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CASE REPORT

Giant pulmonary artery aneurysm in a child: Rare complication of congenital heart disease

Samuel Sisay Hailu[1] | **Hermon Miliard Derbew**[2] | **Abrehet Zeray**[1] | **Tesfahunegn Hailemariam**[1] | **Hansel J. Otero**[2]

[1]Department of Radiology, College of Health Sciences, Addis Ababa University, Addis Ababa, Ethiopia

[2]Department of Radiology, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, USA

Correspondence
Tesfahunegn Hailemariam, Department of Radiology, College of Health Sciences, Addis Ababa University, Addis Ababa, Ethiopia.
Email: tesfa.feleke21@aau.edu.et

Key Clinical Message

This case report aims to increase awareness that pulmonary artery aneurysms may occur as a complication of neglected patent ductus arteriosus and should be sought in children with ill-treated congenital heart diseases.

Abstract

Pulmonary artery aneurysm is a rare anomaly with an autopsy prevalence of 1:14,000. These aneurysms can arise secondary to various etiologies, with congenital causes identified in 25% of cases and congenital heart diseases (CHD) responsible for more than half of these cases. A 12-year-old boy with CHD in the form of patent ductus arteriosus (PDA) and irregular clinical follow-up presented with new onset fatigue of 3 months duration. A physical examination revealed anterior chest wall bulging and a continuous murmur. A chest radiograph showed a smooth left hilar region opacity that has a close relation with the left cardiac border. Transthoracic echocardiogram shows no progression from the previous one; there was a large PDA and pulmonary hypertension, but no further information was available. Computed tomography angiography revealed a giant aneurysm of the main pulmonary artery (PA), with a maximum diameter of 8.6 cm, and dilatation of its branches of 3.4 and 2.9 cm for the right and left PA, respectively.

KEYWORDS

congenital heart disease, patent ductus arteriosus, pulmonary artery aneurysm

1 | INTRODUCTION

Pulmonary artery aneurysm (PAA) is a rare anomaly with an autopsy prevalence of 1:14,000.[1–3] These aneurysms can arise secondary to various etiologies such as congenital heart disease (CHD), connective-tissue disorders, acquired vascular diseases (infections, vasculitis), malignancy, trauma, iatrogenic causes, or, rarely, they can be idiopathic.[2,4] Most patients remain undiagnosed due to its silent course and non-specific clinical manifestations.[1,4] However, lethal complications, including pulmonary artery (PA) dissection, rupture, massive hemoptysis, and acute coronary syndrome from vessel compression, have been reported.[5] PAAs generally occur in a younger age group than aortic aneurysms, with no gender predilection.[6]

PAAs are divided into congenital, acquired (inflammatory, infectious, iatrogenic), and idiopathic causes.[6] Congenital causes are identified in 25% of cases,[5] and CHD is responsible for more than half of these cases,[6] with patent ductus arteriosus (PDA), tetralogy of Fallot (TOF), ventricular septal defects (VSD), atrial septal defects (ASD), and valvular pulmonic stenosis being the most common ones.[5–8]

Although there are multiple case reports of PAAs in the literature, very few are from Africa. A single case was reported previously from Ethiopia, which focused on management aspects.[9] Here, we present the case of a 12-year-old boy diagnosed with giant PAA as a complication of neglected PDA and discuss etiologies and diagnostic imaging approaches in resource-limited settings.

2 | CASE PRESENTATION

A 12-year-old boy with a past medical history of congenital heart disease in the form of a PDA presented to our hospital—Tikur Anbessa Specialized Hospital, Addis Ababa, Ethiopia—with a new onset of fatigue of 3 months duration. He was on a cardiac follow-up, which was symptom-based per national guidelines with no routine imaging. No pertinent additional medical or family history was present.

A physical examination revealed a well-appearing patient with normal vital signs. On inspection, there was anterior chest bulging, with rumbling pathological diastolic and systolic murmurs heard along the right parasternal border radiating to the subxiphoid and apical regions during auscultation. No stigmata of connective tissue disorders were present. Laboratory tests, including complete blood count, organ function tests, and d-dimer, were all in the normal range. Screenings for HIV, tuberculosis, and syphilis were negative.

A chest radiograph (CXR) at presentation showed a smooth left hilar region opacity that has a close relation with the left cardiac boarder, which raised the possibility of aneurysmal dilatation of the PA (Figure 1). The differential included aneurysmal dilatation of the left atrium and a pericardial cyst. Transthoracic echocardiogram does not show progression from the previous one; there was a large PDA and pulmonary arterial hypertension (PAH), with no further information obtained from the report.

A chest computed tomography angiography (CTA) was recommended for definitive characterization. The CTA revealed a giant aneurysm of the main PA, with a maximum diameter of 8.6 cm, and dilatation of both branches, with a diameter of 3.4 and 2.9 cm for the right and left PA, respectively (Figure 2).

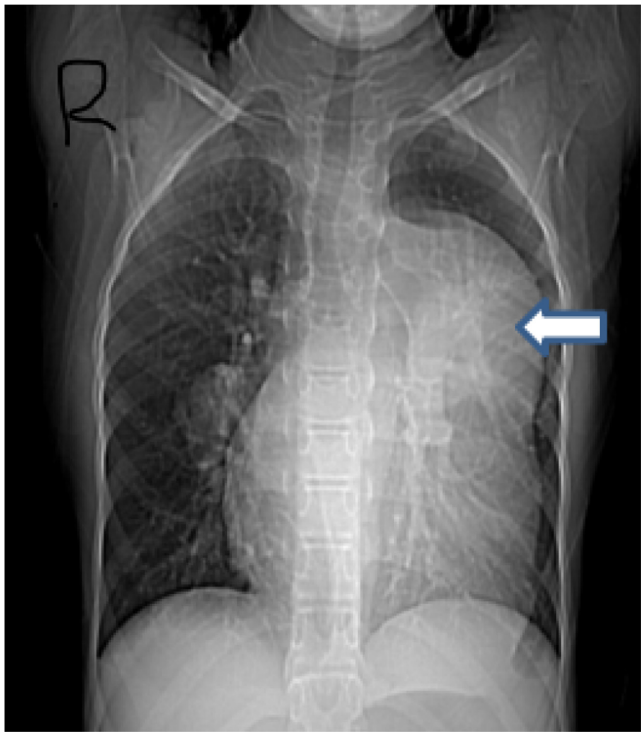


FIGURE 1 Frontal chest x-ray showing smooth left hilar radiopacity.

CTA also showed a PDA with a diameter of 1.1 cm (Figure 3). The patient is currently on medical treatment with a follow-up every 3 months. Definitive surgical management was suggested, but the parents could not afford to take him abroad for treatment.

3 | DISCUSSION

PAA is a rare entity with a wide range of congenital and acquired causes and a strong association with PAH.[6] The exact disease incidence is difficult to estimate due to the lack of comprehensive studies done in recent years.[6] PAAs can be classified into proximal (central) and peripheral, where the former indicate involvement of the pulmonary trunk or, rarely, the right and left major PAs.[4,6,10] The standard diameter size that defines PAA has varied in the literature from 2.9 to 4.0 cm.[5,11] A recent study done in adult cohorts found a mean PA diameter of 3.2 cm (0.46) and suggested a cut-off value of 4.5 cm to define the main PAA.[3] The suggested aneurysmal size to define it as "giant" has ranged from 5 to 8 cm.[11]

Various origins of PAA have been described, including congenital causes, acquired causes, and idiopathic PAA.[6] In younger children, most PAAs are congenital, and more than 50% of those cases were associated with CHD.[5,7] The increased flow caused by the left-to-right shunt in CHD results in increased hemodynamic shear

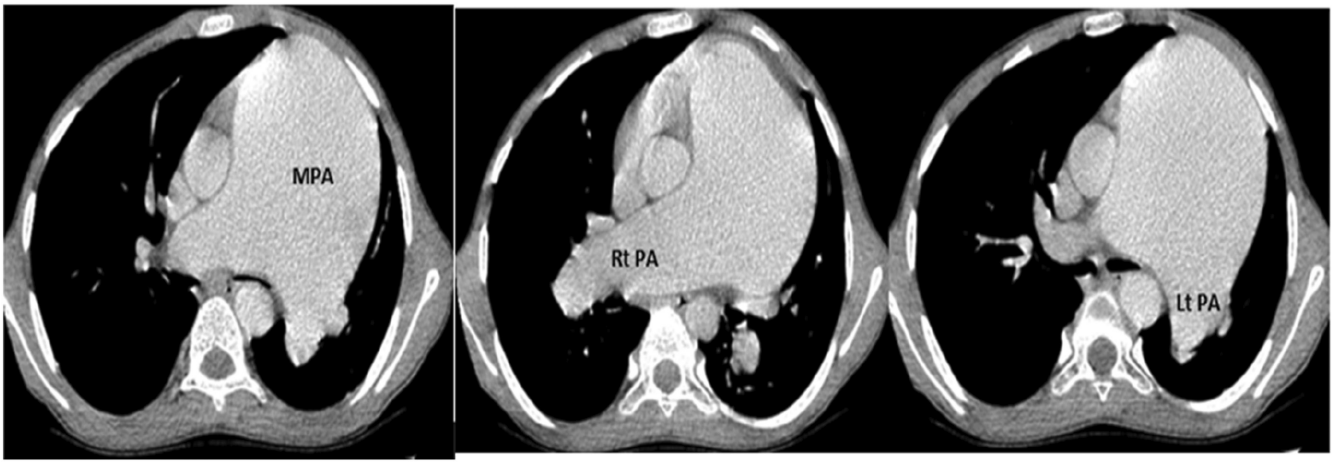
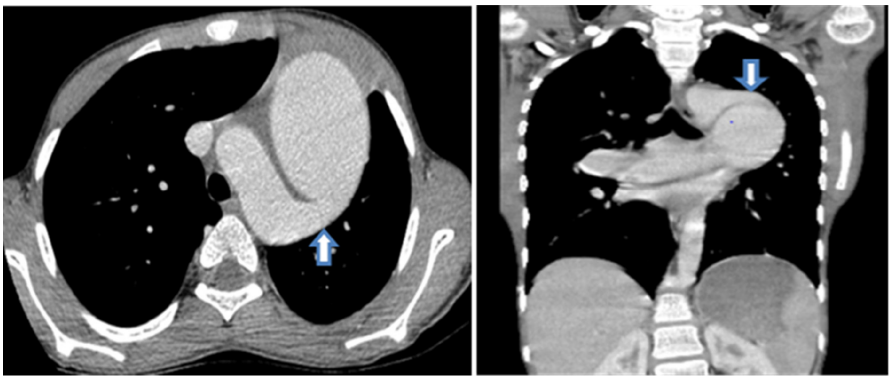


FIGURE 2 Axial CT pulmonary angiography showing dilated main pulmonary artery (MPA), right (Rt PA) and left (Lt PA) pulmonary arteries.

FIGURE 3 Axial and coronal CT pulmonary angiography showing PDA.



stress on the vessel walls and therefore results in aneurysm formation in the PAs.[5,6] Common causes of left-to-right shunts that result in aneurysm formation include PDA, ASD, VSD, and sequelae of congenital heart disease repair.[8] Most PAA dissections occur in the context of CHD, probably related to a longer duration of exposure to elevated PA pressure.[4] Infections and connective tissue disease can coexist with CHD and contribute to aneurysm formation.[12]

Infectious and autoimmune vasculitis are among the acquired causes of PAA.[6] Rasmussen's aneurysms, which occur due to weakening of the pulmonary artery wall from adjacent cavitary tuberculosis, are still reported from developing countries to date.[1,13] Thus, it is important to remember PAA in the differential diagnosis of a lung mass in tuberculosis patients.[10] Pyogenic bacterial infections are an increasingly common cause of PAAs, called mycotic aneurysms, exclusively located in more peripheral branches.[12] Mycotic and Rasmussen aneurysms share similar risk factors and can have indistinguishable clinical and image findings on CXR and CTA.[12] Advanced syphilis used to be a frequent cause of PAA involving large PAs.[6] Schistosomiasis is also known to cause vascular aneurysms in endemic areas, and PA is the most commonly involved arterial district.[14] Autoimmune vasculitis of the PAs, such as Behcet's disease and Hughes-Stovin syndrome (HSS), are associated with cases of PAA.[5] Some of the above cases can be managed conservatively with antibiotics and immunosuppressants, especially when the aneurysm size is small and stable.[12,13]

PAAs are often detected incidentally on imaging.[6] On CXRs, main PAA appears as a hilar enlargement or a rounded bulge that simulates a focal lung mass in the left mediastinal border, as seen in our case.[1] The differentials include giant left atrium and massive pericardial cyst. CTA can confirm the diagnosis and help extract useful information on the size, number, location, and extent of the PAA, and presence of mural thrombus.[1,8] Aneurysms appear as saccular or fusiform areas of dilatation, with homogeneous contrast material filling that occurs simultaneously with that in the PA on CTA.[1,8] CT also has added advantages in that it allows the lung parenchyma and the heart to be evaluated at the same time as the vessels.[2] Recent advances in CT imaging have made it possible to eliminate motion artifacts and markedly reduced radiation exposure, making it an attractive alternative for

imaging children.[15,16] In PDA patients, CT can also be helpful if questions remain after echocardiography due to a limited acoustic window or if there is concern for a PDA aneurysm or thrombus.[16]

A review of cases reported in African literature revealed a number of cases associated with rheumatic valvular heart disease. The most important underlying factor, however, remained PAH, which causes direct pressure on arterial wall, leading to atherosclerosis, medial necrosis, and aneurysmal dilation. Imaging findings were also the same, with suspicious CXR findings confirmed on subsequent CTA. These observations underscore the importance of considering PAA as a differential diagnosis of hilar masses in patients with both congenital and acquired heart diseases.[6] In areas with limited availability of CT scans, echocardiography can be an essential tool to reach a diagnosis of proximal PAA.[9,10]

Currently, there is no agreement on how or when to treat PAA for optimum outcomes, given limited experience from the infrequency of the disease.[3,11] They can be managed conservatively, when possible, by medical treatment targeted at the underlying disease and PAH, especially in patients with CHD,[4,6,7] and radiographic follow-up done with serial echocardiography or CTA.[4,5] Optimal management of giant PAAs is even more difficult to determine, and most authors recommend surgery to relieve symptoms and decrease the risk of complications, mainly mechanical compression of neighboring structures.[11] The risk of PA dissection depends largely on the severity of PAH rather than aneurysmal size.[7] However, surgery carries a high risk of complications in patients with PAH.[3,6]

4 | CONCLUSION

Giant PAAs caused by PDA are extremely rare, without characteristic clinical presentation and often detected incidentally on imaging. In the present case, lack of access to appropriate cardiovascular imaging prevented timely, definitive diagnosis. This case report also highlights the undefined burden of PAA that may exist in African children with ill-treated congenital and acquired heart disease.

AUTHOR CONTRIBUTIONS

Samuel Sisay Hailu: Conceptualization; supervision; validation; visualization; writing – original draft; writing – review and editing. **Hermon Miliard Derbew:** Validation; visualization; writing – original draft. **Abrehet Zeray:** Validation; writing – original draft; writing – review and editing. **Tesfahunegn Hailemariam:** Methodology; visualization; writing – original draft; writing – review and editing. **Hansel J. Otero:** Validation; visualization; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

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DATA AVAILABILITY STATEMENT

The data that supports the findings of this study is available from the corresponding author on reasonable request.

CONSENT

Written informed consent was obtained from the patient's parents for anonymized patient information to be published in this article.

ORCID

Tesfahunegn Hailemariam https://orcid.org/0000-0003-4304-804X

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