Coronary Anomalies

Anomalous Left Coronary Artery

Arising From the Pulmonary Artery in a 21-Year-Old Man

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A 21-year-old man was admitted after fainting abruptly while playing basketball. On finding the patient in cardiac arrest due to ventricular fibrillation, first responders performed cardiopulmonary resuscitation and applied shock to restore sinus rhythm. The patient had no family history of cardiac abnormalities or sudden death; when younger, he had noted that overexertion during sports caused mild chest pain that was relieved with rest. An electrocardiogram showed anterior-wall infarction, and the patient's cardiac troponin T level was elevated. Results of left-sided cardiac catheterization revealed a dominant, massively dilated right coronary artery (RCA) from which collateral vessels arose and supplied the left anterior descending and left circumflex coronary artery territories (Fig. 1). No left coronary artery (LCA) was apparent on the angiogram; however, a computed tomogram showed a small-diameter LCA originating from the pulmonary artery (PA), that is, ALCAPA. An echocardiogram revealed a left ventricular ejection fraction of 43%. To correct the anomaly, we performed uncomplicated surgical reimplantation of the LCA from the PA to the aorta (Fig. 2). Two months later, the patient had resumed normal physical activities.

Comment

The incidence of ALCAPA ranges from 0.25% to 0.5% among congenital heart defects. The mortality rate of 90% associated with untreated ALCAPA is a result of impaired oxygen delivery to left-sided cardiac tissue, which leads to mitral regurgitation, dilated cardiomyopathy, and myocardial infarction.[1] The classic presentation is irritability and dyspnea during the first 4 to 6 weeks of life and progression to shock or death. Adequate collateral flow from the RCA, however, can delay presentation until adulthood, at which time early heart failure or sudden death can occur.[2] Possible clinical findings in ALCAPA are atrial gallop from left ventricular stiffness, mitral regurgitation from papillary muscle dysfunction, or both.[3] In 45% of patients, electrocardiograms indicate anterolateral ischemia in leads I, aVL, V_5, and V_6.[3] The only

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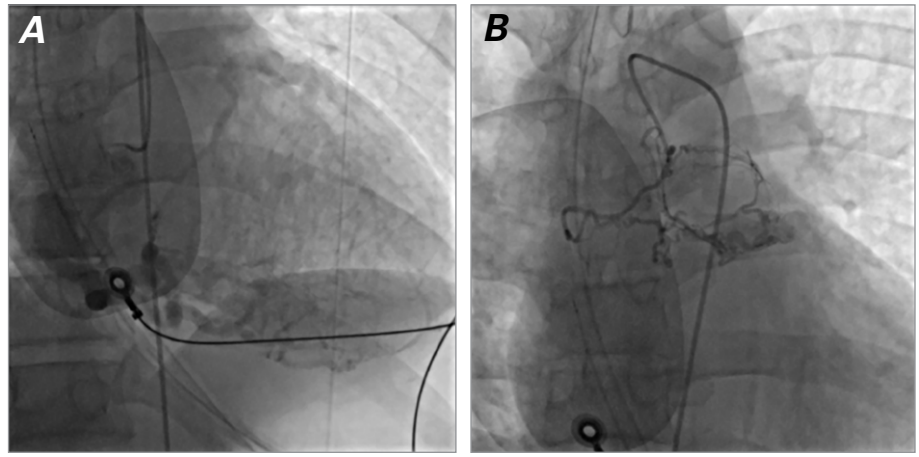

Fig. 1 ***A****) Angiograms during cardiac catheterization show a massively dilated right coronary artery with collateral vessels supplying the territories of* ***B****) the small-caliber left anterior descending and left circumflex coronary arteries.*

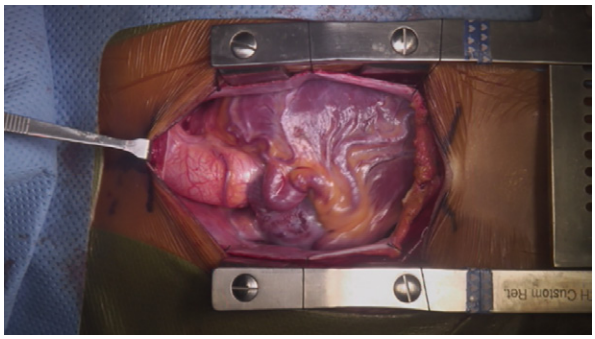

Fig. 2 *Preoperative photograph shows a dilated, tortuous right coronary artery.*

definitive treatment for ALCAPA is surgical correction, with direct LCA reimplantation to the aorta.[4,5]

Our patient, minimally symptomatic until sudden onset, survived 21 years with ALCAPA because his large RCA supplied robust collateral circulation to the LCA territories. His case exemplifies the importance of differential diagnosis in acute cardiac presentations when classic atherosclerotic characteristics are absent.

References

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