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Case Report

Rare malignant anomalous right coronary artery incidentally detected by dual source computed tomography angiography in an adult referred for transcatheter aortic valve implantation ☆,☆☆

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ABSTRACT

Transcatheter aortic valve implantation (TAVI) is a safe treatment option for elderly patients with aortic stenosis (AS) with high or intermediate surgical risk. The anomalous origin of coronaries from a single Valsalva sinus has a higher risk of coronary obstruction during the TAVI procedure, so prior knowledge of these anatomical variants is of paramount importance as it can change the surgical strategy. In this report, we present the case of an adult patient suffering from severe aortic stenosis and incidentally diagnosed with an anomalous malignant variant of right coronary artery (RCA) by pre-procedural Computed Tomography (CT) Angiography. In TAVI planning, the use of electrocardiography (ECG) gated Dual Source CT (DSCT) enables accurate coronary and aorto-iliac vascular study, useful for detecting congenital coronary anomalies.

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Introduction

TAVI (Transcatheter Aortic Valve Implantation) represents a valid alternative to open heart surgery in elderly patients with high or intermediate surgical risk. Computed tomography (CT) is part of the routine TAVI planning as it is useful for determining the size of the aortic root, vascular access sites, and the possible presence of coronary anomalies. Preliminary study

of coronary anomalies is of primary importance to prevent major complications related to the TAVI procedure, such as coronary obstruction and sudden death. The recent advent of ECG-gated DSCT angiography provide accurate images of the coronary tree anatomy, detecting anomalous coronary artery's course with a single CT angiography study [1]. In the present case, we describe a rare case of a 75-year-old male patient with severe AS and abnormal origin of RCA from the left coronary Valsalva sinus, with an inter-arterial course passing between the aorta and the main pulmonary artery.

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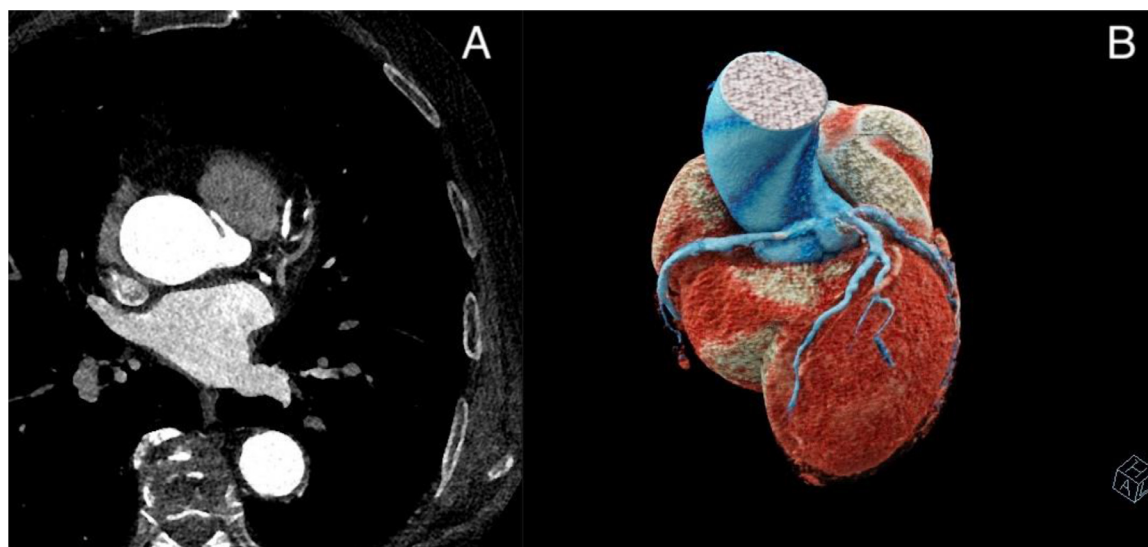


Fig. 1 – Computed Tomography (CT). (A) Axial CT image showing right coronary artery (RCA) originating from left coronary sinus. RCA has a inter-arterial course between aorta (A) and pulmonary artery (PA). (B) Volume-rendered (VR) image of the heart demonstrates both the right and left coronary arteries originating from left coronary sinus

Case report

A 75 year-old male presented with severe AS, suffering from one-year history of exercise-related angina and palpitations with New York Heart Association (NYHA) class III dyspnea. His past medical history revealed hypertension and dyslipidemia both under pharmacological control. The patient did not have experienced syncope and did not have a family history of sudden death. Chest radiograph showed no abnormal findings. An electrocardiogram showed a normal sinus rhythm with non-specific inverted T-wave in the right precordial leads. Transthoracic Echocardiography (TE) showed a calcified aortic valve (area of 0.70 cm²; mean gradient of 52 mmHg) with a preserved left ventricular systolic function (ejection fraction of 58%). The patient was referred to the radiology department to undergo DSCT angiography prior to TAVI procedure. CT angiography was performed using a 384 (192 × 2)-slices third generation DSCT scanner (SOMATOM Force CT; Siemens Healthineers, Forchheim, Germany). Scan protocol consisted in a single prospective ECG-gated ultra-high pitch acquisition. The following parameters were used: 120 kVp, 800 mAs, rotation time: 0.22 s, pitch: 3.2 mm. To optimize timing of scanning a bolus tracking protocol was used, acquisition started when region of interest (ROI) value at the aortic diaphragmatic level reached 240 Hounsfield Unit (HU). Coronary arteries images were acquired at 30 % of R-peak-to-R-peak. Using a dual head pressure injector (EmpowerCTA+ Injector System; Bracco Injengineering, Lausanne, Switzerland) was administered 60 ml of contrast medium (Iomeprol, 400 mgI/ml; Bracco Imaging, Milan, Italy) at flow rate of 5 ml/s followed by 50 ml of saline solution at the same rate. Two reconstructions were performed: a small (200 mm) and large (300 mm) field of view data set for the assessment of the aortic root and coronary tree (0.6 mm slice thickness at 0.4 mm increment) and the ilio-femoral accesses (1.0 mm slice thickness at 0.7 mm increment), respectively.

CT Coronary angiography showed anomalous origin of the right coronary artery (RCA) from the left coronary sinus of Valsalva with an ostium adjacent to that of the left coronary artery (LCA). It was associated with proximal malignant inter-arterial course of the right coronary artery between aorta and main pulmonary artery. The other coronary arteries, including the left main LCA, the left anterior descending (LAD) and the left circumflex (CX) were normal in course with diffused wall calcifications (Fig.1). The patient was subsequently subjected to invasive coronary angiography (ICA) which confirmed the rare congenital anomaly of RCA (Fig.2).

Subsequently, the patient did not undergo the operation because he suffered from stroke during the pre-operative hospitalization. The case was subsequently re-evaluated by a multidisciplinary team and it was decided to postpone the TAVI due to the patient's physical decay.

Discussion

Congenital coronary anomalies are rare conditions that may be found incidentally with an incidence ranging from 0.17 % to 1.2 % of the cases [2]. The clinical presentation in patients with congenital anomalies of coronary arteries includes angina, dyspnea, syncope and sudden cardiac death that is the first and fatal manifestation in more severe cases.

In this study, we reported a case of an aberrant origin of the RCA from the left Valsalva sinus, a rare condition that has an incidence of less than 0.20% in patients undergoing angiography [3].

The anomalous RCA origin may have inter-arterial, retroaortic, prepulmonic or septal course. The inter-arterial course is more frequent and occurs when the coronary artery passes between the ascending aorta and the main pulmonary artery. This course has been defined as malignant because it

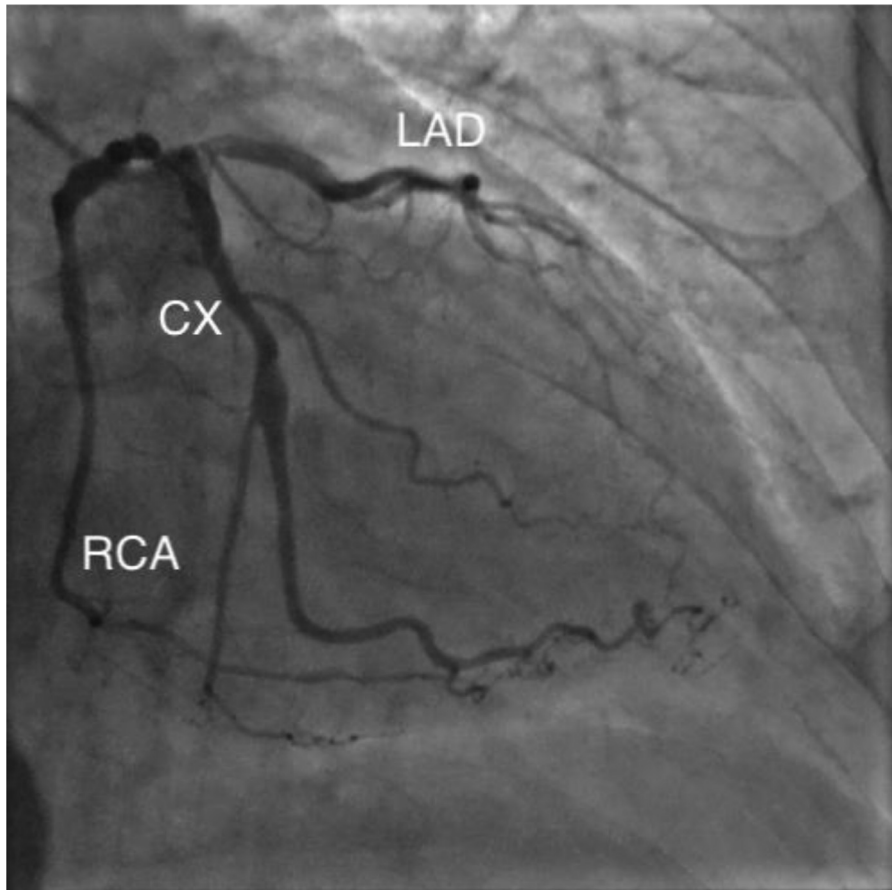


Fig. 2 – Coronary Angiography of the patient showing Right Coronary Artery originating from left sinus of Valsalva. RCA, right coronary artery; CX, circumflex artery; LAD, left anterior descending artery

is associated with a more severe prognosis and an increased risk of sudden cardiac death, the incidence of which is estimated at 25-40% and it is associated with exercise in half of the reported cases [4]. Although some of the symptomatic RCA anomalies are diagnosed in childhood, in other cases the diagnosis is absent due to the asymptomatic state. In these cases the diagnosis may be delayed in adulthood and be completely incidental, as in the case we have described in the present study. However, even if asymptomatic, patients with inter-arterial course of RCA are at risk of sudden cardiac death. For this reason, surgical correction is strongly recommended for all patients, even if asymptomatic [5].

As previously reported, the coronary CT study should be considered as a prime non-invasive imaging tool for suspected coronary anomalies, as compared to ICA, it demonstrates greater sensitivity and specificity and eliminates all the complications related to an invasive procedure [6].

Especially in TAVI planning, DSCT allows, with a single scan, an accurate vascular study of the entire aorta, iliac district and coronary arteries thanks to the large volume coverage and ultra-high pitch factor [1].

Knowledge of the coronary anatomy and their variants is critical, especially because several complications have been reported in TAVI procedure. During and after TAVI, coronary obstruction is a rare and severe complication that was re-

ported to occur in 1% of the cases [7]. The possible cause of coronary artery obstruction is supposed to be the compression of the ostium due to calcified native valve leaflets. In this regard, patients who have anomalous origin of RCA from the left sinus of Valsalva, own an increased risk of a potentially fatal outcome during TAVI. In particular, any obstruction of this single ostium, secondary to stenosis or embolism, could be critical for myocardial perfusion during the deployment of the new aortic valve. During the procedure, acute coronary obstruction may manifest with severe hypotension and with potential sudden cardiac death. The anatomical risk factor to consider to prevent coronary obstruction are low origin of coronary arteries (below 12 mm in height), a shallow sinus of Valsalva (below 30 mm in width), valve misplacement, bulky calcification, coronary embolism, or coronary artery disease itself [7]. Therefore, to predict the risk of coronary obstruction, an accurate CT study is essential for delineate the anatomy of the aortic root. In patients with single coronary ostium, various targeted perioperative strategies are adopted to control and minimize the risk of coronary obstruction [8].

Cases of patients with single coronary artery undergoing TAVI have been previously described, in these cases the operators performed angiography during balloon valvuloplasty to precede the risk of coronary obstruction, or alternatively they placed a coronary wire in the single coronary artery to serve

as protection during the balloon valvuloplasty procedure and bio-prosthesis implant [9,10]. In addition, to reduce the risk of obstruction, the use of self-expanding valves has been proposed, which can be recaptured up to 2/3 of the bio-prosthesis implant: this could have an advantage over preventing coronary obstruction, compared to the balloon expandable valve [11]. However, as there have only been rare cases of coronary anomaly in TAVI, there is currently no evidence or consensus on the choice of prosthesis. Further studies and evidence from further cases are needed to clarify this issue.

In conclusion, we describe a 75-year-old patient referred for TAVI, who was accidentally diagnosed with abnormal RCA originating from the left sinus of Valsalva and with a malignant inter-arterial course. This case highlights the essential role of CT in TAVI planning. That is, making a correct evaluation of the vascular anatomy of the pre-TAVI patient is crucial, as this directly influences the surgical decision for endovascular treatment and allows the adoption of the best operative strategy in reference to anatomy of the patient. In particular, the recent advent of ECG-gating DSCT allows an accurate non-invasive characterization both aorto-iliac anatomy and the entire coronary tree with a single scan. DSCT, in the pre-TAVI evaluation, detect effectively congenital coronary anomalies and rare anomalous coronary artery's course.

Human rights statements and informed consent

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1964 and its later amendments. Informed consent was obtained from all patients for being included in the study.

Animal Rights

This article does not contain any studies with animal subjects performed by the any of the authors.

Patient Consent

All involved persons (subjects or legally authorized representative) gave their informed consent (written or verbal, as appropriate) prior to study inclusion.

All study participants, or their legal guardian, provided informed written consent prior to study enrollment.

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