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High-grade primary myxoid lung sarcoma presenting as recurrent hemorrhagic pleural effusions in a young woman

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Abstract

Primary lung sarcomas are rare but aggressive tumors accounting for less than 0.5% of all lung tumors. The diagnosis of primary lung sarcoma should only be considered after exclusion of other sites. A 32-year-old female presented with recurrent hemorrhagic pleural effusions, shortness of breath and persistent cough. Pleural effusion was drained twice, and each time its analysis was normal. Patient developed atelectasis of left lung with hemothorax for which she underwent video-assisted thoracoscopic surgery. A large mass was found compressing the entire lower lobe of left lung with extension into mediastinum, the biopsy of which showed myxoid sarcoma. The tumor was inoperable and options of chemotherapy or radiotherapy were discussed with the patient. Primary lung sarcoma can rarely present with recurrent hemorrhagic pleural effusion. A high degree of suspicion is required for early diagnosis as large hemothorax on computed tomography or chest X-ray may obscure lung mass and make its diagnosis difficult.

Keywords

Primary lung sarcoma, hemorrhagic pleural effusion, myxoid sarcoma, hemothorax

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Background

Primary lung sarcoma is a very rare but aggressive tumor representing less than 0.5% of all malignant lung tumors.[1] Because of the rarity of lung sarcoma, very little is known about the clinical behavior and treatment of these tumors. Most of the patients affected are young and present with persistent respiratory symptoms such as cough, hemoptysis, chest wall pain and dyspnea. The presence of an infiltrate or a mass is the most common finding on chest X-ray (CXR). Diagnosis of primary lung sarcoma requires careful exclusion of primary sarcoma elsewhere as soft tissue sarcomas can rarely metastasize to lungs. Lung and pleural cancers are known to cause hemorrhagic pleural effusions, but primary lung sarcoma presenting as recurrent hemothorax only has rarely been recognized before. We present a case of 32-year-old female who presented with recurrent hemorrhagic pleural effusions and was diagnosed with lung sarcoma.

Case presentation

A 32-year-old female presented to emergency department with worsening shortness of breath and fatigue. Patient was 1 week postpartum, and while she was pregnant, it was noted that she had left-sided pleural effusion which was drained. Pleural effusion was bloody and culture and cytology were negative at that time. Decision was made to follow up the patient postpartum. Patient ended up presenting to the emergency room (ER) on two separate occasions during pregnancy complaining of shortness of breath, and each time it was noted then that she had recurrent pleural effusions.

After delivery, patient presented again complaining of shortness of breath and fatigue. On examination, vitals were as follows: temperature, 36.8°C; pulse, 93 bpm; respiratory rate, 18/min; blood pressure, 119/81 mmHg; O_2 sat, 95% on

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room air. Chest examination revealed absent breath sounds on left lung base on auscultation. Rest of the physical examination was unremarkable. Following evaluation in the ER, she was found to have recurrence of large left-sided pleural effusion on CXR (Figure 1), so she was admitted on the floor for further evaluation and treatment. Patient did not have any headache, chest pain, palpitations, cough, hemoptysis or weight loss. She also denied any lumps or mass anywhere on her body. Also there was no history of musculoskeletal deformity or pains. Past medical history was significant for gastroesophageal reflux disease (GERD), asthma and depression. The only medications she was on were off and on ibuprofen, prenatal vitamins and zantac. Patient denied use of alcohol and illicit drugs but did have a history of cigarette smoking for 17 years. Family history was positive for breast cancer in sister and diabetes and hypertension in father. There was no history of tuberculosis and exposure of asbestos.

A pigtail catheter was placed for drainage of left pleural effusion. Pleural fluid was again bloody and repeat pleural fluid analysis did not reveal any infectious or malignant etiology. Computed tomography (CT) scan of chest with contrast showed large partly loculated left pleural effusion increasing in volume as compared to previous CT chest done 1 month ago (Figure 2). Diffuse ground-glass opacities and non-specific mediastinal and hilar lymphadenopathy were also seen (Figure 2). Patient was started on empirical levofloxacin and vancomycin on the suspicion of possible pneumonia. Sputum, blood and pleural fluid cultures were all negative. In the mean time, patient developed acute blood loss anemia due to non-resolving hemorrhagic pleural effusions for which multiple blood transfusions were given. During the course of hospital stay, patient developed worsening atelectasis of left lung with large hemothorax (Figure 3). In the setting of above findings, a suspicion of left lung mass obstructing the bronchus was made and patient was scheduled for left video-assisted thoracoscopic (VATS) exploration for hemothorax evacuation and biopsy of possible mass. During the procedure, the left lobe appeared to be completely collapsed with a large tumor appearing to compress and occupy the entire left lower lobe space. Due to the size, extent and location of the tumor being unknown and both the previous imaging scans being negative for a tumor, the procedure was converted to posterolateral thoracotomy as determining the extent and the type of this lesion was of utmost priority. A stat frozen biopsy was taken from the tumor and sent for pathologic examination, which confirmed the specimen was consistent with sarcoma but was

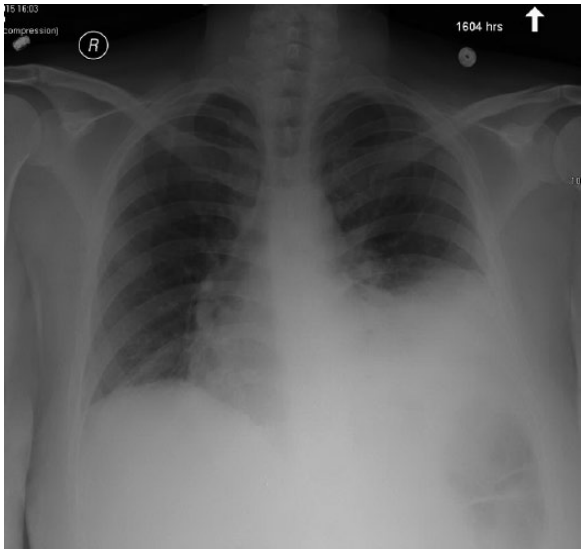

Figure 1. Portable CXR done on the day of admission shows moderate left pleural effusion and left basilar atelectasis.

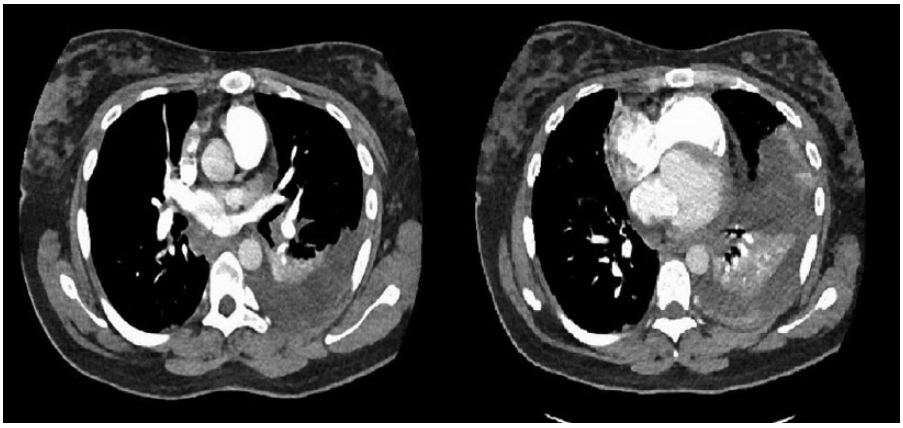

Figure 2. CT scan of chest with contrast showing large left pleural effusion with associated areas of airspace opacity related to consolidation and/or atelectasis. Fluid at the left cardiophrenic sulcus appears to be loculated. Mediastinal and hilar lymph nodes are enlarged. Also note malrotation of heart with the left ventricular apex directed laterally rather than anteriorly, possibly due to mass effect.

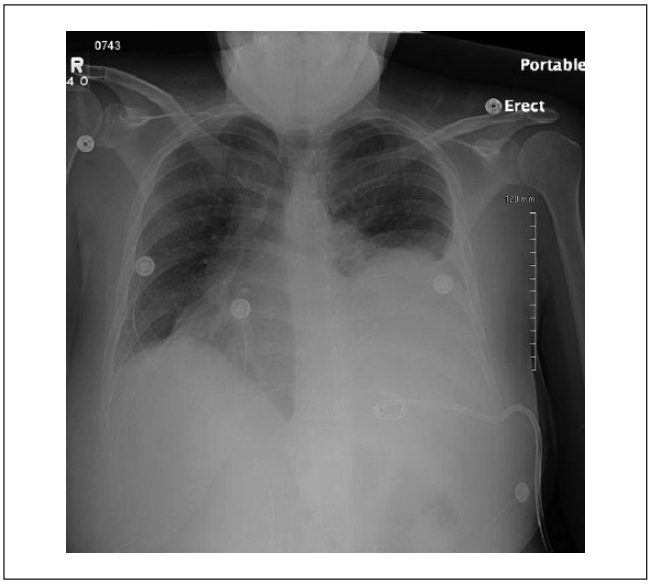

Figure 3. Large left pleural effusion with compressive left lower lobe atelectasis. Chest tube is in place.

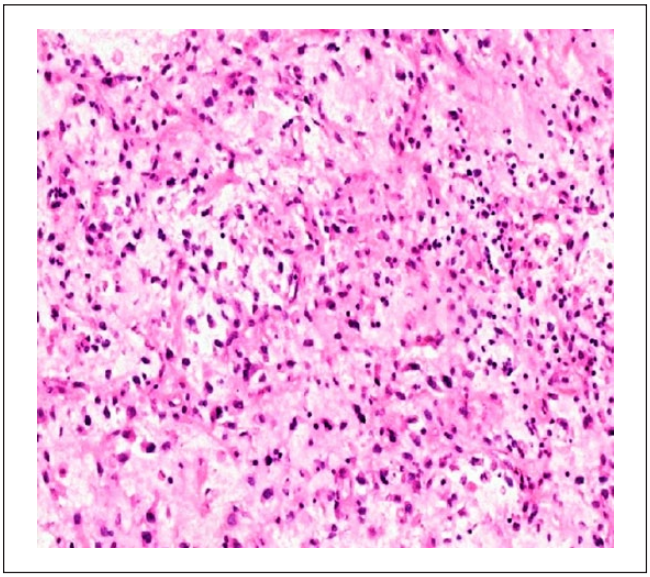

Figure 4. H&E staining of lung biopsy specimen showing myxoid background, poorly differentiated cells and high mitotic figures.

not able to definitively define it without further testing with an additional specimen, which was provided. Significant vascularity was noted and the tumor appeared to be bleeding even with partial manipulation. Due to the large size of the tumor, extension of the tumor into mediastinum as well as the preliminary diagnosis, the decision was made not to proceed with any further attempts at resection due to low chances of successfully completely resecting the tumor and the potential for significant bleeding and complications. Chest cavity was evacuated and hemostasis was achieved.

Pathology results confirmed the diagnosis of high-grade myxoid lung sarcoma (Figure 4). Immunohistochemical staining was positive for Vimentin and negative for S100, desmin, keratin, MUC4 and CD34. CT of abdomen and pelvis and skeletal survey of extremities were done to find any primary sarcoma, but all radiological investigations were normal. Patient was counseled about the inoperable nature of tumor, and discussion of chemotherapy and radiotherapy as possible alternatives was made with the patient. She was started on multi-drug chemotherapy and regularly followed up with an oncologist. After 1 month, the patient presented again with shortness of breath, nausea, vomiting and fever. Repeat CT chest was done which showed progressive mediastinal shift to the right and interval development of nodular hypertrophy or pleural thickening along the left major fissure, suggesting progression of malignant disease (Figure 5). There was also mass effect on heart and trace pericardial effusion (Figure 5).

Discussion

Soft tissue sarcoma is a rare mesenchymal tumor which usually presents in young adults most commonly in the soft tissues of the extremities, but rarely these tumors can arise in neck, lung, heart, mediastinum and abdominal wall sites.[2] Soft tissue sarcomas are classified into various types depending on their tissue of origin. Sarcomas are named according to normal tissue the tumor most closely resembles; however, in some cases, histology is uncertain and sarcoma is designated based on morphological appearance and architectural pattern of cells. The term undifferentiated sarcomas are used for sarcomas which lack specific line of differentiation and do not fit into any other category. Prognosis is determined by location, grade, stage and histological subtype. Primary sarcoma of lung is a very rare but aggressive tumor of lung with varied presentations. The most common presenting complaints are dyspnea (42%), chest pain (39%), cough (32%) and hemoptysis (22%).[3] Primary sarcomas of the lung grow through the parenchyma and in this way can reach a large size without any symptoms. The symptoms are usually limited and majority of pulmonary sarcomas are located within the lung parenchyma and pleura, but due to the aggressive nature of tumor, invasion of mediastinum, heart and chest wall may occur.[4,5] Necrosis and hemorrhage within tumor can rarely occur.

Radiologically, lung sarcoma most commonly presents as a lung mass and mediastinal shift may occur in the presence of an extremely large mass.[5] CT scan can further help to evaluate the borders and content of lung mass. Sputum examination and bronchoscopy are of low yield because tumor usually spreads through parenchyma and rarely invades bronchial wall.[6] This highlights the importance of biopsy (fine-needle aspiration cytology (FNAC) or VATS biopsy) to reach the final diagnosis. Since soft tissue sarcomas can spread to lungs, further clinical and imaging investigation should be done to exclude alternative primary

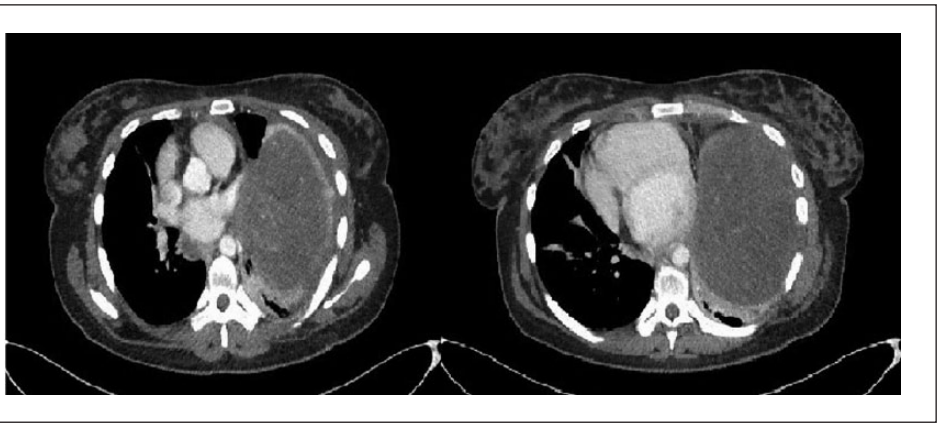

Figure 5. Large heterogeneous enhancing mass arising in the left lower lobe with metastatic nodular pleural thickening along the left hemidiaphragm, left hemithorax and left major fissure, new since the previous examination. There is progressive mediastinal shift and mass effect upon the heart and trace pericardial effusion.

sources.[7] Lung synovial sarcoma (SS) frequently metastasizes to regional lymph nodes which can lead to hilar or mediastinal lymphadenopathy. Distant metastasis via blood is rare and can involve adrenals, brain, bone marrow and spinal cord.[8]

Due to rarity of disease and little data about the presentation and treatment of these tumors, no guideline regarding management of lung sarcoma is available. Surgical resection through either lobectomy or pneumonectomy is considered the treatment of choice.[9] Complete resection with negative margins is usually curative for small, well-differentiated primary lung sarcomas as the main prognostic factor is the ability to achieve a complete resection which can improve survival.[10] Poorly differentiated tumors are less curable, but resection may provide palliation and should be considered. The role of adjuvant radiochemotherapy is unclear since there are no randomized controlled trials available due to rarity of disease. Radiation therapy or chemotherapy is reserved for those who have unresectable or recurrent sarcomas although responses are variable.

Our patient presented with recurrent hemorrhagic pleural effusions, the cause of which was unknown. She was admitted for the evaluation of shortness of breath and had atelectasis and hydrothorax. VATS surgery was performed and a big mass was found obstructing the left bronchus, which was biopsied. Surgical resection was not done due to increased vascularity and locally advanced nature of tumor. It was difficult to differentiate sarcoma on histology into any specific World Health Organization (WHO) subtype due to undifferentiated appearance of cells. Myxoid pattern was appreciated on biopsy with poorly differentiated cells and high mitotic figures (Figure 4), so sarcoma was designated as high-grade myxoid sarcoma. Differential diagnosis of primary lung sarcoma included sarcomatoid carcinoma of the lung, which are poorly differentiated non-small-cell cancers that contain a sarcoma or sarcoma-like component. They are very rare pulmonary neoplasms that exhibit a composition by spindled or pleomorphic tumor cells. Due to this reason, they may be confused easily with true sarcomas diagnostically unless special immunohistological or ultrastructural analysis is performed. Biopsy and immunohistochemical studies helped us to rule out sarcomatoid carcinoma of lung. Extensive workup was done to rule out any primary sarcoma elsewhere in body, but all radiological tests including CT scan of abdomen and pelvis came back normal with no source of primary foci. Based on history and investigations, diagnosis of primary lung sarcoma was made and patient was started on chemotherapy, but unfortunately patient presented again after few weeks and this time in a worst condition. Repeat CT chest showed progression of sarcoma with involvement of pleura and heart, thus revealing a highly malignant nature of primary lung sarcoma.

Conclusion

Primary lung sarcoma is a very rare but aggressive tumor which can be difficult to diagnose due to its parenchymal involvement without invading bronchial wall and its unusual histological features. Recurrent hemorrhagic pleural effusion and hemothorax is a very rare complication which usually occurs when the tumor is well advanced and involves surrounding structures. Surgical resection with tumor-free margins is the treatment of choice, but resection may be difficult in highly malignant and advanced sarcomas. Radiotherapy or chemotherapy may be helpful in such cases. Prognosis is poor, with an overall 5-year survival rate of 50%. Our case not only demonstrates the rarity of the sarcoma found but also stresses the significance of how its clinical presentation may be confounded by the patient's young age, risk factors and even imaging findings.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

Ethics approval

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Informed consent

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

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