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Letter to the Editor

Why can a case report be so important? The case of sudden cardiac death, caused by a coronary anomaly

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Coronary artery anomalies (CAA) are one of the most common causes of sudden cardiac death (SCD) in the young, in athletes and in military recruits [1–3]. Essentially, anomalous origin of a coronary artery from the opposite sinus of Valsalva (ACAOS, when featuring intramural proximal course of the ectopic artery [3]) is the only type of CAA that is a recognized culprit [1–3]. Current understanding of the patho-physiology of these conditions is still tentative, and the students of such active field of clinical investigations are quite interested in the exact clinical circumstances, during the development of such unusual, unexpected tragedies. Effective treatment requires such understanding.

The accompanying article by Kutreti et al. [4] presents in remarkable, unusually available details the case of a 14-year-old girl, who succumbed to one of such cases of SCD.

The main findings in the present case

1. The young teenager was an active jogger, who had suffered a prior event of syncope complicated by seizure that required hospital admission. Work up was negative, at the time, but did not include imaging studies to rule out CAA.
2. She presented to the emergency room of a local hospital, brought by the parents in a private car, following an episode of syncope, preceded by chest pain and dyspnea during brisk jogging.
3. On admission, she was still alert and oriented, but with chest pain, dyspnea and extreme weakness.
4. She was found to be in a state of cardiovascular shock, pulmonary edema, and acidosis. Her rhythm was regular sinus.
5. Echocardiography showed diffuse left ventricular hypokinesia, with an ejection fraction of 38%, but no CAA could be identified.
6. The clinical pattern continued to deteriorate, in spite of aggressive, even though routine treatment (vasopressors and volume expansion), and resuscitation efforts had to be abandoned 4 hr after the onset of the event.
7. Autopsy presented evidence of diffuse and exclusively acute myocardial injury, with contraction band necrosis, that included the right coronary territory (evidently a consequence of prolonged shock state).
8. L-ACAOS was found during the post-mortem study, with the left coronary ostium located slightly to the right of the anterior-right aortic commissure, with a slit-like tangential takeoff, that was followed by

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intramural course, inside the aortic wall during the proximal 10–12 mm of the ectopic vessel. The distal part of this intramural segment was found to be only covered by the aortic adventitia, but not inside the media of the aorta, which is the typical finding in such cases [1–3]. The compelling case of proximal stenosis is well made by Fig. 3 that shows a severe lateral compression of the ectopic vessel, with a longitudinal to transverse diameter ratio of approximately 9:1 (a normal coronary artery is circular in cross-section). This is an unusually severe case of stenosis.

Patho-physiology explained?

The fundamental messages learned by the careful and detailed description of such clinical case seem to indicate:

1. The patient could reach the age of 14 yr of age, being fairly active and fit, until the final 2 episodes (syncope, and then SCD).
2. Sadly, the alarming initial symptom of syncope was not properly diagnosed as being related to such rare condition of CAA. Syncope is the most predictive prodromic symptom in similar cases, even though most cases present SCD as the first recognized evidence of a disease state [1–3,5].
3. As typical in these cases of SCD, no anatomic changes occur during the final SCD event, that can be related to evolution in the anatomic features of CAA (in particular, there was no evidence of intravascular thrombosis, as in unstable ischemic acute syndromes in the adults with atherosclerotic coronary artery disease). No clear scarring suggestive of chronic ischemia was found by the detailed histologic survey.
4. Ischemia most definitely was responsible for the unresponsive shock and heart failure presentation (as commonly seen in cases of atherosclerotic left main coronary critical lesions). Fig. 3, in paper of Kutreti et al. [4], illustrates quite vividly the extreme severity of the left main trunk stenosis. It is quite likely that aggressive exercise precipitated worsening of stenosis in this unfortunate girl, reaching the level of critical severity. Once symptoms of critical ischemia occur, the recovery is unlikely, since hypotension and low cardiac output/acidosis lead to perpetuation of the syndrome, since the intramural stenosis likely worsens in those conditions [3].

In previous, extensive intravascular ultrasound imaging experience in our center [6–8] has consistently proven that all the cases of ACAOS (also named the "between aorta and pulmonary artery" variant) feature intramural course, like shown by necropsy findings [9]. The variability that is critical to recognize in the clinical evaluation of an individual case is still left to be correlated precisely with prognosis, but it is likely related to the cross-sectional stenosis at baseline (in diastole), the intramural versus the sub-adventitial course, the systolic/diastolic phasic pulsation of such section of the ectopic vessel, and its relationship with the level of exercise and aortic root compliance [3,7].

5. Arrhythmias are frequently blamed in the literature for these cases of SCD, but in the present case, the authors had a unique chance to document that this was not the mechanism of death, having a prolonged period of observation in the presence of unresponsive cardiovascular collapse [4]. Knowledge of a definite patho-physiological mechanism is essential for effective treatment in these emergencies: if critical left main stenosis can be confirmed in larger series to be the culprit feature, only artificial mechanical support of the failing heart will be likely effective, in similarly critic circumstances. The Tandem (Cardiac Assist, Inc, Pittburg, PA) or the Impella (Cardiotechnik, Aachen, Germany) artificial left ventricular support percutaneous devices are the most promising means to restore hemodynamic stability in these scenario, when available [3,10].
6. The obvious condition to indicate artificial mechanical support intervention, in a given case of similar nature will be the demonstration of the presence of ACAOS, if not previously available. CTA or MRA are the only reliable imaging technique that can be used, even though they usually require difficult transfer of a critically ill patient to a department of radiology, since echocardiography is rarely diagnostic in adult-size individuals.

We hope this compelling case presentation can improve the understanding and the awareness of the complexity of the problems presented by CAA emergencies, and can encourage both more effective preventing programs (using populations-based screening [3]), and emergency interventional treatment in the unfortunate cases that should be able to arrive to a prepared and qualified hospital during an episode of sudden collapse.

In the rare cases of CAA complicated by SCD, that should arrive to our clinical observation, it is still essential

to publish even individual case reports, (and maybe to create prospective Registries of these rare pathologies), in order to present in-depth documentation of this peculiar pathology. The authors should be commended for the present report, even when we should realize that prevention, by effective and cost/effective screening followed by individualized intervention, should be considered the preferred prospective policy.

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