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Fatal tumour pulmonary embolism

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Abstract

A 30-year-old female with no significant past medical history was referred to our facility with sudden onset of shortness of breath. She had a low clinical probability for pulmonary thromboembolism and a computed tomography angiogram showed enlarged pulmonary arteries but no in situ thrombi. She developed recurrent episodes of hypotension and hypoxia, and was transferred to the intensive care unit where she died despite active resuscitation. An autopsy revealed extensive lymphatic and pulmonary vascular tumour emboli as the immediate cause of death. Pulmonary tumour embolism is a very rare cause of death, but can occur in patients who have an occult neoplasm.

Introduction

Pulmonary tumour embolism is a rare complication of malignant disease and very occasionally presents as the initial and the sole manifestation of a primary malignant disease.

Case Report

A 30-year-old female working as a courier and previously well was referred to our facility with acute onset of severe dyspnoea and central chest pain. She presented with a 4-day history of dry cough and malaise, which had not responded to prescribed antibiotics. There were no known risks for pulmonary thromboembolism; history of preceding loss of consciousness, trauma, weight loss, or fever; or significant past personal and family medical histories.

The findings on examination included a tachycardia (120 beats/min), tachypnoea (32 breaths/min), hypoxia on room air, normal blood pressure, and apyrexia. The chest and cardiac exam findings were unremarkable, apart from an accentuated second heart sound. The arterial blood gas analysis while using an oxygen face mask (providing 40% oxygen) showed a mild respiratory alkalosis (pH, 7.51; partial pressure of carbon dioxide (P_aCO_2), 2.9 kPa; and bicarbonate of 21.9 mmol/L) with a partial pressure of oxygen (P_aO_2) of 18.8 kPa. Her electrocardiogram showed a sinus tachycardia and features of right ventricular strain.

The rapid HIV antibody test was negative and the full blood count, liver function tests, urea, creatinine, electrolytes, and International Normalised Ratio were normal. The d-dimer was 0.34 mg/L (normal < 0.25 mg/L), fibrinogen was 1.2 mg/L (range: 2.0–4.0 mg/dL), and the C-reactive protein was mildly elevated. An echocardiogram showed severely dilated right heart chambers with elevated right ventricular systolic pressures of 115 mmHg (range: 15–25 mmHg) and moderately impaired right ventricular systolic function. There was paradoxical interventricular septal bowing which was consistent with the right ventricular overload and/or elevated right ventricular end-diastolic pressure. The pulmonary artery was severely dilated. The left heart was normal. Doppler ultrasound of the lower limbs and abdomen did not show evidence of thrombi; however, lymph nodes were noted in the porta hepatis and para-aortic region.

The chest radiograph showed features of pulmonary hypertension, namely, an enlarged right atrium, prominent pulmonary outflow tract, enlarged pulmonary arteries, and

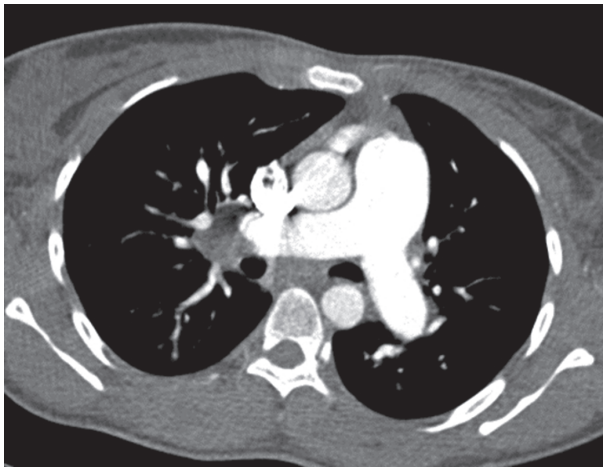

Figure 1. An axial contrasted computed tomography scan of the chest showing enlarged main pulmonary trunk suggesting pulmonary hypertension and extensive mediastinal lymphadenopathy.

pruning of peripheral pulmonary vessels with widening of the mediastinum. The computed tomography (CT) pulmonary angiogram confirmed features of pulmonary hypertension, while also revealing extensive mediastinal adenopathy. Significantly, the CT scan did not show evidence of intraluminal pulmonary thrombi or features of parenchymal lung disease (Fig. 1).

During the next 24 h, she developed cardiovascular instability characterized by recurrent discrete episodes of hypotension and hypoxia without arrhythmia, necessitating intermittent non-invasive positive pressure ventilation and fluid resuscitation with good inter-episode clinical response. After the second such episode, she was thrombolysed for suspected new massive pulmonary thromboembolism and admitted to the intensive care unit. On the 65th hour from hospital admission and after a period of clinical stability, she suffered a recurrent episode of hypotension and hypoxia, and demised despite prolonged resuscitation.

A post-mortem examination macroscopically showed generalized lymphadenopathy and prominent small pulmonary vessels. Microscopic examination of the enlarged mediastinal lymph nodes showed a poorly differentiated adenocarcinoma, which was also found in the peripancreatic and peri-adrenal soft tissue and invading the diaphragm. Examination of the lungs demonstrated multiple organized tumour emboli occluding the pulmonary vessels (Fig. 2).

Immunohistochemistry performed was positive for CK20 and negative for CK7, which is typical for colorectal adenocarcinomas. Macroscopic examination of the gastrointestinal system, however, failed to localize a macroscopic lesion in the gastrointestinal tract. In addition, CDX-2 (usually positive in colorectal adenocarcinomas) tested negative in this patient. The post-mortem was unable to identify conclusively a primary location for this disseminated malignancy.

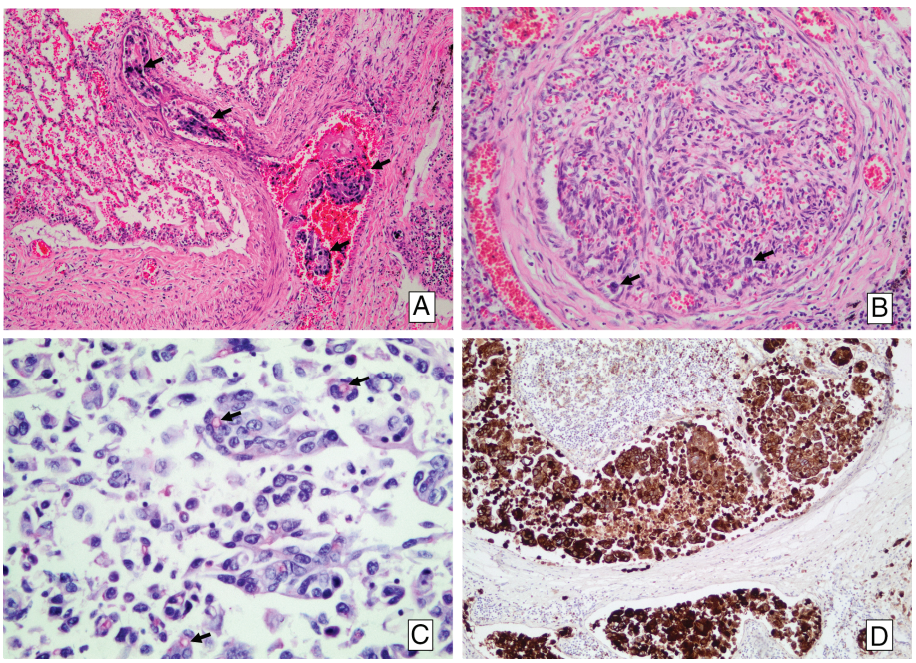

Figure 2. Microscopic slides of sections of the lungs showing intravascular collections of fibrin and markedly anaplastic tumour cells consistent with poorly differentiated adenocarcinoma. (A) A slide showing tumour cells with associated thrombus in the pulmonary vessels (arrows). Haematoxylin and eosin stain. Original magnification 100×. (B) A slide showing recanalization of a thrombosed pulmonary blood vessel. Note the individual tumour cells (arrows). Haematoxylin and eosin stain. Original magnification 200×. (C) A slide illustrating intracytoplasmic mucin vacuoles in tumour cells (arrows). Periodic acid-Schiff stain treated with diastase. Original magnification 400×. (D) A slide illustrating positive staining (brown stain) of tumour cells with an immunohistochemical stain (cytokeratin cocktail) for AE1/AE3 in keeping with a carcinoma rather than a lymphoma or melanoma. Original magnification 100×.

Discussion

Pulmonary tumour embolism refers to the identification of tumour within pulmonary blood vessels on pathological lung samples. Invasion of the surrounding interstitium is not typically seen, a feature that distinguishes it from pulmonary metastases. It remains a rare and poorly described complication of malignant disease, and its underlying pathophysiology is not completely understood [1].

The risk of tumour emboli appears to be greatest with renal cell and hepatocellular carcinoma as well as mucin-secreting adenocarcinomas of the breast, lung, stomach, and colon [2]. It usually, but not exclusively, occurs in patients with an established diagnosis of malignant disease.

Antemortem diagnosis is challenging in clinical practice and most information on tumour emboli have been obtained from autopsy series or case reports [3]. Clinical features are non-specific. Findings on special investigations are also non-specific. Patients may demonstrate electrocardiographic signs of right-sided heart strain and right ventricle hypertrophy. Arterial blood gases may reveal hypoxia with an associated respiratory alkalosis due to hyperventilation. Most imaging studies are insensitive for making a diagnosis, particularly in the early phase. Chest radiographs and CT are frequently normal, and pulmonary arteriography is usually non-diagnostic. Ventilation–perfusion lung scans may be normal or may demonstrate multiple, small, peripheral, subsegmental perfusion defects with normal ventilation. The optimal diagnostic approach is not known. In suspected cases, histological diagnosis has been pursued with a transbronchial lung biopsy, surgical lung biopsy, or aspiration of pulmonary artery blood using a right heart catheter [4].

Treatment is directed at the underlying malignancy and supportive care. Pulmonary tumour emboli is typically an end-stage manifestation of malignancy, and the prognosis with or without therapy is generally poor after onset of symptoms [5].

The patient was a young, previously healthy female with no suggestive medical or family history. She presented with rapid onset fatal cor pulmonale due to pulmonary tumour embolism from an occult carcinoma of an unknown origin, which is extremely rare. Most cases of pulmonary tumour embolism occur in patients with established malignancies.

Disclosure Statements

No conflict of interest declared.
Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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