CASE REPORT

An unusual case of peripartum pulmonary oedema

Yogesh N V Reddy,¹ Varun Sundaram,² Jonathan S Stamler^{2,3}¹Department of Internal Medicine, University Hospitals Case Medical Center, Cleveland, Ohio, USA²Division of Cardiovascular Medicine, Harrington Heart and Vascular Medicine Institute, University Hospitals Case Medical Center, Cleveland, Ohio, USA³Harrington Discovery Institute, University Hospitals Case Medical Center, Cleveland, Ohio, USA**Correspondence to**

Dr Yogesh N V Reddy, yogesh.reddy@uhhospitals.org

SUMMARY

A previously healthy woman in her early 20s presented in week 38 of pregnancy with a 2-week history of haemoptysis, positional dyspnoea and an examination suggestive of pulmonary oedema. A chest CT showed a left lower lobe lung mass invading through the inferior pulmonary vein into the left atrium and abutting the posterior mitral leaflet. Pulmonary oedema consequent on left inferior pulmonary vein occlusion was initially restricted to the left lower lobe, but progressed to involve the contralateral lung following extension of the mass to the mitral orifice, resulting in functional mitral stenosis. A biopsy was positive for poorly differentiated spindle cell neoplasm. This is the first reported case of a pregnant patient with spindle cell neoplasm of lung and of a spindle neoplasm causing mitral stenosis, as well as the youngest known case of this disease.

BACKGROUND

We presented an unusual case of a young pregnant woman with spindle cell neoplasm of the lung invading the left atrium, causing functional mitral stenosis and pulmonary oedema. The interest in the case is manifold: (1) spindle cell neoplasm of the lung is thought to be a disease of elderly men, whereas our patient is a young pregnant woman; (2) the cause of spindle cell cancer and determinants of its aggressiveness are unknown; pregnancy suggests a possible hormonal link; (3) our case provides an instructive correlation between anatomy of disease and symptoms: positional dyspnoea reflecting localised (ipsilateral) pulmonary oedema caused by left inferior pulmonary vein occlusion against orthopnea reflecting bilateral pulmonary oedema caused by functional mitral stenosis; (4) an invasive tumour should be considered in the differential diagnosis of peripartum pulmonary oedema; (5) spindle cell neoplasm is identified as a rare cause of functional mitral stenosis and (6) our case presents the youngest and only known pregnant patient with this cancer.

CASE PRESENTATION

A young woman in her early 20s with no medical history presented in her 38th week of pregnancy with a 2-week history of rapidly progressing dyspnoea on exertion. Symptoms included haemoptysis, positional dyspnoea on her left side and increasing orthopnea. On admission she was afebrile with a respiratory rate 24 and heart rate 100. Pulse oximetry was 95% at room air. Her physical examination was notable for posterior left lower lobe rales and bilateral pedal oedema.

Figure 1 Left lower lobe mass with localised pulmonary oedema caused by inferior pulmonary vein occlusion. Early cephalisation is suggestive of venous hypertension.

INVESTIGATIONS

Laboratory values were unremarkable on admission. Initial imaging included a chest X-ray, which showed a left lower infiltrate or mass and signs of venous hypertension (mild cephalisation of vasculature; figure 1). Her N-terminal pro-brain natriuretic peptide (NT-proBNP) level was low, making left ventricular failure unlikely. A chest CT was negative for pulmonary embolism (PE) (excluding PE/infarct as the cause of dyspnoea and haemoptysis), but showed a large left lower lobe heterogeneous mass invading through the inferior pulmonary vein into the left atrium and prolapsing over the posterior mitral leaflet (figure 2). Ground-glass changes around the mass were consistent with localised pulmonary oedema from inferior pulmonary vein occlusion.

An urgent cesarean section was performed to enable a full workup. Following surgery, the patient became increasingly dyspnoeic with oxygen saturations in the 80s. Her clinical examination showed bilateral rales and an elevated jugular venous pressure (figure 3). A chest X-ray showed bilateral perihilar pulmonary oedema and an echocardiogram confirmed the diagnosis of functional mitral stenosis.

DIFFERENTIAL DIAGNOSIS

Severe dyspnoea and cardiogenic pulmonary oedema in pregnancy suggested the diagnosis of peripartum cardiomyopathy. Left-sided infiltrates

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Figure 2 CT scan showing a left lower lobe mass invading through the inferior pulmonary vein into the left atrium and abutting the posterior mitral leaflet (arrow).

raised the possibility of pulmonary infarction (hypercoagulable state of pregnancy). However, immediate caveats with the case included the absence of cardiomegaly on X-ray and a normal NT-proBNP. A chest CT (figure 2) and echocardiogram (figure 4) clarified the situation. Bronchoscopic and CT-guided biopsies were inconclusive before a repeat CT-guided biopsy returned the diagnosis of high-grade spindle cell neoplasm of the lung (figure 5). Analysis of placental tissue revealed metastatic spindle cell neoplasm.

The high-grade tumour had lost histological characteristics and molecular signatures of mesenchymal versus epithelial origins and was thus designated as a neoplasm of unclear origin having spindle cell morphology. Immunohistochemistry was diffusely positive for vimentin, CD99, BCL2 and WT1; focally positive for calretinin and CD10; and negative for cytokeratins, smooth muscle, skeletal muscle, neural sheath and neuroendocrine differentiation markers and germ cell markers. Synovial as well as Ewing sarcomas have been known to present in a similar fashion, but fluorescence in situ hybridization (FISH) studies were negative for SYT (18q11) translocation (synovial sarcoma) and EWRS1 rearrangement (Ewing sarcoma).^{1 2} There have been reports of mycobacterial spindle cell pseudotumour of the

Figure 3 Bilateral pleural effusions and perihilar pulmonary oedema as the mass enlarged to cause functional mitral stenosis and bilateral obstruction of pulmonary veins.

Figure 4 Transthoracic echocardiogram with four chamber and parasternal long axis views showing left atrial mass prolapsing across the mitral valve, causing functional mitral stenosis.

lung in immunosuppressed patients, but acid-fast stain was negative.³ The differential included leiomyosarcoma with spindle cell morphology and spindle cell carcinoma (a rare form of non-small cell lung cancer), either of which may have de-differentiated into a high-grade neoplasm (losing their original sarcomatous or carcinomatous features, respectively).

Figure 5 High-grade spindle cell neoplasm ($\times 40$ H&E stain).

TREATMENT

The patient was treated with diuresis and emergent radiation therapy. An objective decrease in chest mass size was noted by the third treatment. The response, however, was temporising and further staging with PET and CT revealed widespread metastases to the spleen, abdominal wall, liver, peritoneal cavity, left breast and brain.

OUTCOME AND FOLLOW-UP

The patient's course was complicated by disseminated intravascular coagulation and thrombocytopenia, leading to severe perisplenic bleeding and haemorrhagic conversion of the brain metastases with uncal herniation. An emergent craniotomy was performed for decompression. Owing to the severity of complications and dire prognosis, care was transitioned to hospice; the young woman passed away shortly thereafter.

DISCUSSION

Our case of spindle cell neoplasm of the lung in a young pregnant woman and of functional mitral stenosis as the cause of pulmonary oedema is highly unusual. To our knowledge this is the first reported case of a spindle cell lung tumour causing mitral stenosis as well as the youngest patient with this disease. The pathophysiology of dyspnoea in our patient is also noteworthy as it was caused by invasion of vascular and cardiac structures rather than of lung parenchyma. The initial left-sided positional dyspnoea (trepopnea) reflected localised pulmonary oedema caused by left inferior pulmonary vein occlusion. The mechanism underlying trepopnea is a positional increase in hydrostatic pressure (to the left lower lobe), aggravating pre-existing pulmonary oedema and ventilation-perfusion mismatch. Severe orthopnea that ensued once the cancer had invaded the left atrium was a consequence of functional mitral stenosis.

Spindle cell carcinomas are usually observed in the elderly (mean age 60), in males and in heavy smokers and thus represent an unlikely diagnosis in a young, pregnant, female non-smoker.^{4–6} However, there have been several reported cases of leiomyosarcomas originating in the pulmonary vein and invading the left atrium, with some having spindle cell morphology.^{7–9} Leiomyosarcomas occur predominantly in women and have been observed to express oestrogen and progesterone receptors, suggesting a hormonal role in pathogenesis. One possibility in our patient is that the malignancy originated as a spindle cell leiomyosarcoma and then de-differentiated into a high-grade spindle cell neoplasm. This interpretation raises the possibility of hormonal therapy for at least some of these highly aggressive malignancies. Unfortunately, we have little else to offer.¹⁰

Learning points

- ▶ Cardiogenic pulmonary oedema in the absence of left ventricular dilation and cardiomegaly on X-ray may be explained by diastolic dysfunction, mitral valve stenosis or pulmonary vein obstruction.
- ▶ Lung tumours invading through the pulmonary vein into the left atrium may present initially with trepopnea (reflecting unilateral pulmonary oedema) and later with worsening orthopnea (reflecting mitral stenosis).
- ▶ Tumour invasion of the lung should be considered as a rare cause of peripartum pulmonary oedema.
- ▶ Lung tumours that invade the pulmonary veins can be a rare cause of mitral stenosis.
- ▶ Spindle cell neoplasm, a rare cancer of elderly males, may also be observed in younger women.

Contributors All three authors were involved in manuscript preparation, editing and literature review.

Competing interests None.

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