Angiosarcoma of the Lower Extremity presenting as Bilateral Pneumothoraces

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Angiosarcoma of the Lower Extremity Presenting as Bilateral Pneumothoraces
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Abstract

Spontaneous pneumothorax usually presents as unilateral disease. Bilateral spontaneous pneumothoraces are less common and are more likely to be secondary than primary. We describe a case of bilateral spontaneous pneumothoraces that were resistant to conventional treatment, and found ultimately to be secondary to angiosarcoma of the lower extremity. A previously well 49 year old white female was referred to our institution in London with bilateral pneumothoraces. A left video assisted thoracoscopic procedure was undertaken, and the wedge resection of the lung specimen contained an area of metastatic tumor. Talc was placed into the left pleural cavity; talc was also placed at the bedside into the right pleural cavity which was slow to heal, and required a second chest tube and Heimlich valve.

The pneumothoraces were eventually found to be secondary to metastatic angiosarcoma of the thigh. A groin mass was biopsied and found to be the source of the metastatic deposits. This was not present on initial examination. The patient was referred for chemotherapy to a specialized sarcoma unit. Spontaneous pneumothorax may be the first and only evidence for pulmonary metastases. This report highlights the importance of considering an underlying pathology in patients presenting with spontaneous bilateral pneumothoraces.

Keywords

Bilateral pneumothorax, angiosarcoma

Introduction

Spontaneous pneumothorax usually presents as unilateral disease and may be primary (where there is no obvious underlying lung disease) or secondary (underlying lung disease present). Most cases resolve with conservative management, thoracocentesis or tube thoracostomy. Guidelines and indications for VATS surgical treatment include recurrent pneumothorax, because the recurrence rate increases significantly after one pneumothorax, persistent air leak or if there is pathology identified on a CT scan such as a bleb. Where surgical treatment is performed for recurrence or persistent pneumothorax, the results are often excellent with only a small incidence of recurrence. Operations include thoracoscopy using VATS or robotic techniques. This is the standard of care for almost all cases. Thoracotomy is usually not indicated. All incisions are therefore less than 2cm, and post operative pain and discomfort is therefore avoided. The affected area of lung tissue is resected using endoscopic staplers, through port sites, and this is combined with either a mechanical or chemical pleurodesis. This is also performed as an endoscopic technique. Pleurectomy is reserved for more resistant cases.

Bilateral spontaneous pneumothoraces are however far less common and are more likely to be secondary or iatrogenic than primary. Treatment of the underlying condition is necessary in addition to surgical treatment. Management is more challenging, since the patient often has bilateral chest tubes, and the anesthetic technique may be more intricate. We describe a case of bilateral spontaneous pneumothoraces that were of unknown etiology, and were resistant to conventional conservative surgical treatment. Even surgical treatment of one side (the left in this case), was difficult to accomplish. The pneumothoraces were eventually found to be secondary to metastatic angiosarcoma of the thigh, which in itself posed problems with pathological diagnosis.
Case Report

A previously well 49 year old white female was referred to our institution with bilateral pneumothoraces. She had been treated with bilateral chest tubes at the outside institution. Upon arrival, the patient was spontaneously ventilating, neurologically intact and was haemodynamically stable. There were small air leaks from the chest tubes bilaterally. A chest CT scan showed small bilateral pneumothoraces with no evidence of foreign bodies, mass lesions, bullae or pulmonary infiltrates. There was a history of one prior pneumothorax treated successfully by tube thoracostomy two months previously, which had resolved without incident. The patient had also been treated for a DVT four months previously. Clinical examination of the lower extremities was negative. The patient had a recent CT scan of the abdomen and pelvis that had showed no abnormality.

The patient was initially managed conservatively, however on successive chest X rays, the left lung had collapsed further and the air leak had become larger. A decision was made to explore the left side using a VATS technique. During the operation, a double lumen tube was placed and the right lung was ventilated in a stable manner with only a small air leak. The left lung looked grossly normal with a small area of torn scar tissue at the lung apex which was resected. Parietal pleural abrasion subsequently was performed, chest tubes were placed and the patient was extubated.

The air leaks from both lungs resolved over time with full lung expansion, and the chest tubes were removed. Prior to removal of the right chest tube, talc slurry was instilled into the right pleural cavity to achieve pleurodesis. However this failed to prevent a recurrence of pneumothorax on this side and a new chest tube had to be inserted. The patient was discharged home with the right chest tube and Heimlich valve. She was seen as an outpatient when the right tube was finally removed without incident. A right sided VATS surgical approach was being considered. However the right sided pneumothorax healed well after placement of the second chest tube therefore avoiding another operation with further positive pressure ventilation.

The initial pathology of the resected lung apex had shown an area of carcinoma or sarcoma, and needed further clarification. Further characterization of the pathology specimen by outside review finally showed metastatic angiosarcoma.

The patient was referred to a sarcoma oncologist for chemotherapy, during which she noted a new mass in her left groin. Biopsy confirmed the diagnosis of primary angiosarcoma. Following chemotherapy, the groin mass lesion regressed, and the patient progressed well with no complications and well expanded lung fields bilaterally. There was no effusion or pneumothorax at this point.

Discussion

The differential diagnosis of bilateral spontaneous PTX includes rupture of apical blebs, lymphangioleiomyomatosis (LAM), tuberculosis, emphysematous bullae, diffuse metastatic cancer, trauma, HIV disease and infiltrative pulmonary disease. In our case, the patient’s diagnosis was unclear until her pulmonary issues had largely resolved. There was little to suggest the actual diagnosis at the time of presentation. There was no CT evidence of pulmonary pathology, and the initial pathology report from the resected pulmonary specimen was tumor of unknown etiology. There was also no obvious primary tumor on clinical or CT
examination at the time of the patient’s initial presentation. Thus the diagnosis of metastatic angiosarcoma was not readily apparent.

The treatment of bilateral spontaneous pneumothorax is usually dictated in part by the underlying etiology. Surgery may help resolve the pneumothorax by resecting blebs or bullae and creating pleural adhesions, but at the same time, the diagnosis can be attained from the histological examination of the resected lung tissue. Medical treatment should be instigated for the underlying disease process where appropriate. In the case described, the preferred treatment may have been more conservative if diffuse pulmonary metastases had been known to be present preoperatively. Studies have shown that the treatment of pleural disease from metastatic cancer is more appropriately treated by a conservative approach, and this would have been the plan in our patient. Interestingly there are reports of the occurrence of bilateral spontaneous pneumothorax during pazopanib therapy for pulmonary soft tissue sarcoma metastases. Pazopanib has been recently introduced into the treatment plan for metastatic soft tissue sarcomas that are relatively resistant to conventional chemotherapy. Spontaneous pneumothorax is a rare but well recognized complication of such therapy. In our described patient, no therapy had been implemented; however, it could be seen how easily such therapy may bring out the development of pneumothorax pathology. An accelerated treatment response however is not necessary for a pneumothorax to develop.

The final pathological diagnosis in our patient was angiosarcoma. This is a rare tumor which arises from endothelial cells. It is an aggressive tumor and tends to recur locally, spread widely and have a high rate of lymph node and systemic metastasis. Median survival is between ten and twenty months and patients should be managed in specialized oncology centers with an interest in sarcomas where treatment may include surgical excision, chemotherapy and/or radiotherapy. Our patient was referred appropriately and responded well to chemotherapy.

Bilateral PTX associated with metastatic angiosarcoma from the scalp and breast have been described previously. In three of these reports, bilateral pneumothorax was the presenting symptom which subsequently led to the diagnosis of the primary scalp angiosarcoma. In one case it was disseminated angiosarcoma that presented as a hydropneumothorax. There are however no previous reports of angiosarcoma from extremeties presenting as bilateral pneumothorax. The primary tumor in our case had metastasized from the thigh to the lungs, although was extremely small and therefore not discovered at the time of or prior to presentation. No abnormality was seen on the presenting chest radiograph or CT scan, although histological analysis confirmed the presence of micrometastatic lesions, suggesting that chest radiographs and CT scan may be suboptimal in such situations. Taken together the etiology of the pneumothorax was less clear in the present case than in previous other cases described. In this case, the treatment response was also more difficult to attain. Spontaneous pneumothorax may be the first and only evidence for metastases from angiosarcoma or any other tumor. Pneumothorax may also be the only symptom with infectious or infiltrative pulmonary disease. Our report highlights the importance of carefully considering an underlying pathology in patients presenting with spontaneous bilateral pneumothoraces.
References


