An Unusual Case of Persistent Pneumothorax

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We herein report a case of a 45-year-old white male who referred to the emergency department for a right pneumothorax. A chest tube was emergently placed. Due to incomplete lung reexpansion, the patient underwent a right thoracoscopy disclosing the presence of several kinky vessels consistent of localized pleural angiomatosis, and a talc pleurodesis was performed. Computed tomographic scan and angiography confirmed an anomalous vascular connection between systemic and pulmonary circulation. Thus, a vascular percutaneous transcatheter embolization of the abnormal vessel was successfully executed and the patient was discharged without consequence.


Benign proliferation of pulmonary vascular tissue is a rare lesion usually presenting as solitary nodules (hemangioma). In particular, diffuse pulmonary capillary hemangiomatosis (PCH), described mainly in neonates as part of a generalized process affecting the skin, mucous membranes, and visceral organs is even less common [1]. Pleural angiomatosis (PA) is a sporadically reported condition in the English literature. We herein report a challenging case of localized PA resulting from a vascular malformation and discovered by video-assisted thoracic surgery (VATS).

A 45-year-old white male referred to the emergency department after a 1-month history of progressive dyspnea and cough. With the exception of an active smoking habit, his medical history was unremarkable. At emergency department admission, the patient referred a moderate to severe shortness of breath associated with chest pain localized to the right hemithorax. Vital signs consisted of blood pressure of 130/80 mm Hg, heart rate and respiratory rate, respectively, of 99 bpm and 20 per minute, and temperature of 37°C. Thoracic examination evidenced a right hyper-expanded hemithorax associated with limited respiratory movement and recruitment of accessory muscles. Breath sounds on the right side were partially suppressed.

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Standard chest X-ray (CXR) revealed a right pneumothorax without mediastinal shift (Fig 1). In emergency room, a chest tube was promptly placed and up to 300 mL of serous pleural fluid was drained. Subsequently, his general and respiratory conditions gradually improved. The following CXRs showed an incomplete reexpansion of the lung. In order to define the cause of the persistent pneumothorax, the patient underwent a right thoracoscopy. After introducing the 10-mm video camera, the presence of several kinky vessels, crossing the surface of the upper third of the right upper lobe, was immediately evidenced. In particular, angiomatosis was consistent of vessels of cranio-caudally increasing size and localized into visceral pleural only; as well, a vascular connection with upper mediastinum was clearly identified (Fig 2). Moreover, some localized pleural adhesions were disclosed. Lung evaluation did not evidence any parenchymal lesion. The air-leakage hydropneumatic test was unremarkable. In order to obtain an acceptable lung expansion, pleural adhesions were removed and a large-particle asbestos-free talc pleurodesis was performed. A chest tube with external pleural suction was finally placed. Postoperative CXR showed a complete reexpansion of the right lung. The chest tube was removed on postoperative day 3 without consequences. Subsequently, computed tomography scan (Fig 3) and angiography (Fig 4) confirmed an anomalous vascular connection between systemic circulation (right thyrocervical artery trunk) and pulmonary circulation (PA vessels). No other vascular abnormalities were reported. A vascular percutaneous transcatheter embolization of abnormal vessel was then successfully performed and patient was discharged postoperative day 7. One year after surgery, the patient was clinically free from recurrence.
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Pulmonary angiomatosis usually refers to an extremely rare condition (PCH) of unknown etiology due to abnormal blood vessel infiltration of the lung parenchyma, interlobular septa, bronchial walls, and pleura. This condition led to a pulmonary veno-occlusive disease caused by involvement of the pulmonary veins and venules and resulting in progressive precapillary hypertension. It is not clear whether PCH is a neoplastic process or a congenital abnormality [2]. Some authors suggested that these lesions could arise from non-neoplastic micro-vascular proliferation [3, 4] (angiogenic etiology). Especially in pediatric patients, diffuse hemangiomatosis involves mucocutaneous surfaces as well as multiple internal organs. The prognosis is generally poor because of the development of complications such as congestive heart failure, consumptive coagulopathy, severe bleeding and compression of vital organs [1]. The PCH should be differentiated from pulmonary hemangiomatosis (usually congenital or as a part of von Hippel-Lindau disease) that occurs in young adults. In this condition, a majority of such hemangiomas are discovered incidentally or when spontaneous rupture results in hemorrhage and hemorrhagic pleural effusion [5].

In the case we report, the patient underwent a VATS because of right pneumothorax associated with persistent air leak. We observed the presence of a hemangiomatosis-like lesion consistent with several large kinky vessels localized on the pleural surface of the right upper lobe only. Moreover, a singular communication between these vessels and upper mediastinum was
detected. Computed tomography and angiography confirmed an unusual communication between the right thymocervical artery trunk and the vascular malformation. Due to this evidence, a diagnosis of diffuse hemangiomatosis or pulmonary angiomatosis was definitely excluded. In fact, our finding consists in a localized PA due to a vascular abnormality (connection between systemic and pulmonary vascular system).

In literature, PA is sporadically reported. Nanaware and colleagues [6] described a case of spontaneous hemothorax caused by a pleuropulmonary hemangioma discovered during surgery. Saito and colleagues [7] detected pulmonary hemangiomatosis-like foci (caused by a thoracic endometriosis) by thoracoscopy. Although reports on pleuropulmonary hemangiomatosis are rarely described in the English literature, localized PA due to vascular malformation was never described before. As well, to our knowledge, this is the first reported case in which a diagnosis of PA was carried out by VATS. However, further studies are needed for investigating the role of VATS in diagnosing pleuropulmonary angiomatosis.

In conclusion, pulmonary angiomatosis is a rare condition in which the pleural involvement is outstanding. It should be ruled out in all cases where spontaneous hydropneumothorax or persistent pneumothorax is detected. In selected case, VATS could lead to an accurate diagnosis of PA; as well, talc pleurodesis should be performed in order to reduce the risk of recurrence. Finally, when an abnormal vascular connection is confirmed, embolization of such vessels is mandatory.

References