediastinum caused by repeated Muller's maneuvers: complications of marijuana smoking. Ann Emerg Med 2001;38:694-7.
9. Mutlu H, Silit E, Pekkafali Z, et al. 'Ecstasy'(MDMA)-induced pneumomediastinum and epidural pneumatosis. Diagn Interv Radiol. 2005;11:150-1.
10. Eesa M, Kandpal H, Sharma R et al. Spontaneous pneumorrhachis in bronchial asthma. Acta Radiol 2006;47:672-4.
11. Ristagno RL, Hiratzka LF, Rost RC Jr. An unusual case of pneumorrhachis following resection of lung carcinoma. Chest 2002;121: 1712-4.
12. Drevelengas A, Kalaitzoglou I, Petridis A. Pneumorrhachis associated with spontaneous pneumomediastinum. Eur J Radiol 1994; 18: 122–3.
13. Goh BK, Yeo AW. Traumatic pneumorrhachis. J Trauma 2005;58: 875-9.
14. Pooyan P, Puruckherr M, Summers JA et al. Pneumomediastinum, pneumopericardium, and epidural pneumatosis in DKA. J Diabetes Complications 2004;18:242-7.
15. Goh BK, Tan HK, Tan FL et al. Epidural emphysema associated with spontaneous pneumomediastinum. Am J Surg 2006;192: 355-6.
16. Bernaerts A, Verniest T, Vanhoenacker F et al. Pneumomediastinum and epidural pneumatosis after inhalation of "Ectasy". Eur Radiol 2003;13:642-3.
17. Laman EN, McClesky CH. Supraclavicular subcutaneous emphysema following lumbar epidural anesthesia. Anesthesiology 1978: 48:219-221.