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Carmen Niño-Taravilla *

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Case Report

Fibromuscular Dysplasia of the Coronary Arteries: An Infrequent Cause of Sudden Death

Carmen Niño-Taravilla [1,2,3], Hugo Otaola-Arca [4,5], Natalie Lara [1], Carla Palavecino [1], Luis Veloso-Papez [6], Maryel Riquelme [1]

[1] Pediatric Intensive Care Unit. Hospital de Niños Roberto del Rio. Santiago, Chile.

[2] Pediatric Intensive Care Unit. Clinica Indisa. Santiago, Chile.

[3] Faculty of Medicine. Andres Bello University. Santiago, Chile.

[4] Urology Department. Clinica Alemana. Santiago, Chile.

[5] Faculty of Medicine. Clinica Alemana-Universidad del Desarrollo. Santiago, Chile.

[6] Department of Pathological Anatomy. Hospital de Niños Roberto del Rio. Santiago, Chile.

***Corresponding Author:** Carmen Niño-Taravilla, Hospital de Niños Roberto del Rio. Profesor Zañartu 1085, Independencia, Región Metropolitana, Chile.

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Abstract

Sudden cardiac death (SCD) is defined as a sudden and unexpected death secondary to a cardiovascular cause. In many cases, cardiopulmonary resuscitation (CPR) maneuvers can achieve recovery of the patient, which is then called resuscitated SCD. We present the case of an adolescent with resuscitated SCD, who finally died of refractory cardiogenic shock, secondary to fibromuscular dysplasia of the coronary arteries.

Key words: coronary arteries; sudden death; fibromuscular dysplasia

Introduction

Sudden cardiac death (SCD) is defined as a sudden and unexpected death secondary to a cardiovascular cause. In many cases, cardiopulmonary resuscitation (CPR) maneuvers can achieve recovery of the patient, which is then called resuscitated SCD. Studies estimate an incidence of 0.8 to 6.2 cases per 100,000 inhabitants per year and up to 25% of these deaths occur during sports practice. Hypertrophic cardiomyopathy is the most common cause of SCD in adolescents, the second leading cause being coronary abnormalities (mainly the anomalous origin of the left coronary artery from the sinus venosus) [1].

We present the case of an adolescent with resuscitated SCD, who finally died of refractory cardiogenic shock, secondary to fibromuscular dysplasia (FMD) of the coronary arteries.

Clinical Case:

A 12-year-old adolescent with a history of syncope at the age of 9 after impact with a ball in the abdomen, with a normal cardiological study at that time and no other history of interest. He fainted while playing soccer. He was transferred by his father to a primary care center, where he arrived 20 minutes later, in cardiopulmonary arrest (CPA) with a rhythm of ventricular fibrillation. Advanced CPR was started, requiring 5 discharges of 4 J/kg, recovering sinus rhythm after 20 minutes. He was intubated by the out-of-hospital emergency team and transferred to the Pediatric Intensive Care Unit (PICU). He was admitted hypotensive and tachycardic, with 80% oxygen saturation despite 100% fiO2, with poorly reactive miotic pupils. Tests revealed mixed acidosis (pH 7.16, pCO2 55 mmHg, pO2 64 mmHg, base excess -9.8 mmol/L, bicarbonate 19.4 mmol/L) with lactic acid 64 mg/dL, compatible chest X-ray with acute pulmonary edema (figure 1), electrocardiogram with sinus rhythm at 188 bpm, no other findings and echocardiogram without pericardial effusion, structurally normal heart, with severe global dysfunction of the left ventricle, with significant apical dyskinesia, ejection fraction by Simpson of 25%, with right ventricle with adequate function. High-dose epinephrine and norepinephrine support was started, and the use of milrinone was not tolerated due to hypotension. He evolved with refractory cardiogenic shock and acute pulmonary edema with severe hypoxemia in addition to airway bleeding. Support with an extracorporeal membrane oxygenation system was proposed, but it was ruled out due to prolonged out-of-hospital CPA. Four hours after admission to the PICU, he presented a new CPA, and it was decided to suspend advanced

CPR maneuvers at 30 minutes. An autopsy was performed, finding FMD of the coronary arteries (figure 2), with no other significant findings.

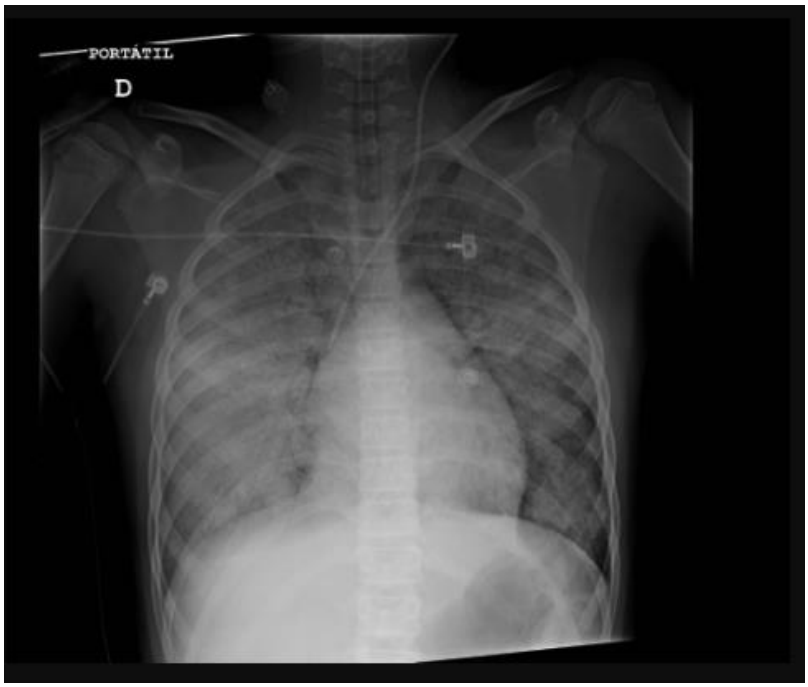

Figure 1: Chest X-ray Generalized interstitial infiltrate compatible with acute pulmonary edema. Left jugular venous catheter with a tip in the superior vena cava. Endotracheal tube 2 cm from the carina. Nasogastric tube in gastric chamber.

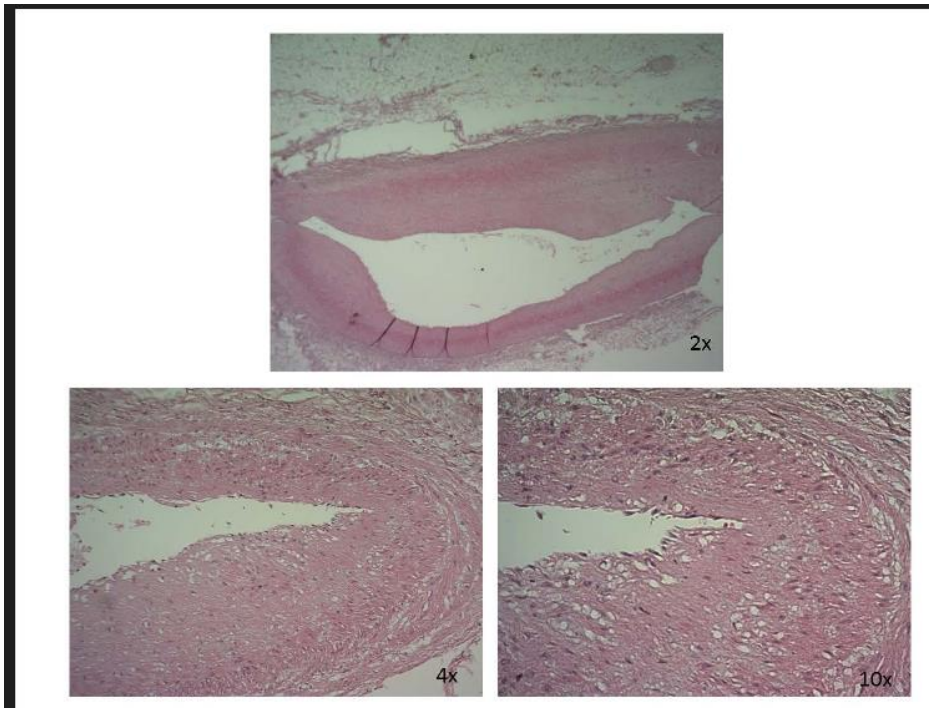

Figure 2: Cross section of the anterior descending coronary artery (hematoxylin and eosin staining) Fibrous thickening of the intima in the anterior descending coronary artery.

Discussion:

FMD is a non-inflammatory, non-atheromatous vasculopathy characterized by multifocal segmental fibrosis, usually affecting small to medium-sized arteries. The prevalence is unknown and it occurs more frequently in women, its presence in childhood being extremely rare. Its etiology is unknown, but a genetic component is suggested by the increased risk of FMD among first-degree relatives of affected individuals. Association has been found with some diseases such as pheochromocytoma, α1-antitrypsin deficiency

syndrome, Ehlers-Danlos syndrome type IV, Alport syndrome and Marfan syndrome [2].

From a histopathological point of view, it can be classified into three subgroups, according to the location of the fibroblastic changes in the tunica media (more frequent, 80-90% of cases), in the intima or in the adventitia [2].

The most common location is the renal artery (60-80% of cases), followed by cerebrovascular circulation (25-30%), with coronary involvement being rare [2].

Regarding the clinical manifestations, in adults with coronary FMD, cases of dissection have been described, leading to unstable angina, acute myocardial infarction (AMI), left ventricular dysfunction or SCD, mainly secondary to fatal arrhythmias due to disturbance in the arteries of the heart's conduction system. Diagnosis is made by coronary angiography and management follows the principles of acute coronary syndromes [2].

In childhood, there are few reported cases. Two of the cases described occurred in newborns, manifesting as cardiogenic shock at birth, both died and the diagnosis was made at autopsy [3,4]. The other two cases occurred in two 12-year-old adolescents, one of them presented SCD, diagnosed after autopsy [5] and the other presented AMI, surviving after performing percutaneous transluminal coronary angioplasty [6].

In conclusion, coronary FMD is a rare but probably underestimated cause of SCD. Diagnosis in asymptomatic patients is currently a challenge. This case highlights the importance of studies post-mortem in SCD cases to be able to provide answers and follow-up to the family.

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Conflict of interest: none

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