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Teaching cases

Pneumothorax-associated fibroblastic lesion in combination with localized pleural angiomatosis: A possible cause of juvenile spontaneous hemopneumothorax

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ABSTRACT

Spontaneous hemopneumothorax is an uncommon but potentially life-threatening condition, with a potential for a rapid ventilatory collapse and a large collection of hidden blood loss into the pleural cavity. Here, we report the first case in the literature on pneumothorax-associated fibroblastic lesion in combination with localized pleural angiomatosis in a 19-year-old Caucasian male, resulting in massive spontaneous hemopneumothorax and hypovolemic shock. Our findings support a causal link between this condition and pneumothorax. The possible superimposed hemothorax is explainable by the pleural involvement of large angiomatous vessels, prone to rupture.

Introduction

Spontaneous hemopneumothorax (SHP) is an uncommon but potentially life-threatening condition with a potential for a rapid ventilatory collapse and a large collection of hidden blood loss into the pleural cavity. It is one of the most important causes for which a patient is admitted to the emergency room with unexplained signs of significant hypovolemia. It can be the result of primary vascular events, spontaneous pneumothorax, coagulopathies, thoracic malignancies and infectious diseases, such as tuberculosis [8]. Males are between 8 and 30 times more likely to develop SHP than females, for a reason which remains unclear [12]. SHP occurs as the first episode in about the 80% of patients, while the remaining 20% have previously suffered from pneumothorax recurrences or contralateral pneumothorax [12]. The chest X-ray remains the most useful investigation in the diagnosis of SHP [4]. As to the source of bleeding, it can firstly be a small non-contractile vessel in an area of torn adhesions between the two pleural layers. Secondly, it can originate from the rupture of a vascular bulla or lung parenchyma at the apex of the lungs and, finally, it can take its origin from an aberrant thin-walled vessel, devoid of muscular fibers and, therefore, not able to contract itself [5]. Recent pathological studies have revealed that a degenerative sclerosis, as well as an intima-media fibrosis, should be at the basis of vasoconstriction failure in these aberrant vessels [7].

Case report

A 19-year-old Caucasian man was admitted to the emergency room for sudden shortness of breath with clinical signs of pleural effusion and hypovolemic shock. A chest X-ray and CT-scan showed a right hemopneumothorax extending from the apex to the base, 5 cm in maximum thickness (Fig. 1). The patient was promptly submitted to an apical resection of the right upper lobe, the site of the bleeding origin. On histopathological examination (Fig. 2), a distinct pattern of pneumothorax-associated fibroblastic lesion (PAFL) was revealed, together with a reactive surface process, characterized by the presence of eosinophils and macrophages (reactive eosinophilic

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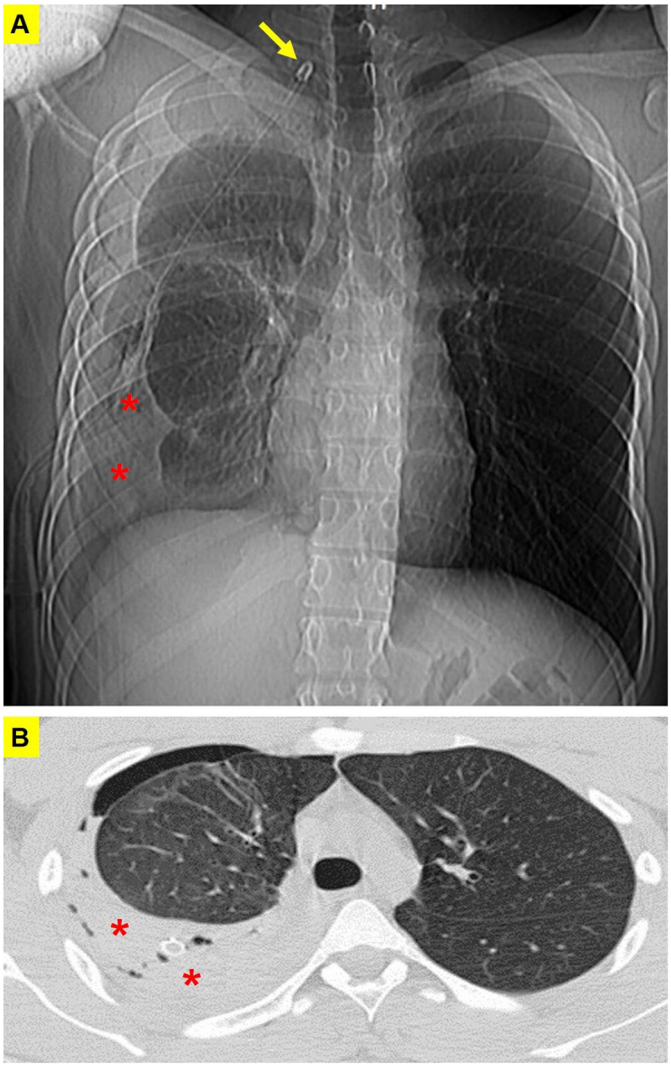

Fig. 1. The supine chest X-ray shows massive pleural effusion and right pneumothorax (A, red stars). A pleural drainage is also visible (A, yellow arrow). The CT scan confirms the co-presence of a copious right-sided hemothorax (B, red stars). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

pleuritis) and related to the pneumothorax [1]. In combination with PAFL, a localized pleural angiomatosis was clearly ascertained by hematoxylin/eosin staining (Fig. 2D).

Discussion

This is the first case in the literature on PAFL in combination with localized pleural angiomatosis, resulting in conspicuous SHP. Previously, Kawamukai and colleagues had described an unusual case of persistent pneumothorax due to a localized pleural angiomatosis in a 45-year-old white male [6]. Both these reports suggest a causal relationship between this vascular abnormality and pneumothorax. In our case, the superimposed hemothorax is explainable by the pleural involvement of large angiomatous vessels associated with PAFL. Their very large size could be the result of adhesions from an antecedent episode (or episodes) in the context of a persistent/recurrent pneumothorax.

PAFL is a unique histologic lesion which can be incidentally found in the lung apices of young, predominantly male patients affected by primary spontaneous pneumothorax and without significant smoking history. It consists of a wedge-shaped fibrotic area at the pleural–subpleural interface with fibroblastic foci at the leading parenchymal edge. Therefore, histologically, it is not seen in all patients with pneumothorax, and its significance is unclear at this time [2,3]. PAFL has been retrospectively identified in 11 of the 43 cases (26%) of a large multi-institutional cohort of primary spontaneous pneumothorax [2]. Currently, primary spontaneous pneumothorax is believed to derive from a combination of pleural anomalies and disruptive changes in alveolar spaces [9]. By analyzing some diseases which are commonly associated with secondary spontaneous pneumothorax, such as Marfan's syndrome, Ehlers–Danlos syndrome, rheumatoid arthritis, scleroderma, ankylosing spondylitis and idiopathic pulmonary fibrosis, it has been found that there is a crucial alteration of the fibro-connective substrate [9]. It is possible that PAFL is the lung manifestation of a connective disorder, whose peripheral subpleural occurrence leads to an early pneumothorax. As for exuberant type-2 pneumocytes hyperplasia, in which a strong lepidic-like proliferation of type-2 pneumocytes is associated with spontaneous pneumothorax [10,11], PAFL could be reconsidered as the cause, and not only the mere reactive consequence, of this pathology. Surely, the presence of PAFL identifies a distinct subpopulation of primary spontaneous pneumothorax that warrants further research.