ANOMALOUS AORTIC ORIGIN OF THE LEFT MAIN CORONARY ARTERY ASSOCIATED WITH ARTERIAL COMPRESSION - CASE REPORT

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ABSTRACT
Anomalous Aortic Origin of Coronary Artery (AAOCA) is a rare finding, with varied presentation and symptomatology. Increasingly recognized by cardiac imaging, when found it raises questions about the appropriate approach and management. We present a case of an 11-year-old female who presented with episodes of shortness of breath, angina and syncope during exercise. Further investigation demonstrated episodes of nonsustained ventricular tachycardia on Holter and coronary angiotomography revealed that the left coronary artery had an anomalous origin from the right cusp with initial short intramural segment and significant external compression in its initial course between the aorta and the pulmonary artery. Patient was submitted to surgical correction with dissection of left coronary artery posterior to the pulmonary artery, coronary arteriotomy, roof ampliation with autologous pericardium and creation of neo-ostium in aorta. Patient had satisfactory postoperative recovery, was discharged on the fifth day post op, and remains asymptomatic after six months follow-up. Herein we present surgical video and postoperative echo and CT scan.

ABBREVIATIONS, ACRONYMS & SYMBOLS

AAOCA - Anomalous Aortic Origin of Coronary Artery
ALCA - Anomalous Left Coronary Artery

CABG - Coronary Artery Bypass Grafting
CT - Computed Tomography
EKG - Electrocardiogram
LMCA - Left Main Coronary Artery
PA - Pulmonary Artery
POD - Postoperative Day
SCD - Sudden Cardiac Death

INTRODUCTION

The second most prevalent cause of SCD in young athletes (<35 years) during vigorous exercise is coronary artery anomaly [2]. The AAOCA from the opposite sinus of Valsalva has received special attention due to its relationship with SCD in healthy individuals, with prevalence in the population being around 0.1 to 0.3%, and the right coronary artery arising from the left sinus of Valsalva is estimated to be 6 to 10 times more common than anomalous left main coronary artery (ALCA) from the right sinus Valsalva. Despite the lower incidence, this presentation is more related to SCD due to the narrow pathway between the ascending aorta and the pulmonary artery, explained by the increased blood pressure of the aorta and the pulmonary artery during periods of high myocardial metabolic need that leads to compression of the ALCA and progress with myocardial ischemia, which symptoms during exercises covers chest pain, dyspnea, ventricular dysrhythmias and SCD, while the pathophysiology is not clearly elucidated yet [3-6].

In addition to compression, the mechanisms of SCD during ischemia includes: obstruction of the left coronary artery in its intramural segment and narrowing of the ALCA slit-like ostium with increased aortic pressure; and its acute take-off angle [4]. Reports suggest numerous risk factors for SCD linked to this anomaly, such as symptoms (mostly syncope; less often chest pain during or after exercises); age <29 years; signs of myocardial ischemia and ALCA with proximal path associated with intramural path, especially when the latter one has coronary length >1.5cm. [4, 5] Up to 15% of SCD and syncope could be linked to anomalous coronary artery and the etiology of SCD is likely to be ischemia that can course with pump failure [5].

Congenital Heart Surgeons' Society Registry (CHSS) and other authors have included all pathologic variants in a single group called generically "anomalous aortic origin of a coronary artery" (AAOCA) without subclasses or strict definitions. Eventually, interarterial course was added to the definition of AAOCA. Recently proposed nomenclature "anomalous origin of a coronary artery from the opposite sinus (ACAOS)," to this term is added an "L" or "R" prefix to indicate the affected coronary artery (LCA or RCA) and a suffix to indicate the abnormal proximal course: "IM" for intramural, "PP" for prepulmonic, "SP" for subpulmonary, "RA" for retroaortic, "RC" for retrocardiac, or "WA" for wrapping around the apex. L-ACAOS-IM constitutes the most serious group of LCA anomalies. And this is exactly the presentation of the patient discussed in this case-report [7].

Data determined by screening magnetic resonance imaging (s-MRI), in a continuous series of more than 5,000 school children from the general population suggest that close to 0.45% of the US population has ACAOS: either R-ACAOS-IM (prevalence, 0.35%) or L-ACAOS-IM (prevalence, 0.1%) [8].

This article presents a case report of a L-ACAOS-IM successfully managed with surgical treatment.

CASE-REPORT

PATIENT INFORMATION

11-year-old female patient. Child was previously healthy and no family history is known as she was adopted as a newborn.

CLINICAL FINDINGS

Patient presented to the clinic with a history of syncope after physical effort. Symptoms were also associated with intense emotions, and the last episode occurred during a school dance. She complained of episodes of shortness of breath, angina and syncope during exercise for the last year.

TIMELINE

(Fig 1)

DIAGNOSTIC ASSESSMENT

Concerned with this situation the patient's mother was able to schedule an transthoracic echocardiogram and a Holter even before any medical evaluation. The echocardiogram didn't disclose any abnormalities and the Holter revealed episodes of nonsustained ventricular tachycardia and sinus tachycardia in moments when the patient complained of chest discomfort.

With this in hand, and after initial medical evaluation, the patient was sent to our hospital to perform an cardiac stress test, and during the exam she experienced the same symptoms she previously reported during exercise. The test was interrupted and she was transferred to the emergency unit for further care. Initial EKG was normal and a new echocardiogram displayed normal ventricular function with no dyskinesia, right coronary had normal path and origin, however the left coronary however had an interarterial path and its origin could not be visualised through the echocardiogram.

A coronary angiotomography was then ordered and revealed that the left main coronary artery had an anomalous origin in the right valsalva sinus of the aortic valve, in the plane of the sinotubular junction (Fig 2). Initial path was interposed between the aorta and pulmonary artery, where there was relevant compression and significant luminal reduction, with a short intramural course (Fig 3).

Due to the repeated symptomatology, the worrisome characteristics and possible malignant evolution of the anomaly surgical treatment was indicated by the Heart Team.

THERAPEUTIC INTERVENTION

The operation was performed through a median sternotomy using aortic and bicaval cannulation for cardiopulmonary bypass and cooling to 30oC, Followed by aortic cross-clamping and administration of antegrade cold blood Del Nido cardioplegia.

Aorta and pulmonary artery were transected, during the process of dissection a small punctiform structure that corresponded to the left coronary artery ostium in the right sinus of Valsalva was identified, right next to the commissure with an initial intramural path and presenting with an strained course in its proximal portion.

At that moment the coronary path was dissected posterior to the pulmonary artery and a longitudinal section of the left coronary performed, proximal to its take-off from aortic wall taking extra care not to damage the artery. Aortic wall incision was made at the point of most contact with LCA take-off.

Finally the arteriotomy was closed by creating a non-tension roof with fixed autologous pericardium roof that extended until the aortic incision, thus creating a new ostium, but preserving the previous ostium (Figs 4-5). Pulmonary artery was re-anastomosed with section of ligamentum arteriosum to relieve any tension and provide better mobilization. The aortic cross-clamp was released and the operation completed in the standard manner.

Postoperative transthoracic echocardiogram exhibited good biventricular function and presence of neo-ostium with good blood flow.

FOLLOW-UP AND OUTCOMES

Initial postoperative recovery was uneventful, patient was extubated after five hours of the surgery, vasoactive drugs were discontinued after 24 hours and the patient was transferred to the infirmary on the POD. Surgical drains and pacing wires were removed on the third POD and the patient was discharged home on the fifth POD.

Control coronary angiotomography revealed the neo-ostium from the left coronary sinus and the maintenance of the previous short intramural path (Fig 6). The ostium measured 2x4mm , with an ovoid conformation and without any significant luminal reduction. Control echocardiogram displayed the left coronary neo-ostium connected to the left coronary sinus, completely pervious, with good flow by color mapping on the left main coronary artery and the anterior descending artery, and good ventricular function.

Patient remains asymptomatic after six months follow up, new cardiac stress was performed without any symptoms during exercise, arrhythmias or any other abnormality suggestive of ischemia.

DISCUSSION

Transthoracic echocardiography is used in young patients, but it is operator-dependent to localize the coronary ostia, limiting his accuracy [9]. The electrocardiogram is abnormal in only 50% of symptomatic patients and often normal in asymptomatics, but a ST-elevation in lead aVR, that could suggest left coronary artery disease, would increase anomalous left coronary artery suspicion in young patients without risk factors for coronary artery disease; while coronary CT scan conclude diagnosis [5,10].

Surgery is indicated for all patients with ALCA from the right sinus of Valsalva due to the high risk of SCD. There are a range of surgical techniques to correct this anomaly, such as CABG which is usually performed with an internal mammary artery as the conduit, but it is not recommended in young patients due to the unrestricted flow through the anomalous coronary artery at rest, leading to decreased patency with stenosis secondary to competitive flow [6,9]. In this option, ligation of the coronary artery proximal to the suture graft has been advised [6,9]. Percutaneous coronary intervention is not seen as a routine option to treat these patients, as few evidence exists, and the American College of Cardiology/ American Heart Association Task Force advises surgical procedures as the only path to correct those defects [9].

Lateral pulmonary translocation is a technique where the main pulmonary artery is transected just before its bifurcation and an opening is made in the left pulmonary artery; the surgery continues with closure of previous bifurcation and the main pulmonary artery is then translocated laterally to the left, aiming to reduce the potential compression of the anomalous vessel path [6].

The target of this technique (lateral or anterior translocation) is to distance the pulmonary artery from the ascending aorta, so the risk of compression of the anomalous coronary artery is reduced, but does not involves potential etiologies of coronary insufficiency, as slit-like deformation of the ostium, or punctiform as in our case, or stenosis of the intramural course [6]. So, this technique should be restricted to patients where all other alternative techniques are not possible, as in rare conditions such as single coronary artery or interarterial course, where anomalous coronary artery runs between aorta and pulmonary artery, without an intramural course.

In the last scenario, excision of the anomalous ostium followed by reimplantation into the correct sinus promotes ‘anatomical’ repair [6]. Despite lateral pulmonary translocation provides good results in certain anatomical variants, it should be avoided when intramural course is present because it is left intact and promotes the substrate for residual obstruction, so unroofing of the intramural course could be used in those patients [6].

Unroofing is obtained by excising the common wall between aorta and coronary artery, but here we have two scenarios: (1) the intramural segment is above the valvar commissure, so entire course is unroofed, but usually (2) the intramural segment is below the sinotubular junction, with close proximity to valvar commissure, in which case a neo-ostium is made in the aortic wall of the correct sinus, opposite to the site where the anomalous vessel borns from the aortic wall and the ostium is enlarged towards the commissure, which is meticulously preserved and once this is done, the intima tunic common to the aorta and the coronary is sutured in the neo-ostium to prevent dissection and then it is created with a more suitable take-off angle [6]. However, the most distal part of the intramural segment, at the site where the coronary exits the aortic wall, is left intact and this area could be harshly stenosed that could lead to ischemia; moreover, this technique have good results in short-term, but in some cases affects the commissure, so it compromises the aortic valve competency that probably results in aortic insufficiency in mid-term [4,6].

Karl et al proposed in 2008 a more physiological technique by opening the anomalous coronary artery from the ostium down to the normal epicardial artery path, then suturing a patch over the opened parts, the ostium is wided to amplify the proximal diameter and as the interarterial course is left in place, the pulmonary artery is translocated to amplify space between the basal arterial vessels, making this procedure as one that considers all potential mechanisms that can promote ischemia [6].

The technique employed in our case, as suggested by Gaudin et al, proposes a more anatomical correction where the anomalous segment maintains its integrity but bypassed and a new wided coronary ostium is created in the correct sinus of Valsalva, excluding completely the intramural segment and recomposes the normal angle of take-off, allowing this method to all anatomical variants, with or without intramural segment, and the patch of choice should be autologous fresh or glutaraldehyde treated pericardium, instead of saphenous vein that may be affected by gradual dilatation, where reoperation could be needed [6]. The short and mid-term results are convincing, while the long term is not yet determined.

INFORMED CONSENT

The patient and her parents have given their informed consent for this case report to be published.

FINANCIAL SUPPORT AND CONFLICT OF INTEREST

No financial support. No conflict of interest.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

AUTHOR'S ROLES & RESPONSIBILITIES

DFT Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; final approval of the version to be published

ACABF Substantial contributions to the conception or design of the work; or the acquisItion, analysis, or interpretation of data for the work; final approval of the version to be published

GPG Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; final approval of the version to be published

MRGC Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; final approval of the version to be published

VM Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; final approval of the version to be published

LAM Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; final approval of the version to be published

MBJ Drafting the work or revising it critically for important intellectual content; final approval of the version to be published

FBJ Final approval of the version to be published

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