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LETTER TO THE EDITORS

Malignant anomalous origin of circumflex artery in an elite athlete

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Sirs,

Coronary artery anomalies are rare congenital conditions with a prevalence of 0.3–1.3% of patients [1]. Depending on the affected vessel and the course of the vessel, they can be coincidental findings with no symptoms, or, in a worst-case scenario, they can lead to cardiac ischemia. In this case, these coronary anomalies are considered to be malignant, as they constitute a high risk for sudden cardiac death [1–5]. Although such malignant coronary anomalies are rare [1–3], they are a major cause of sudden cardiac death (SCD) in young athletes during exercise [2].

Individuals with coronary artery anomalies often have no severe symptoms, and sudden cardiac death constitutes the first and only manifestation in athletes [3, 4]. Therefore, the detection of coronary artery anomalies in pre-participation screening of athletes is very difficult [3–5]. Competitive sports are prohibited if coronary anomalies are detected that could lead to myocardial ischemia [3, 4]. Thus far, rare coronary anomalies that do not cause ischemia are considered to be a benign finding that does not necessitate exclusion from competition [3, 4]. However, in cases of very rare, high-risk coronary anomalies, as discussed in a few case reports in the literature, the decision to forbid or allow competitive sports is difficult.

We describe the case of a 30-year-old professional athlete with a history of 12 years in professional sports. The results of his medical examinations throughout the years were normal. After changing his sports club, he was examined in our clinic for the first time in a regular pre-season medical assessment. The physical examination did not show any abnormalities. ECG, blood pressure measurement at rest, and blood tests were unremarkable. Stress testing revealed a high maximum power output (5 W/kg) without any clinical symptoms or changes in the electrocardiogram. However, echocardiography revealed hypokinesia of the inferior and inferolateral wall sections (Fig. 1a), which has not been previously described. Though, global left ventricular ejection fraction was measured normal due to an excellent contractility of the other wall sections.

For further investigation, cardiac magnetic resonance imaging (MRI) was performed. It confirmed the echocardiographic findings and additionally a late gadolinium enhancement in the hypokinetic wall sections with a subendocardial, non-transmural scar was observed (Fig. 1b). No other pathologies were detected.

We decided to perform coronary computed tomographic angiography (CTA). The right coronary (RCA) and the left anterior descending coronary (LAD) arteries appeared to be normal, and coronary artery disease was excluded (Fig. 2a). However, the left circumflex artery had no connection to the left main artery or the LAD: its origin was a convolution of vessels arising from small branches of bronchial arteries (Fig. 2b). The course of the left circumflex artery was normal, leading to the inferior and inferolateral wall sections in which the late gadolinium enhancement and the non-transmural scar were observed in the MRI. Additional cardiac stress MRI with dobutamine revealed the existence of ischemia beyond these wall sections.

Therefore, we diagnosed a malignant coronary anomaly with persistent exercise-induced myocardial ischemia and recommended exclusion from participation in competitive sports. The detected unique coronary artery anomaly constitutes an increased risk of sudden cardiac death during exercise. Furthermore, after heart team consultation, a surgical approach with anatomic correction of the origin of circumflex artery was recommended.

To the best of our knowledge, we are the first to describe this specific coronary artery anomaly with the circumflex artery originating from a convolution of vessels arising from small branches of bronchial arteries. In the completely asymptomatic athlete, it was detected in a pre-participation screening program and, as a consequence, participation in competitive sports was not recommended.

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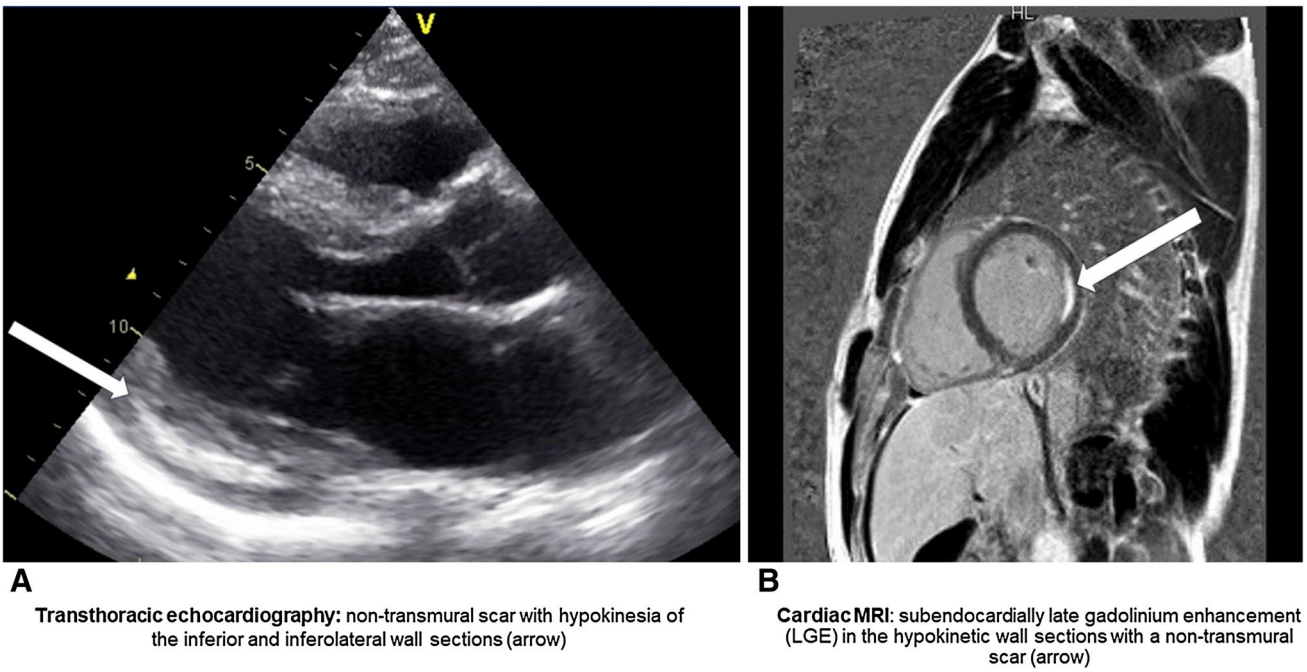

Fig. 1 Cardiac imaging studies, consisting of transthoracic echocardiography (**a**) and cardiac MRI (**b**)

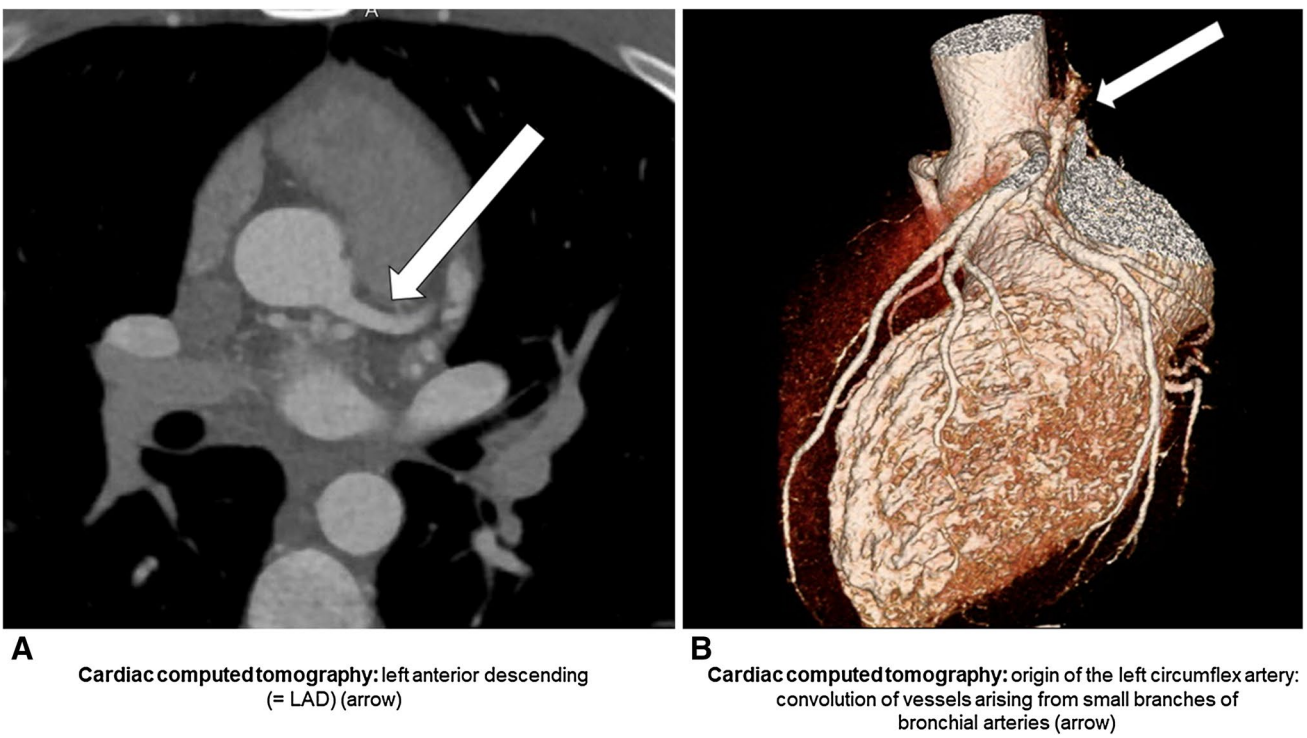

Fig. 2 Cardiac computed tomography, showing the coronary anomaly

The risk of SCD is 2–3 fold higher during intensive exercise, which is called the "sports paradox" [3]. Its incidence in athletes, however, is low at 1–3 per 100,000 [3]. In young athletes (< 35 years), congenital cardiac diseases such as hypertrophic cardiomyopathies and coronary artery anomalies are the major causes of SCD [3–5]. In contrast, SCD

in older athletes (≥35 years) is mainly caused by coronary artery disease (CAD) [3].

In Germany, most competitive athletes undergo regular medical testing and this pre-competition screening is recommended in Europe [6]. It has been proven to decrease the incidence of SCD in athletes [6]. The routine pre-competition screening consists of a medical history, a physical examination, and ECG screening [6]. These tests significantly improved the detection of potentially life-threatening cardiomyopathies and arrhythmias, decreasing the rate of SCD in young competitive athletes by nearly 90% [4]. However, some congenital cardiac diseases such as hypertrophic cardiomyopathies might remain undetected in this setting, especially when they cause no symptoms.

In our case, the recommended routine pre-participation screening would have failed to detect the potentially life-threatening cardiac disease, which constitutes a major risk for SCD. Our athlete was free of symptoms and both physical examination and ECG were normal. Furthermore, his physical performance was excellent.

Echocardiography, regularly performed in competitive athletes in our sports cardiology department, revealed hypokinesia of the inferior and inferolateral wall sections. It was the key diagnostic tool in diagnosing the coronary artery anomaly in our case. Further investigation with MRI confirmed the echocardiographic finding and detected ischemia and a transmural scar in the in the hypokinetic wall sections. Because CAD was unlikely in this specific case and we suspected a coronary artery anomaly, we decided to perform coronary CTA. This was in line with the recently published guidelines, which recommend the use of a coronary CTA to diagnose coronary anomalies [5]. An invasive approach with coronary angiography would have been possible for further investigation, although, today, coronary CTA is considered to be the first-choice approach [5]. In addition, coronary angiography would have been not able to display the anomalous origin of the circumflex artery in our case, as it arises from a convolution of vessels from small branches of bronchial arteries and has no connection to the aorta, left main, or LAD. Coronary CTA was less invasive and detected the abnormal origin and the complete course of the circumflex artery. Furthermore, we were able to exclude coronary artery disease [7–10].

In conclusion, coronary artery anomalies are rare and difficult to diagnose but constitute a major risk for SCD in athletes. The routine pre-participation screening for athletes often does not reveal pathologies such as malignant coronary anomalies. Therefore, other imaging methods are necessary for detection. Transthoracic echocardiography (TTE) was the key tool for further investigation in our asymptomatic athlete. Though, the value of TTE as a first-line screening tool for conditions predisposing to SCD remains to be proven, largely because of additional cost, lack of expertise and infrastructure, and no evidence for incremental diagnostic value compared to ECG alone [6].

Coronary CTA is currently the recommended method for diagnosing suspected coronary anomalies. If a malignant coronary artery anomaly is detected, participation in competitive sports should be contraindicated.

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