Haemoptysis: just another case of endocarditis?
A case report

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Background
Pulmonary arteriovenous malformations (PAVM) are rare, and most cases are congenital. They require prompt recognition and management particularly in patients presenting with hypoxia and haemoptysis. We describe a unique case of recurrent endocarditis causing pulmonary artery aneurysms (PAAs) and formation of PAVM.

Case summary
A 60-year-old woman presented with dyspnoea, haemoptysis, and severe hypoxia. Her background was significant for previous pacemaker lead infection, refractory heart failure secondary to severe tricuspid valve distortion by her pacemaker lead, tricuspid and mitral valve replacements complicated by recurrent endocarditis over several years. Two years prior to her current presentation computed tomography (CT) scanning revealed new small PAAs thought possibly to be mycotic in origin. After her current presentation, prompt high-resolution CT scanning of her chest with contrast revealed significant pulmonary haemorrhage and new clusters of PAVM. Urgent pulmonary angiography confirmed PAVM and was successfully treated with coil embolization. Her dyspnoea, pulmonary haemorrhage, and hypoxia resolved.

Discussion
Acquired causes account for a very small percentage of PAVM and the mechanism of their development is unknown. As she had recurrent right-sided endocarditis and her PAAs developed following this, with new PAVM developing 2 years later; we hypothesize that they were causally related. We believe this is the first case of recurrent left- and right-sided endocarditis leading to formation of PAAs and development of PAVM presenting with significant hypoxia and haemoptysis requiring prompt intervention.

Keywords
Endocarditis • Heart failure • Pulmonary arteriovenous malformations • Valvular disease • Case report

Learning points
• Understand common aetiologies of pulmonary arteriovenous malformations, diagnosis, and treatment.
• Consider recurrent endocarditis as a rare cause of acquired pulmonary arteriovenous malformation that may be life-threatening and amenable to definitive therapy.

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Introduction

Pulmonary arteriovenous malformations (PAVM) are rare and 80% of cases are congenital, with an estimated prevalence of 38 per 100,000 individuals.\textsuperscript{1,2} PAVM are abnormal communications between pulmonary arteries and veins without an intervening capillary bed.\textsuperscript{1,3} The presence of PAVM can be life-threatening due to severe hypoxia and large volume haemoptysis.\textsuperscript{4} We describe what is, to our knowledge, the first case of acquired PAVM secondary to recurrent endocarditis.

Timeline

| Year | Event |
|------|-------|
| 1999 | Dual-chamber pacemaker (PPM) inserted for sick sinus syndrome. Antibiotic treatment for lead infection |
| 2012 | Recurrent heart failure with severe tricuspid regurgitation and mitral regurgitation |
|      | Tricuspid valve (TV) replacement and mitral valve (MV) repair complicated by \textit{Staphylococcus aureus} bacteremia and prosthetic TV infective endocarditis (IE) |
|      | Redo TV and MV replacement, box change, and lead extraction |
| 2015–2017 | Two separate episodes of IE treated with intravenous antibiotics |
|      | New pulmonary artery aneurysms (PAAs) noted, presumed mycotic but stable |
| 2018 | Haemoptysis and cyanosis |
|      | Right lower lobe pulmonary arteriovenous malformations (PAVM) diagnosed on contrast computed tomography requiring urgent coil embolization |
|      | Aetiology of PAVM proposed to be secondary to development of PAA on a background of recurrent left- and right-sided valvular endocarditis |

Case presentation

A 60-year-old female presented in 2018 with dyspnoea and haemoptysis.

In 1999, a dual-chamber pacemaker (PPM) was inserted overseas for sick sinus and intermittent atrial fibrillation (AF) and was complicated by lead infection, treated with antibiotics without lead extraction. She remained well until 2012 where she was treated in a heart failure clinic for recurrent exacerbations. She presented with dyspnoea, in florid right heart failure and clinically severe tricuspid regurgitation (TR). A transthoracic echocardiogram (TTE) revealed normal left ventricular function, torrential TR, moderate mitral regurgitation (MR), and raised pulmonary pressures. Cardiac catheterization showed normal coronary arteries, pulmonary pressure, and pulmonary vascular resistance. Her heart failure was considered secondary to severe TR and moderate MR. She proceeded to tricuspid valve (TV) replacement with a bioprosthetic valve, mitral valve (MV) repair, and epicardial lead insertion. The original generator and lead were left in situ. Intraoperatively the PPM lead was tangled at the base of the anterior leaflet papillary muscle with severe distortion, fibrosis, and retraction of the TV.

Three weeks postoperatively she presented with sepsis and blood cultures positive for \textit{Staphylococcus aureus}. A transoesophageal echocardiogram (TOE) confirmed vegetations on the prosthetic TV (Videos 1 and 2). Following treatment with intravenous (IV) antibiotics, blood cultures were initially negative but there was a subsequent relapse of bacteremia and worsening heart failure. A redo TV replacement, MV replacement, PPM system extraction, and connection to the tunneled epicardial lead were performed.

In 2015, she was admitted with worsening heart failure, fevers, and \textit{Streptococcus salivarius} bacteremia. A TOE revealed MV vegetations; successfully treated with 6 weeks of intravenous antibiotics. In December 2016, she was admitted with a dental abscess and sepsis which was drained with benzylpenicillin coverage. One week later she presented with fevers. Blood cultures were negative, but a TOE revealed new MV vegetations. She was treated as culture-negative endocarditis. Computed tomography (CT) scan of her chest revealed two new pulmonary artery aneurysms (PAAs) in the left upper lobe, presumably mycotic aneurysms, stable on serial imaging (Figure 1). These were not present on prior CT scans in 2012.

In December 2018, she presented with haemoptysis in the context of warfarin for AF, severe dyspnoea, and fever. Examination revealed tachypnoea with a respiratory rate of 25 breaths/min with an oxygen saturation of 88% on room air. Blood pressure was 114/61 mmHg, and heart rate was 75 b.p.m. She was cyanosed with new clubbing (Figure 2), jugular venous pressure was elevated to 7 cm. Heart sounds were normal with previously noted soft systolic and diastolic murmurs across the tricuspid region. Chest auscultation showed right crepitations worse than the left with reduced air entry bilaterally at the lung bases.

Differential diagnoses included recurrence of infective endocarditis with septic emboli to the lungs causing infarction, pneumonia,
pulmonary haemorrhage with her known history of aneurysms and pulmonary embolus.

Arterial blood gases revealed Type 1 respiratory failure and respiratory alkalosis with pO$_2$ 56 mmHg, pCO$_2$ 28 mmHg, and pH 7.48. Haemoglobin (Hb) was 132 g/L, INR 2.7, and creatinine 57 mL/min/1.73 m$^2$. Chest X-ray revealed right lower zone consolidation (Figure 3).

A TTE did not reveal vegetations. Haemoptysis continued and the patient’s Hb dropped to 98 g/L. High-resolution CT chest identified two large clusters of PAVM associated with extensive ground glass changes in the right middle and lower lobes consistent with pulmonary haemorrhage (Figure 3).

She was referred for urgent percutaneous pulmonary angiography with coil embolization (Figure 4, Video 3, and Supplementary material online, Video S1). The two largest clusters were coiled successfully; however, the procedure was terminated due to radiation exposure time and contrast use. Following her coil embolization, her arterial oxygen saturation had improved to 97%, pO$_2$ 91 mmHg, and pH 7.38. She declined further treatment of the remaining PAVM and discharged well.

Video 2 Transoesophageal echocardiogram showing tricuspid valve vegetation.

Video 2 Transoesophageal echocardiogram showing tricuspid valve vegetation.

Figure 2 Insidious clubbing.

Figure 1 Computed tomography chest (A): 2012 absence of left upper lobe aneurysm. Computed tomography chest (B): 2017 development of a small left upper lobe aneurysm.
Discussion

We report a case of recurrent endocarditis as a rare, acquired cause of PAVM. The most common aetiology of PAVM are congenital and associated with hereditary haemorrhagic telangiectasia. PAVM occurs more frequently in women than men (1:1.5–1.8). Acquired causes of PAVM are uncommon and their mechanism of development is unknown. Reported aetiologies include hepatic cirrhosis.
actinomycosis and schistosomiasis infections, mitral stenosis, chest trauma, metastatic carcinoma, and thoracic surgery.1

PAVM present from asymptomatic hypoxaemia to haemorrhage. Complications of PAVM include polycythaemia, pulmonary hypertension, paradoxical myocardial infarction, stroke, and cerebral abscesses.1,5 Our patient presented with near life-threatening PAVM requiring immediate management. In patients suspected to have PAVM, the first-line test is a transthoracic contrast echocardiogram (TTCE) to detect a left to right shunt.6 TTCE has the highest sensitivity of close to 100% and lowest risk of complications amongst all the screening tests of PAVM.1,2,6 One large cross-sectional study demonstrated a strong association between shunt grade and the incidence of stroke and abscess.7 Once a shunt is identified, PAVM is confirmed with contrast-enhanced CT.4 First-line treatment of PAVM is with percutaneous pulmonary angiography and embolization, with surgical excision reserved for unsuccessful cases.1,3

Our patient had refractory heart failure with preserved ejection fraction and was medically managed for several years until severe pacemaker-related TR was documented. Surgical replacement of her TV resolved her heart failure. The frequency of developing significant TR after implantation of a cardiac device is ~10–39%.8 TV dysfunction can be lead related due to direct damage of the leaflets, papillary muscles or chordae tendinae during implantation and mechanical disruption of coaptation, and fibrosis of the TV over time.8

Our patient initially had right-sided pacemaker-related endocarditis. The next decade of her life saw recurrence of infection on both left- and right-sided heart valves, and development of new PAA. There was no evidence of PAVM on serial imaging of her chest during this time. She presented to us with insidious clubbing, hypoxia, and haemoptysis. Urgent imaging revealed large clusters of PAVM in the right lower lobe of the lung. The exact pathogenesis and mechanisms of developing PAVM secondary to infection is unknown.7 Infective foci can cause mycotic aneurysms due to extension and erosion into adjacent vascular structures; this has been described in pneumonias.9,10 In the context of endocarditis, it is postulated that septic emboli causes endovascular seeding into the lumen of the pulmonary vasculature leading to PAA formation.9–11 There are early descriptions of mycotic aneurysms causing PAVM in the literature in the context of tuberculosis and Rasmussen aneurysms rupturing into adjacent veins.12 We hypothesize that her history of recurrent endocarditis led to the development of PAA and with time, erosion and penetration of her PAAs into contiguous veins leading to formation of her PAVM. This, to our knowledge, has not been previously described.

Conclusions

Acquired PAVM are rare and the aetiologies described in the literature do not include recurrent endocarditis. Our patient had right-sided endocarditis with subsequent recurrent episodes of endocarditis leading to mycotic PAAs and we hypothesize erosion into adjacent veins leading to PAVM. Prompt management of patients with PAVM presenting with haemoptysis and haemorrhage is crucial, with pulmonary embolization being the mainstay of treatment.

Lead author biography

Ronald Huynh completed his medical studies at the University of Notre Dame in Sydney, Australia in 2013. He completed his Cardiology Advanced Training at Concord Repatriation General Hospital in Sydney, Australia in 2020.

Supplementary material

Supplementary material is available at European Heart Journal - Case Reports online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.
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