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Spontaneous secondary pneumothorax with concomitant malignant pericardial effusion in the context of metastatic synovial sarcoma

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SUMMARY

Synovial sarcoma is a rare malignancy that commonly metastasises to the lungs, lymph nodes and more infrequently to the heart. It is associated with an elevated risk of pneumothorax. In this case, we report a case of dual pathology in a metastatic synovial sarcoma patient. The patient not only presented with a pericardial effusion but also with a secondary pneumothorax. A bedside echocardiogram was performed quickly, and the pericardial effusion was diagnosed early. Diagnosing the pneumothorax was delayed as the chest X-ray was not expedited but the patient was treated with an intercostal catheter before complications ensued. In the context of chest pain in patients with metastatic synovial sarcoma, we argue that conducting an early bedside echocardiogram and chest X-ray is paramount to avoid potential life-threatening complications. Concurrent lung disease and recent chemotherapy administration should also raise the clinician's suspicion of pneumothorax in such cases.

BACKGROUND

Synovial sarcoma is a rare soft tissue sarcoma that has an incidence of 1.42 cases for every 1 million adults in the United States.¹ Synovial sarcoma is a misnomer; the cancer cells do not originate from intra-articular synovium but from primitive mesenchymal cells from any part of the body.² The most common site of metastases are regional lymph nodes and the lung³ but metastases have been reported in the heart as well.⁴ These metastatic sites are especially relevant for clinicians to know, so site specific complications can be monitored and assessed adequately. The patient in this case study suffered from a pericardial effusion, in addition to a pneumothorax as a complication of synovial sarcoma metastases. First-line treatment for synovial sarcoma is complete surgical resection of the primary tumour with negative margins and additional radiotherapy based on tumour characteristics.⁵ Chemotherapeutics such as doxorubicin and ifosfamide are often used to help control distant metastases, while pazopanib (an immunotherapeutic) is used if chemotherapeutic agents fail.^{5,6} The disease has a poor prognosis, with a recent study suggesting that the survival rate is approximately 60% 5 years after diagnosis.⁷ It should be noted that in addition to lung metastases, the drugs used in the treatment of synovial sarcoma are also associated with an increased risk of pneumothorax.^{8,9}

CASE PRESENTATION

A male ex-smoker in his 30s presented to a regional emergency department with worsening chest pain, presyncope and dyspnoea on exertion on a background of treatment-resistant metastatic synovial sarcoma. The patient was undergoing palliative chemotherapy with gemcitabine/docetaxel on presentation.

His synovial sarcoma diagnosis was diagnosed 2 years prior after a biopsy was taken from a slow growing right abdominal wall mass. Histology from the biopsy revealed poorly differentiated synovial sarcoma, while a fluorescence in situ hybridisation assay revealed the presence of a known cytogenetic hallmark of synovial sarcoma, the SS18-SSX rearrangement.¹⁰ A whole-body positron emission tomography (PET) scan 2 months later confirmed that the abdominal mass was the primary location of the cancer, as there was no evidence of other avid mass lesions. The primary tumour was surgically resected 1 month after the PET scan but computed tomography (CT) surveillance imaging revealed multiple lung metastases 6 months after the surgery.

CT imaging 1 month prior to presentation demonstrated progressive disease despite immunotherapeutic treatment with pazopanib (figure 1). Further, the patient underwent a CT pulmonary angiogram 2 weeks prior to presenting which excluded a pulmonary embolus.

On examination, dual heart sounds were present but were muffled on chest auscultation. There was reduced air entry in the left lung globally and further, in the right lung base. ECG revealed sinus tachycardia.

In view of the muffled heart sounds and history of pericardial disease, a bedside echocardiogram was performed (figure 2). This showed a large pericardial effusion. Due to lack of specialist staff and facilities at the hospital, the patient was subsequently referred to an external cardiothoracic surgical team for consideration of pericardial drainage.

The chest radiograph was requested but was delayed. It demonstrated a large secondary pneumothorax with a small pleural effusion in keeping a hydropneumothorax (figure 3).

The patient subsequently underwent an urgent left intercostal catheter insertion via pigtail insertion for drainage of the hydropneumothorax.

INVESTIGATIONS

See the section case presentation.



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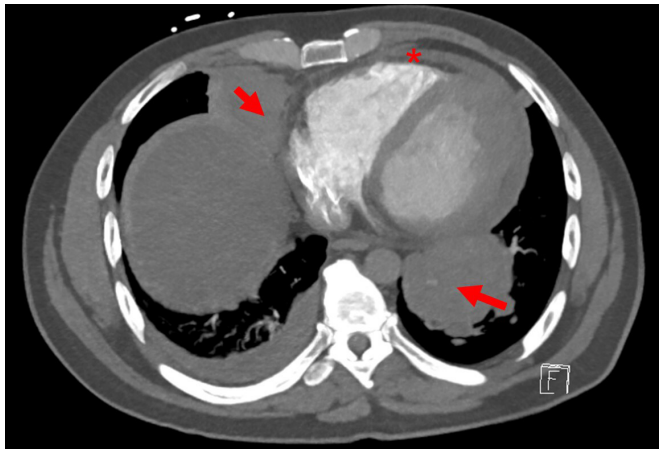


Figure 1 Axial CT chest 1 month prior to presentation showing large bilateral lower lung metastatic lesions (arrows), a small pericardial effusion (asterisk). Multiple small left lower lobe metastases can also be identified.

DIFFERENTIAL DIAGNOSIS

See the section case presentation.

TREATMENT

See the section case presentation.

OUTCOME AND FOLLOW-UP

The patient was admitted to the medical ward prior to being transferred to another facility where he underwent a surgical pericardial window and had 950 mL of fluid drained. Just prior to the procedure, the pericardial effusion was confirmed via a formal echocardiogram which demonstrated severe left ventricular systolic dysfunction, right ventricular diastolic free wall collapse and moderate to large sized pericardial effusion with early signs of tamponade.

After the procedure, the patient was then transferred back to the medical ward. However, days after he became acutely short of breath due to a newly developing right malignant pleural effusion detected on CT. The patient subsequently underwent a right intercostal catheter insertion via pigtail insertion but continued to be symptomatic after the procedure. He was subsequently referred to the palliative care unit. During his stay in the palliative care unit, the patient went to the toilet and collapsed

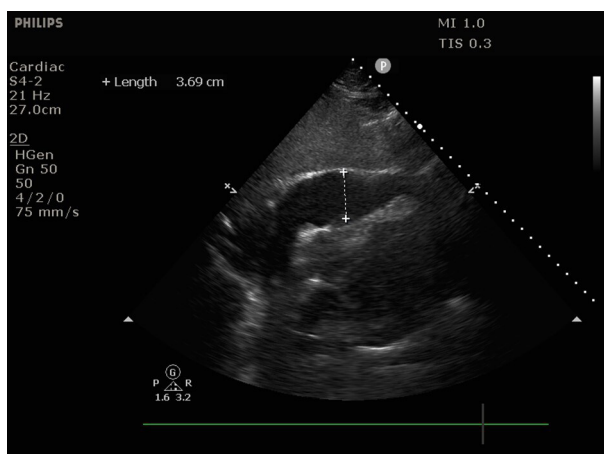


Figure 2 Bedside echocardiogram (subxiphoid approach) showing a 3.69 cm pericardial effusion.

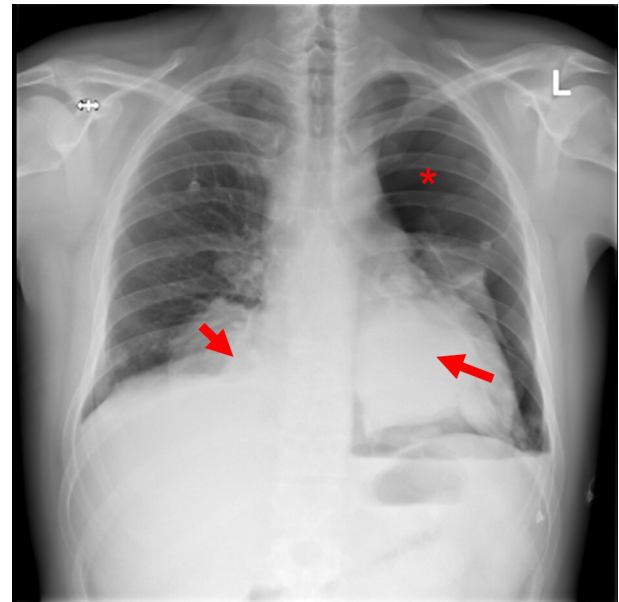


Figure 3 Chest radiograph (posterior-anterior projection) showing left and right large metastatic lesions (arrows) and large left hydropneumothorax (asterisk).

suddenly. He was pronounced deceased at the scene. Up until his last moment, he was comfortable with anxiolytics and analgesics.

DISCUSSION

It is well known that metastases from soft tissue sarcomas such as synovial sarcoma are associated with a risk of pneumothorax. The prevalence of a spontaneous pneumothorax occurring in patients with soft tissue sarcoma and lung metastases is 1.9%.¹¹ There are several ways that a pneumothorax can happen in this patient population.

1. Spontaneous rupture of a small air-filled lesion just under the pleural surface.
2. Lung disease: the most common lung disease associated with pneumothoraces is chronic obstructive pulmonary disease (COPD), which accounts for 70% of cases.¹²
3. Malignancies: there are three mechanisms postulated by the literature whereby malignancies can lead to a pneumothorax. First, a pneumothorax could occur from the spontaneous rupture of a necrotic tumour, causing a broncho-pleural communication to form. Oncological treatment could accelerate this process.⁸ Second, over distension of a tumour at the periphery of the lung could lead to the formation and rupture of subpleural bullae and last, the spread of the tumour to the pleura itself could cause a pneumothorax to form by rupturing the pleural membrane. The immunotherapeutic, pazopanib has also been associated with an increased risk of secondary spontaneous pneumothorax, and a retrospective study showed that up to 10.3% of soft tissue sarcoma patients treated with the drug developed a pneumothorax at some point.⁹

All these mechanisms could have played a part in the development of the pneumothorax in the patient. Given that the patient was an ex-smoker, he likely would have had some level of COPD in addition to known lung metastases. The patient was also on chemotherapy and consequent tumour lysis from the therapy could have caused a broncho-pleural fistula to form and precipitate a pneumothorax. However, the most likely mechanism in this patient would have been the large left metastatic lesion

invading both the left pleural and pericardial spaces causing a hydropneumothorax and effusion in each cavity to form respectively. While clinicians continue to use the bedside ultrasound to look for pericardial effusions in the context of chest pain, dyspnoea and muffled heart sounds, clinicians would do wisely to remember that soft tissue sarcomas commonly metastasise to the lung and have an increased risk of pneumothorax associated with them. A chest radiograph for this patient should have been performed at the same time as the bedside ultrasound for assessment and prevention of life-threatening illness.

Learning points

- ▶ Metastatic synovial sarcomas commonly spread to the lung and are associated with an elevated risk of pneumothorax.
- ▶ Concurrent lung disease and chemotherapeutic drugs are associated with an elevated risk of pneumothorax.
- ▶ In the context of chest pain, patients with a metastatic synovial sarcoma should have a prompt chest X-ray, in close association with a bedside echocardiogram on presentation.

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Contributors LK was the primary author of this manuscript and the guarantor, while JC acted as the supervisor of the case study. LK wrote the manuscript, designed the figures in their entirety, collected the data and conducted the literature search. JC was responsible for initially taking on the case study, and he also reviewed the manuscript multiple times while providing many useful suggestions to improve how the case study was presented. JC and LK would like to thank the patient for agreeing to partake in the case study and for allowing us to present this case to the public shortly before his death.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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