CASE REPORT

An unusual pulmonary embolus

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Abstract

Due to advances in paediatric congenital heart surgery in recent years, the number of patients who survive into adulthood with complex congenital heart disease has increased remarkably. When these patients present to non-specialist hospitals with apparently specific symptoms, the diagnosis may not be as straightforward as initially thought. Here we highlight a case which demonstrates this.

Key words: stented valves, pulmonary embolus, congenital heart disease.

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Introduction

Increasing numbers of patients with complex congenital heart disease are surviving into adult life. Patients may present to non-specialist hospitals with new problems. In this report we describe an unusual case of pulmonary embolus.

Case report

A 30-year-old woman presented to the Emergency Department of her local hospital with a 24-hour history of chest pain and shortness of breath. The chest pain was sharp, centrally located and was radiating to her neck and back.

At 12 months of age the patient had been diagnosed with a congenital heart abnormality consisting of a complex pulmonary atresia, multiple aorto-pulmonary collaterals and a ventricular septal defect (VSD). She underwent numerous surgical procedures throughout childhood, including operations to ligate the aorto-pulmonary collaterals, insertion of a left Blalock Taussig shunt (later closed) and attempted closure of her VSD. A Rastelli operation was performed when she was 16 years old; a homograft conduit was inserted from the right ventricle (RV) to the pulmonary artery (PA). Subsequent to that, she had further surgery to reconstruct the right ventricular outflow tract and RV/PA conduit using a vascular graft.

Significant pulmonary valve regurgitation had led to progressive right-sided heart failure. Nine months prior to this admission she had had a stented valve implanted within the RV/PA conduit, and this resulted in an improvement in her symptoms.

She had developed a deep vein thrombosis 13 years previously and was taking warfarin. Until this presentation she had been fully independent, with an exercise tolerance greater than one mile.

Initial examination found her to be apyrexial, tachypnoeic (respiratory rate 28 per minute), peripherally cyanosed and distressed. She had a heart rate of 110 beats per minute (bpm) in sinus rhythm and a blood pressure of 110/60 mmHg. An ejection systolic murmur was found, with a laterally displaced apex beat, right ventricular heave and inspiratory bibasal crepitations. Pulse oximetry revealed an oxygen saturation (SpO_2) of 86% on air, rising to 94% on 15 litres of oxygen delivered via face mask.

Her ECG showed sinus rhythm with a rate of 84 bpm and (old) right bundle branch block. Chest X-ray showed cardiomegaly and the presence of a metallic prosthesis in the right ventricle.

Arterial blood gas analysis on air was pO_2 6.08 kPa, pCO_2 3.95 kPa and SaO_2 85.4%. Haemoglobin was 13.8 g/dL, white

Figure 1. Ventilation perfusion scan, showing abnormal filling defects

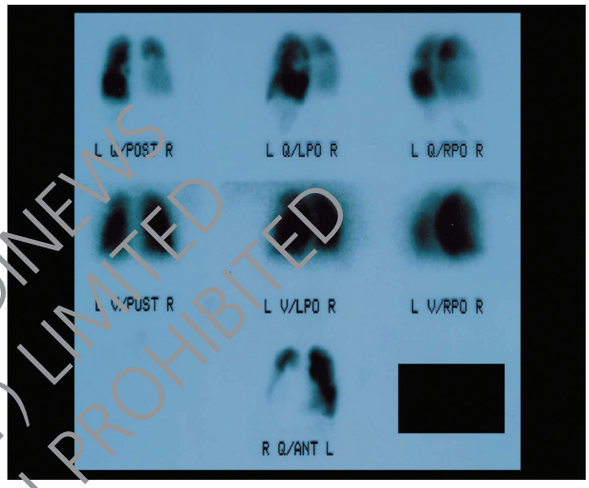

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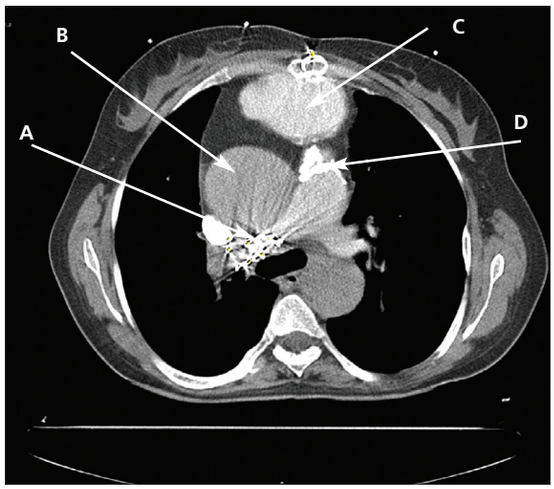
Figure 2. CT scan of patient's chest on admission



Key: CT = computerised tomography; A = stented valve shown displaced into right pulmonary artery; B = left ventricle; C = right ventricle; D = calcification within right ventricle/pulmonary artery conduit

Key messages

- Increasing numbers of patients with complex congenital heart disease are surviving to adult life
- An element of lateral thinking must be applied if they present with cardiorespiratory problems
- Consult a cardiac centre at an early stage

cell count 4.7 x 10^9/L, platelets 156 x 10^9/L and cross-linked fibrin degradation products (FDP) 300 ng/ml (normal range 0–500 ng/ml). Urea, electrolytes and creatinine concentrations were in the normal range. The international normalised ratio was 1.8 (on warfarin), lactate dehydrogenase 532 IU/L (normal range 100–190 IU/L) and C-reactive protein < 5 mg/L.

A diagnosis of pulmonary embolus was made based on the history and clinical findings. This was confirmed with a ventilation perfusion (V/Q) scan, which demonstrated V/Q mismatching in both lung fields (figure 1). She was prescribed tinzaparin and her warfarin dose was increased. She remained hypoxaemic over the next 48 hours, with an SpO_2 of 89% on 10 litres of oxygen via face mask. Her symptoms improved over the following 24 hours but she still remained hypoxaemic.

Transthoracic echocardiography showed a possible thrombus in the right ventricle. Four days after her initial admission, she was transferred to this hospital (Royal Brompton Hospital) for further investigation and management.

On admission, her transthoracic echocardiogram was repeated: it demonstrated a dilated right ventricle and right atrium with poor right ventricular function. Doppler studies of both lower limbs and the inferior vena cava did not reveal any thrombus. Computed tomogram (CT) of her thorax was performed (figure 2).

The CT scan revealed sternotomy wires and a partially calcified conduit. The stented valve which had been placed nine months previously was within the right pulmonary artery.

After allowing for suboptimal opacification, differential opacification was observed between the left and right pulmonary arteries distal to the stent (not shown). In addition, several filling defects were identified within the sub-segmental arteries. The heart was enlarged and a small VSD was identified, with contrast seen passing into the left ventricle and outlining one of the aortic valve leaflets. The appearances were highly suggestive of stent migration with associated obstruction of the right pulmonary artery.

Her management occurred in two stages. First, in the catheter laboratory, the stented valve was snared with biopsy forceps (via the right femoral vein) and repositioned for easier access in the right ventricle. The patient was then transferred to the operating theatre, where she underwent repeat sternotomy, removal of the PA conduit and the retrieval of stented valve. A new biological valved conduit was inserted (18 mm in size, [coronary stents range from 2–5 mm]) and the VSD was closed. She made a quick post-operative recovery and was discharged 10 days following her operation.

Discussion

Right ventricle to pulmonary artery homografts and bioprosthetic conduits have been used to palliate various types of complex congenital heart disease, including truncus arteriosus, tetralogy of Fallot, pulmonary atresia and complete transposition of the great arteries with VSDs.

The principal benefit of stent implantation lies in prolonging conduit life span (limited by the development of progressive lumen obstruction) and increasing the interval between the almost inevitable re-operations.[1] Conduit stenting may, however, aggravate pulmonary insufficiency. Bonhoeffer *et al*. developed a system for percutaneous stent implantation combined with valve replacement in the pulmonary position.[2] The patient described in this report had a stented valve inserted within the RV/PA conduit that subsequently migrated distally and caused partial obstruction of the right pulmonary artery.

Powell *et al.* performed a retrospective review of 44 patients who underwent placement of stents in obstructed RV-PA conduits.[3] Of the 44 patients, 16 returned for follow-up catheterisation because non-invasive assessments of RV pressures suggested progressive re-stenosis. Seven were found to have stent fractures and all were asymptomatic. In three of these patients, stent fragments embolised to the pulmonary circulation: no evidence of significant pulmonary blood flow obstruction was seen and all embolised fragments were left in place. Proximal displacement of a stent into the RV cavity was

discovered in one patient. There were no cases of distal displacement into the pulmonary artery.

In another series by Ovaert *et al.* distal migration was observed but this was apparent at the time of insertion and another stent was implanted across the obstruction.[4]

In the case presented here, displacement of the stented valve proved to be symptomatic and potentially life-threatening, presenting as a pulmonary embolus with normal levels of D-dimers. Displacement and embolisation of part or all of a bioprosthesis should be considered in such patients.

Conflict of interest

None declared.

References

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