CASE REPORT

A patient with severe hypoxia secondary to a large iatrogenic pulmonary artery to pulmonary vein fistula

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SUMMARY

Pulmonary arteriovenous malformations are uncommon communications between the pulmonary arteries and veins, most commonly associated with hereditary haemorrhagic telangiectasia. They can also be associated with a variety of other conditions, and can be single or multiple. We present a case of a female patient with a history of coronary artery bypass grafting and mitral valve repair, who presented to the hospital with severe hypoxia. She was found to have a large pulmonary artery to superior pulmonary vein fistula that was successfully repaired using a septal occluder. To our knowledge, this is the first case of a large pulmonary artery to superior pulmonary vein fistula following mitral valve repair.

BACKGROUND

Pulmonary arteriovenous malformations (PAVMs) are uncommon communications involving pulmonary arteries and veins. They are most commonly congenital, and can be associated with a variety of conditions, including hereditary haemorrhagic telangiectasia, liver cirrhosis, mitral stenosis, trauma, schistosomiasis, actinomycosis, Fanconi's syndrome and metastatic thyroid carcinoma.[1] Iatrogenic causes have rarely been reported. We present the following case report of a patient with an iatrogenic symptomatic single large right pulmonary artery to superior pulmonary vein fistula following mitral valve repair that was treated successfully with a septal occluder. To our knowledge, this is the first case of a large pulmonary artery to vein fistula that likely developed and was acquired post mitral valve repair surgery.

CASE PRESENTATION

A 71-year-old woman with a history of mitral valve repair for mitral stenosis, associated with coronary artery bypass grafting (CABG) 10 years prior, presented with a 2-week history of progressively worsening dyspnoea associated with chest pain to the emergency department. She had a limited functional status at baseline with dyspnoea on minimal exertion and hypoxaemia requiring 3 L/min of oxygen by nasal cannula continuously. The patient's medical history was significant for chronic kidney disease stage 2 and peripheral vascular disease, which was managed with both medical therapy and stenting of the femoral arteries. She was an ex-smoker who had quit 10 years prior at the time of her surgery. Vitals showed a blood pressure of 170/101 mm Hg, heart rate 90 bpm and respiratory rate 25 breaths/min. Oxygen saturation was 80% on a 100% non-rebreather mask. Physical examination was notable for cyanosis without respiratory distress; her jugular vein was 8 cm above the sternal angle; cardiovascular and pulmonary examination showed a 3/6 holosystolic murmur over the left lower sternal border (she had a 1–2/6 holosystolic murmur reported in the past) with clear lungs and 1+ lower extremity oedema. There was no hepatomegaly, splenomegaly or finger clubbing.

INVESTIGATIONS

Pertinent laboratory results were as follows: arterial blood gas showed a pH of 7.48, an arterial pressure of CO_2 28 mm Hg, an arterial pressure of O_2 38 mm Hg on 100% via non-rebreather mask; haemoglobin was 14.7 g/dL, brain natriuretic peptide 2590 pg/mL and serum creatinine 1.1 mg/dL. Chest roentgenogram and CT are shown in figure 1A, B. No pulmonary embolism or aetiology of the hypoxia was reported on initial read of the chest CT. No parenchymal lung abnormalities were seen. The patient was diuresed aggressively with Lasix 40 mg intravenously twice daily for 48 h, and started on a nitroglycerine patch based on cardiology recommendations, and given her elevated brain natriuretic peptide, jugular venous distention and lower extremity oedema. Oxygen saturations did not improve despite extensive diuresis and remained in the 80–85% range. A transthoracic echocardiogram demonstrated normal left ventricular systolic function with moderate right ventricular dilation and preserved bioprosthetic mitral valve function. Bubble study was positive after the third heartbeat. Transoesophageal echocardiogram revealed the absence of cardiac shunting. Right heart catheterisation showed normal pulmonary artery pressures, with no evidence of shunting.

DIFFERENTIAL DIAGNOSIS

This is a patient with severe hypoxia in the absence of parenchymal lung abnormalities. She had a positive bubble study after the third heartbeat. Differential diagnosis included a chronic obstructive pulmonary disease (COPD) exacerbation, cardiac or pulmonary right-to-left shunt, hypoventilation, pulmonary hypertension or pulmonary embolism. COPD exacerbation was ruled out by the absence of parenchymal lung abnormalities on CT of the chest and the absence of wheezing with clear lung sounds on chest examination. Hypoventilation, pulmonary

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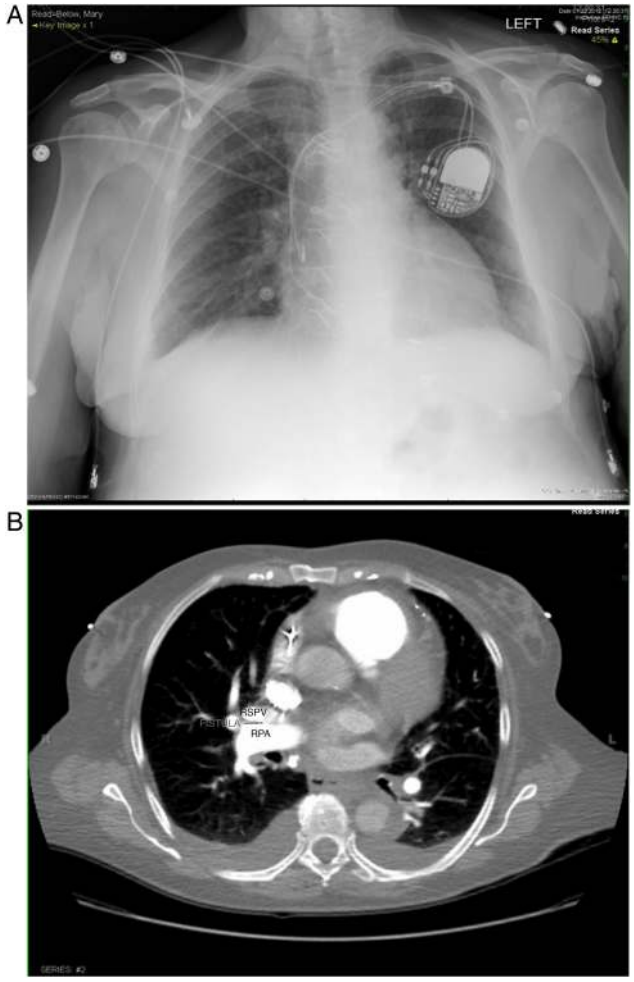

Figure 1 (A) Chest roentgenogram showing clear lungs. (B) Chest CT with pulmonary embolism protocol. The fistula is labelled (RSPV, right superior pulmonary vein; RPA, right pulmonary artery).

embolism, pulmonary hypertension and cardiac right-to-left shunting were respectively ruled out by the respiratory alkalosis on the arterial blood gases, the negative contrast-enhanced CT scan of the chest, the normal right heart catheterisation and the absence of a cardiac shunt on transoesophageal echocardiography. Review of the CT scan images with interventional radiology revealed a 1.1 cm right pulmonary artery to superior pulmonary vein fistula, which was not detected on initial CT scan read (figure 2). This was confirmed with angiography.

TREATMENT

Surgery and embolotherapy were deemed high risk given the size and location of the PAVM and the fact that the patient had undergone a previous chest surgery. Therefore, the patient underwent placement of an Amplatzer septal occluder (figure 3) with marked improvement in dyspnoea symptoms and hypoxaemia.

OUTCOME AND FOLLOW-UP

The patient required 3 L of supplemental oxygen continuously by nasal cannula on discharge to maintain oxygen saturations above 90% and remains stable 2 years later.

DISCUSSION

This case illustrates the importance of considering a PAVM as a diagnostic consideration in patients with unexplained hypoxia.

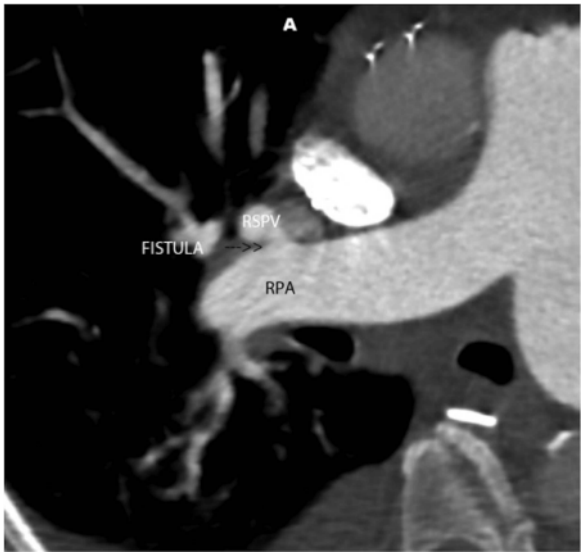

Figure 2 Close-up view of the pulmonary artery to superior pulmonary vein fistula (RSPV, right superior pulmonary vein; RPA, right pulmonary artery).

There may be several explanations for the development of the large PAVM in this patient. The patient could have had a small congenital PAVM that progressively enlarged because of several factors: mitral stenosis leading to increased pulmonary venous resistance, stress of her cardiac surgery or Swan-Ganz catheter placement.[1–3] Alternatively, the PAVM could have been iatrogenic with progressive enlargement over time, secondary to the CABG and mitral valve repair surgery or to intraoperative pulmonary artery catheter placement with pseudoaneurysm formation.[2 3]

The patient had been becoming progressively more hypoxaemic and dyspnoeic over the past years. Her most recent oxygen requirement 2 months before her acute presentation was 3 L/min by nasal cannula. The acute worsening of her hypoxaemia could be explained by enlargement of her PAVM to a critical diameter leading to acute dilation of her fistula. This could have been exacerbated by a stressful event.

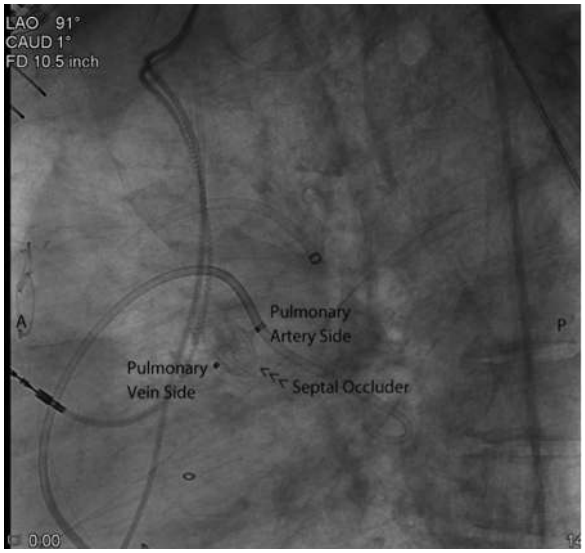

Figure 3 Amplatzer septal occluder placed closing the fistula.

While embolotherapy remains the mainstay of treatment for PAVMs, it can be associated with significant embolic complications, including migration of the embolic device, transient ischaemic attacks, angina pectoris and cerebral infarction, in large PAVMs (>1 cm in diametre).[4 5] Surgery may be high risk in certain patient populations, especially in a patient who had previous cardiac surgery and has limited functional capacity and multiple comorbidities. Placement of a septal occluder, which acts to occlude the fistulous connection between the pulmonary artery and vein, may be successful in these situations.[6 7] This is demonstrated in our case.

Learning points

- A pulmonary arteriovenous malformation (PAVM) should always be in the differential diagnosis of unexplained hypoxaemia with a normal chest X-ray.
- Most PAVMs are congenital; however, a history of previous mitral stenosis, cardiac surgery or Swan-Ganz catheter placement can lead to increased vascular resistance and progressive enlargement of PAVMs.
- PAVMs can rarely occur after coronary artery bypass and valvular repair surgeries, and should be considered in cases of unexplained hypoxia following these surgeries.
- Embolotherapy is the mainstay of treatment for PAVMs, but can lead to significant embolic complications in large PAVMs.
- Amplatzer septal occluders may be effective non-surgical options in cases of large PAVMs.

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Contributors KD has been involved in the diagnosis and management of this case and has written and edited the manuscript for submission. RG was instrumental in the diagnosis and management of the case and has provided the images for this article, and has contributed to its review. KD is the guarantor.

Competing interests AC has been involved in this case and has presented it as an oral presentation at the American College of Chest Physicians conference in 2012.

Patient consent Obtained.

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