An unusual cause of dyspnoea in an 83-year-old woman

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An 83-year-old woman was referred for assessment of rapidly worsening dyspnoea, limiting mobility to 10 yards. She had a history of multiple potential sources of her dyspnoea: coronary artery disease (previous stenting of the right coronary artery, a chronic total occlusion of the mid left anterior descending artery, and 50% stenosis in the circumflex with normal left ventricular function), chronic obstructive airways disease requiring inhaled bronchodilators and steroids, and chronic renal impairment with a mild anaemia. She had also been noted to have a stable aneurysm of the ascending aorta (4.5 cm in diameter) at last angiography 2 years before.

Examination at 45° revealed central cyanosis with oxygen saturation of 88% on room air but normal jugular venous pressure and clear lung fields to auscultation. On lying flat, the oxygen saturation improved to 92%. Ventilation perfusion lung scan indicated a low probability for pulmonary emboli. Chest X-ray showed widening of the superior mediastinum with the enlargement of the cardiac silhouette. Computed tomography (CT) aortogram showed an ascending aortic aneurysm with a maximum diameter of 6.8 cm (Panel A), substantially larger than the last measurement 2 years before. Transthoracic echocardiography showed a markedly dilated ascending aorta impinging on the right atrial free wall resulting in a functional tricuspid stenosis (mean gradient 5 mmHg), with contrast bubble study demonstrating a right-to-left shunt through a patent foramen ovale (PFO) (Panels B and C). Hyperoxic pulmonary function testing showed a shunt of 24%. The patient declined aortic surgery and was discharged on home oxygen, but agreed to consider palliative percutaneous PFO closure. However, 2 days later, she presented with chest pain. Repeat CT now showed dissection of the enlarging ascending aortic root aneurysm. The patient requested active measures be withdrawn and she died a few hours later.

This case illustrates an unusual mode of presentation for an ascending aortic aneurysm: marked hypoxia with cyanosis due to right-to-left intracardiac shunting resulting from right atrial compression. There were multiple other possible causes of dyspnoea including coronary ischaemia, left ventricular systolic or diastolic dysfunction, worsening of airflow limitation, anaemia, or fluid overload from chronic renal impairment. However, hypoxia with clear lung fields on examination and on chest X-ray suggested intracardiac right-to-left shunting as the cause.

Intracardiac right-to-left shunting can be interventricular but usually occurs across the atrial septum through an atrial septal defect or more commonly through a PFO and only when the right atrial pressure exceeds the left atrial pressure. Isolated elevation of the right atrial pressure is usually due to pulmonary hypertension but may also occur with obstruction to right atrial outflow (due to right atrial myxoma, tricuspid stenosis, or localized right ventricular dysfunction) and extrinsic compression of the right atrium (due to tumour, localized pleural or pericardial effusion, or, as in this case, due to aortic root enlargement).

Patapnoea-orthodeoxia syndrome (where dyspnoea and cyanosis are characteristically worse in the upright posture and improved by lying supine) is a rare condition usually where there is distortion of the interatrial septum, opening a PFO, which is exacerbated by erect posture. Distortion of the interatrial septum may result from right pneumonectomy/lobectomy or abnormal aortic root anatomy. Enlargement of the ascending aorta rotates the heart anti-clockwise and may result in anterior displacement of the superior limbus of the foramen ovale into the right atrial cavity, causing a PFO to be held open while compression of the right atrium raises pressure, promoting right-to-left shunting. The effects of gravity in the erect position cause further anterior and inferior displacement of the aortic root increasing PFO aperture size and right atrial compression resulting in increased shunting and worsening of hypoxia.

Panel A Computed tomography scan of the thorax in the sagittal plane demonstrating the ascending aortic aneurysm (arrow) with a maximum diameter of 6.8 cm.

Panel B Transthoracic echocardiogram from the subcostal view before bubble contrast injection showing the atrial pacing lead (arrow) in the right atrium, but no contrast in either atrium.

Panel C Similar view on transthoracic echo after bubble contrast injection showing right-to-left shunting across a patent foramen ovale (white arrow), as a result of the right atrial compression, with contrast filling the right atrium (RA) but also seen in the left atrium (LA).

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