Breathlessness and a widened mediastinum

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An 82-year-old woman with ischaemic heart disease presented with gradually increasing breathlessness. She had no clinical signs of heart failure. Her chest radiograph showed an aneurysmal proximal aorta. Echocardiography showed slightly impaired left ventricular function and aortic valve thickening with trivial reflux. There were widespread electrocardiographic changes characteristic of ischaemia. She was treated with an angiotensin-converting enzyme inhibitor.

She presented again five years later with progressive tiredness, breathlessness and dizziness culminating in acute severe dyspnoea and palpitations. She was in atrial fibrillation (heart rate 136 beats/min), had an elevated jugular venous pulse and bilateral basal pulmonary crepitations. There were no cardiac murmurs. Despite control of the atrial fibrillation and cardiac failure with digoxin and diuretics her dyspnoea steadily worsened. She became breathless at rest, had a dry cough and lost weight. There were now no signs of cardiac failure. Results of blood gas analysis were: pH 7.47, pO₂ 8.34 kPa, pCO₂ 5.13 kPa. Her chest radiograph (figure 1) showed further widening of the mediastinum. Echocardiographic findings were unchanged. Thoracic computed tomography (CT) (figure 2) and lung perfusion scintigraphy (figure 3) were performed. VDRL was negative.

Figure 1 Chest radiograph

Figure 2 Contrast enhanced thoracic CT

Figure 3 Lung perfusion scintigram

Questions

- 1 Describe the CT findings.
- 2 List 10 causes of a widened mediastinum.
- 3 What is the most likely diagnosis and what is the differential diagnosis?

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Answers

QUESTION 1

The thoracic CT showed a large aneurysm of the aortic arch and root with a calcified rim and multiple layers of organised mural thrombus. The right pulmonary artery was stretched around the aneurysm posteriorly and greatly compressed. There was also minor compression of the right main bronchus. Lung windows showed hypoperfusion of the right lung which was confirmed by lung perfusion scintigraphy.

QUESTION 2

Causes of a widened mediastinum are listed in box 1. These are classified conventionally into anterior (to the heart), middle and posterior.

QUESTION 3

This patient's symptoms are explained by hypoperfusion of the entire right lung due to compression of the right pulmonary artery by an enlarging ascending aortic aneurysm. Such an aneurysm may also cause dyspnoea by bronchial compression (a minor feature in this case) or rupture into the vena cava or pulmonary artery creating a fistula and left-to-right shunt. Pulmonary embolism should also be considered.

Discussion

Thoracic aortic aneurysms may produce symptoms by compression or erosion of various

structures including the superior vena cava, trachea and bronchi, oesophagus, pulmonary and coronary arteries. The right pulmonary artery is particularly prone to compression by ascending aortic aneurysms because, at its origin, it is situated immediately posterior to the aortic root and is enclosed with it in a common extension of the pericardium.^{1,2}

At the time of previous British reports,³ *ante-mortem* diagnosis was difficult and relied upon invasive pulmonary and aortic angiography. Digital subtraction angiography, CT, magnetic resonance imaging and isotope studies may be used, but CT is now generally regarded as the imaging method of choice. Unlike angiography, it shows the full size of the aneurysm and any involvement of other structures, as well as being less invasive and allowing sequential monitoring. Transoesophageal echocardiography has been advocated⁴ but this is more invasive and less readily available than CT in the UK.

Although rare, pulmonary artery compression by thoracic aortic aneurysms has been reported in specialist literature. It may complicate ascending (usually), arch or descending limb aneurysms, dissecting or non-dissecting, and those of atherosclerotic, syphilitic and post-stenotic aetiology. The presentation depends on the rate at which the compression develops. If this is acute, such as after aortic dissection, it may mimic massive pulmonary embolism. A more slowly enlarging aneurysm may produce progressive dyspnoea and cough. Signs of right heart failure may develop in either case. Most reported cases have had acute symptoms resembling pulmonary embolism.^{2,5,6} Many presenting subacutely have displayed features of right heart failure^{4,7} but, as this case demonstrates, these features are not always present.^{1,3}

A 'high probability' ventilation/perfusion lung scan indicates pulmonary embolism in the vast majority of cases (91% positive predictive value if no previous history of pulmonary embolism),⁸ but as seen here, it may have other causes. Unilateral absent lung perfusion with normal ventilation and contralateral perfusion would conform to this classification but this picture is uncommon in pulmonary embolism which usually produces bilateral abnormalities.^{2,5} It is more likely to be due to pulmonary artery compression. The causes of unilateral pulmonary artery oligoemia are given in box 2.

Causes of a widened mediastinum

Anterior

- thyroid tumour
- thymic mass
- germinal cell neoplasm
- sternal tumour
- pericardial fat pad
- diaphragmatic hernia
- pericardial cyst

Middle

- carcinoma of bronchus
- bronchogenic cyst

Posterior

- neurogenic tumours
- paravertebral masses: abscess, myeloma, reticuloses, metastases
- anterior thoracic meningocoele
- dilated oesophagus
- neurenteric cyst
- hiatus hernia

Any region

- aortic aneurysm
- lymphadenopathy

Diffuse

- superior vena cava obstruction: mediastinal tumour, chronic inflammatory mediastinitis, idiopathic mediastinal fibrosis
- extensive tumour: lymphomas, metastases
- oedema: cardiac tamponade, tricuspid stenosis, leaking aneurysm, dissecting aneurysm

Box 1

Causes of unilateral pulmonary oligoemia¹

- external compression of pulmonary artery: aortic aneurysm, bronchial carcinoma, fibrosing mediastinitis, constrictive pericarditis
- massive pulmonary embolism
- unilateral pulmonary emphysema (Macleod's syndrome)
- obstructive hyperinflation
- congenital pulmonary artery hypoplasia

Box 2

Pulmonary artery compression by an aortic aneurysm should be suspected when there is:

- apparent pulmonary embolism with a widened mediastinum,⁵
- lack of improvement in the perfusion scan following anticoagulation,⁶
- right ventricular hypertrophy combined with aortic insufficiency,⁶ or
- dyspnoea out of proportion to signs of cardiac failure accompanied by a widened mediastinum and/or evidence of aortic valve regurgitation.

Improved detection creates a potential for surgical correction and success has been reported.^{1,3,4} However, the major thoracic

surgery required was not considered a realistic option for our patient. She was treated with warfarin anticoagulation and palliative oxygen. Five months later she is housebound but otherwise well and functionally independent.

Final diagnosis

Hypoperfusion of right lung due to compression of right pulmonary artery by a large atherosclerotic ascending aortic aneurysm.

Keywords: thoracic aortic aneurysms, pulmonary artery compression, dyspnoea

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Retroperitoneal cystic mass

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A 48-year-old woman was admitted to hospital with continuous pain in her left flank radiating to the left groin and leg. Her blood pressure was 135/75 mmHg, and pulse rate 80 beats/min. Her chest and abdominal X-rays and electrocardiogram were normal. On clinical exploration, a subcutaneous mass was palpable in the left lumbar region. It was about 10 cm in diameter, smooth, not painful and non-adherent to skin. Abdominal ultrasound showed a cystic and polylobulated mass in the retroperitoneal space. No evidence of a hepatic lesion was found. An abdominal computed tomography (CT) scan was performed (see figure).

Figure CT scan showing left retroperitoneal cystic mass

Question

What is the most likely diagnosis?

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