Ectopic connection of left coronary artery from right coronary sinus, is the treatment always surgical?

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SOUHRN

Kontext: S anomálním odstupem kmene levé koronární tepny z protilehlého sinu (anomalous left coronary artery aortic origin from the right sinus, ALCA) se lze setkat vzácně. Anomální odstup koronárních tepen (anomalous origin of coronary arteries, AOCA) představuje druhou nejčastější příčinu náhlé srdeční smrti mladých sportovců v USA. Léčba těchto anomálií je předmětem sporu a dosud nebyla vypracována doporučení pro stratifikaci rizika.
Kazuistika: Pětačtyřicetiletý muž bez kardiovaskulárních rizikových faktorů byl přijat do naší nemocnice pro svíravou, silnou bolest za hrudní kostí během intenzivní fyzické aktivity. Jeho vitální známky byly normální. Výsledek EKG vyšetření byl normální a laboratorní vyšetření odhalilo vysoké hodnoty troponinů. Echokardiogram prokázal nedilatovanou levou komoru srdeční se zachovanou ejekční frakcí levé komory. Koronarografie zjistila abnormální odstup levé věnčité tepny z pravého koronárního sinu bez významné stenózy a vyšetření koronárních tepen výpočetní tomografií prokázalo preaortální počáteční průběh levého kmene věnčité tepny. Zátěžové scintigrafické vyšetření myokardu neprokázalo reziduální ischemii. U pacienta byla zahájena nefarmakologická léčba formou úpravy životosprávy a pravidelného monitorování srdeční funkce. Při kontrole po jednom roce byl pacient naprosto bez symptomů.
Závěr: S anomálním odstupem kmene levé koronární tepny z protilehlého sinu se lze setkat velice zřídka, vzhledem k vyššímu riziku srdečních příhod je však nutno mít tuto možnost na paměti. K přesnějšímu zjištění spojení a počátečního průběhu lze použít zobrazovací techniky. Rozhodování o tom, zda usilovat či neusilovat o korekci ALCA, je i nadále obtížné. Naše případy byly řešeny odlišně, přičemž úspěšné dlouhodobé sledování prokázalo, že léčbu je nutno individualizovat a ideálně posoudit poměr přínosu a rizik samostatně pro každého jednotlivého pacienta.

ABSTRACT

Background: Anomalous left coronary artery aortic origin from the right sinus (ALCA) is rare. Anomalous origin of coronary arteries (AOCA) is the second leading cause of cardiac death among young athletes in the US. Their management remains controversial and there are still no pre-established recommendations for risk stratification.
Case presentation: A 45-year-old male with no cardiovascular risk factors, was admitted to our hospital for constrictive, intense, retrosternal chest pain during intense activities. His vital signs were normal. ECG was normal and biology revealed high troponin levels.
Echocardiogram revealed a non-dilated left ventricle with a preserved left ventricle ejection fraction. Coronary artery angiography showed an abnormal left coronary artery origin from the right coronary sinus with no significant coronary stenosis and coronary computed tomography described a pre-aortic initial course of the left main artery.
Stress myocardial scintigraphy did not show residual ischemia. The patient was medically treated with changes in his lifestyle and regular monitoring. A one-year follow-up showed a perfectly asymptomatic patient.
Conclusion: ALCA is very rare but must be particularly considered given the higher risk of cardiac events. It is possible to use imaging techniques to better analyze the connection and the initial course. The decision to correct surgically or not to correct an ALCA remains difficult. Our cases were treated differently, with satisfactory long-term follow-up showing that the treatment should be individualized with a benefit/risk ratio assessed preferentially for each patient separately.

Keywords:
Chest pain
Congenital heart disease
Coronary anomaly
Sudden death

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Background

Anomalous aortic origin of the coronary arteries (AOCA) are still a challenge in the field of cardiology. Often accidentally discovered, their incidence is estimated at 1.2% of the general population.[1,2] This pathology grows in view of the risk of sudden death associated mainly with left coronary artery originating from the right sinus.

Despite decades of research, we still have a limited understanding of the mechanisms of sudden death and there is incomplete data on risk stratification and management.

Case presentation

A 45-year-old male, was admitted to our hospital for chest pain. His history noted no diabetes, no hypertension and no active smoking.

The patient described the pain as constrictive, intense, retrosternal chest pain during intense activities during 3 to 5 minutes and disappearing at rest.

His vital signs at presentation were as follows: blood pressure at 130/65 mmHg, heart rate at 90 b.p.m. and respiratory rate of 18 cycles/min. Lung sounds were normal; and there were no signs of heart failure.

ECG was normal and biology revealed high troponin levels at 0.1 ng/ml. The rest of the results were normal.

The patient was therefore hospitalized.

Echocardiogram revealed a non-dilated left ventricle (LV) with a preserved left ventricular ejection fraction (LVEF) at 55%. Left coronary artery birth anomaly was also seen (Fig.1).

Coronary artery angiography showed an abnormal left coronary artery departure from the right coronary sinus with no significant coronary stenosis (Fig. 2).

A coronary computed tomography scan (Figs 3–5) confirmed the diagnosis and described a pre-aortic initial course of the left main artery.

Stress myocardial scintigraphy did not show residual ischemia. The patient was put on aspirin, beta-blockers and isosorbide dinitrate with changes in his lifestyle and regular monitoring.

A one year follow-up showed a perfectly asymptomatic patient.

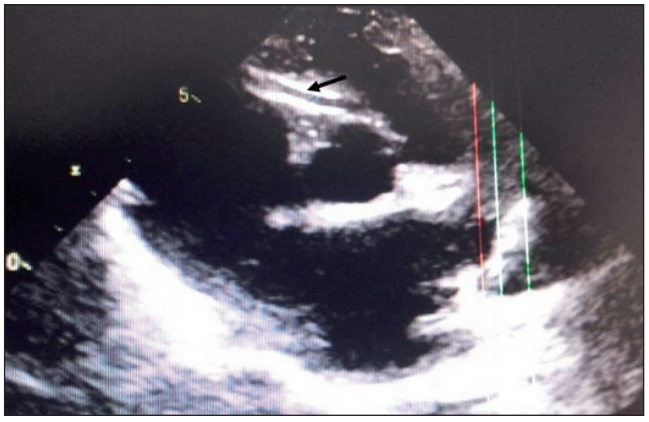

Fig. 1 – TTE showing a left coronary artery of ectopic birth.

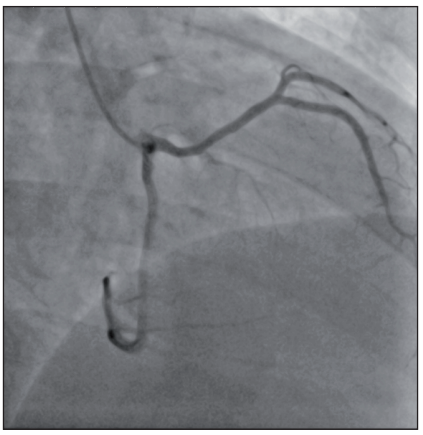

Fig. 2 – Angiographic image of the left coronary artery arising from the right sinus.

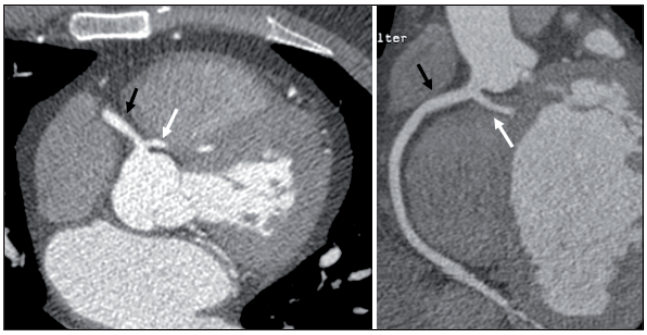

Fig. 3 – Coronary computed tomography: Axial and coronary tomographic images of an ectopic connection of the left coronary artery (white arrow) in the anterolateral sinus, close to the birth of the right coronary artery (black arrow).

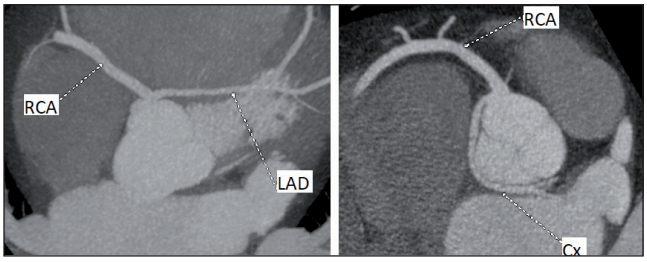

Fig. 4 – Coronary computed tomography: Axial images of an ectopic connection of the left coronary artery in the anterolateral sinus, close to the birth of the right coronary artery. Cx – circumflex artery; LAD – left descending artery; RCA – right coronary artery,

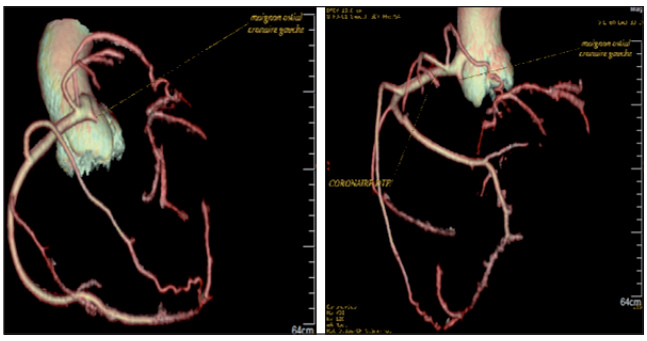

Fig. 5 – Coronary computed tomography: 3D images showing the ostial stump of the left coronary artery from the right coronary sinus and its pre-aortic initial course.

Discussion

Although incidence and natural history of anomalous aortic origin of the coronary arteries (AOCA) are difficult to determine, it is estimated to range from 0.6 to 1.2% of the general population according to large angiographic studies.[1,2] In a Tunisian study, we found it at 0.27%.[3] But these series are likely to underestimate the true incidence as many people never undergo coronary angiography. The abnormal origin of the left coronary artery from the right sinus (ALCA) is quite rare (with an estimated incidence of 0.03–0.05%),[4–12] it is the second most common cause of cardiac death among young athletes in the United States.[13]

Basso et al.[14] examined 2 large registries in the United States and Italy reporting sudden cardiovascular death in young competitive athletes over a period of 10 to 20 years. Twenty-seven deaths were recorded in patients who had no symptoms, 14 patients had ALCA. It is mostly the anomalous aortic origin of the left coronary artery that is often associated with early sudden death, especially during vigorous exercise.

The symptoms are usually absent in everyday life. The most common mode of discovery remains a "recovered" sudden death, occurring while or immediately after a very intense physical effort. Sometimes warning signs prior to the acute episode exist such as: chest pain, syncope, or cardiac arrhythmia during exercise. Increasingly, the anomaly is detected during an echocardiographic assessment performed for another cardiovascular reason.[15]

Sudden death can result from one or more mechanisms that compromise coronary blood flow. Several episodes of ischemia are probably necessary creating a substrate for the catastrophic event.[14] Although there is no clear consensus on the mechanism of ischemia and the risk factors leading to sudden death in these patients.[16] It has been suggested that this risk is increased in young patients (< 35 years) with an abnormal left coronary artery origin, after intense physical activity, and in the presence of some anatomical features summarized in: an inter-aortopulmonary course, an intra-mural aortic course, a slit-like ostium and an acute angle of the coronary artery departure from the aorta.[17,18] A number of studies have been conducted attempting to isolate a key factor, but no single component of coronary artery anatomy has been identified as the definitive cause of ischemia. A slit-like ostium and a narrow intramural course might be the most involved, but absolute proof has not yet been advanced.[19]

Atherosclerotic disease has not been described in these abnormalities.[1] In the absence of revealing symptoms, the diagnosis can be made on a variety of imaging modalities, including echocardiography, computed tomography (CT), magnetic resonance imaging (MRI) and cardiac catheterization, but a protocol of standard imaging has not been yet decided.[2,19]

Echocardiographic examination supports diagnosis, especially in children or adolescents. Technological advances in transthoracic echocardiography (TTE) now allow vision of coronary artery anatomy in many patients. The identification of the abnormal origin of the left coronary artery has been described using TTE.[20–25] TTE in two-dimensional mode must often be combined with color Doppler to successfully identify the abnormal origin of a coronary artery.[20] In adults, the diagnosis may be more difficult and any suspicion must be confirmed by an imaging technique (coronary angiography, CT scan or MRI). In asymptomatic patients, additional examinations are necessary to reveal an eventual on exertion ischemia (stress echocardiography or myocardial scintigraphy).[15]

The management of AOCA remains controversial. Although patients with symptoms of ischemia or arrhythmia are candidates for urgent management, decisions about asymptomatic patients are less well defined and there is still no pre-established consensus for risk stratification.[26] Noninvasive functional imaging is commonly used to stratify risk and emerging data from invasive cardiac catheterization with intravascular ultrasound (IVUS) may be useful in a selected group of patients.[27]

The North American guidelines recommend surgical revascularization of all ALCAs with an inter aortopulmonary course, regardless of the existence of myocardial ischemia and age.[28]

The top professional athletes must stop their activities and completely change their life goals. Other patients must change their lifestyle and accept a constant risk of sudden death from physical exertion. This can be difficult to bear in patients especially in teenagers and young adults. Behavior and psychosocial development can be severely impaired.

Our patient was medically treated, with satisfactory long-term follow-up, this shows that the treatment should be individualized with a benefit/risk ratio assessed preferentially for each patient apart.

Conclusion

AAOCA is a rare anomaly but is gaining more and more importance in current cardiac practice given the high risk and difficulty in assessing sudden death in young and athletic patients. Despite decades of research, we still have a limited understanding of the mechanisms of sudden death and there is incomplete data on risk stratification. The ideal treatment does not yet exist and a case by case management is necessary.

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Conflicts of interest
The authors declare that they have no competing interests.

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Ethical statement and consent to participate
Not applicable.

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

Availability of data and materials

Available upon request.

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