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CONUS BRANCH FISTULA TO MAIN PULMONARY ARTERY: A RARE CORONARY ANOMALY THAT MAY CAUSE CHEST PAIN IN A PEDIATRIC PATIENT

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ABSTRACT

Coronary artery fistulas (CAFs) are rare congenital coronary anomalies where abnormal communication develops between coronary arteries and cardiac chambers or a thoracic great vessel. CAF is usually diagnosed incidentally with cardiac imaging. Although the majority of fistulas are congenital, iatrogenic CAFs are increasingly observed due to multiple cardiac interventions or direct chest trauma. CAFs in children may result in hemodynamically significant problems due to vascular shunting. We report an 11-year-old boy who presented with atypical chest pain and was found to have a rare variant of CAF; fortunately, he had no significant hemodynamic complications.

KEYWORDS

Echocardiography, cardiac computed tomography, conus branch, fistula

LEARNING POINTS

- Coronary artery fistula is a rare anomalous connection between the coronary artery and any segment of the systemic or pulmonary circulation and is usually an incidental finding on cardiac imaging.
- Echocardiography, computed tomography coronary angiography, cardiac magnetic resonance imaging, and conventional coronary angiography are the tools that provide detailed anatomical visualization to confirm vascular connections in addition to the assessment of hemodynamic consequences resulting from the shunt.
- A coronary fistula between a separately originating conus branch and the main pulmonary artery is a very rare variation in paediatric practice that may be a cause of chest pain in children.

INTRODUCTION/BACKGROUND

Coronary artery fistulas (CAFs) occur in 0.1 to 0.2% of the general population. Usually, the abnormal vessel connection lacked from an outlet bed of capillaries results in minimal flow resistance, which, in the case of fairly large fistulas, predisposes to marked shunting and progressive dilatation

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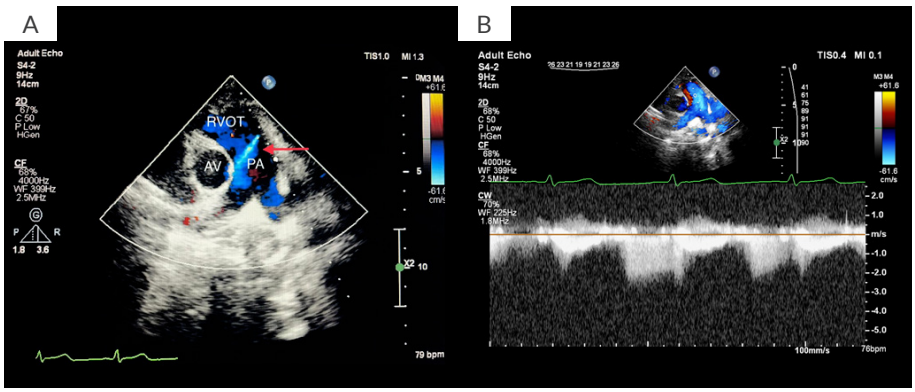


Figure 1. A) Color flow Doppler imaging showed laminar flow in main pulmonary artery (red arrow). B) Continuous wave Doppler showed continuous flow more prominent in diastolic phase.
Abbreviations: AV, aortic valve; RVOT, right ventricular outflow tract; PA, pulmonary artery.

of the fistula. The conus artery, a branch of the right coronary artery (RCA), supplies blood to the right ventricular outflow tract. Our patient had a rare anatomic variation of CAF where a conus branch that originated separately from the right sinus of Valsalva communicated with the main pulmonary artery (MPA).

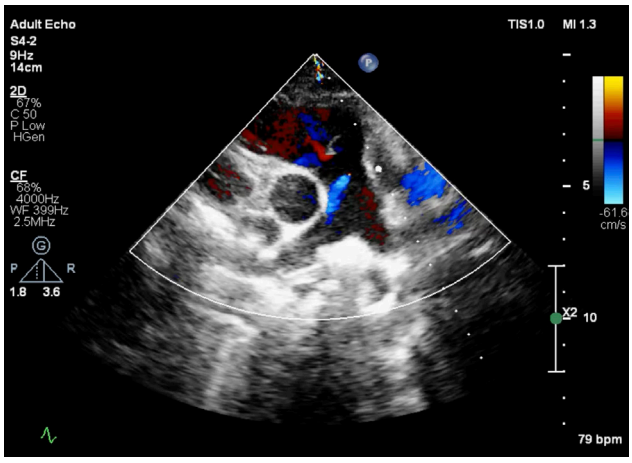
CASE DESCRIPTION

An 11-year-old boy presented to the cardiology clinic with atypical chest pain on and off for a few months. He was not known to have significant medical problems, and his family history was negative for genetic disease. The physical examination, including vital signs, was normal, as was his standard 12-lead electrocardiogram. Transthoracic echocardiography revealed normal cardiac chamber size with a left ventricle ejection fraction of 65%, no notable valvular abnormalities, and a pulmonary systolic pressure of 23 mmHg. Colour Doppler imaging in the short-axis view at the pulmonary level identified continuous laminar flow within the MPA that was more pronounced during diastole in continuous Doppler flow tracing *(Fig. 1, Video 1)*, raising the suspicion of a coronary communication with the MPA versus patent ductus arteriosus (PDA). Computed tomography coronary angiography (CTCA) revealed a conus artery (usually a branch of RCA) with a separate origin from the right sinus of Valsalva that coursed anteriorly in a tortious pathway terminating in the anterolateral aspect of the MPA *(Fig. 2)*. The CTCA showed patent main coronary arteries and no evidence of PDA. Given the absence of a significant shunt and the low risk of ischemia, a decision was made to monitor the patient with echocardiography. The case was diagnosed as a rare variant of CAF between a conus branch with a separate origin and the MPA.

This case highlights the role of echocardiography in identifying coronary communication through Doppler imaging and providing a hemodynamic assessment of the effects of the shunt. CTCA is vital in delineating the exact anatomical variation in coronary communication. Furthermore, this case highlights that a CAF could be a potential cause of chest pain in children.

DISCUSSION

The prevalence of CAFs on conventional coronary angiography is 0.05–0.25%, whereas it reaches 0.9% on CTCA[1]. CAFs occur in 0.1 to 0.2% of the population[2]. They are usually congenital, but iatrogenic/acquired forms have been reported due to multiple cardiac interventions or direct chest trauma. Nearly half of the CAFs originate from the right coronary system, and 35% originate from the left-coronary system, whereas in only 5% of cases, the site of origin is bilateral[3]. The terminal sites of CAFs can be in any cardiac cavities or major thoracic vessels. The pulmonary artery, coronary sinus, and superior vena cava are the most common vessels as terminal sites in decreasing frequency[4]. Although most patients have a single CAF, 20% have fistulas originating from two or more coronary arteries Usually, the abnormal vessel connection lacked from an outlet bed of capillaries results in minimal flow resistance, which, in the case of fairly large fistulas, predisposes to marked shunting and progressive dilatation of the fistula. CAFs can be characterized as small, medium, or large if the fistula diameter is <1, ≥1 to 2, or >2 times the largest diameter of the coronary vessel not feeding the coronary fistula, respectively. An important issue in the pathophysiology of CAFs is the shunt which is usually determined by the fistula's size and the pressure difference between the coronary artery and the chamber or vessel into which the fistula drains. Many CAFs remain asymptomatic[5], especially if the shunt is small and does not compromise coronary blood flow. However, some may lead to clinical manifestations such as chest



Video 1. Transthoracic echocardiography. Color Doppler imaging in the short-axis view at the pulmonary level identified continuous laminar flow within the main pulmonary artery.
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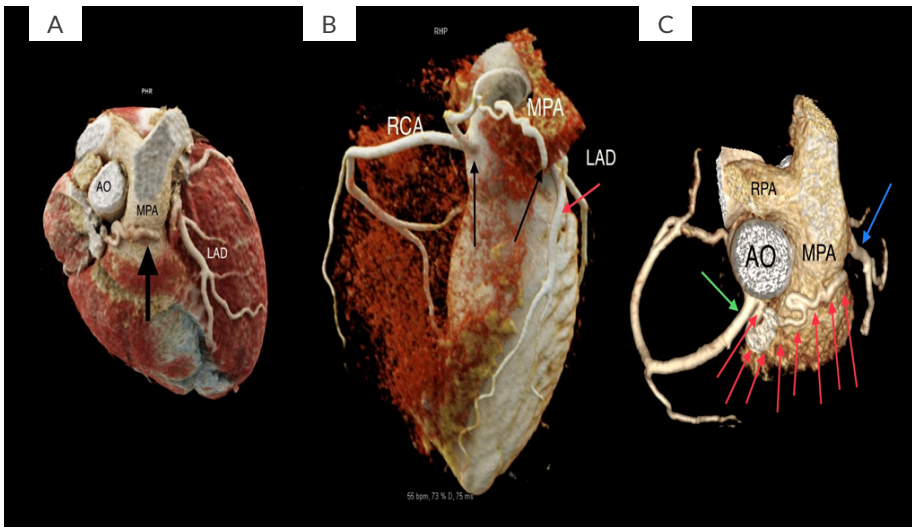


Figure 2. Three-dimensional volume rendered reconstruction images of cardiac computed tomography angiography showing a separately originated conus branch with tortious pathway and termination inti anterolateral aspect of main pulmonary artery (MPA) (black and red arrows).
Abbreviations: LAD, left anterior descending artery, RCA, right coronary artery, AO, aorta, RPA, right pulmonary artery.

pain, arrhythmias, or heart failure due to volume overload. Small fistulas have the potential for spontaneous closure over time[6], whereas larger CAFs may require surgical or transcatheter closure. Echocardiography is a valuable non-invasive initial screening tool where, in two-dimensional mode, a dilated coronary artery can be visualized, and its drain site can be detected through colour Doppler imaging. CTCA emerged as an important non-invasive tool in diagnosing CAFs, and it is capable of showing the complex geometry of coronary artery anatomy in two-dimensional and three-dimensional images. Moreover, volume-rendered images give an excellent overview of cardiac and vascular anatomy. Additional imaging modalities, such as magnetic resonance imaging and coronary angiography, may be utilized for further assessment. There is no high-quality evidence beyond small case series to guide the management of CAFs. Treatment strategies depend on symptom severity and fistula characteristics. Small, asymptomatic fistulas can be managed conservatively with regular follow-up. In contrast, larger or symptomatic fistulas may require percutaneous closure or surgical intervention to mitigate risks, such as aneurysm formation, thromboembolism, or myocardial ischemia[7,8]. In paediatric patients, unexplained chest pain should prompt further investigation for congenital anomalies once common causes have been excluded.

CONCLUSION

The case highlights the importance of recognizing rare coronary anomalies as potential contributors to chest pain in pediatric patients. Advanced imaging techniques facilitate accurate diagnosis, guiding individualized management plans.

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