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CASE REPORT

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# Paediatric haemoptysis from multiple pulmonary artery aneurysms

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**Abstract**

The case of multiple peripheral pulmonary artery aneurysms in children presenting with haemoptysis following an episode of bacterial endocarditis is presented. They are extremely rare and clinically non-specific but critical because early diagnosis is crucial for preventing sudden death from their rupture. Computed tomography pulmonary angiography remains the investigation of choice. Despite no consensus recommendation on the management, a multidisciplinary team should plan the beneficial approach with minimal procedure-related morbidity and mortality to improve survival. This case report aimed to emphasize the recognition of this rare cause of haemoptysis in children.

**KEYWORDS**
children, haemoptysis, paediatric, pulmonary artery aneurysms

## INTRODUCTION

Paediatric haemoptysis from multiple pulmonary artery aneurysms (PAAs) is extremely rare but clinically important because their rupture can be lethal. Early diagnosis is critical for preventing sudden death. The aetiologies may be congenital associated with congenital heart diseases, acquired from vascular anomalies, vasculitis, pulmonary hypertension, post-infection or idiopathic.[1] Their natural history remains largely unknown. Here, we report a case of PAAs presented with haemoptysis after an episode of bacterial endocarditis to emphasize this rare fatal condition in children.

## CASE REPORT

A 5-year-old girl with ventricular septal defect (VSD) was admitted with acute non-massive haemoptysis and breathlessness 30 min prior to admission. She had history of recurrent non-massive haemoptysis on two occasions, about 50 ml 12 days ago and 100 ml 8 days ago. At that time, she did not have fever or respiratory distress.

She had a history of bacterial endocarditis presented with prolonged fever and congestive heart failure 2 months prior to this admission. At that time, echocardiography showed $6 \times 4$ mm$^2$ vegetation at the posterior leaflet of mitral valve with severe mitral valve regurgitation. Haemoculture was positive for *Streptococcus anginosus*, which was sensitive to penicillin G and gentamicin. Haemoculture for fungus was negative. After receiving PGS for 8 weeks combined with gentamicin on the first week, she improved and was discharged without dyspnoea or haemoptysis.

At admission, her vital signs were as follows: temperature 37°C, blood pressure 108/68 mmHg, pulse rate 100/min (full), respiratory rate 40/min and peripheral capillary oxygen saturation ($SpO_2$) 86% on room air. Physical examination showed no digital clubbing, no oedema, intercostal and subcostal retraction, coarse crepitation at both basal lungs and systolic ejection murmurs (Grade 2/6) at apex. Her complete blood count and coagulogram were normal. Her haemoglobin level was 11.8 g/L.

Chest x-ray revealed cardiomegaly with increased pulmonary vasculature and bilateral perihilar opacities with air-bronchogram (Figure 1). Regarding the vascular causes of haemoptysis, computed tomography (CT) angiogram showed multiple outpouching lesions at bilateral segmental pulmonary arteries, six aneurysms of right pulmonary artery with the largest size of $1.7 \times 1$ cm$^2$ at lateral-basal segment of right lower lung and four aneurysms of left pulmonary artery with the largest size of $1.1 \times 0.7$ cm$^2$ at apico-posterior segment of left

lower lung. Internal eccentric filling defect suggested partially thrombosed aneurysm without active contrast extravasation or calcification (Figure 2).

An echocardiogram revealed small VSD (5 mm) with left to right shunt, moderate mitral regurgitation with 6 mm gap mitral valve cleft and prolapse of anterior mitral valve leaflets; mild left atrium enlargement, left ventricular enlargement; trivial tricuspid regurgitation, right ventricular systolic pressure (RVSP) 44 mmHg; mild pulmonic regurgitation, no atrial septal defect, no patent ductus arteriosus, good left ventricle systolic function left ventricular ejection fraction (LVEF) 65%.

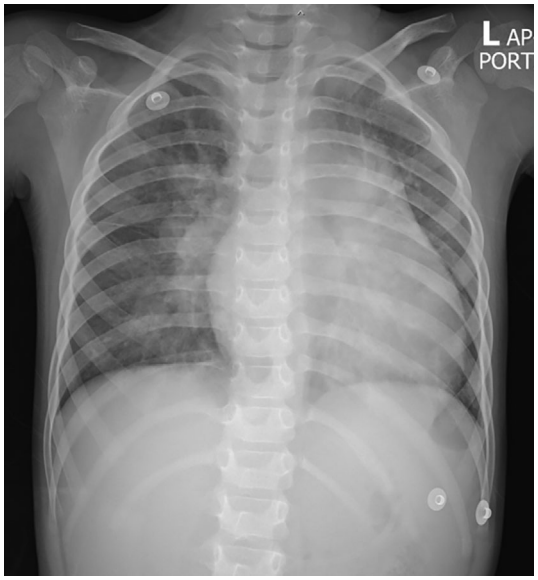


**FIGURE 1** Chest x-ray revealed cardiomegaly with increased pulmonary vasculature and bilateral perihilar opacities with air-bronchogram

The most likely cause of PAAs in this case is mycotic aneurysms after an episode of bacterial endocarditis, although there was no vegetation in the right-sided cardiac chamber or less likely as consequences from VSD. We gave her intravenous antibiotics and respiratory support with high-flow nasal cannula for 2 days. Her vital sign was stable and she did not have haemoptysis after admission. Owing to small multiple lesions, embolization was not done concerning high risk of complication.

The follow-up CT chest 1 month later showed overall slightly decreased size of multiple bilateral PAAs without active contrast extravasation. Neither new outpouching lesions nor definite conglomerate vessel, nidus or gross AV shunt was seen. It also revealed improvement of pulmonary hypertension and pulmonary haemorrhage.

During COVID pandemic, she was referred to her community hospital to continue antibiotic treatment. We plan to repeat CT chest to follow size, number and complication of her PAAs. If CT chest shows increase in size, our multidisciplinary team of intensivist, pulmonologist, cardiologist, interventional radiologist and thoracic surgeon will consider surgical intervention to prevent their rupture.

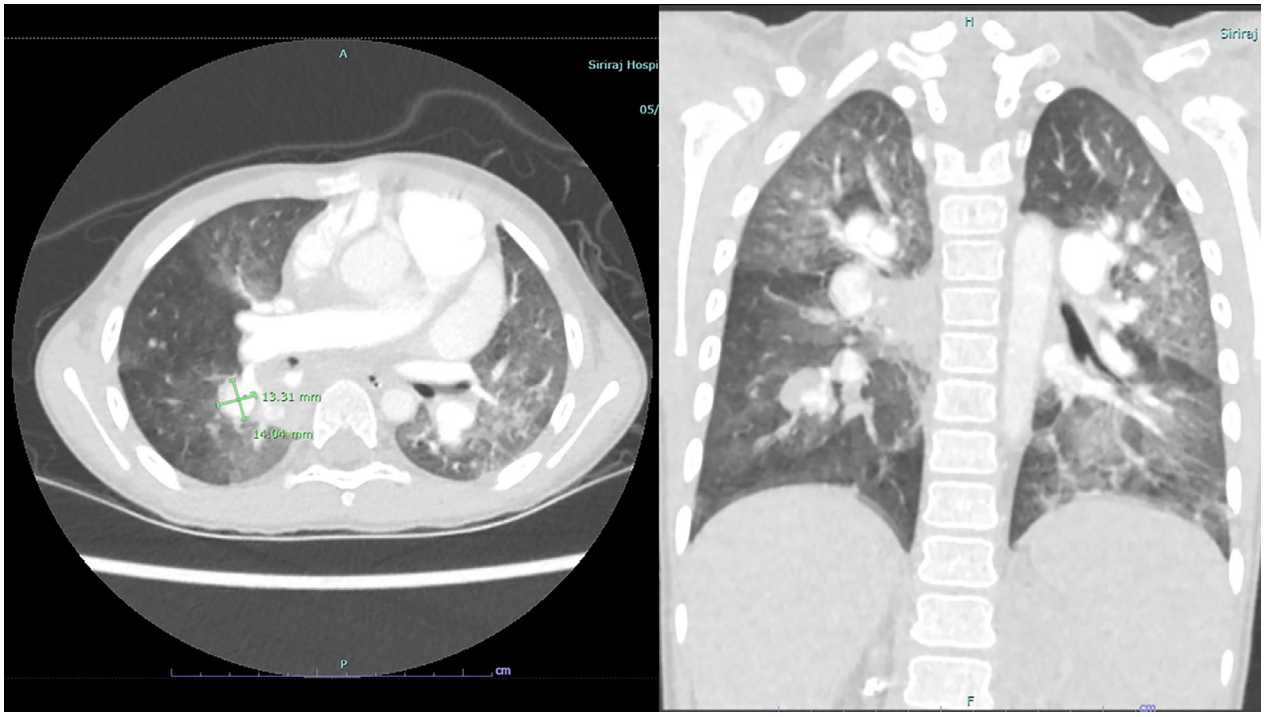


**FIGURE 2** Computed tomography angiogram showed multiple outpouching lesions at bilateral segmental pulmonary arteries; internal eccentric filling defect suggested partially thrombosed aneurysm without active contrast extravasation or calcification

## DISCUSSION

PAAs are rare with a reported incidence of 0.007%.[2] Classified by location, they are more central (89% involve pulmonary trunk or main pulmonary arteries) than peripheral (11% do not involve pulmonary trunk or main pulmonary arteries).[2] It could be congenital, acquired from many conditions including congenital heart diseases, connective tissue disorders, systemic vasculitis, infections or idiopathic.[1]

Mycotic PAA is a rare complication of right-sided infective endocarditis. Most cases were associated with congenital heart diseases or intravenous drug addiction. There was a variety of causative organisms such as bacteria including *Staphylococcus aureus* and Streptococcus species, fungi including Candida and Aspergillus, *Mycobacterium tuberculosis*, *Treponema pallidum* or Actinomyces.

The exact aetiopathogenesis of PAAs is not clearly understood, and no clear guidelines on their management are available.

The proposed pathogenesis includes a direct extension of an endoluminal septic thromboembolism into the arterial wall, as seen in cases of bacterial endocarditis as this patient. Virulent microorganisms cause extensive destruction of the layers of the arterial wall and usually result in the formation of a false aneurysm or pseudoaneurysm. On the other hand, indolent organisms often cause a true aneurysm as the focus of suppurating or cavitating pulmonary artery in tuberculosis, which is termed as Rasmussen aneurysm. Other causes include ischemic insult to the arterial wall due to infection of the vasa vasorum, as seen in syphilis.

Altered flow dynamics and increased hemodynamic shear stress on the vessel walls by the left to right shunt can also be proposed as pathogenesis of PAAs from congenital heart diseases. The most common congenital cardiac diseases associated with PAAs are patent ductus arteriosus, VSD, atrial septal defect, hypoplastic aortic valve, bicuspid aortic valve and pulmonary valve stenosis.[2,3]

Clinical manifestations vary related to the underlying aetiologies, location and size of these PAAs. Haemoptysis is the most common presenting symptom resulting from rupture of these PAAs and maybe fatal. Cough and dyspnoea are also observed when PAAs compress the surrounding airway. Fever is seen mainly in mycotic aneurysms. Sometimes, a harsh systolic murmur maybe heard over the left second or third intercostal space from pulmonic valve abnormalities. As mentioned, the most catastrophic complication of PAAs is aneurysmal rupture or dissection.

Due to its non-specific clinical presentation, awareness of this rare condition in cases with pre-disposing risk factors, requesting and detecting from CT angiogram, is important to reach the diagnosis. However, pulmonary angiography remains the gold standard for diagnosis and may be needed for therapeutic intervention. Around 20%–60% of patients with PAAs may die from aneurysm rupture while other serious complications including airway compression, intravascular thrombosis and lung infarction are also common.

The plain radiographic appearance of a mycotic pulmonary aneurysm is a pulmonary opacity, which may be either well-defined or ill-defined. There may be parenchymal consolidation as well, which is generally non-specific and cannot be differentiated from infections. Although conventional angiography was previously considered the gold standard of diagnosis, cross-sectional imaging such as contrast-enhanced CT and magnetic resonance imaging are now important alternative imaging modalities that clearly demarcate the vascular origin of the lesion from the pulmonary vessels. In our patient, CT angiogram showed multiple outpouching lesions connected to a pulmonary artery on the arterial phase and some lesions showed surrounding area of low attenuation, which was the thrombosed component.

Experience in the management of mycotic pulmonary aneurysms is limited as their diagnosis is rare and infrequently reported in the literature. Management is usually surgical and involves aneurysmorrhaphy, lobectomy, aneurysmectomy, pneumonectomy, banding, embolization and alternative endovascular techniques, such as occlusion of aneurysm with steel coils or detachable balloons.[4] Surgical resection carries a high risk of intrapulmonary haemorrhage, especially in cases with pulmonary hypertension. Endovascular therapy may best serve saccular PAAs, both central and peripheral. Meanwhile, bronchopulmonary shunting has to be ruled out before embolization. However, conservative management can be considered when there is no evidence of acute haemoptysis or other emergency symptoms, or when a patient is not a candidate for intervention.

Spontaneous resolution has been previously reported for small mycotic PAAs, but regular follow-up CT scans are still mandatory. Surgical treatment should always be undertaken to prevent catastrophic consequences such as rupture of large aneurysms. Direct ligation of the feeding vessel and endoaneurysmorrhaphy with preservation of lung tissue has been advocated as the ideal surgical technique; however, it is rarely feasible and practicable. Although the prognosis of mycotic PAAs is not well established, high mortality rates of 42%–80% without intervention have been previously reported.[5]

In conclusion, this case report highlights the clinical manifestation of multiple mycotic PAAs after bacterial endocarditis. Although rare and clinically non-specific, they warrant prompt diagnosis and treatment. CT pulmonary angiography remains the investigation of choice. Despite no consensus recommendation on their management, a multidisciplinary team approach that would prove beneficial with minimal procedure-related morbidity and mortality can improve survival.

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### CONFLICT OF INTEREST

None declared.

## AUTHOR CONTRIBUTION

Prakarn Tovichien and Phatthareeda Kaeotawee drafted the manuscript and contributed to patient care. Prakarn Tovichien contributed to revise the manuscript as corresponding author. Both authors read and approved the final manuscript.

## ETHICS STATEMENT

The authors declare that appropriate written informed consent was obtained for publication of this manuscript and accompanying images.

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