

The RAC, bleb, and crossed aorta signs: retroaortic anomalous coronary artery visualization by transthoracic echocardiography

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SOUHRN

Transtorakální echokardiografie představuje užitečný nástroj pro screening vrozených anomálií koronárních tepen. Jednoznačně vyvrátila názor, že anatomii koronárních tepen nelze zkoumat neinvazivně. Uznávají se tři echokardiografické znaky související s anomáliemi levé koronární tepny. Tyto znaky jsou: znak retroaortálního r. circumflexus (retroaortic circumflex, RAC), znak bublinky (bleb sign) a znak překřížené aorty (crossed aorta sign). Znak RAC je vysoce specifický a na popisu echokardiogramu jednoznačně ukazuje na anomální koronární tepnu. Je však naprosto nezbytné ho odlišit od podobně vypadajících struktur. Proto se doporučuje jako primární metody pro ověření známých anomálií koronárních tepen nebo podezření na ně použít koronární CT angiografii.

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ABSTRACT

Transthoracic echocardiography is a useful screening tool for identifying congenital coronary artery (CA) anomalies. It has effectively dispelled the notion that CA anatomy cannot be identified noninvasively. There are three recognized echocardiographic signs associated with anomalous left coronary artery. These signs are the retroaortic circumflex (RAC) sign, the blew sign, and the crossed aorta sign. The RAC sign is highly specific and should strongly indicate an anomalous coronary artery on echocardiogram reports. However, it is crucial to distinguish it from similar-looking structures. Therefore, coronary computed tomography angiography is recommended as the primary method for assessing known or suspected coronary anomalies.

Introduction

Identification of congenital coronary artery (CA) anomalies using transthoracic echocardiography (TTE) as a screening tool has become routine and has helped dissipate the notion that CA anatomy cannot be identified noninvasively. In the last years, coronary computed tomography angiography (CCTA) has replaced cardiac catheterization as the primary tool for the characterization of most congenital CA anomalies.^{1,2}

TTE is a highly recommended modality in clinical practice due to its versatility, safety, cost-effectiveness, and non-invasive nature. It can also be performed easily in any clinical setting, making it a widely available option for many patients.^{3,4} Three signs are recognized echocardiographic features well linked with anomalous left coronary artery, specifically associated with left circumflex (Lcx) artery abnormalities. These signs are: 1) The retro-aortic course of the Lcx (RAC) sign, described as a “tubular shape”³ in TTE, 2) the “bleb sign” described by Wierzbowska–Drabik et

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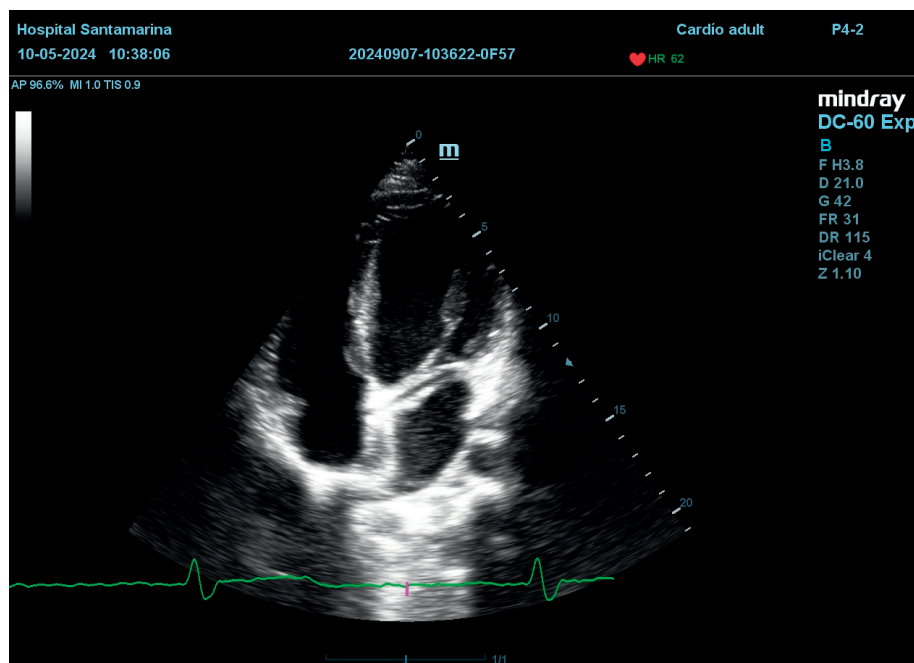


Fig. 1 – Apical four chambers view showed the retroaortic circumflex (RAC) sign.

al. by transesophageal echocardiography (TEE) studies, as a round structure in the mitro-aortic angle in the aortic long-axis view,⁵ and 3) the 'crossed aorta sign', a binary structure that appears to cross the aorta perpendicular to its longitudinal axis in an apical five-chamber view.^{6,7}

Case presentation

An asymptomatic 16-year-old male underwent a routine cardiac evaluation. RAC sign was incidentally detected by TTE. In the apical 4 chambers (A4C) view, tilting anterior demonstrated the RAC sign, a highly echogenic tubular structure in the retro-aortic region above the mitral valve (Fig. 1, and Video 1). Additionally, in the parasternal long-axis view, a round structure next to the aorto-mitral curtain, known as the bleb sign, was also identified (Video 2). The RAC sign was also evident in the short-axis (SAX) as a tunnel-shaped structure located behind the aortic root. This structure is also an orthogonal view of the "bleb sign". An anomalous LCx was confirmed with CCTA (Figs 2, and 3).

Discussion

Anomalous origin and course of the LCx from the right sinus of Valsalva is the most frequent anomaly (0.39%).⁸ Traditionally, coronary angiography has been required for diagnosis; however, these tests are expensive and invasive. Currently, CCTA is considered superior to conventional angiography in diagnosing and classifying coronary anomalies^{9,10} since it provides useful information regarding the anatomy and course of anomalous coronary arteries.

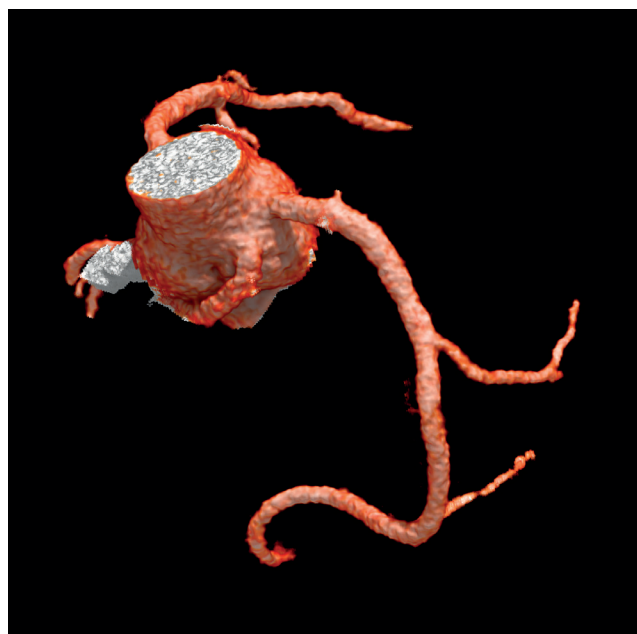


Fig. 2 – Coronary computed tomography angiography (CCTA). Three-dimensional reconstruction showing the origin of the coronary arteries.

In 2018 Witt et al. reported that RAC sign was seen in 63.3% of patients with confirmed coronary anomalies and in 6.1% of patients with normal coronary anatomy.³ They also reported that the RAC sign had a sensitivity of 63.3% and a specificity of 93.9%. Additionally, these authors demonstrated a significant association between the RAC sign and the presence of a retro-aortic anomalous coronary artery ($p < 0.001$).³ When RAC and bleb

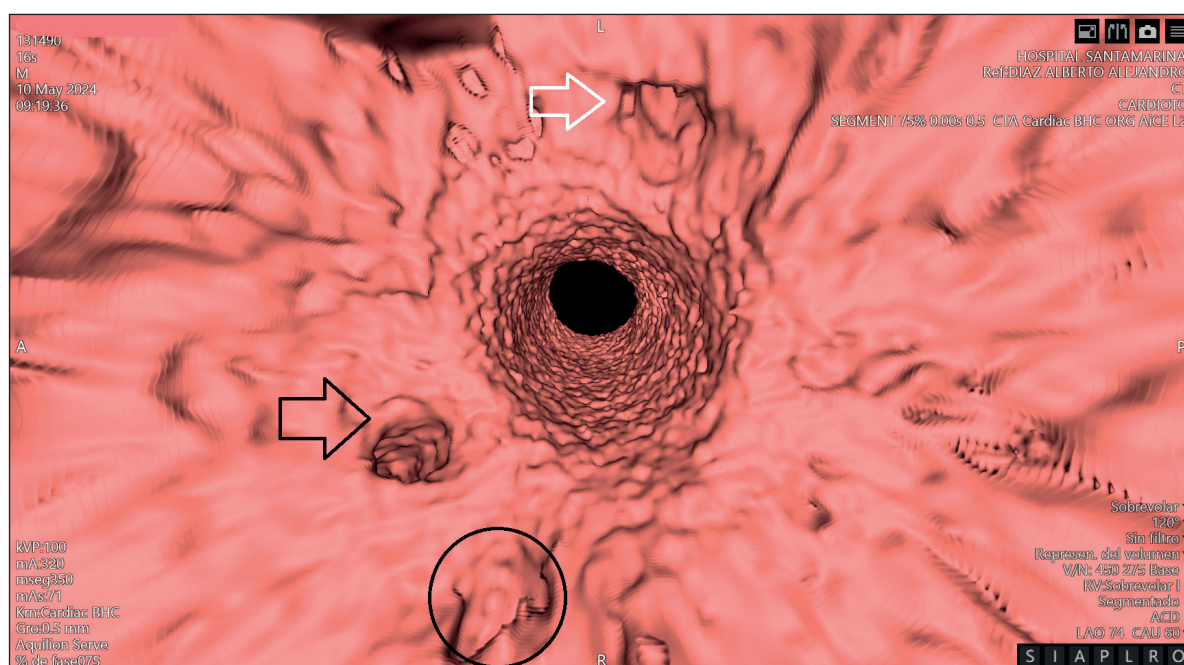


Fig. 3 – Virtual coronary endoscopy created by computed tomography angiography. Visualization of right coronary ostium (white arrow) left coronary ostium (black arrow), and independent circumflex ostium (black circle).

signs are present in an echocardiographic routine study, it's important to consider other more common findings as possible differential diagnoses. In the A4C view, the RAC sign may be mistaken for the coronary sinus, which is typically located further back. Other possible causes of confusion include catheters, coronary calcifications, and calcifications of the aortic valve or mitral annulus. In addition, the bleb sign can be differentiated from an abscess of the mitral-aortic fibrosa by observing the retro-aortic tubular appearance of the structure in the SAX view.^{3,4,6,7} The use of CCTA is a reliable noninvasive tool for defining anomalous coronary arteries and provides detailed three-dimensional anatomic information that may be difficult to obtain with invasive coronary angiography.⁹

Conclusion

The RAC, bleb, and crossed aorta signs offer a noninvasive means of identifying an anomalous LCx. The RAC sign also has a high specificity, signifying that its presence could be documented as strongly suggestive of an anomalous coronary artery on echocardiogram reports. This case emphasizes the significance of maintaining a high level of suspicion of coronary anomalies when RAC and/or bleb signs are present in routine echocardiographic studies. The usefulness of these signs should not be overlooked or misinterpreted, as being aware of their significance can prevent confusion. CCTA has been recommended as the first-line method for the assessment of known or suspected coronary anomalies.

Conflict of interest

The authors declare that they have no conflicts of interest.

Funding

None.

Ethical statement

All ethical matters have been fully considered to protect the rights of the patient. This case report was exempted from approval by the Institutional Ethics Committee.

Informed consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

Credit authorship contribution statement

Diaz Alejandro: conceptualization, writing original draft, review & editing. Victoria Vannoni and Sofia Inclán: conceptualization, writing original draft.

Jose Guillen and Daiana Cattaneo: methodology, writing, review & editing.

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