Case study

LATE AND INCIDENTAL DISCOVERY OF ACAOS AFTER STEMI

ABSTRACT :

Coronary artery anomalies (CAAs) comprise a diverse group of congenital malformations with widely varying expressions and pathophysiological mechanisms. The most notable group of CAAs has been termed 'Anomalous coronary artery originating from the opposite sinus of Valsalva' (ACAOS), a rare congenital heart disease that is associated with sudden cardiac death and ischemia.

We present the case of an 80-year-old man presenting with inferior STEMI having a single coronary ostium and a rare variant of the coronary artery origin belonging to type A4d with an interatrial LAD course according to Angelini's classification. This abnormal finding was managed conservatively and the patient underwent successful drug-eluding stent implantation in the culprit right coronary artery in its middle portion.

Keywords: STEMI, ACAOS, CTA, AAOCA

1. INTRODUCTION :

Coronary artery anomalies represent a heterogeneous entity with diverse manifestations and pathophysiological mechanisms. The field of CAA is witnessing a genuine evolution regarding the definition, morphogenesis, clinical presentation, diagnostic workup, prognosis and management of these anomalies [1].

While most CAAs do not produce signs, symptoms, or complications, with incidental discoveries at the time of catheterization, a fraction of them, particularly AAOCAs, may be associated with a risk of sudden cardiac death [2]. They require accurate identification, careful screening for high-risk features, and sometimes surgical correction [3].

In this article, we will present a case of late and incidental discovery of ACAOS after STEMI.

2. PRESENTATION OF THE CASE :

We present the case of an 80-year-old man with no known cardiovascular history other than hypertension (HTN), for which he has been taking ARB2 for 7 years, who does not smoke and has no recorded family history of cardiac disease, especially no syncope or sudden cardiac death (SCD).

He self-presented to our emergency department after 7 hours of chest pain. On admission, the patient presented with significant angina, hemodynamics were stable, there were no symptoms of heart failure, the ECG; revealed an inferior ST-segment elevation. The diagnosis was inferior STEMI, a loading dose (LD) of heparin and double antiplatelet therapy was given.

A TTE was performed immediately and revealed inferior wall hypokinesis with intact systolic function (LVEF 55%). Troponins were overwhelmingly positive.

Given this clinical picture, a coronary angiography with a radial approach was carried out (Figure 1 and 2). The latter revealed a very particular coronary anatomy with a left main coronary artery (LMCA) originating along with the right coronary artery (RCA) from the right aortic sinus of Valsalva, the latter was stenotic in its middle portion, a pre-dilatation and then a drug-eluding stent implantation were performed. This examination was completed by a computerized tomography angiography (figures 3 and 4) which confirmed these data by specifying the proximal course of the left anterior descending (LAD) artery as inter-arterial, and by individualizing a circumflex artery (CX) originating from the LAD. The patient thus has a coronary anatomy of type "single coronary ostium", with a left Anomalous origination of a Coronary Artery from the Opposite Sinus of Valsalva (ACAOS).

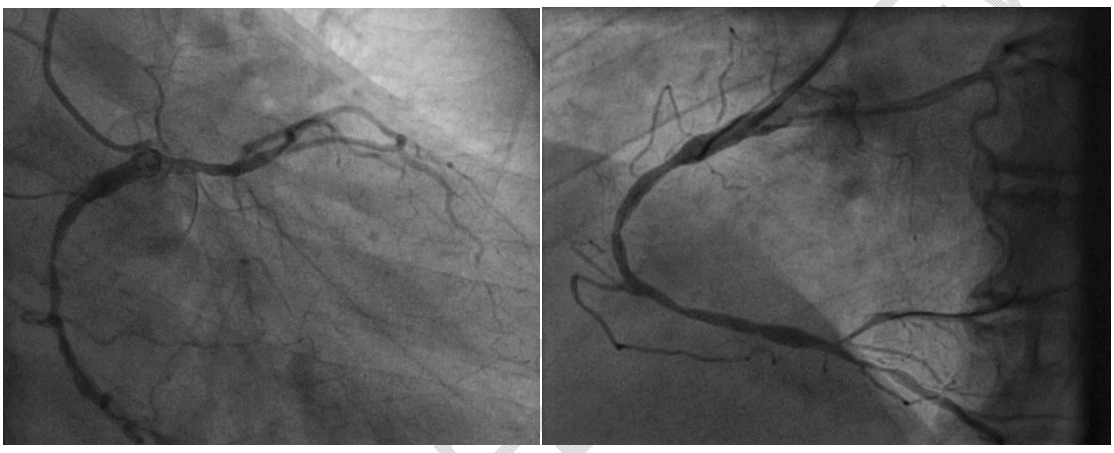
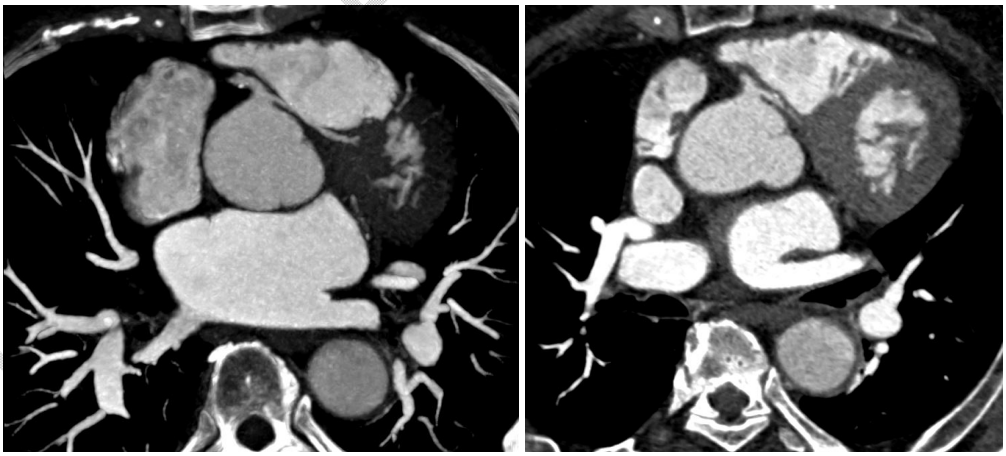


Figure 1 and 2 : coronary angiography showing ACAOS



Figures 3 and 4 : CCTA showing a single ostium and coronary course

3. DISCUSSION

Congenital anomalies of the coronary arteries (CAA) are rare and can be broadly categorized as anomalies in the origin, pathway, destination, size, or number of coronary artery vessels [1], the major challenge in order to comprehend their full clinical implications is to establish a

clear pattern of involvement for a particular type of CAA in compromising the coronary circulation.

Anomalous aortic origin of a coronary artery (AAOCA) is a congenital abnormality in the origin or course of a CA originating in the aorta. Nearly half of patients with AAOCA may present with cardiac symptoms suggestive of myocardial ischemia, while others may be discovered fortuitously following an abnormal ECG or a pre-sports testing [2]. The approach to management is controversial and long-term follow-up information is scarce [2].

The normal features of coronary anatomy in humans were described by Angelini [1] in terms of quantitative and qualitative criteria, and once the normal features have been excluded, the remaining characteristics must be considered to define the anomaly. He proposed a comprehensive classification taking into account all possible coronary anatomical variations independently of clinical and hemodynamic status.

Our patient's CAA can be classified as 'A4d' ACAOS according to this classification [1] meaning: (A) Anomaly of origination and course, (4) Anomalous location of coronary ostium at improper sinus (single coronary pattern), (d) LCA that arises from right anterior sinus, with anomalous course; with an additional LAD course anomaly between aorta and pulmonary artery.

Anomalous origin of the coronary artery from the opposite sinus of Valsalva (ACAOS) is a rather exceptional inborn coronary artery anomaly [1]. The prevalence of ACAOS in the general population is about 1% [3].

ACAOS does not have a classic clinical presentation, and in some cases, SCD or resuscitated/aborted SCD is the initial event. Although some individuals present with cardiac symptoms or arrhythmia, the vast majority are asymptomatic and ACAOS is discovered coincidentally on invasive or non-invasive cardiac imaging performed for other causes, such as ECG abnormalities or heart murmur. Routine screening for ACAOS is not recommended [4].

The conventional definition of ACAOS distinguishes benign and malignant variants. Malignant ACAOS has an interarterial course (IAC). Benign variants account all other pathways, such as the pre-pulmonary or retro-aortic course [5]. It is suggested that individuals with ACAOS and IAC may be in danger of myocardial ischemia and subsequent arrhythmia, even in the absence of atherosclerotic lesions, as the anomalous vessel is prone to dynamic compression upon exertion [6] ; However, the absolute risk of SCD for ACAOS remains unknown[7].

While our patient initially presented with a myocardial infarction, the culprit artery found at coronary angiography had an almost normal anatomy, we cannot then determine if the ACAOS is fortuitous or contributing to the infarction.

As shown in several studies, patients with ARCA (71%) tended to be seen more often than those with ALCA (15%) [2]. But inter-arterial ALCA is rare in frequency compared with inter-arterial ARCA [7].

The allocation to high- and low-risk groups is arbitrary. Patients are considered high-risk (nearly one-half) if they had any symptoms attributable to ischemia, positive functional tests for ischemia, or worrisome anatomy on angiography [2]. In a series of patients with AAOCA followed prospectively [2], statistical analyses were used to define factors predictive of high risk: these were higher age at diagnosis, black race, intramural course, and exercise syncope.

Transthoracic (TTE) is a widely used method to evaluate young individuals with known or suspected heart disease, given that it is non-invasive, efficient, and cost-effective. However, TTE is not very accurate in detecting AAOCA, requiring skilled operators to visualize coronary ostia [7].

Transesophageal echocardiography (TOE) has been used to identify for perioperative visualization of CAA. The addition of 3-dimensional TOE may improve visualization of AAOCAs and their surrounding anatomy. However, it is not yet considered a mainstream diagnostic technique [7].

Advanced imaging is key to identifying the anatomic features of AAOCA, and computed tomography angiography (CTA) is the key imaging modality for assessing high-risk anatomic features, it can accurately delineate the intramural and intramyocardial course and the ostial conformation [2,4]. Currently, coronary CTA and MRA (Magnetic Resonance Angiography) are the only Class I–indicated tests for imaging AAOCA [4].

Invasive coronary angiography (ICA) (Class IIa) [7] has an improved spatial and temporal resolution, with the possibility to use of IVUS to assess the underlying mechanisms responsible for ischemia and the valuation of proximal narrowing [1].

Compared with CTA, MRI provides functional imaging of the coronary arteries without radiation or iodine contrast agents, but has lower spatial resolution, longer scan times, and much higher cost [7].

Studies using non-invasive functional tests, both treadmill exercise testing and stress myocardial perfusion imaging, have reported false-positive and false-negative results [7]. Therefore, the absence of ischemia on functional testing cannot be taken as a guarantee, particularly when high-risk anatomic features are present. Further studies are required to assist risk stratification and to compare the performance of different tests used to detect ischemia in AAOCA [7].

The three steps of an optimal diagnostic process are [8]: 1. initial screening of high-risk populations with preferred reliance on magnetic resonance imaging; 2. assessment of severity (symptoms of chest pain, syncope, or sudden death) and performance of a stress test; 3. in patients identified as having an ACAOS with an intramural course, assessment of the severity of coronary obstruction by intravascular ultrasound (IVUS).

Surgical correction, percutaneous coronary intervention, medical and conservative treatment with or without sports restriction are all management options for people with ACAOS. Because there is no consistent approach to stratify these patients, estimating risk and deciding on treatment is problematic. Symptomatic patients with ACAOS and IAC, with or without ischemia on stress testing, should quit competitive sports and undergo surgical or percutaneous treatment [6].

However, elderly patients with newly detected ACAOS appear to have a lower risk of adverse cardiac events and, in most cases, can be treated conservatively [6]; Management strategies for asymptomatic patients are also less well codified [9].

Fortunately, in our patient's instance, the delayed onset of his myocardial infarction at the age of 80 with a culprit artery other than the anomalous one prompt us to take a cautious approach and treat only the right coronary artery with percutaneous angioplasty.

Three major gaps in knowledge persist; First, the interplay of various anatomic variations and the circumstances that lead to ischemia and sudden death are not clearly understood. Second, the adequacy of the different investigations used to inform decision making remains to be ascertained in this setting. And finally, long-term outcomes, including complications of surgical repair, are barely emerging [9,10].

4. CONCLUSION

As imaging advances, the incidence of anomalous coronary arteries is increasing. Depending on the course, certain variants of coronary anatomy, such as ACAOS, can carry a significantly high risk. Management guidelines for ACAOS remain an evolving area of debate, with a particular focus on the indications for surgical repair, in the elderly patient, especially when asymptomatic, a conservative strategy may be adopted

CONSENT (WHERE EVER APPLICABLE)

All authors declare that written informed consent was obtained from the for publication of this case report and accompanying images.

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