Case study

Heart arrest revealing a killer coronary artery

Abstract:

Anomalous origin of the left main coronary artery is a congenital heart defect in which it arises from the right coronary sinus. This anomaly carries a high risk of sudden death. The diagnosis can be made by ultrasound examination. Silent myocardial ischemia must be sought. Various surgical techniques have been described. We report the case of a child who presented with cardiac arrest occurring at rest, recovered by external cardiac massage and which has benefited from a surgical intervention with a good evolution.

Keywords:

Congenital coronary artery anomalies; interarterial pathway; computed tomography angiography; sudden death.

Introduction:

Congenital anomalies of the coronary circulation are an important issue in cardiology and cardiovascular surgery. It is an important entity because of its clinical impact, the risk of sudden death, its prevalence and the complexity of practical management. They concern 0.3 to 1.5% of the general population[1].

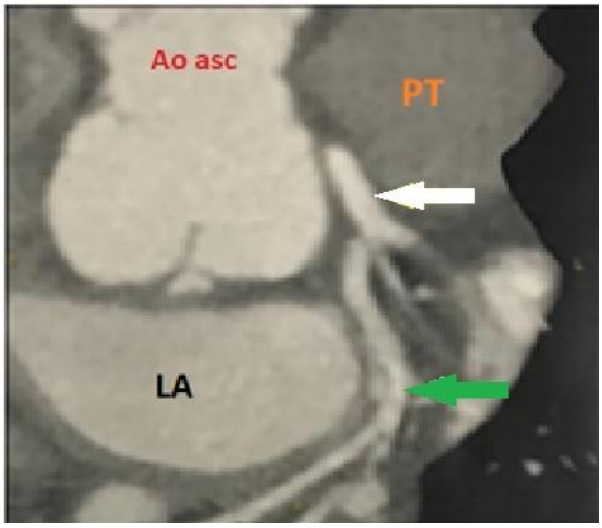
The abnormal origin of the left main coronary artery is a rare birth defect, typically found in infants or young adults when exploring for syncope or sudden death occurring during or after exercise[2].

We report the clinical situation of the abnormal origin of the left coronary artery with an inter-arterial path, revealed by sudden death, concerning a clinical case presented in our hospital.

Case presentation:

15-year-old patient, with no particular pathological history and no cardiovascular risk factor, admitted for cardiac arrest, recovered in the school after 3 minutes of cardio-pulmonary resuscitation.

On admission, after conditioning the patient, physical examination and an ECG were normal. Cardiac ultrasound showed heart chambers of preserved size and systolic function. Coronary angiography revealed birth defects of the left coronary and right coronary artery which both originate from the antero-right sinus with an inter-aorto-pulmonary malignant course. The coro-scanner confirmed the diagnosis showing the birth of the left coronary artery of a common trunk with the right coronary from the right anterolateral sinus and describes a malignant inter-aorto-pulmonary path without athero-calcific coronary infiltration (Figure 1). Magnetic resonance imaging demonstrated the same angiographic and CT findings, moreover, it found no argument in favor of interstitial fibrosis, or myocardial infarction or myocarditis (Figure 2).



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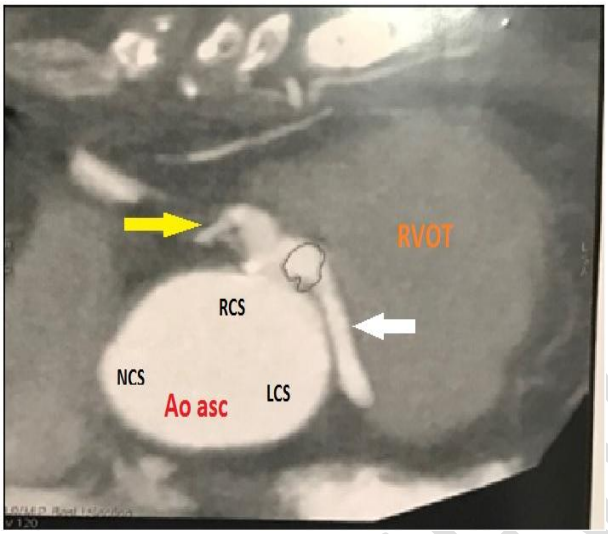

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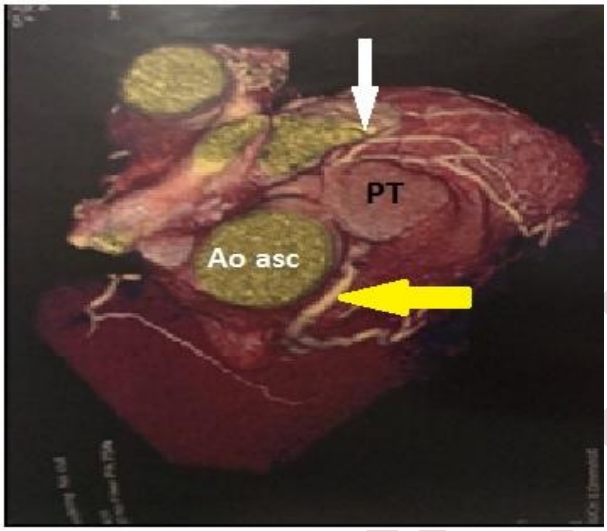

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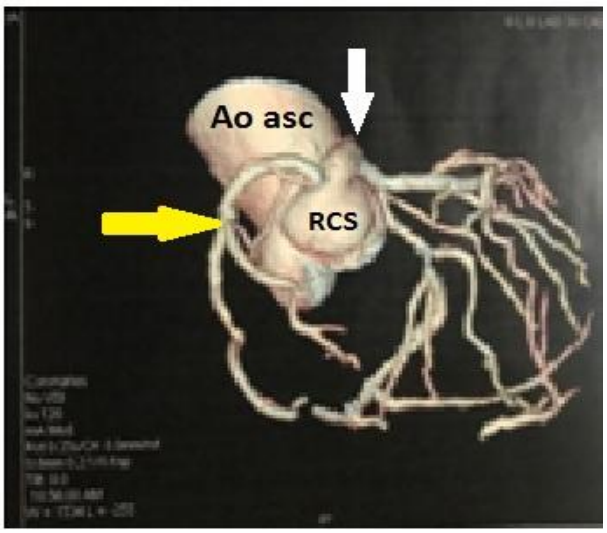

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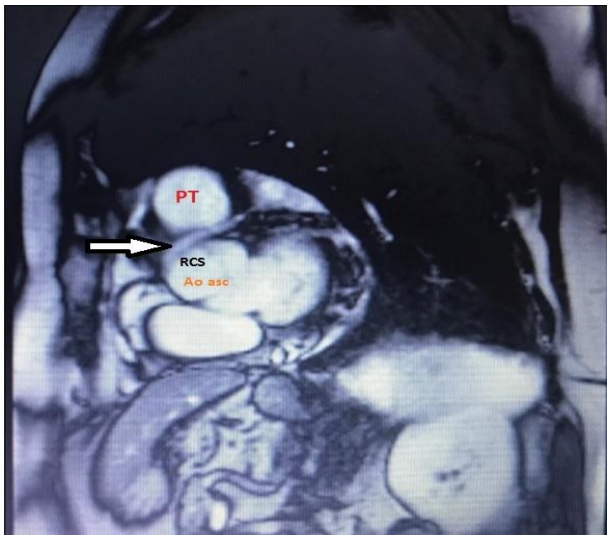
- **Figures 1:**

The coro-CT scan (a- coronal plane; b- axial plane; c and d - Reconstructed 3-D VRT) showing the origin of the left coronary artery from a common trunk with the right coronary artery from the right coronary sinus and describes a malignant inter-aorto-pulmonary path. **Ao asc :** ascending aorta; **PT** : pulmonary trunk; **LA** : left atrium; **RCS** : right coronary sinus; **LCS** : Left coronary sinus; **NCS** : non-coronary sinus; **white arrow** : left main trunk; **yellow arrow** : right coronary artery; **green arrow :** left circumflex artery.



- **Figure 2:**

The coro-MRI sagital plane, showing the origin of the left coronary artery from a common trunk with the right coronary artery from the right coronary sinus and describes a malignant inter-aorto-pulmonary path. **Ao asc :** ascending aorta; **PT** : pulmonary trunk; **white arrow** : left main trunk.

The case was presented in a multidisciplinary session; it was decided to proceed with surgery with the creation of a neo-ostium. In surgery, we repositioned the left coronary artery, creating a new ostium in the appropriate aortic sinus with good evolution.

Discussion:

The left coronary artery is a coronary artery that arises from the aorta above the left cusp of the aortic valve, and supplies blood to the left side of the heart. The origin in the left sinus of Valsalva is therefore not an essential condition to identify this artery, but in cases where this does not occur, we would speak of a situation of abnormal origin of the left coronary artery[3,4]. In these situations, the description of the arterial path in relation to the large vessels (pulmonary and aortic arteries) is important, in order to exclude the presence of an inter-arterial path [3,5,6].

The categorical method that is used the most frequently is the ectopic origin site, which involves ectopic origin of coronary arteries from 1) the aorta, either from a wrong sinus or beyond the sinuses; 2) the pulmonary artery; 3) as a branch of another coronary artery; 4) other arteries; and 5) the ventricular chamber. Ectopic aortic origin is the most common type, of which an anomalous origin from a wrong sinus of Valsalva predominates[7,8].

Several types of aberrant pathways are described: inter-arterial, retro-aortic or retro-cardiac, pre-pulmonary or pre-cardiac, intra-septal or sub-pulmonary. Apart from the inter-aortico-pulmonary path which can be the cause of sudden death, these pathway abnormalities of the coronary branches have no clinical consequences (the retro aortic path may lead to an early atheroma)[3,5,9].

They may present clinically with atypical exertional chest pain with normal ultrasound. Sudden death can also occur on exertion in young people[5,10]. They represent 12 to 19% of the causes of sudden death in young athletes[2,11,12].

The mechanism most often mentioned is an increase in pressure and dilation in the aorta and the trunk of the pulmonary artery during exertion, responsible for compression of the coronary artery which has an aberrant path between these two vessels with myocardial ischemia[13].

The second hypothesis put forward to explain myocardial ischemia is the bypassed path, by kinking of these coronary arteries running between the aorta and the pulmonary artery, potentially responsible for a plication of the coronary artery. This kinking may be responsible for a myocardial hypoperfusion in the territory of the affected coronary artery [13].

Finally, some pathologists suggest the presence of a fibrous fold in the ostium of ectopic coronary arteries whose implantation is tangential to the aortic wall[14,15]. This fibrous fold, to be distinguished from submucosal pathways, is potentially responsible for stenosis of the ostium of the coronary artery during an increase in pressure at the level of the root of the aorta, resulting in myocardial ischemia, especially during effort.

Anomalous left coronary arteries originating from the RCA appeared to be more susceptible to atherosclerosis with a narrower proximal section when it came to anomalous coronary arteries. This is most likely caused by proximal narrowing, which may increase endothelial shear stress[16,17]. Blood flow friction on the endothelial surface is responsible for the tangential force applied to the vessel wall [18], [19], which increases the risk of atherosclerosis in this anomaly.

The coronary angiography allows the diagnosis of this type of coronary anomaly in the majority of the times[9]. Nevertheless, a detailed analysis by the multislice coronary scanner with ECG synchronization has many advantages in the detection and exploration of coronary artery path abnormalities[20,21].

MRI appears to be a promising test for exploring these abnormalities because it is a non-irradiating test, not requiring the injection of iodinated contrast medium. Some authors suggest performing a 3D angio-MRI with respiratory synchronization for the assessment of these ectopic arteries[20]. But MRI, in this indication, remains, despite these advantages, a longer and less reproducible imaging technique than the multisection scanner[9].

The inter-arterial path says the killer coronary presents risks [5,22,23], we note:

- Arterial compression between the two large vessels
- Arterial spasm by closing the angle between the coronary path and its ostium or ostial valve on exertion
- Paroxysmal rhythm disturbances on exertion
- Chronic ischemic heart disease

Thus, an accurate and early diagnosis of this risky path is essential in order to assess the need for surgical treatment which, although heavy and aggressive, alone can prevent the inherent risk of sudden death[21]. CT is therefore invaluable in selecting possible candidates for cardiac surgery in patients with such malformations.

A special case that must also be mentioned in the case of an inter-arterial path is that which follows the surgical repair of a congenital heart disease. This is the case of patients who have had a transfer of the coronary arteries after an arterial switch for transposition of the great vessels or of patients in whom a tube has been interposed between the right ventricle and the pulmonary artery for a right obstacle.

The management of these coronary abnormalities requires a multidisciplinary approach, which includes the clinical cardiologist, the cardiovascular surgeon and other specialists such as interventional cardiologists and cardiovascular radiologists.

It is crucial to carefully stratify the risk when treating patients, taking into account both the risk of planned procedures and the risk of sudden cardiac death. Surgical treatment should be taken into consideration for patients who have a high risk of sudden cardiac death: CABG, coronary reimplantation, or coronary unroofing[24,25].

Given the possibility for competing flow from native vessels to result in graft failure when choosing to pursue surgical treatment, data suggests that coronary artery bypass graft should be avoided in the absence of concurrent obstructive coronary artery disease. In terms of surgical repair methods, coronary unroofing is typically chosen in patients with an early intramural course, when practical. Other possibilities include coronary reimplantation, fenestration, neo-ostia creation, or combination methods. Aortic valve commissure and its support must always be protected against iatrogenic injury[9].

Conclusion

Coronary artery birth and path abnormalities are rare. Among these anatomical variations, those with the greatest risk of complications are those whose aberrant path runs between the trunk of the pulmonary artery and the aorta. They can be responsible for myocardial ischemia on exertion and sudden death in young subjects. It therefore appears necessary to search for this anomaly systematically, at least in all young adults who engage in strenuous sports activities.

Ethical Approval:

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

Consent

As per international standards, parental written consent has been collected and preserved by the author(s).

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