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Anomalous origin of left main coronary artery from the right sinus of Valsalva leading to sudden death

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Abstract. A 14-year-old female was brought to the emergency room with chest pain, shortness of breath and cyanosis. She was previously well with the exception of one previous post-exertion seizure-like event. On this day, she had been jogging when she complained of chest pain and collapsed. Her initial vital signs were heart rate 58/min, blood pressure 70/40 mmHg, respiratory rate 50/min, temperature 37 °C, and SaO2 68%. Electrocardiogram showed significant ST changes. She received multiple fluid boluses and dopamine was initiated (5–20 μg/kg/min). She was intubated and started on norepinephrine (0.05–0.5 μg/kg/min) for refractory hypotension. During the resuscitation, echocardiography showed poor left ventricular function with an ejection fraction of 38%. The coronary arteries could not be visualized clearly. To maintain cardiac output, epinephrine by infusion (0.1–3.0 μg/kg/min) was added, and she received multiple epinephrine boluses. Despite maximum ventilatory support and escalating inotropes, cardiac output rapidly deteriorated, and she developed an agonal rhythm with non-reactive pupils. Resuscitation was discontinued. Autopsy demonstrated an anomalous origin of left coronary artery from the right aortic sinus of Valsalva with acute myocardial ischemia. We describe the sudden coronary death of a young patient, and we review congenital coronary artery pathophysiology, screening difficulties and potential interventions.

Keywords: Anomalous coronary artery, sudden cardiac death, coronary sinus

1. Introduction

Sudden cardiac death is rare in pediatrics with an estimated incidence of less than 1:200,000 [1,2]. Congenital and acquired abnormalities can lead to sudden cardiac death, and these causes can be further broken down into structural heart disease, cardiomyopathies, coronary artery abnormalities, arrhythmias, primary pulmonary hypertension, and commotio cordis. A coronary etiology is found in approximately 10% of all sudden cardiac deaths in the young. A significant family history or a past history of palpitations, chest pain or syncope, particularly with exertion, requires prompt attention by care givers [1].

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In this article, we describe the sudden coronary death of a young patient, and we review congenital coronary artery pathophysiology, screening difficulties and potential interventions.

2. Case report

The patient was a physically active 14-year-old female who had experienced one previous seizure-like episode following intense physical exertion. Previous investigations, which were interpreted as normal, included computed tomography head imaging, electroencephalogram and electrocardiogram (ECG). She was not taking any medications. On this presentation, she was brought by a family member to the emergency department in a Children's Hospital. She had been jogging with a friend when she complained of chest pain and collapsed. She was awake on arrival and her initial pulse rate was 58/min, blood pressure 70/40 mmHg, respiratory rate 50/min, temperature 37 °C and SaO_2 was 68%. There was no evidence of arrhythmia. An intraosseous line was rapidly placed and multiple fluid boluses were given for hypotension. She was started on dopamine, which was quickly elevated to 20 μg/kg/min. A venous blood gas taken on arrival returned with a mixed respiratory and metabolic acidosis (pH 7.03, PCO_2 60, HCO_3- 16, BE -14.7), and lactate was 5.4 mmol/L. The initial troponin-T was normal at <0.01 μg/L. A chest X-ray demonstrated a normal cardiac silhouette with bilateral pulmonary edema (Fig. 1A). A 12 lead ECG showed bifascicular block (complete right bundle branch block and left posterior fascicular block) as well as significant ST changes suggestive of ischemia (Fig. 1B). Given her refractory hypotension, mixed acidosis and low SaO_2, she was intubated and norepinephrine by infusion (0.05–0.5 μg/kg/min) was initiated. A subsequent cardiac echocardiogram showed significant left ventricular dysfunction, particularly the posterior wall, with an ejection shortening fraction of 38% (Fig. 1C). The left ventricle dimensions were normal (LVIDd [left ventricular internal dimension in diastole] 4.5 cm [normal: 3.9–5.6], IVSd [intraventricular septum thickness in diastole] 0.95 cm [normal: 0.46–1.08], LVPWd [left ventricular posterior wall thickness in diastole] 0.77 cm [normal: 0.46–0.95]). Right ventricular function was normal. Additional intravenous access was established with a femoral central venous sheath. An epinephrine infusion (0.1–3.0 μg/kg/min) was also started to maintain cardiac output with detectable systolic blood pressures ranging from 53 to 80 mmHg. Possible insertion of an intra-aortic balloon pump was considered, but could not be organized prior to the rapidly worsening left ventricular function shown on echocardiogram. The SaO_2 could not be maintained with bag mask ventilation and a positive end-expiratory pressure valve. She was therefore transferred to pediatric critical care unit for advanced ventilation and further management. Despite maximum ventilation and inotropic support, cardiac output could not be maintained. She subsequently developed an agonal rhythm with no detectable blood pressure and required cardiopulmonary resuscitation. Further examination showed fixed and dilated pupils. Resuscitation was discontinued with her family at the bedside. She died in less than four hours of her initial presentation.

A complete autopsy was performed. Heart weight was normal at 258 g, and her body weight was 62 kg. The normal heart weight for a 60 kg female is 214 g (confidence interval [95%]: 140 to 326 g) [3]. Examination of the major epicardial coronary arteries showed anomalous origin of the left main coronary artery (LMCA) from the right sinus of Valsalva (Figs 2A and 2B). The ostium of the LMCA was widely patent but partly slit-like and associated with an acute angle take-off of the vessel. Indeed, the LMCA appears to closely appose the aortic wall and is at least partially sub-adventitial (Figs 3A and 3B). Further, the LMCA was partly incorporated into the wall of the aorta within the first centimeter of its origin. The LMCA passed between the aorta and pulmonary artery before branching into left anterior descending and left circumflex branches. All of these vessels were patent along their entire length, with no areas of stenosis and no intravascular thrombi. The right coronary artery (RCA) showed normal origin from within the right sinus of Valsalva. The RCA followed a normal course and was the dominant vessel forming the posterior descending branch. The RCA showed no areas of narrowing and no thrombi. The remainder of the heart was grossly normal and without any additional congenital anomalies. Microscopically, myocardial sections showed widespread acute ischemic necrosis throughout the left ventricle. These ischemic areas were characterized by hypereosinophilia, myofibre waviness, and widespread contraction band change (Fig. 2C). No inflammatory cell reaction had developed. There was no evidence of previous ischemic injury. Right ventricle sections showed only acute ischemic changes, but not necrosis.

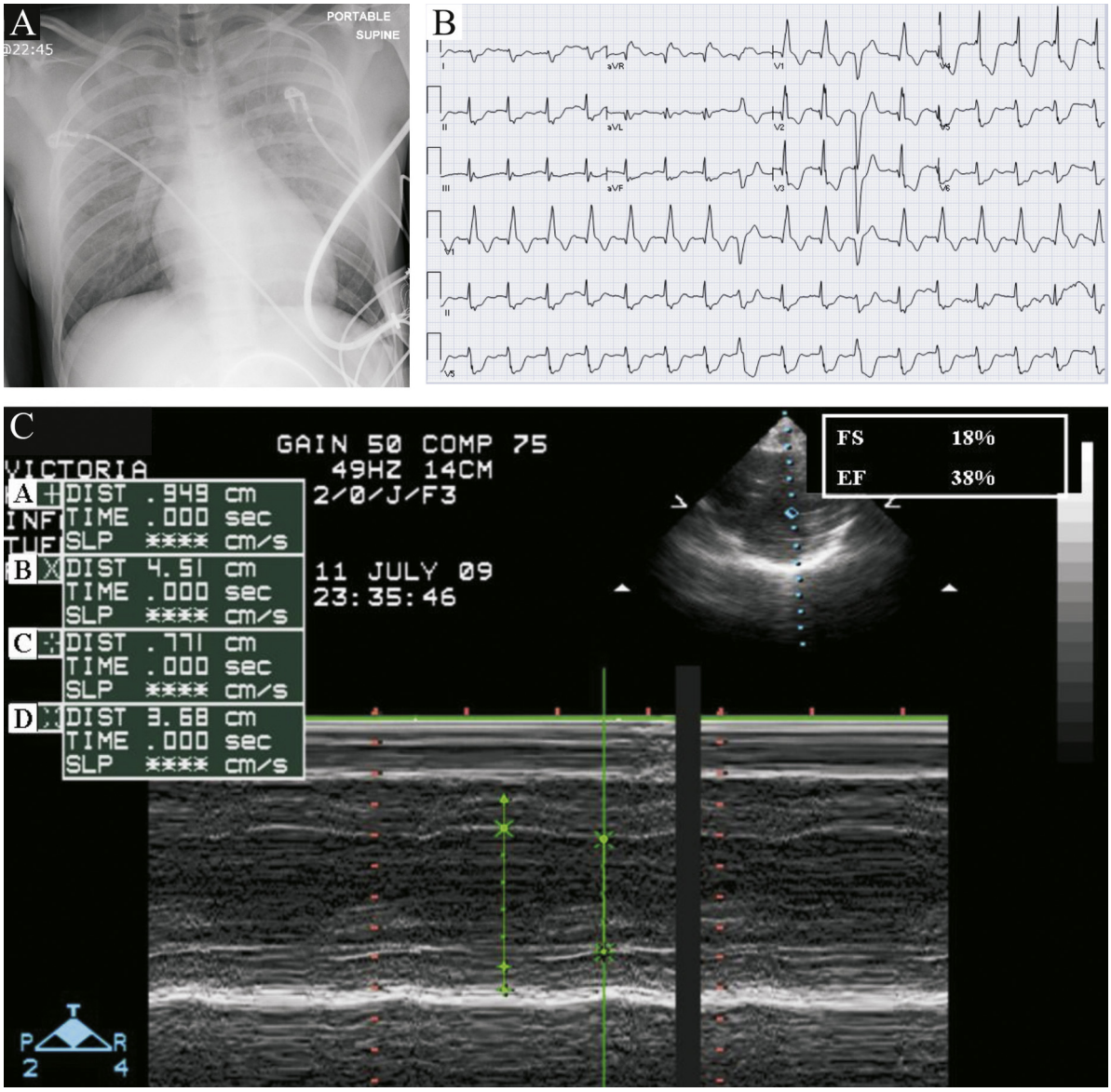

Fig. 1. Admission investigations on the reported patient obtained shortly after presentation. (A) A chest radiograph illustrating a normal cardiac silhouette with severe pulmonary edema. (B) An electrocardiogram demonstrating acute ischemia. Note the bifascicular block (complete right bundle branch block and left posterior fascicular block) and significant ST changes inferior-lateral. (C) An M-mode echocardiography of the left ventricle in the para-sternal short axis (after fluid administration, inotropes and intubation). FS = Fractional shortening; EF = Ejection fraction; Inset: A = Thickness of interventricular septum; B = End-diastolic dimension of left ventricle; C = Thickness of left ventricle free wall; D = End-systolic dimension of left ventricle.

3. Discussion

Congenital coronary artery anomalies are a rare cause of sudden death in children, as well as young adults, mostly during or immediately after intense exertion on the athletic field [4]. The reported incidence of coronary artery anomalies is 0.6% to 1.3% in angiographic studies [5,6] and 0.3% in an autopsy series [6]. Although the RCA arising from the left coronary sinus (anomalous right coronary artery [ARCA]) is four times as common as the left coronary artery arising from right sinus of Valsalva (anomalous left coronary artery

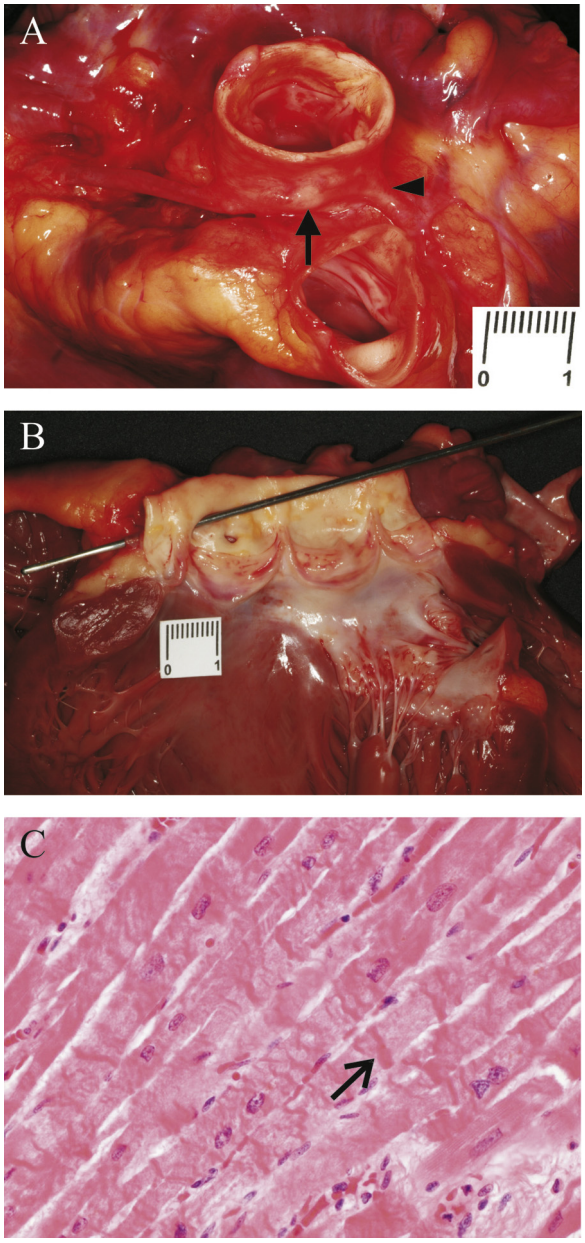

Fig. 2. Post-mortem cardiac examination. (A) External view of the heart base. Arrow indicates the site of origin of the left main coronary artery, between the aorta and pulmonary trunk. Arrowhead shows the left main coronary artery as it courses away from the aorta approximately 1 cm beyond its origin. (B) Internal view of left ventricular outflow tract. Probe is inserted in the left coronary ostium which is situated in the right sinus of Valsalva. (C) Microscopic image of left ventricular myocardium showing changes of early ischemic necrosis including prominent contraction bands (arrow).

[ALCA]), it is the ALCA, which is more commonly associated with sudden death after vigorous physical activity. Of the anomalies associated with sudden death,

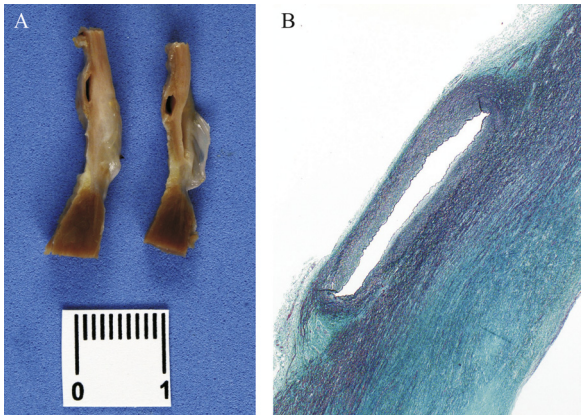

Fig. 3. Intramural course of the proximal left main coronary artery. (A) The cross sectional relationship of the anomalous left coronary artery in relation to the aortic wall. The artery is closely apposed the aortic wall and appears to be at least sub-adventitial. (B) A microscopic image of the left main coronary artery partly incorporated into the wall of the aorta within the first centimeter of its origin (Movat's elastic trichrome stain).

ALCA was present in 0.017% to 0.03% of cases [5,7]. Of the four types of ALCA, the interarterial type, where the left coronary artery passes anteriorly between the aorta and pulmonary artery, is the only type that places the patient at risk of sudden death [2]. The mechanism by which there is sudden occlusion of the interarterial coronary artery is unknown. A number of hypotheses have been proposed involving the oblique passage of the vessel as it leaves the aorta [8].

Recommendations for cardiovascular screening of young athletes have been developed and endorsed by several medical associations, and consist of a careful personal and family history and a comprehensive physical examination [9]. Screening with 12-lead ECG is generally not recommended, as it is limited in its ability to detect coronary artery anomalies in young competitive athletes [9–11]. Consequently, such coronary anomalies are not recognized during routine screening and are usually first recognized at autopsy. Indeed, coronary anomalies are a great challenge to identify and determine their clinical relevance so that proper treatment can be instituted [12]. Approximately 4% to 7% of all patients with acute myocardial infarction do not have atherosclerotic coronary artery disease by coronary arteriography or at autopsy, and such prevalence is nearly four times greater in patients with acute myocardial ischemic events, under 35 yr of age [13,14].

Nearly 1% to 2% of general population has anomalous coronary anatomy [15]. After hypertrophic cardiomyopathy, coronary artery anomalies of origin from

the wrong sinus of Valsalva are the second most common cause of death on an athletic field [11,16–21]. Sudden death during or immediately after athletic or vigorous physical activity can occur at any age, but since most competitive athletes are under age 50, sudden death secondary to a cardiac anomalous artery occurs more frequently in the younger age group [20]. The physiological demands of physical exercise depend on type of exercise. Dynamic or isometric exercise involves the use of large muscle masses that increase venous return, increase left ventricular end diastolic volume and together with adrenergic stimulation increase heart rate, blood pressure, cardiac output and myocardial contractility. These responses increase myocardial oxygen demand and this is met by increased myocardial blood flow. In the presence of coronary artery obstructive disease, sudden death is related to either sudden marked myocardial ischemia or myocardial scarring resulting in malignant ventricular arrhythmia [8].

In our case, the patient was in apparent good health until presentation; however, her parents recount a previous episode of syncope post-exertion. On the day of her death, her clinical history is unremarkable except that she was jogging intensely. The coronary artery abnormality remained undiagnosed until revealed by autopsy. The precipitating cause of death in this case is myocardial infarction was likely decreased myocardial oxygen delivery, secondary to an anomalous origin of left coronary artery from the right aortic sinus of Valsalva and the oblique passage of the artery as it exited the aorta. Hyper-acute myocardial infarction was observed globally in the left ventricle, also involving the RCA territory, and was likely secondary to the global hypotension resulting in poor myocardial perfusion throughout her prolonged resuscitation.

Coronary anomalies predisposing to sudden death are those with an anomalous left coronary artery from the right sinus of Valsalva (ALCA) and those with an anomalous RCA from the left sinus of Valsalva (ARCA). ALCA is considered more serious than ARCA because of larger amount of left ventricular myocardium at ischemic risk [22]. It has been reported that 100% of cases with an anomalous left coronary artery died suddenly as compared with 43% of cases with an anomalous RCA [22]. There are other coronary artery anomalies that very occasionally can be the etiology of sudden death with physical activity. Patients with a single coronary artery and ectopic origin have died suddenly during athletic activity [23,24]. A small minority of patients with the ALCA arising from the pulmonary artery live to adulthood before being diagnosed [25].

There are four types of ALCA defined by the path the left coronary artery takes after arising from the right coronary sinus [26]. The path of the first type is anterior to right ventricular outflow tract before reaching the anterior sulcus, the usual area of bifurcation. The second type courses behind the right ventricular outflow. The third one courses dorsal to the ascending aorta. These three types in absence of atherosclerotic plaque obstruction are benign. The fourth type arises from the right sinus of Valsalva and passes obliquely between the aorta and pulmonary trunk. This latter type is the only one predisposing to sudden death [26].

There are several possible mechanisms for ischemia in fourth type, when a LMCA arises from the right sinus of Valsalva. Normally, the coronary ostia are round to oval in shape, but in this anomaly, the artery has an acute angulation at the origin that could make a slit-like ostium. With increased cardiac output, the aorta dilates with stretching of the wall making the ostium severely narrowed [5,27]. Additionally, compression of the artery between the aorta and pulmonary root has been postulated [12]. The diagnosis of coronary anomalies can be made through different imaging modalities. Transthoracic echocardiography often suggests the diagnosis; however, transesophageal echocardiography is useful in identifying an intra or extramural course of the abnormal vessels [28]. In addition, coronary angiography and magnetic resonance have been found useful for making the diagnosis [28,29].

This report emphasizes that it is rare to diagnose a young patient with an anomalous left coronary artery arising from the right sinus of Valsalva and that sudden death during or shortly after vigorous physical activity is often the first indication of the presence of these coronary anomalies. Her previous seizure-like event post-exertion may have been a critical warning of the coronary abnormalities. Surgery is indicated in symptomatic ALCA (or ARCA) patients with non-lethal episodes of syncope, angina, arrhythmias, or resuscitation after cardiac [8]. Surgical intervention usually involves the direct repair of the anomalous origination in the aortic root or coronary artery bypass grafting. The management of asymptomatic patients remains controversial. The use of extracorporeal membrane oxygenation or a left ventricular assist device have been suggested for patients with severely reduced ventricular function if surgical re-implantation of the anomalous artery is feasible, or as a bridge to cardiac transplantation [30].

However, in centers where extracorporeal membrane oxygenation/left ventricular assist device are not available, such as ours, an alternative bridging solution might be insertion of an intra-aortic balloon pump that is often used in cases of left ventricular failure in adults secondary to coronary artery disease [31]. This case illustrates that anomalous origin of the left coronary artery from the opposite sinus of Valsalva (inter-arterial type) is a rare congenital cardiac anomaly, and may present at young age with catastrophic life threatening myocardial infarction.

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