Cardiology in the Young

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Rare case of anomalous origin of the left coronary artery from the pulmonary artery in a 4-year-old child

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Brief Report

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Abstract

Anomalous origin of the left coronary artery from the pulmonary artery is an exceedingly rare and potentially fatal congenital coronary anomaly that typically presents early in infancy. We report an unusual case of anomalous origin of the left coronary artery from the pulmonary artery in a 4-year-old child who presented later in life with vague respiratory and gastrointestinal symptoms and was found to have severe global cardiac dysfunction with evidence myocardial ischaemia.

Anomalous origin of the left coronary artery from the pulmonary artery is an exceedingly rare congenital coronary anomaly that occurs in 1 in 300,000 live births and comprises approximately 0.26–0.5% of all CHD.[1–4] Anomalous origin of the left coronary artery from the pulmonary artery usually presents early in infancy with symptoms of congestive heart failure and myocardial ischaemia. These symptoms start to manifest around 8 weeks after birth, near the end of the physiologic decline in pulmonary arterial pressure. This drop-in pulmonary arterial pressure promotes reversal of flow in the left coronary artery, leading to abnormal left ventricular perfusion and subsequent myocardial ischaemia.[1] Anomalous origin of the left coronary artery from the pulmonary artery is a potentially fatal diagnosis, with death occurring in 90% of neonates who are left untreated.[1–3]

Rarely, patients with anomalous origin of the left coronary artery from the pulmonary artery survive to adulthood secondary to the development of significant inter-coronary collaterals.[5–6] The symptomology in this group varies and is largely dependent on the degree of collateralisation; however, maortopulmonary windowany remain asymptomatic until the third decade of life.[1,5] We report a rare case of anomalous origin of the left coronary artery from the pulmonary artery in a 4-year-old child who presented with vague respiratory and gastrointestinal complaints and was found to have severe global cardiac dysfunction.

Methods: case report

Case

A 4-year-old male with no significant medical history presented to the emergency department with complaints of intermittent nausea, emesis, cough, and low-grade fever that started 2 days earlier. On examination, he had fever with temperature up to 103.1 °F, tachycardia with heart rate in the mid-160's, and tachypnoea with accessory muscle usage. His initial workup revealed cardiomegaly on chest X-ray, elevated pro–B-type natriuretic peptide (1.391 pg/mL), and positive testing for respiratory syncytial virus. Quantitative troponin T level was normal (<0.01 ng/mL). A bedside echocardiogram showed severely depressed left ventricular systolic function with an estimated left ventricular ejection fraction of 23% and moderate left-sided heart dilation. The origin of the left main coronary artery was not definitively delineated. A 12-lead electrocardiogram showed normal sinus rhythm, left ventricular hypertrophy, and Q-waves in leads I and AVR. Due to the severely depressed cardiac function, he was admitted to the cardiac ICU for further workup and management.

On arrival to the ICU, he was started on a heart failure regimen including oral Lasix, Aldactone, Carvedilol, and intravenous Milrinone infusion. He was also started on a prophylactic dose of aspirin for the prevention of atherothrombosis. On a subsequent echocardiogram, left ventricular function was slightly improved (moderately depressed with an estimated left ventricular ejection fraction of 33%); however, the origin of the left main coronary artery remained questionable. A cardiac CTA was subsequently obtained and suggested anomalous origin of the left coronary artery from the pulmonary artery. Further clarification via catheter-based angiography was recommended and this confirmed the suspected diagnosis. Aortic root angiography showed a right dominant coronary system with dilated right coronary artery (Fig 1a), and multiple collateral vessels supplying the left coronary artery system (Fig 1b and 1c). Direct haemodynamic measurements were obtained and revealed an

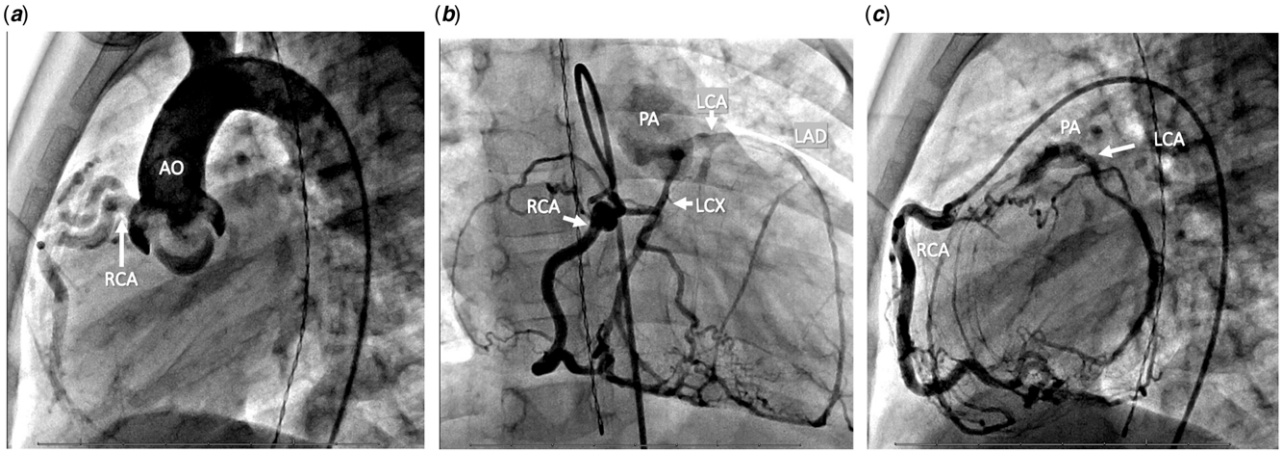

Figure 1. Conventional angiography demonstrating anomalous left main coronary artery arising from the pulmonary artery (PA), with dilated right coronary artery (RCA) and extensive inter-coronary collateralization to the left coronary system – specifically, the left anterior descending artery (LAD), left circumflex artery (LCX), and left main coronary artery (LMCA). (***a***) Aortic root injection. Anteroposterior (***b***) and lateral (***c***) projections of direct right coronary artery injection.

elevated left ventricular end-diastolic pressure of 12–13 mmHg, with normal estimated cardiac index of 3.8 L/minute/m^2.

He remained in the cardiac ICU on a continuous Milrinone infusion and oral heart failure therapy for approximately 1 week before undergoing surgical repair. During that time, his gastrointestinal symptoms completely resolved and he was able to be weaned off of all respiratory support. He eventually underwent Takeuchi repair, which involved the creation of an aortopulmonary window and intrapulmonary tunnel to baffle the aorta to the ostium of the anomalous left main coronary artery.[7–8] Due to the leftward and distal positioning of the left main coronary artery orifice, coronary reimplantation was not possible. The patient tolerated the procedure well with no major complications.

His post-operative course was largely uneventful, aside from the development of a moderate-size posterior pericardial effusion. This effusion eventually resolved in the outpatient setting, following a 4-week course of colchicine therapy. He was discharged home and continued on oral heart failure medications due to persistent ventricular dysfunction. He was followed closely in the outpatient setting but continued to display moderately depressed systolic and diastolic function on serial echocardiograms.

Discussion

Conventionally, anomalous origin of the left coronary artery from the pulmonary artery syndrome has been divided into two types: the infant type and adult type. Both types are well-described; however, very few cases of anomalous origin of the left coronary artery from the pulmonary artery presenting in early childhood have been recounted.[1,5]

We report an unusual case of anomalous origin of the left coronary artery from the pulmonary artery in a 4-year-old child who presented with acute heglobal cardiac dysfunction art failure after a recent viral illness. This patient had likely remained asymptomatic up until this point, owing to the development of extensive coronary artery collateralisation. It is likely that the increase in metabolic demands from the acute viral illness offset his haemodynamic equilibrium, leading to inadequate left ventricular perfusion and myocardial ischaemia. Once the diagnosis of anomalous origin of the left coronary artery from the pulmonary artery was confirmed, he underwent surgical repair utilising the Takeuchi method. Despite successful surgical intervention and medical therapy, he continued to have a moderate degree of left ventricular systolic and diastolic dysfunction.

Early diagnosis and prompt medical treatment are crucial in preventing adverse outcomes and sudden cardiac death related to anomalous origin of the left coronary artery from the pulmonary artery. It is imperative that cases associated with congenital coronary anomalies continue to be reported so that we may learn more about the pathogenesis and epidemiology of this entity. As clinicians, the more we understand about this disease, the better-equipped we can be when caring for this high-risk patient population.

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Conflicts of interest. None.

References

1. Peña E, et al. ALCAPA syndrome: not just a pediatric disease. Radiographics 2009; 29: 553–565. DOI 10.1148/rg.292085059.
2. Zacharias M, et al. A late presentation of an anomalous left coronary artery originating from the pulmonary artery (ALCAPA): a case study and review of the literature. J Cardiol Cases 2014; 11: 56–59. DOI 10.1016/j.jccase.2014.10.006.
3. Chen P. Diagnosis of ALCAPA in a 5-year-old presenting with atrial arrhythmias. J Pediatr Pediatr Med 2018; 2: 1–4. DOI 10.29245/2578-2940/2018/2.1118.
4. Wesselhoeft H, et al. Anomalous origin of the left coronary artery from the pulmonary trunk. Its clinical spectrum, pathology, and pathophysiology, based on a review of 140 cases with seven further cases. Circulation 1968; 38: 403–425. DOI 10.1161/01.cir.38.2.403.
5. Jinmei Z, et al. Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) diagnosed in children and adolescents. J Cardiothorac Surg 2020; 15: 90. DOI 10.1186/s13019-020-01116-z.
6. Cankurt T, et al. ALCAPA syndrome and atrial septal defect In a 68-year-old woman: an extremely rare congenital association. Acta Cardiol Sin 2017; 33: 447–449. DOI 10.6515/acs20160608c.
7. Ginde S, et al. Late complications after Takeuchi repair of anomalous left coronary artery from the pulmonary artery: case series and review of literature. Pediatr Cardiol 2012; 33: 1115–1123. DOI 10.1007/s00246-012-0260-5.
8. Takeuchi S, et al. New surgical method for repair of anomalous left coronary artery from pulmonary artery. J Thorac Cardiovasc Surg 1979; 78: 7–11.

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