Anomalous origin of the left coronary artery from pulmonary artery misdiagnosed as postpartum cardiomyopathy

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ABSTRACT

A 30-year-old woman with a history of presumed postpartum cardiomyopathy presented to her primary care physician's office with crescendo angina and shortness of breath. She was found to be in supraventricular tachycardia, managed with the Valsalva maneuver, and was discharged home without any further workup. She again presented with exertional symptoms. Her electrocardiogram revealed abnormal T waves and her stress test was strongly positive. Cardiac catheterization revealed an absent left coronary ostium and a dilated right coronary artery, with large collaterals supplying the left coronary system; coronary computerized tomography revealed the anomalous origin of the left main coronary artery arising from the posterolateral aspect of the pulmonary artery, highly suggestive of the anomalous origin of the left coronary artery from pulmonary artery (ALCAPA) anomaly (Bland-White-Garland anomaly).

KEYWORDS ALCAPA; anomalous origin of the left coronary artery from pulmonary artery; cardiac catheterization; coronary computerized tomography

Anomalous origin of the left coronary artery from pulmonary artery (ALCAPA) is a rare but hemodynamically significant congenital heart disease. ALCAPA syndrome is reported to be present in 1 per 300,000 live births, accounting for 0.25% to 0.5% of all congenital heart diseases.¹ Krause and Brooks first reported ALCAPA in 1865 and 1885, respectively.² Because it is predominantly present in newborns and has a high mortality rate without surgery, diagnosis in adults is rare and information is derived mainly from isolated case reports or extrapolated pediatric cases. Two types of ALCAPA syndrome have been described, and the classic manifestation is symptom onset in the first or second months when the pulmonary artery pressure falls after the closure of the ductus arteriosus.³ Many die from ischemic cardiomyopathy and endocardial fibrosis.⁴ Although exertional dyspnea and angina pectoris are the predominant symptoms in adults, sudden cardiac death and ventricular fibrillation have also been reported.⁴

Our patient survived to adulthood due to the formation of an extensive network of collaterals from her right to left coronary arteries, became symptomatic in the postpartum period, and presented with exertional angina pectoris, palpitation, and dyspnea.

CASE PRESENTATION

A 30-year-old woman presented to her primary care physician with squeezing retrosternal chest pain, shortness of breath, dizziness, and palpitations. She was sent to the emergency department and was found to be in supraventricular tachycardia, with heart rates of up to 214 beats per minute. Rate and rhythm control was achieved with the Valsalva maneuver. Her symptoms resolved with stable vital signs and negative troponin levels. An electrocardiogram revealed T wave inversion in V1, V2, V3, V4, V5, V6, I, and aVL, which was her baseline, and she was discharged without any further workup. She later reported being short of breath,

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Figure 1. (a) 3D reformat image from computed tomography angiography (CTA) showing an enlarged right coronary artery (arrow) arising from the aorta (asterisk). The right coronary artery measured 8 mm in greatest dimension. (b) Cardiac catheterization showing tortuous dominant right coronary artery. (c) Axial image from CTA showing an enlarged right coronary artery (arrow) arising from the aorta (arrowhead). The right coronary artery measured 8 mm in greatest dimension. (d) An axial image from a CTA showing an arterial contrast blush (arrows) extending from the left main coronary artery into the main pulmonary artery (asterisk), which is suggestive of reversal of flow from the anomalous left coronary artery into the main pulmonary trunk.

lightheaded, and dizzy every time she exerted herself. A treadmill stress test was strongly positive. Her past medical history was significant for postpartum cardiomyopathy diagnosed in 2010 at 6 months postpartum with normal delivery and an abortion at 10 weeks. She was taking metoprolol and losartan, and her symptoms and cardiac contractility improved with medications. She avoided becoming pregnant ever since due to cardiomyopathy.

A transthoracic echocardiogram was normal, and she underwent cardiac catheterization. A selective coronary angiogram showed a dominant and large right coronary artery, which was ectatic throughout and supplied the entire left system with collaterals (*Figure 1b*). The left main artery was not visualized. Coronary computerized tomography showed the anomalous origin of the left main coronary artery, which was seen in direct contact with and arising from the left posterolateral aspect of the pulmonary artery; that is, a dilated and tortuous right coronary artery (*Figures 1a, 1c*). The arterial phase of the study revealed an arterial contrast blush extending from the left main coronary artery into the main pulmonary artery, suggestive of reversal of flow and coronary steal phenomenon (*Figure 1d*). The short and atretic left main artery branched into the left circumflex and left anterior descending arteries. The right coronary artery was diffusely ectatic and tortuous, measuring 8 mm in

Table 1. Characteristics of published cases of ALCAPA

Category	Variable	N
Age ^a	Adults	
	Men (mean 48 ± 2 years)	78
	Women (mean 47 ± 2 years)	115
	Neonates (<1 month)	10
	Infants (>1–12 months)	74
	Toddler (12–36 months)	6
	3–17 years	20
Clinical manifestations	Asymptomatic	14 (7%)
	Dyspnea on exertion	37 (18%)
	Stable angina	34 (16%)
	Sudden cardiac death	34 (16%)
	Successfully resuscitated	17
	Decompensated heart failure	48 (23%)
	Cardiogenic shock	3 (1%)
	Palpitations	12 (8%)
	Atrial fibrillation	2
	Monomorphic ventricular tachycardia	1
	Atrioventricular nodal reentrant tachycardia	1
	Unexplained	8
	Bradycardia	1
	Syncope	3 (1%)
	Fatigue	6 (3%)
	Failure to thrive in pediatric patients	9 (4%)
ALCAPA and cardiac ischemia in the pediatric population	Acute myocardial infarction	
	Neonate ^b	3
	Infant	11
	Cardiac arrest	
	Neonate	2
	Infant	2
	Early adolescent	1
	Angina: early adolescent	1
	Shone's complex	1
	ASD, hypoplastic LV, and coarctation of aorta	1
Association with other congenital heart diseases and other organ anomalies in the pediatric population ^c	ASD and ventricular septal defect	1
	Patent ductus arteriosus	1
	Coarctation of aorta, coronary sinus orifice	1

(Continued on next page)

Table 1. Continued

Category	Variable	N
	atresia with left superior vena cava	
	Tetralogy of Fallot	1
	Interrupted aortic arch type B and type 3 aortopulmonary window	1
	Klinefelter's syndrome and ASD	1
	Thymomegaly	1
	Scimitar syndrome	3
	Hypoplastic left heart syndrome	1

ALCAPA indicates anomalous origin of the left coronary artery from pulmonary artery; ASD, atrial septal defect; LV, left ventricle.

^aOther cases—7 “pediatric,” 2 “children,” and 1 “boy”—were reported in abstracts without mention of ages.

^bTwo patients were diagnosed after an autopsy that revealed ALCAPA and extensive anterior wall myocardial infarction.

^cThree patients with isolated anomalous origin of the right coronary artery from the pulmonary artery and three with additional congenital heart diseases were also described.

diameter. Multiple septal branches were seen arising from the left main artery along the course of the interventricular septum, which could suggest a network of collateralization.

The patient underwent a Takeuchi procedure, in which the aortopulmonary window and intrapulmonary tunnel were created using pulmonary artery baffle and the left coronary ostium was anastomosed with the aorta. The patient had an uncomplicated postoperative recovery, and a repeat stress test was negative.

DISCUSSION

Most cardiac anomalies are benign, with a good prognosis. We searched case reports from MEDLINE and retrieved 254 case reports and case series about ALCAPA (Table 1). Our literature review found that the most common presentations were decompensated heart failure, stable angina pectoris, sudden cardiac death, arrhythmias, and cardiac ischemia in the pediatric population. The common echocardiographic findings were reduced left ventricular ejection fraction and mitral regurgitation (56%), dilated and tortuous right coronary artery with septal collaterals and flow from left coronary artery to pulmonary artery (15%), and normal echocardiographic findings (4%). No cases of postpartum cardiomyopathy attributed to ALCAPA have been reported. Studies have reported an association of ALCAPA with ischemic heart disease and sudden cardiac death syndrome, particularly in young adults and athletes.^{5–11} The Sudden Death Committee of the American Heart Association stated that

19% of sudden cardiac deaths can be attributed to coronary artery anomalies.¹²

In conclusion, ALCAPA syndrome is a rare congenital heart disease found predominantly in newborns, and limited case reports have been found in adults. Clinical presentation varies from no symptoms to angina, tachyarrhythmias, and unexplained cardiomyopathy. ALCAPA is diagnosed by cardiac catheterization and coronary artery computerized tomography scan as the gold standard. Timely surgery helps to prevent myocardial ischemia and sudden cardiac death with a good long-term prognosis.

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