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Electrocardiographic evidence of ischemia in a young male with anomalous origin of a coronary artery from the opposite sinus

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ABSTRACT

Congenital coronary artery anomalies are a well-known cause of life-threatening syncope, but usually present with either cardiac arrest, or after resolution of the episode. We discuss a young man with exertional syncope, who upon evaluation in the field, had electrocardiographic evidence of severe but transient myocardial ischemia. This case provides rare insight into the pathophysiology of sudden cardiac death in patients with anomalous left coronary artery from the right aortic sinus.

1. Case report

A 12 year old African American male developed chest pain and syncope while playing basketball at home with friends. He regained consciousness without intervention while awaiting EMS arrival. EKG en route to the hospital demonstrated evidence of left main coronary occlusion with significant ST elevation in leads aVR and V1 and ST depression in leads I, II, III, aVF, V3-V6 (Fig. 1a) [1]. Repeat EKG 15 min later showed resolution of the ischemic changes (Fig. 1b). Upon arrival in the emergency room, his chest pain was persistent, albeit reduced. Troponin I peaked at 6.720 ng/mL (ref range: 0.012–0.034 ng /mL) on his second set of cardiac enzymes within ten hours of the event, indicating the patient had suffered a myocardial infarction. Repeat measurements of Troponin I returned to normal over the next 48 h. Following admission, the patient received an echocardiogram which demonstrated anomalous origin of the left coronary artery (LCA) from the right aortic sinus with suspected intramural course. A subsequent cardiac magnetic resonance (CMR) imaging study confirmed the finding of anomalous origin of the LCA from the right aortic sinus with an intramural course (Fig. 2). CMR at that time also showed no myocardial perfusion defects at rest, no late gadolinium enhancement and normal biventricular chamber dimensions and systolic function. The patient underwent operative unroofing of the LCA, where the intramural course was confirmed and was later discharged to home in good condition.

2. Discussion

Congenital coronary artery anomalies (CCAAs) are a well-known cause of sudden cardiac death (SCD) in the pediatric population. While the prevalence of CCAAs may be as high as 5%, the majority of these pose no clinical significance [2]. Among those that have been shown to predispose to SCD, the most important group includes anomalous origin of a coronary artery from the opposite aortic sinus (ACAOS). The prevalence of ACAOS is generally thought to be 0.1–1%, with the group being further classified by the course which the anomalous vessel travels to reach its perfusion area: interarterial, prepulmonic, retroaortic, and intraseptal [2–5]. With few exceptions, the latter three pathways are typically thought to be benign; only the anomalous vessels that travel between the aortic root and pulmonary trunk with an intramural course have been consistently shown to be associated with SCD [3,4]. The mechanism by which SCD occurs with these lesions is unclear, but several theories have been proposed. The ostia from which anomalous coronaries take off from the aorta are often slit-like, thereby limiting flow. In addition, the anomalous coronary arises at an oblique angle from the aortic root often with an associated ostial ridge, which may reduce coronary blood flow during exercise. Finally, given that most interarterial ACAOS follow an intramural course, traveling within the wall of the aorta for a variable length, intramural compression of the coronary vessel during exercise may lead to myocardial ischemia [2,3,5].

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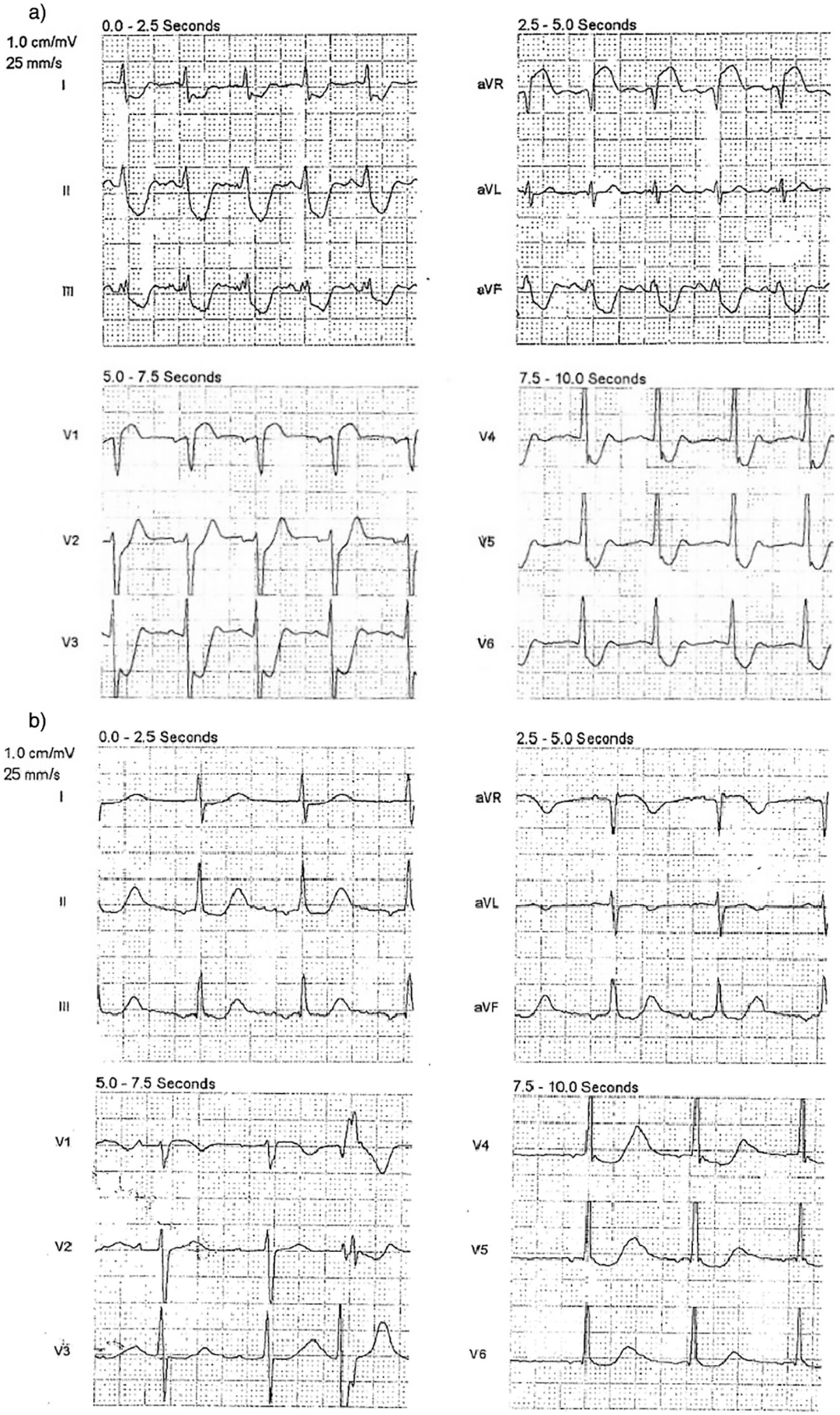

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Fig. 1. a Preliminary EKG with ST segment depression in leads I, II, III, aVF, V3-V6 and ST segment elevation in leads aVR and V1.
b Subsequent EKG 15 min later showing resolution of ischemic event, albeit with QT prolongation (QTc = 549 ms).

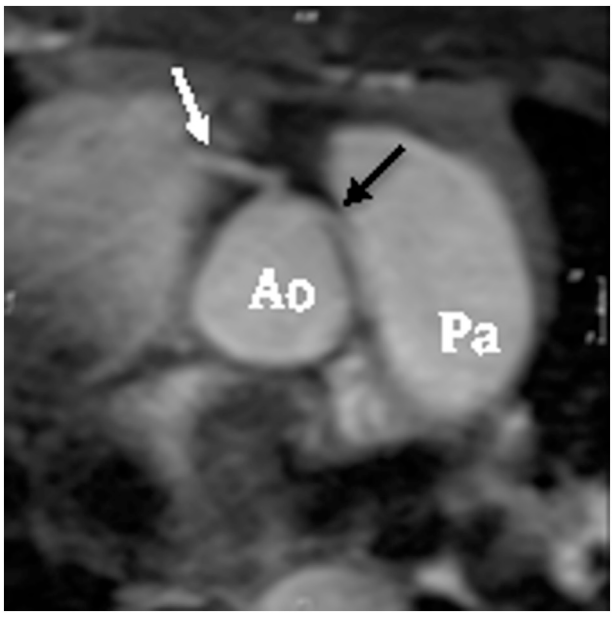

Fig. 2. Axial FLASH 3D respiratory navigated sequence (3T Skyra magnet) showing anomalous origin of the left main coronary artery from the right aortic sinus with interarterial course and probable intramural course. Ao: Aorta, Pa: Pulmonary artery, White Arrow: Right coronary artery, Black Arrow: Intramural LCA arising from right aortic sinus and coursing through the aortic wall between the aorta and main pulmonary artery.

This case illustrates coronary ischemia in a patient with ACAOS. Reports of angina, dyspnea, and syncope are not uncommon in those that experience exercise-induced SCD in the setting of ACAOS; yet, many are often asymptomatic prior to the fatal event as well. While the patient in question survived this event, preliminary EKG testing by EMS personnel demonstrated myocardial ischemia which spontaneously resolved. Capturing this transition from ischemia to event resolution within a small window of time provided a rare opportunity to visualize an episode of myocardial ischemia in a patient with ACAOS.

Declaration of Competing Interest

None.

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