Case study

Proximal coronary artery connection anomaly revealed by acute coronary syndrome: A case report and review of the literature.

Abstract:

Anomalous aortic origin of the coronary artery is rare with an angiographic prevalence of approximately 1%.Its symptomatology is very varied, from incidental discovery to sudden death by myocardial ischemia, especially during exercise, hence the importance of early diagnosis and management.Angiographic diagnosis is generally easy, but it is sometimes difficult to specify the initial course of the ectopic vessel, hence the interest of non-invasive imaging, in particular the Coroscanner. This latter represents the innovative imaging of choice for assessing the anatomical risk and selecting candidates for surgical treatment. We report a case with an anomaly of birth and path of the left main trunk revealed by an acute coronary syndrome and we present a review of the literature on the different anatomical forms of the pathology and its management.

Keywords: Anomalous aortic origin, sudden death, Coronary angiography, Coroscanner.

INTRODUCTION:

-Anomalies of proximal coronary artery connection (ANOCOR) are rare and polymorphic with an angiographic prevalence of approximately 1% . Their clinical manifestations are diverse from incidental discovery to sudden death by myocardial ischemia especially when the abnormal coronary artery arises from the opposite sinus of Valsalva. Thus, the initial course of the ectopic coronary is important to consider, because the prognosis of these proximal connection anomalies depends on the anatomic shape.

The contribution of non-invasive imaging is described for the positive diagnosis of these anatomical variations, sometimes delicate in coronary angiography, but also for the distinction of benign forms and malignant forms potentially responsible for myocardial ischemia. Thus, the difficulties encountered in their management are explained by the numerous anatomical forms described and the lack of data on their follow-up whether treated or not.

In the present article, we report a case of proximal left common trunk connection anomaly in the right anterior sinus revealed at the age of 80 years on the occasion of a high-risk NSTEMI and we present a review of the literature on the different anatomical forms of the pathology by highlighting the management which remains poorly codified.

CASE REPORT:

A 80 years old patient, with cardiovascular risk factors other than age and gender, balanced hypertension under ACE inhibitor for 7 years, dyslipidemia of incidental discovery, not known to be diabetic or a smoker, with no particular personal or family pathological history, in particular no history of angina pain, no notion of sudden death in the family or of coronary heredity. Hospitalized in a cardiology intensive care unit for the management of an acute coronary syndrome without high-risk ST-segment elevation, he presented at H20 with constrictive, intense, prolonged retrosternal chest pain radiating to the left upper limb without any notion of dyspnea, palpitations, loss of

consciousness or other associated signs. The clinical examination shows a patient in good general condition, conscious, with a correct hemodynamic and respiratory state, the blood pressure is 139/85 mmHg, the heart rate (HR) is 86b/min, the SaO2 is 97% on room air. There were no signs of heart failure. The rest of the clinical examination was unremarkable.

His electrocardiogram (ECG) was in sinus rhythm and showed a transient ST-segment elevation in the inferior part without Q wave of necrosis with a mirror image in the high lateral part (Figure 1).

Trans-thoracic echocardiography (TTE), performed in the intensive care unit, showed mild inferoseptal and inferolateral hypokinesia at the basal and medial levels with a left ventricle of preserved size and systolic function (LVEF 55% by Simpson biplane).Troponins came back strongly elevated with positive kinetics without any other detectable anomalies.

Coronary angiography, performed urgently through the right radial approach, showed a coronary connection anomaly with the origin of the left common trunk (CT) from the right anterior sinus (Figure 2) associated with an intermediate stenosis of the middle CT, short tight stenosis of the distal circumflex artery (CX), Tight stenosis of the middle right coronary artery (DC), double intermediate stenosis of the distal DC and tight stenosis of the posterior interventricular artery, who underwent angioplasty of the middle right coronary artery with placement of an active stent after pre-dilation. The patient was put on medical treatment combining double antiplatelet aggregation, beta-blocker, ACE inhibitor, gastric protection and statin.

The Coro scanner confirms the anomaly of connection of the coronary arteries with birth of the anterior interventricular artery (IVA) and the right coronary in the right sinus and the birth of the CX from the IVA with inter-arterial pathway (between the aorta and the pulmonary artery) of the proximal part of the IVA.

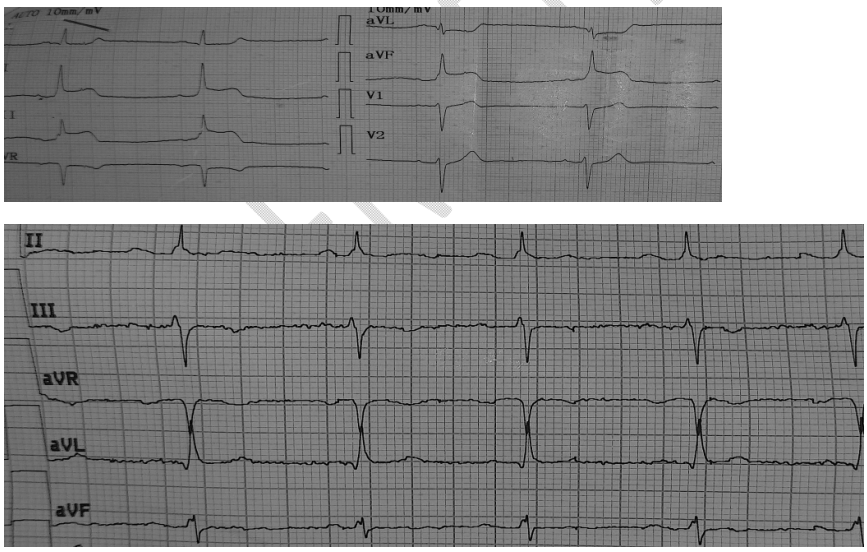

Figure 1: ECG showing a transient ST-segment elevation in the inferior leads without Q wave of necrosis.

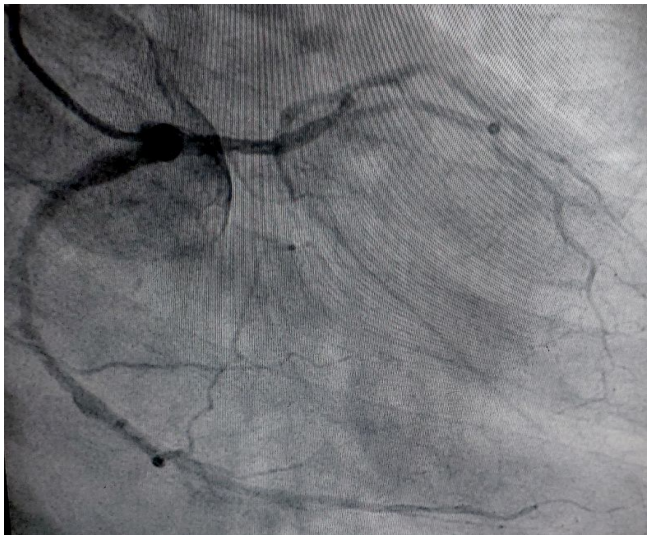

Figure 2: Coronary angiography showing a coronary connection anomaly with the origin of the left common trunk from the right anterior sinus.

DISCUSSION:

Anomalies of proximal coronary artery connection (ANOCOR) are the most common congenital coronary anomalies. Their angiographic prevalence is close to 1% (1). The ectopic connection of the Cx artery is the most concerned (50% of cases) versus 30% for that of the right coronary and 20% for that of the common trunk or the IVA artery. Indeed, the recognition of an ANOCOR requires to define beforehand what is a normal coronary network by including the anatomical variants, in order not to make an excessive diagnosis. The normal number of coronaries, usually two, may increase to three if the VIA and circumflex arise separately at the level of the left sinus. The place of connection may be debatable when it involves the normal sinus but with an unusual location (abnormally low or high or very close to the contralateral sinus). This makes the angiographic diagnosis between an anatomical variant and a connection anomaly difficult.

The main ANOCORs are represented by the connection in the contralateral sinus or artery, the connection in a normal sinus but in an eccentric position, the abnormally high aortic connection and the so-called single coronary. The majority of these patients are asymptomatic with incidental discovery. The symptomatology consists most often of chest pain on exertion, palpitations, syncope on exertion and sudden death which is the most dreadful complication. The revelation of ANOCORS by an acute coronary syndrome remains rare apart from an associated atheromatous disease after the age of 40.

The angiographic diagnosis of this pathology is generally easy, but it is sometimes difficult to specify the initial path of the ectopic vessel. Non-invasive imaging techniques are now essential to determine the relationship of the ectopic vessel with the adjacent structures, mainly the aorta and the pulmonary artery. In our patient, the ANOCOR concerns the ectopic connection of the common trunk in the right anterior sinus, and several paths of the CT are possible (Figure 3), whose relationship with the arterial trunks defines four anatomical types:

- type A with the trunk passing in front of the pulmonary artery (pre-pulmonary path)

- type B with the trunk passing between the aorta and the pulmonary artery (inter-arterial tract)

- type C with the trunk passing in the infundibular septum (intra septal path);

- Type D with the trunk passing behind the aorta (retro aortic path).

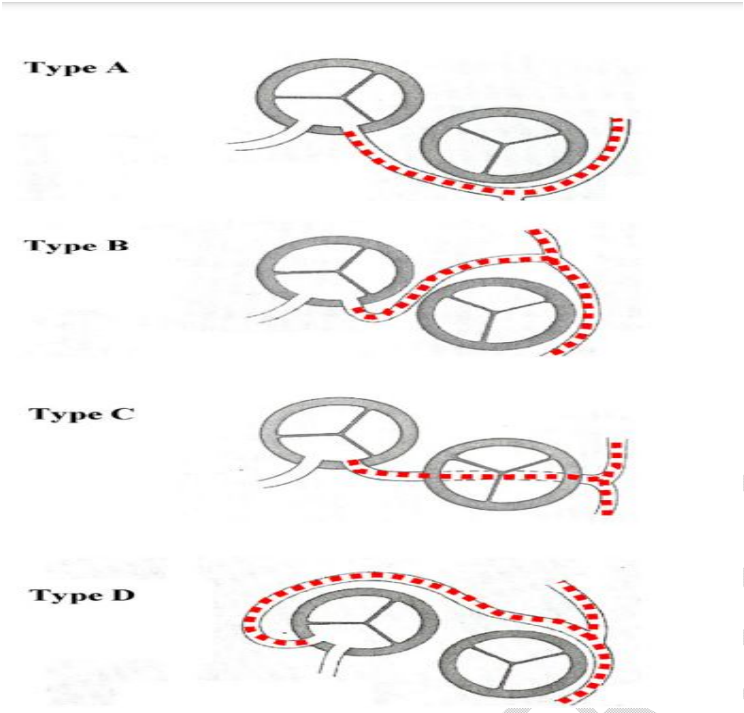

Figure 3: Diagram of the 4 anatomical types of common trunk connection in the right sinus.

The recommendations of the European Society of Cardiology have well defined the anatomical forms at high risk of myocardial ischemia evaluated by coroscanner, which are represented by an intramural pathway, an interarterial pathway, hypoplasia of the proximal coronary artery, orifice abnormalities (cleft deformity, acute angle of departure of the coronary artery, high orifice located > 1 cm above the sinotubular junction). Recommended noninvasive ischemia tests are stress echocardiography, stress cardiac MRI, and stress scintigraphy.

The surgical indications for coronary connection anomalies according to the European Society of Cardiology recommendations published in 2020 are represented in Table 1.

Anomalous aortic origin of the coronary artery		
Surgery is recommended for AAOCA in patients with typical angina symptoms who present with evidence of stress-induced myocardial ischaemia in a matching territory or high-risk anatomy.[c]	I	C
Surgery should be considered in *asymptomatic* patients with AAOCA (right or left) and evidence of myocardial ischaemia.	IIa	C
Surgery should be considered in *asymptomatic* patients with AAOLCA and no evidence of myocardial ischaemia but a high-risk anatomy.[c]	IIa	C
Surgery may be considered for symptomatic patients with AAOCA even if there is no evidence of myocardial ischaemia or high-risk anatomy.[c]	IIb	C
Surgery may be considered for *asymptomatic* patients with AAOLCA without myocardial ischaemia and without high-risk anatomy[c] when they present at young age (<35 years).	IIb	C
Surgery is not recommended for AAORCA in asymptomatic patients without myocardial ischaemia and without high-risk anatomy.[c]	III	C

Table 1: Surgical indications for anomalous coronary connections according to the recommendations of the European Society of Cardiology .

CONCLUSION:

Proximal coronary artery connection anomalies are a rare entity. Their symptomatology is very varied, from incidental discovery to sudden death by myocardial ischemia. According to the recommendations of the European Society of Cardiology published in 2020, the coroscanner is currently the recommended imaging technique for the evaluation of high-risk anatomical forms of myocardial ischemia with the aim of codifying surgical management.

Indeed, the pooling in the form of a national registry of these anomalies with their anatomical, clinical, morphological, and therapeutic particularities could eventually provide some answers to the questions still raised by these congenital coronary anomalies.

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