Pulmonary artery sarcoma masquerading as pulmonary thromboembolism

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INTRODUCTION

Pulmonary thromboembolic disease is a common acute medical problem. Several investigative tools are now available which assist in diagnosis. It is important to remember that both common and uncommon disorders may mimic pulmonary embolism. This article describes a case where the initial presentation was suggestive of pulmonary thromboembolic disease, but where this was subsequently found not to be so.

DISCUSSION

Primary pulmonary artery sarcoma is an uncommon condition, rarely diagnosed ante mortem. It was first described by Mandelstamm (1923), and there have subsequently been about one hundred case reports. Clinical features mimic those of pulmonary thromboembolic disease but include weight loss as in this patient. Plain chest radiography, perfusion scanning and computed tomography do not provide discriminating diagnostic features. It has been observed that perfusion defects remain static in time in cases of pulmonary artery sarcoma (Myerson et al, 1976), whereas one would expect this not to be the case. Histologically the tumour shows inconsistent degrees of differentiation. It is postulated that the dysplastic cells originate from primitive mesenchymal cells of the bulbus cordis (Baker and Goodwin, 1985).

Spiral computed tomography (CT) is becoming more widely available and assists in the diagnosis of pulmonary thromboembolism. Perfusion scanning is often used as a primary investigation, although its usefulness is sometimes questioned, especially in patients with intrinsic lung pathology (Holbert et al, 1999). When compared with the 'gold standard' of pulmonary angiography, the specificity and sensitivity of spiral CT is greater than 90% (Van Rossum et al, 1996). It is, however, operator dependent and is less useful in detecting peripheral emboli.

In this case an uncommon condition was incorrectly but confidently diagnosed as pulmonary thromboembolism. The disparity between the CT findings and the post-mortem findings are of interest. The extent of the disease may have been overestimated on CT, or possibly thrombus in the superior vena cava was effectively thrombolysed.

The prognosis for pulmonary artery sarcoma remains universally poor. Analyses have looked at combinations of surgery, radiotherapy and chemotherapy. Although it has been suggested that surgical intervention gives some increase in survival time, the overall prognosis, despite intervention, is nearly always fatal (Anderson et al, 1995).

CONCLUSION

Pulmonary artery sarcoma is an uncommon and diagnostically challenging condition. This case mimicked a presen-

CASE REPORT

A 46-year-old Caucasian female presented as an emergency with a 6-month history of dyspnoea. This had become much worse over the preceding 3 weeks. There was also a history of lethargy, right-sided pleuritic chest pain and 7 kg weight loss. She had suffered an anterior myocardial infarction 4 years before admission with minimal angina subsequently.

In the weeks before admission, cyanosis had been noted and she had been commenced on an angiotensin-converting enzyme inhibitor for what was felt to be deteriorating left ventricular function. This was in addition to aspirin, atenolol and a long-acting nitrate preparation. There was no personal or family history of thromboembolic disease.

On arrival she was unwell with severe dyspnoea, blood pressure of 130/60 mmHg and central cyanosis. Her jugular venous pressure was elevated to over 10 cm, she had a right ventricular heave, bilateral pleural effusions and late inspiratory crackles in the right mid-zone. On room air her arterial partial pressure of oxygen was 6.45 KPa. The electrocardiogram showed sinus rhythm, right axis deviation, prominent P waves and anterior Q waves. Chest radiograph showed a prominent right hilum and bilateral pleural effusions.

A clinical diagnosis of extensive pulmonary thromboembolic disease was made and a spiral computed tomogram (CT) of the thorax with contrast was undertaken (*Figure 1*). This showed filling defects in the right and left lower pulmonary arteries, in the right main pulmonary artery partially occluding the middle lobe branches and extending into the posterior segmental branches of the right upper lobe. A large amount of material was noted in the superior vena cava.

Immediate thrombolysis with tissue plasminogen activator was administered, with low molecular weight heparin, warfarin and high flow oxygen. The patient remained stable for the next 48 hours, but then became obtunded with lateralizing neurological signs. A CT scan of her brain confirmed a large frontal haematoma with intraventricular extension, blood tracking to the brainstem and evidence of coning. She died within 12 hours of this event.

At post-mortem the main pulmonary arteries and pulmonary trunk were occluded by soft yellow mucoid tissue, with similar tissue throughout the intraparenchymal branches of the pulmonary arteries. Histological examination of this tissue showed appearances consistent with pulmonary artery sarcoma. The post-mortem also confirmed a primary spontaneous intracerebral haemorrhage. Examination of the heart showed right ventricular enlargement and dilatation, left ventricular dilatation, coronary artery atherosclerosis, and anteroseptal left ventricular scarring. There was no material in either the inferior or superior vena cavae.

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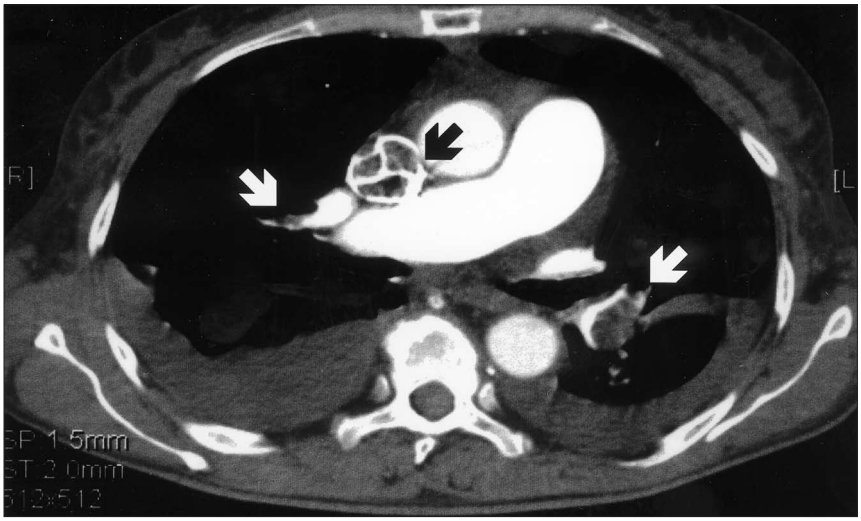

Figure 1. Spiral computed tomogram of the thorax showing filling defects in pulmonary arteries (white arrows) and superior vena cava (black arrow).

tation of pulmonary thromboembolism and highlights the fact that while imaging modalities can expedite administration of definitive treatment, they are not without limitations. It also reminds us that in an age where clinicians may hesitate to request a post-mortem, had this not been carried out an incorrect final diagnosis would have been made. **HM**

Anderson MB, Kriett JM, Kapelanski DP, Tarazi R, Jameson SW (1995) Primary pulmonary artery sarcoma: A report of six cases. *Ann Thorac Surg* **59:** 1487–90

Baker PB, Goodwin RA (1985) Pulmonary artery sarcoma. *Arch Pathol Lab Med* **109:** 35–9

Holbert JM, Costello P, Federle MP (1999) Role of spiral computerised tomography in the diagnosis of pulmonary embolus in the emergency department. *Ann Emerg Med* **33**(5): 520–8

Mandelstamm M (1923) Ueber primaere Neubildungen des Herzens. *Virchow Arch Pathol Anat* **245:** 43

Myerson PJ, Myerson DA, Datz R, Lawson JP (1976) Gallium imaging in pulmonary artery sarcoma mimicking pulmonary embolism: case report. *J Nucl Med* **17:** 893–95

Van Rossum AM, Pattynama PM, Ton ER et al (1996) Pulmonary embolism: validation of spiral CT in 149 patients. *Radiology* **201:** 467–70