Cardiology in the Young

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Occlusion of left main coronary artery presenting with ventricular fibrillation in teenagers: an unrecognised cause of sudden death?

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Abstract

In young patients with unexplained ventricular fibrillation, coronary occlusion may be missed by echocardiogram and misinterpreted by CT. We report two patients presenting with ventricular fibrillation and initially negative workup, later identified to have occlusion of left main coronary artery. We demonstrate the importance of angiography to rule out coronary occlusion in patients with unexplained ventricular fibrillation.

Brief Report

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Ventricular fibrillation and sudden death are rare in children and usually secondary to cardiac causes such as cardiomyopathies, channelopathies or coronary anomalies. Coronary anomalies are rare, occurring in <1% of the population.[1] While coronary artery originated from the opposite sinus with an intramural or an inter-arterial course has received much attention,[2] occlusion of the left main coronary artery has not. An occluded or atretic left main coronary artery is extremely rare with only 60 cases described in the literature prior to 2017 and few recent publications.[3] Imaging modalities such as echocardiography and CT angiography can identify coronary artery anomalies; however, they may also provide false reassurance of normal coronary anatomy.[4–6] Early diagnosis and surgical intervention lead to improved outcomes.[4] We report two adolescent patients who presented with syncope and ventricular fibrillation secondary to occlusion of the left main coronary artery. Our cases demonstrate the difficulty of accurate diagnosis with non-invasive modalities (echocardiography and CT) with confirmatory diagnosis made by coronary angiography.

Case report

Patient 1 presented at 12 years of age with multiple episodes of exertional syncope. His initial workup included an unrevealing electrocardiogram and normal coronary origins seen on echocardiogram as well as CT. An implantable loop recorder was placed and showed intermittent tachycardia. This prompted an electrophysiology study which was also negative. The patient was asymptomatic for 3 years, when he presented at age 15 with ventricular fibrillation during exertion. Echocardiogram again demonstrated normal coronary artery origins and function. He was mistakenly diagnosed with possible catecholaminergic polymorphic ventricular tachycardia after an Arrhythmia Comprehensive Genetic panel detected a variant of uncertain significance (RYR2 gene). He was started on metoprolol plus flecainide, and a subcutaneous Implantable Cardioverter-Defibrillator was placed. He continued to have episodes of ventricular fibrillation and intermittent exertional chest pain, pallor, and diaphoresis. A repeat exercise stress test revealed ST depression, and CT suggested severe stenosis of left coronary artery. Coronary angiography showed complete occlusion of the proximal left main coronary artery with collateral filling from the right coronary artery system (Fig 1). He underwent coronary artery bypass surgery with no further ventricular fibrillation episodes in 17 months of follow-up.

Patient 2 was a previously healthy 16-year-old who presented with ventricular fibrillation during exercise. His post-arrest diagnostic workup included an echocardiogram with normal coronary origins and electrocardiogram with mild ST elevation. CT suggested an anomalous origin of the left main coronary artery arising from the right coronary sinus. Coronary angiography demonstrated complete occlusion of the left main coronary artery with retrograde flow from the right coronary artery system (Fig 2). He underwent coronary artery bypass surgery without complications and is doing well 12 months since presentation.

Discussion

Our cases demonstrate that left main coronary artery atresia may be a rare, under-reported cause of sudden death in teens, and non-invasive modalities such as echocardiography and

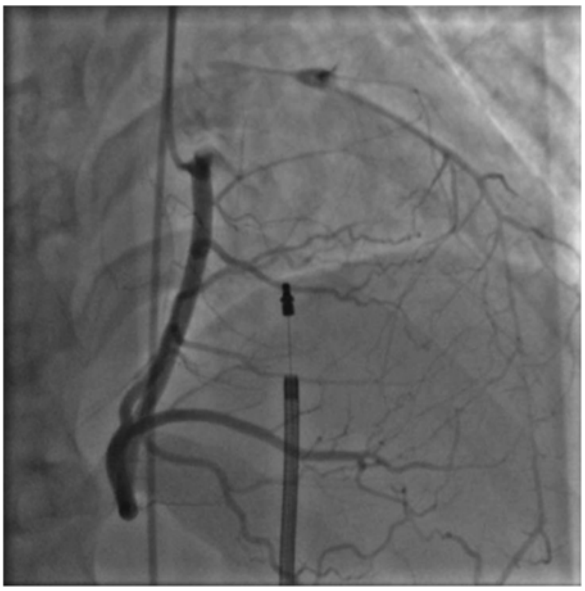

Figure 1. Cardiac catheterisation angiography, complete occlusion of proximal left main coronary artery. Collateral filling of left anterior descending from right coronary artery with retrograde flow into tiny circumflex and possibly ramus intermedius branches.

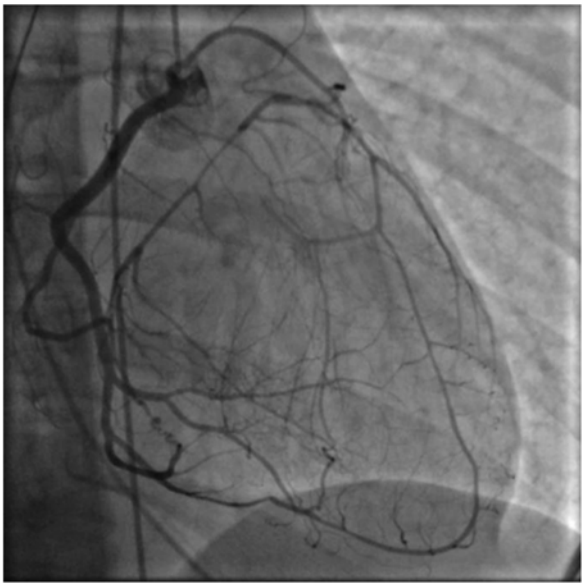

Figure 2. Cardiac catheterisation angiography, complete occlusion of the left main coronary artery. Retrograde flow from the right coronary artery reconstitutes the left coronary artery system to the level of the circumflex and left anterior descending artery. The left coronary system beyond the left main coronary artery is intact and hypoplastic. A second, smaller coronary artery arises from a separate orifice within the right coronary sinus.

CT may be inadequate for diagnosis. Coronary angiography may be the only method to make a definitive diagnosis.

An anomalous coronary artery origin from the opposite sinus with an intra-mural or an inter-arterial course is well described as a cause of sudden death,[2] but occlusion of the left main coronary artery has not, presumably due to its rarity.[3] Left main coronary artery atresia differs from an intramural or inter-arterial course of an anomalous coronary artery, as the blood flows retrograde from smaller calibre collateral arteries to larger calibre left-sided vessels.[4] Musiani et al hypothesise the collateral vessels are inadequate to meet the myocardial demand which leads to cardiac and coronary insufficiency.[4]

Both our patients underwent echocardiography in a well-equipped laboratory with sonographers and faculty experienced in CHD. Despite this, the echocardiograms obtained were thought to show normal origins of both left and right coronary arteries. Similarly, the CT studies were performed by radiologists with significant CHD experience. Although both the CT studies obtained suggested an abnormal course of the coronary arteries, they did not accurately identify the diagnoses.

The presentation of left main coronary artery atresia varies based on age. The symptomatic infantile form presents with failure to thrive and myocardial infarction, while older children and adolescents present with syncope and tachyarrhythmias.[4,7] A review of literature reveals numerous other case studies with delayed diagnosis of left main coronary artery atresia.[3-6] Amaral et al described two paediatric cases confirmed via coronary angiography or direct inspection in the operating room, despite normal coronaries on echocardiogram. Shah et al described a paediatric case with recurrent exertional syncopal episodes, an echocardiogram with prominent right coronary artery (otherwise normal), normal exercise testing and diagnosis confirmed by cardiac catheterisation.[8]

These papers and our experience suggest that in patients presenting with sudden death or ventricular fibrillation, a high index of suspicion for coronary anomalies (including congenital left main coronary artery atresia) is paramount for efficient and accurate diagnosis.

In conclusion, in young patients with unexplained ventricular fibrillation, coronary occlusion must be considered as a possible cause. Echocardiography and CT may falsely reassure providers of normal coronary anatomy. We recommend performing coronary angiography in all patients with suspicion of coronary artery anomalies, especially if they present with sudden death or ventricular fibrillation. Our cases demonstrate the importance of coronary angiography to definitively rule out total occlusion of the left main coronary artery.

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Conflict of Interest. Mayme Marshall declares that she has no conflict of interest. Seshadri Balaji declares that he has no conflict of interest.

Ethical Standards. This article does not contain any studies with human participants or animals performed by any of the authors. This study is a retrospective report of two cases and, as such does not require IRB approval or informed consent per our institutional standards.

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