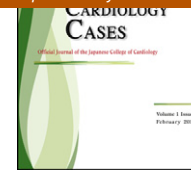




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Case Report

Acute myocardial infarction caused by an anomalous left main coronary artery in a 16-year-old boy

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KEYWORDS

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Summary A variety of structural cardiovascular abnormalities have been implicated in deaths of athletes, particularly congenital coronary arteries of anomalous origin, which are rare but major causes of myocardial ischemia and sudden death in young people. We present here the case of a rare congenital coronary artery anomaly in a 16-year-old boy who suffered from acute myocardial infarction due to occlusion of the left main trunk coronary artery, providing specific intravascular ultrasound findings for this anomaly.

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Introduction

Because trained athletes are often regarded as the healthiest members of the society, any medical catastrophes affecting them are highly publicized. A variety of structural cardiovascular abnormalities have been implicated in deaths of athletes, particularly congenital coronary arteries of anomalous origin, which are rare but major causes of myocardial ischemia and exercise-related sudden death at ages under 35 years [1]. However, they are only very rarely identified before a cardiac event because of the scarcity

of symptoms and inadequacy of routine diagnostic testing. Coronary anomalies can in fact be treated by surgical intervention, so that an accurate diagnosis early in life is very important.

We present here the case of a rare congenital coronary artery anomaly in a young athlete who suffered from acute myocardial infarction due to occlusion of the left main trunk coronary artery (LMT), providing specific intravascular ultrasound (IVUS) findings for this anomaly.

Case report

A 16-year-old boy without previous relevant medical history but with a family history of unknown juvenile sudden death was transported by ambulance to an emergency department with acute-onset syncope after running. The patient, who

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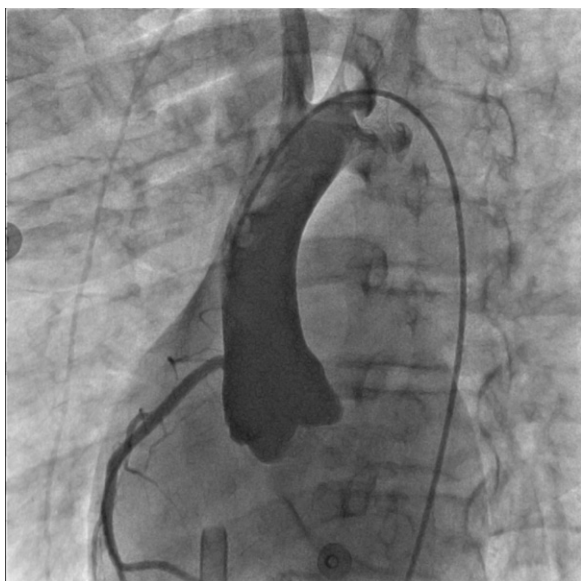


Figure 1 Aortography showed normal right coronary artery and total occlusion of the left main trunk.

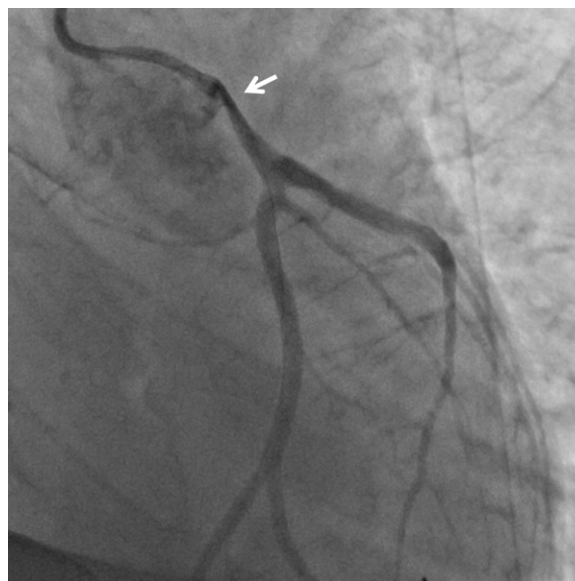


Figure 2 Left coronary angiography in the right anterior oblique-caudal view. It revealed an asymmetric narrowing of the left main trunk (arrow).

had experienced exertion-related syncope on 4 other occasions during the previous 5 months, complained of severe chest pain and was in a state of shock. Electrocardiography (ECG) revealed a complete left bundle-branch block and ST-segment elevation in the lead aVR. The patient lost consciousness again and went into cardiac arrest. Since he did not recover despite cardiopulmonary resuscitation, we immediately performed percutaneous cardiopulmonary support and cardiac catheterization. Coronary angiography showed a normal right coronary artery, but aortography demonstrated total occlusion of the LMT (Fig. 1). Finding that his LMT originated from the right sinus of Valsalva, we diagnosed the case as an anomalous LMT. With support of intra-aortic balloon pumping, left coronary flow was restored, resulting in achievement of thrombolysis in myocardial infarction (TIMI) trial grade 3, and an asymmetric narrowing of the LMT was detected (Fig. 2). Because hemodynamic stability was obtained by mechanical support, he subsequently underwent IVUS to determine the treatment strategy. Continuous imaging by IVUS showed pulsating compression of the anomalous LMT with pressure from the outside in addition to atherosclerotic changes and a slit-like orifice due to the acute takeoff of the anomalous LMT (Fig. 3). We performed percutaneous coronary intervention and successfully implanted a bare-metal stent (S-STENT4.0/11mm, Biosensors International) in the LMT. However, the patient's post-operative course was unfortunately complicated and he died 3 days after his admission. Autopsy was not enforced because there was no approval of the family.

Discussion

Congenital anomalous origin coronary arteries are rare but major causes of myocardial ischemia and sudden death in young people [1]. After hypertrophic cardiomyopathy, the presence of a congenital coronary artery anomaly is the

second leading etiology of death on the athletic field in the USA, occurring in about 20% of all cases [2]. However, it is difficult to make a diagnosis before a cardiac event because of the general lack of symptoms. Premonitory symptoms such as chest pain, dyspnea, syncope, and/or dizziness on effort have been reported in less than 40% of patients [3].

Sudden death most likely occurs because of a reduction in blood flow in the anomalous vessel, resulting in myocardial ischemia and infarction. Although the exact mechanisms for this decreased blood flow are not fully understood, autopsy studies have revealed that sudden death in young athletes is frequently associated with this anomaly [3].

Several mechanisms are possible to explain the cause of sudden cardiac death in anomaly cases. First, the affected vessel might be compressed between the aorta and the pulmonary trunk, especially during exercise-induced dilatation of the sinus of Valsalva and the pulmonary artery. Second, acute takeoff of the anomalous vessel might produce a narrowed slit-like orifice, which along with the intramural course of the proximal segment of the anomalous vessel can cause compression, particularly because this segment and the aorta share the same muscular tunica media. Third, sporadic spasm of the anomalous coronary artery might be induced by endothelial injury [4]. A previous study showed that endothelial injury resulting in induction of coronary spasm is frequently seen with congenital anomalies of coronary arteries [5]. Although autopsy was not performed, underlying mechanisms were possible to explain the etiology of the present case. At first, aortography showed total occlusion of the LMT ostium, and IVUS images revealed acute takeoff of the anomalous vessel and its narrowed slit-like orifice, along with the pulsating compression of the anomalous LMT, which also showed atherosclerotic change. Furthermore, a family history might be related to prevalence of anomalous LMT or atherosclerosis, and exercise is clearly a major factor in creating the conditions for events to occur.

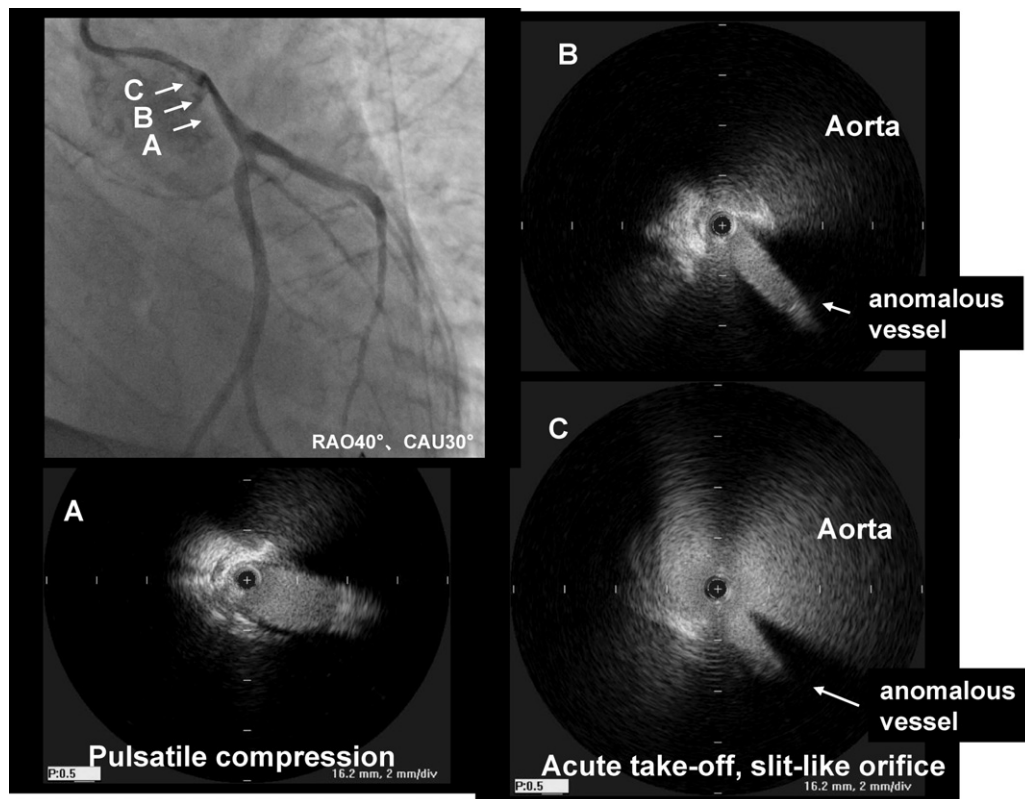


Figure 3 Intravascular ultrasound findings of the anomalous vessel. (A) Pulsating compression of the anomalous left main trunk with pressure from the outside. (B) Atherosclerotic changes. (C) Slit-like coronary orifice due to the acute takeoff of the anomalous left main trunk.

In conclusion, it is important to detect and diagnose these anomalies early to prevent such catastrophic occurrences. The possibility of a congenital coronary anomaly should always be considered in a young individual with a history of chest pain and/or syncope as well as a family history, particularly if the episode is triggered by exercise.

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