CASE REPORT

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Sudden Cardiac Death During Exercise due to an Isolated Multiple Anomaly of the Left Coronary Artery in a 12-Year-Old Girl: Clinicopathologic Findings

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ABSTRACT: A 12-year-old girl was taken to the Virgen Macarena Universitary Hospital in Seville, Spain, complaining of abdominal pain, nausea, palpitations, and difficulty in breathing. Her school teacher informed that the girl collapsed after a 100-m race at sports in school.

Emergency rescue personnel found the patient pale, in a cold sweat, with low peripheral perfusion. Initial ECF revealed supraventricular tachycardia (150 bpm), right bundle branch block and left anterior hemiblock that, eventually, progressed to full cardiac arrest. At autopsy, an isolated multiple congenital anomaly of the left coronary artery was found (initial segment anomaly; course anomaly and tunneled coronary artery anomaly).

In our opinion, this appears to be the first reported case of exercise-related sudden cardiac death, due to an isolated multiple congenital anomaly of the left coronary artery (at the origin, on the course and at the termination).

KEYWORDS: forensic science, forensic pathology, sudden death, congenital coronary anomaly, adolescent, exercise-related sudden death

Congenital coronary anomalies are associated with sudden death and exercise related death. Clarification of the risk and mechanisms of sudden death in patients with coronary abnormalities may aid in decisions on interventions (1). The clinicopathologic correlations in exercise-related sudden death, especially in younger people and the pediatric population, are very important in order to obtain detailed information on ECG alterations, mode of death and abnormalities of coronary arteries (initial segment, course and tunneled artery abnormalities).

We report a case of exercise-related sudden cardiac death, due to a multiple congenital anomaly of the coronary artery of a 12-year-old girl. A good clinicopathologic correlation was achieved including ECG and autopsy findings.

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Case Report

V.F.M., a previously healthy 12-year-old girl, was taken to the Virgen Macarena Universitary Hospital in Seville by her school teacher, on 21 March, 1994. The teacher explained that the girl had presented a syncope after a 100-m race at sports in school.

At the hospital, the girl was concious and exhibited great anxiety. She stated that she had abdominal pain, nausea, palpitations, and difficulty in breathing.

Emergency rescue personnel found the patient pale, in a cold sweat, with low peripheral perfusion and an important peripheral vasoconstriction that made the catheterization of the femoral vein necessary. After a few minutes, the girl showed tachypnea and dyspnea making tracheal intubation necessary. A great deal of pink fluid flowed from the cannula.

The initial ECG (Fig. 1) revealed supraventricular tachycardia (150 bpm), right bundle branch block and left anterior hemiblock that, eventually, progressed to advanced A-V block. The chest X-ray showed important right parahiliar pulmonary edema which was only slight in the left lung. Blood analysis showed metabolic acidosis and hypoxia. The patient was transferred to the intensive care unit and went, eventually, into full cardiac arrest. Advanced resuscitation efforts were unsuccessful. A final clinical diagnosis of cardiogenic shock with pulmonary edema, of unknown etiology, was emitted by the Cardiology Department.

Autopsy Examination

A complete autopsy, including toxicological screening, was performed 24 h after the death. This provided the following pathological findings: Pulmonary edema and vascular congestion with terminal regurgitation and aspiration of gastric contents, especially into the right lung. The remaining organs, with exception of the heart, showed unremarkable findings. Detailed routine toxicological investigation, habitual in our Institute, was negative.

The 160 g heart was of normal size and configuration, and we did not find valve pathology. The macroscopic parameters were: TV:7.0 cm, PV:4.0 cm, MV:7.0 cm, AV:4.0 cm, LV:1.0 cm, and

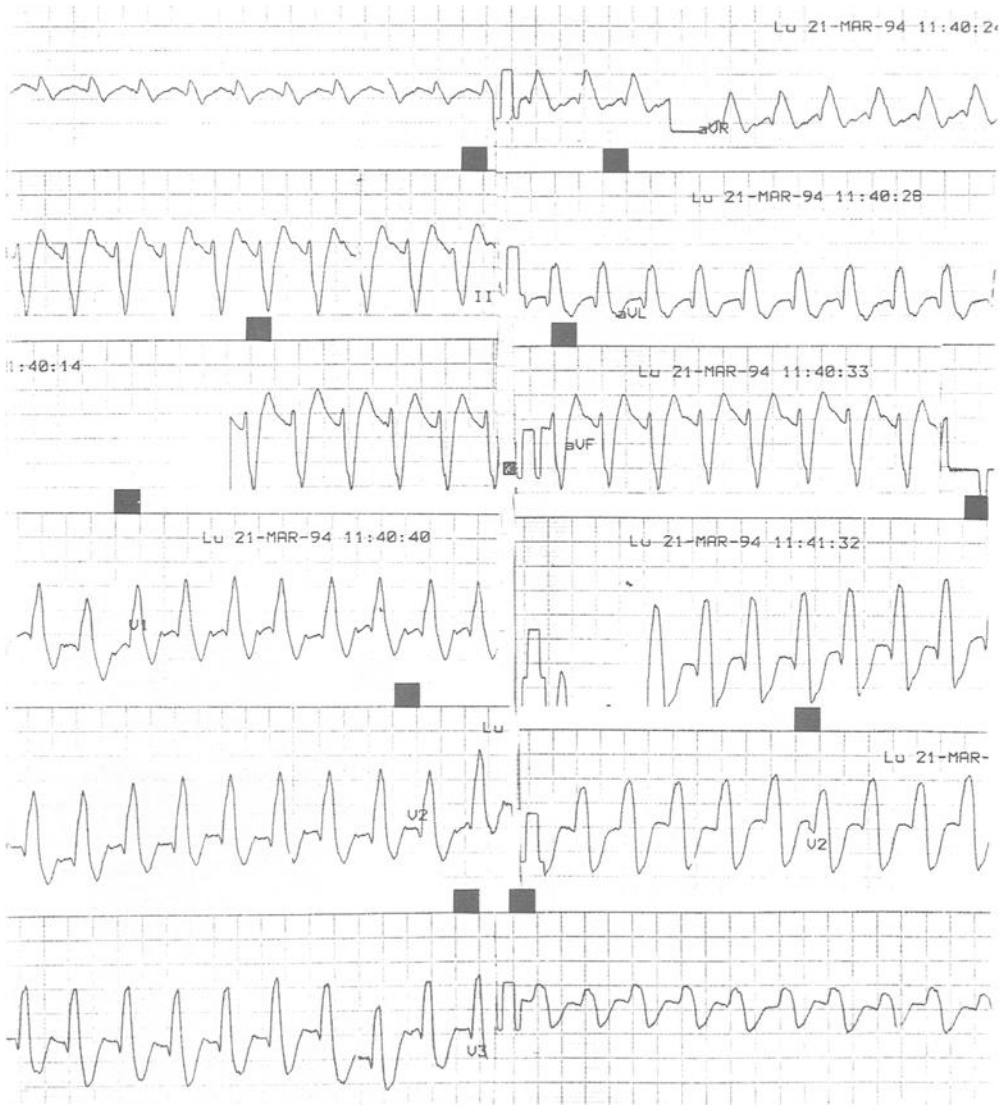

FIG. 1—*The ECG showed supraventricular tachycardia (150 bpm) with aberrant conduction and bifascicular block (right bundle branch block and left anterior hemiblock), P waves (positives in I, II and aVF, and positive-negatives in aVR, V1–V2). PR: 0.12 s; QRS: 0.12 s in III and 0.20 s in precordial leads. QT: 0.32 s with a QTc of 0.29 s. Q waves can be seen in aVL, with rS in II, III, aVF; rsR' in V1–V3 and rS in V6. The conduction disorders were attributed to an acute episode of ischemia.*

RV:0.2 cm. After opening the aortic valve, we found two separate coronary ostia located 0.5–0.7 cm above the cusp of the right aortic sinus of Valsalva (Fig. 2A). The ostium of the right coronary artery (RCA) was located nearest to the right aortic valve cusp. From this point, the artery followed a normal course. In contrast, the left main coronary artery (LMCA) originating from the superior ostium, abruptly turned towards the left ventricle and passed between the trunks of the pulmonary artery and aorta (Figs. 2B–2C). The initial segment of the coronary artery was located inside the ventral aortic wall (aortic intramural segment in Fig. 2B), immediately after its origin. Thereafter the LMCA originated the left anterior coronary descending artery (LADA), the left diagonal artery and the left circumflex artery (Fig. 2C). The LADA followed an intramural course beginning at 4.0 cm from its origin. The intramural LADA followed a course deeply placed in the interventricular anterior septum wall (0.5 cm tunnel depth in Fig. 2D). We found some foci of pale gray areas of myocardial fibrosis in the vicinity of the intramural LADA (arrow in Fig. 2D).

Histological Study—Samples from the right ventricle, left ventricle and from the intramural LADA were removed for microscopical study. The samples were included in paraffin, sectioned at 5μ, and stained with Haematoxylin-Phloxin-Eosin and Masson trichrome stain.

Microscopical Examination—The microscopical study confirmed the existence of areas of interstitial fibrosis, resulting in separation of neighboring myocardiocites (Fig. 3), and numerous ondulating fibers together with the presence of contraction band necrosis (Fig. 4), the histological hallmark of myocardial necrosis produced by temporary occlusion of a coronary artery and reperfusion (2,3), providing morphological evidence of the underlying nature of the myocardial lesions.

Discussion

Sudden death can occur occasionally in the course of certain congenital heart diseases. The most important of these are coronary

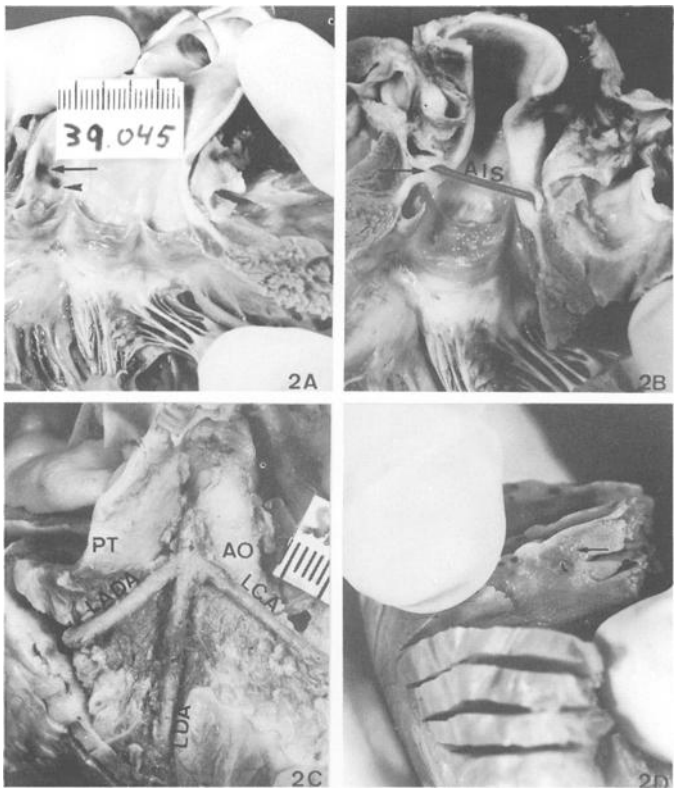

FIG. 2—*A: Both coronary arteries from the anterior sinus of Valsalva. RCA (small arrow), LMCA (large arrow), B: Aortic intramural segment [AIS] of the left coronary artery, C: Left main coronary artery [LMCA] passing between the pulmonary trunk [PT] and the aorta [AO]. LADA (left anterior descending artery). LDA (left diagonal artery). LCA (left circumflex artery), and D: Tunneled LADA. Pale gray zone of fibrosis (arrow).*

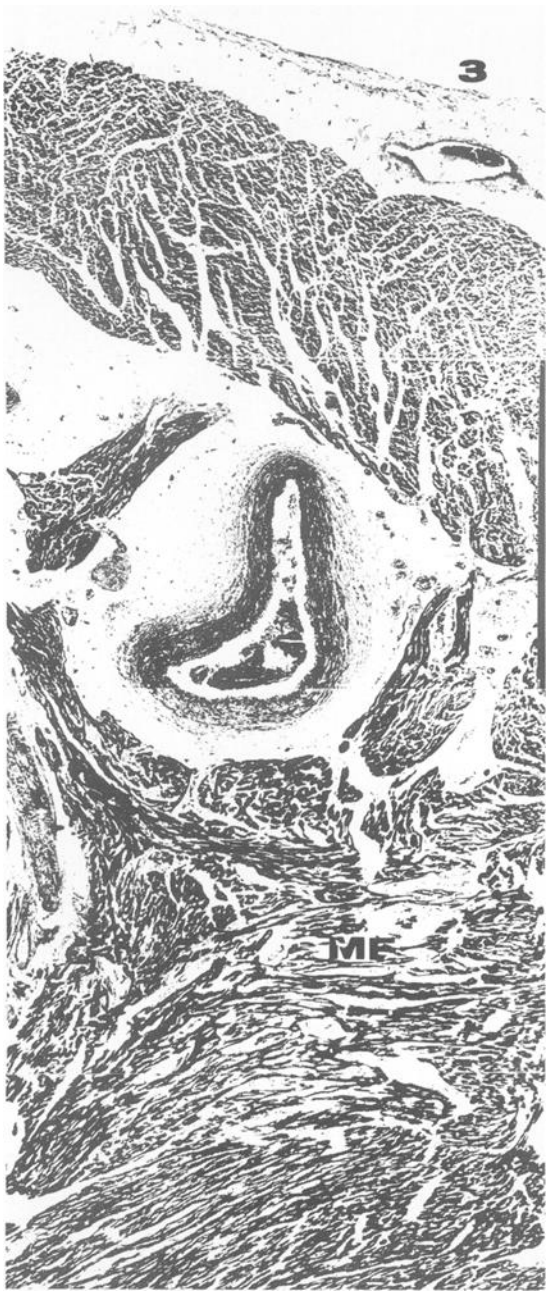

FIG. 3—*Microscopic section of the LADA. MF (myocardial fibrosis). 40 ×.*

artery anomalies, Ebstein's disease, cardiomyopathies, primary pulmonary hypertension, and the post-operative stage of Tetralogy of Fallot, interventricular septal defect, complete atrioventricular canal, transposition of the great vessels, interauricular septal defect and coarctation of the aorta (4).

Congenital coronary anomalies are an uncommon cause of sudden death, and such cases usually occur during stress. This is the second most common cause of sudden death in young athletes. Although more commonly described in the pediatric population, it has been recently recognized in adults (5). The anomalous origin of the left coronary artery (LCA), on the right Valsalva sinus or noncoronary sinus seems to be associated with a greater incidence of sudden death (especially in cases in which the anomalous artery lies between the aortic root and the pulmonary trunk) (6). Some cases of sudden death in patients with anomalous origin of the right coronary artery (RCA) on the left Valsalva sinus have been reported (7,8).

Congenital isolated coronary anomalies can be classified on the basis of their origin (ostium coronary location, ostial valvelike ridges), initial coronary artery segment (acute angle of take-off, aortic intramural segment), coursing of the anomalous artery (between the pulmonary artery and aorta), and termination (9).

In our opinion, this case illustrates a complete four-fold congenital anomaly of the left coronary artery: (a)—origin anomaly—The ostium was located above the sinotubular junction of the right aortic sinus of Valsalva. This anomaly can be considered a "positional ostium anomaly." (b)—initial segment anomaly—The coronary artery showed an aortic intramural segment, so that the initial portion of the main coronary artery was incorporated in the wall of the aorta. (c)—trunk coronary course anomaly—The LMCA lay between the pulmonary artery and the aorta. (d)—major epicardial coronary artery anomaly—The presence of an intramural left anterior descending coronary artery (LADA), deeply placed within the anterior portion of the interventricular septum (IVS), has been considered a pathological variant when LADA can be associated with myocardial ischemia (10). In this case we have found chronic ischemic pathological lesions in the territory supplied by the artery (interstitial fibrosis in the immediate vicinity of the deeply located

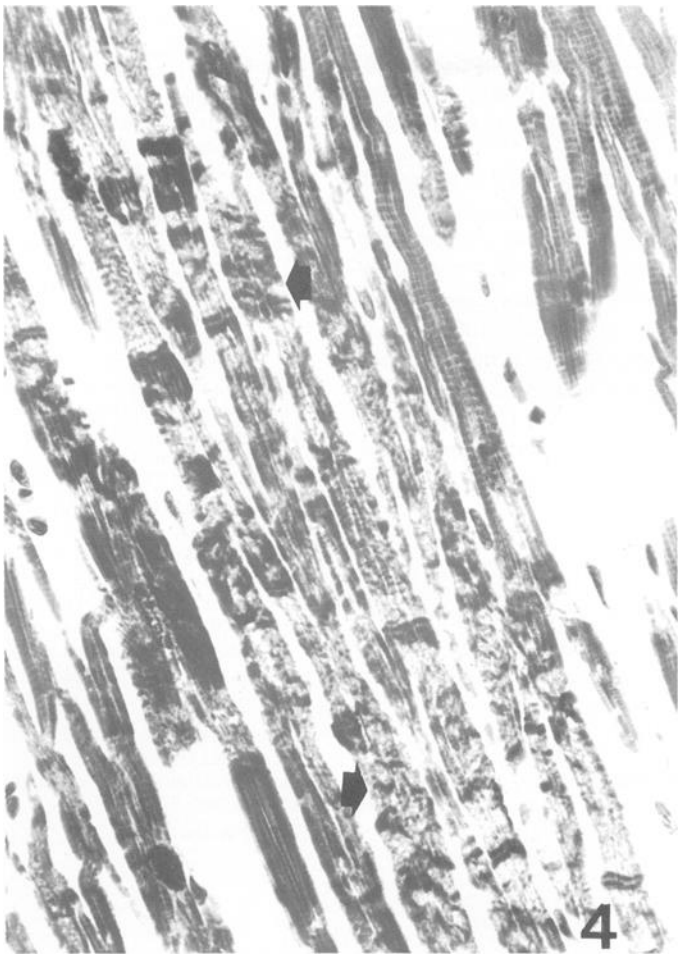

FIG. 4—*Contraction band necrosis located at the territory supplied by LADA (anterior septum wall) (arrows). 400 ×.*

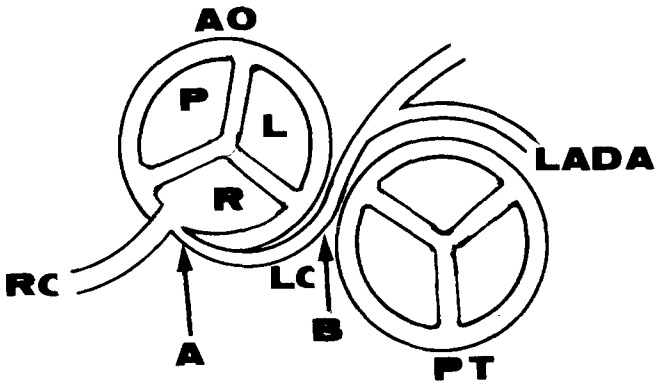

FIG. 5—*Diagram of the anomalous origin of the left coronary artery on the right Valsalva sinus. Note the existence, in this diagram, of two of the three suggested mechanisms of coronary closure (arrows): (A) ostial closure with expansion of the aorta and pull on the left coronary artery. Flap-like closure of the ostium by the coronary arterial wall, and (B) compression of the left coronary (main stem) between the trunks of the pulmonary artery [PT] and aorta [AO] (due to the increased systolic volume), P (posterior), R (right), L (left), RC (right coronary artery), LC (left coronary artery. LADA tunneling, the third suggested pathophysiological mechanism of arterial compression during exercise related sudden death, is not represented in the diagram.*

segment of the intramural LADA) together with contraction band necrosis (expression of acute myocardial ischemia).

For these reasons, we agree with Morales et al. (10), when they considered that the presence of myocardial lesions, in the territory of the intramural vessel, is the key to identifying an abnormal intramural LADA from a simple anatomic variant.

The group of Virmani (9) have reviewed 242 patients with isolated coronary artery anomalies in order to obtain information on: Mode of death, abnormalities of the initial segment, and the course of the anomalous coronary artery. They found that sudden death and exercise-related death were most common when the left main coronary artery originated from the right coronary sinus. High risk anatomy defects include: Abnormalities of the initial coronary artery segment or coursing of the anomalous artery between the pulmonary artery and aorta. Unfortunately this case confirmed the conclusions of the authors that younger patients with an isolated coronary artery anomaly are at risk of dying suddenly and with exercise.

In this case, exercise-related sudden death could be due to, at least, three pathophysiological mechanisms (see diagram Fig. 5): (a) Kinking at the origin (Flap-like closure of the ostium by hyperdistention of the aorta wall due to the increased systolic volume), (A in Fig. 5), (b) trunk coronary compression during coursing of the artery between the pulmonary artery and the aorta, (B in Fig. 5), and (c) compression and/or diastolic time lag, for reestablishing

blood flow, in the intramural LADA. These facts together with enhanced oxygen requirements during exercise, produce an imbalance between oxygen supply and demand, which facilitates the appearance of malignant ventricular arrhythmias and sudden death (10).

As pointed out by some authors (5), the congenital anomalies of the coronary arteries are an important cause of sudden death in otherwise healthy adolescents and young patients. Actually this is considered the second most common cause of sudden death in young athletes who die during exercise. For this reason, it is important to note in forensic pathology that in all cases of exercise-related sudden death occurring in young people, it must be born in mind that during routine postmortem examination of the heart, there are some important characteristics to consider: Ostium coronary location, valvelike ridges, the aortic intramural segment, the angle of take-off, the arterial course, and termination (tunneled coronary artery) of the coronary vessels.

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