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Perfusion Study Helps in the Management of the Intraseptal Course of an Anomalous Coronary Artery

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Abstract

Anomalous origin of a coronary artery from the opposite aortic sinus of Valsalva can present in various ways, ranging from a benign and incidental finding to sudden cardiac death. The variant with an intraseptal subpulmonary course (sometimes referred to as intraconal), is widely perceived to carry a low risk of ischemia and has been considered to be a benign variant, not requiring surgical treatment. In one of our recent patients, however, nuclear scintigraphy highlighted a myocardial perfusion deficit in the territory supplied by the allegedly benign anomalous coronary artery, prompting the need for a more aggressive surgical approach.

Keywords

anomalous aortic origin coronary artery (AAOCA), anomalous coronary artery arising from the opposite sinus (ACAOS), intramyocardial course, intraseptal course, myocardial perfusion, stress test, asymptomatic ischemia, sudden cardiac death, congenital heart disease

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Introduction

Anomalous origin of a coronary artery from the opposite aortic sinus can present in various ways, ranging from a benign incidental finding to sudden cardiac death (SCD).[1] Although the risk of sudden death is widely recognized,[1-3] with hazards ranging up to one-quarter in cases involving the right coronary artery, and up to three-fifths in cases involving the left coronary artery,[3-5] the indications for surgery in asymptomatic patients remain a matter of debate.[6-8] In this regard, the risks of sudden death are increased in symptomatic patients, and in those with involvement of the left coronary artery. They are then known to be compounded by certain anatomical features, such as an interarterial course, a longer intramural tract, a slitlike ostium, an acute angle of take-off.[1-2,5] These risks have then to be weighed against the risks of the surgical intervention itself.[6-8] As far as is currently known, however, surgical management providing anatomical repair achieves good results, and can be undertaken at low risk.[6,8] The procedure seems to be effective in preventing sudden death,[8] provided that normal coronary anatomy and function are restored, and all potential culprits are addressed.[8] To endorse the notion that such surgery is effective in the prevention of ischemia, we report the case of an 11-year-old boy with an incidental diagnosis of an anomalous left coronary artery that took a subpulmonic intramyocardial course.

Case Report

An asymptomatic 11-year-old boy underwent transthoracic echocardiography as a screening for sport practice. The examination raised suspicions for the presence of a coronary

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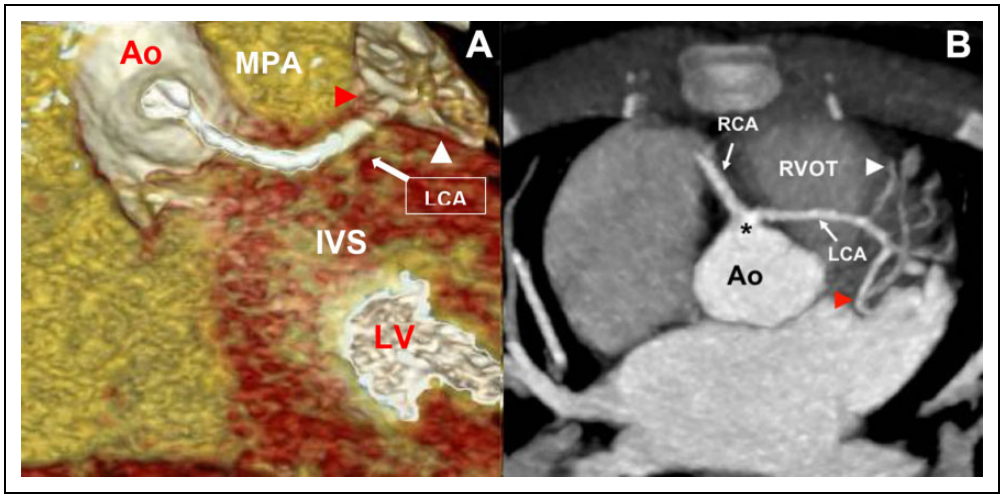

Figure 1. Coronary computed tomography angiography with volume rendering reconstruction (Panel A) and transaxial maximum intensity projection view (Panel B) show single shared coronary ostium with anomalous left coronary artery (LCA) from the right coronary sinus of Valsalva. The anomalous LCA main-stem courses within the subpulmonic outlet septum with a long intramyocardial segment which then emerges at epicardial level in the anterior interventricular groove, where it bifurcates into left anterior descending artery (white arrowhead) and circumflex artery (red arrowhead).

arterial abnormality, showing both coronary arteries arising from a single common orifice located in the right aortic sinus of Valsava. The family and personal history were negative for sudden death, chest pain, syncope, or other nonspecific symptoms. An ergometric test proved negative for inducible ischemia. A nuclear perfusion study, in contrast, performed under treadmill strain, highlighted narrow perfusion deficits in the apical third of the anteroseptal wall of the left ventricle. Left ventricular dimensions, however, were within normal limits, and the ejection fraction, both at rest and after exertion, was greater than 60%.

A computed tomographic study, using the electrocardiographically gated multidetector sequential technique, with an intravenous infusion of 50 mL of organo-ionic contrast agent (Iomeron 400), confirmed the presence of a single coronary arterial orifice within the right aortic sinus of Valsalva. The solitary coronary artery gave rise to the left coronary artery, which crossed between the arterial roots, taking an intramyocardial course within the crest of the muscular ventricular septum. Beyond this intramyocardial (intraconal) segment, the left coronary artery emerged as an epicardial vessel in the anterior interventricular groove. The right coronary artery, having branched from the solitary artery, coursed through the right atrioventricular groove and terminated as the inferior interventricular artery (Figure 1).

Following the examinations, and considering the presence of the myocardial ischemia highlighted by the perfusion study, it was decided to proceed with cardiac surgery. The operation was conducted under normothermic cardiopulmonary bypass, lasting 96 minutes, with aortic cross-clamping of 72 minutes, and providing myocardial protection with "Del Nido" crystalloid cardioplegia. After transection of the pulmonary trunk, the pulmonary root was completely harvested (as for the Ross procedure) in order to achieve a better exposure of the coronary intraseptal retropulmonary course and to ease the process of coronary debridging. The left coronary artery was completely freed from its tight muscular surroundings up to the anterior face of the ventricular septum, where it became epicardial The pulmonary root was then reimplanted and anteriorly translocated, with a posterior pericardial patch extension, to avoid any compression, twisting or distortion of the restored epicardial course of the coronary artery. The postoperative recovery was uneventful, with rapid normalization of myocardial enzymes, good biventricular function, and no evidence of ischemia. The patient was discharged on the seventh postoperative day. Six months after cardiac surgery, he underwent a control myocardial perfusion study with treadmill protocol. The study showed complete normalization of the perfusion deficit in the left ventricle, with the ejection fraction documented with Gated-SPET technique found to be 74% after exertion (Figure 2).

Discussion

Anomalous origin of a coronary artery from an inappropriate aortic valvar sinus is a concerning anomaly, especially because it can involve young and active population with catastrophic consequences.[3] The dimensions of the problem, furthermore, seem to be greater than expected.[1] Recent studies, in fact, have estimated the prevalence of this anomaly in the general population to be of 1% to 2%, more than double than previously assumed.[2] Although retroaortic, prepulmonic, and intraseptal courses of such anomalous arteries have been considered benign, recent reports show that up to one-quarter of patients with an intramyocardial septal course have developed serious cardiac events.[9] Anomalous origin through a single orifice in the valvar sinus, as in our patient, is particularly rare, with a reported incidence of 0.0024% to 0.044%.[9,10] According to a

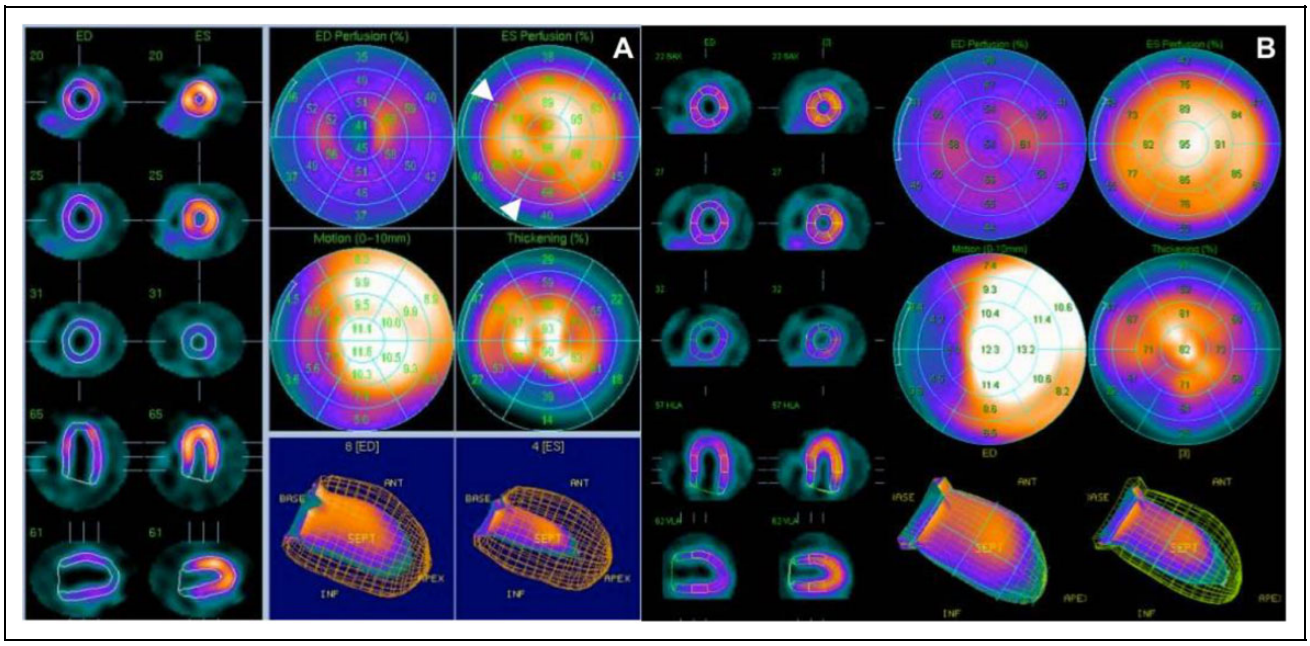

Figure 2. Nuclear myocardial stress perfusion test reveals inducible basal septal myocardial perfusion defect (arrowheads panel A). At the postsurgical six-month follow-up scan, there is complete normalization of the myocardial stress perfusion (panel B).

recent review, there are only five reports of such a variant, all involving older and symptomatic patients.[9]

The decision whether to intervene in asymptomatic patients with anomalies other than the recognized interarterial "malignant" course can be difficult. It is dependent on the type of lesion, the course of the coronary artery, and the age of the patient.[2,5-8] A transseptal intramyocardial course was initially thought to have the same risk for developing myocardial ischemia as for myocardial bridging.[11] More recent studies, however, have indicated that this should be considered a high-risk variant.[12]

When making the decision with regard to intervention, nuclear perfusion tests have been shown to be a helpful tool, and have gained growing importance over time.[2,8-9] This is confirmed by our own experience. Our patient was asymptomatic, with a negative ergometric stress test. The recognized anomaly, according to conventional wisdom, should not have been considered malignant. Myocardial scintigraphy, nonetheless, revealed the presence of ischemia of the anterior-septal wall of the left ventricle, thus providing the indication for surgical intervention.[13,14] Surgical management of this rare anomaly is challenging. The surgeon is confronted not only by the anomalous origin through a single orifice, potentially compressed by the pulmonary trunk, but also by the aberrant intraseptal and intramyocardial course. Alternative approaches have been proposed to address the distortion produced by the pulmonary root, pulmonary artery translocation has been proposed in two different ways. On the one hand, the pulmonary trunk can be translocated to the left, as described for treatment of anomalous origin of the main stem of the left coronary artery from the wrong sinus of Valsalva, with the artery extending anomalously course between the aorta and the pulmonary trunk.[15] On the other hand, the right pulmonary artery can be translocated anterior to the ascending aorta, as used in infants with tetralogy of Fallot and absent pulmonary valve syndrome to relieve airway compression.[16]

In our patient, the coronary artery was located deep in the muscular septum, posteroinferior to the pulmonary trunk. Isolated translocation of the pulmonary trunk was not feasible in order to address all the mechanisms of ischemia. Accordingly, we performed an additional extended myocardial debridging. The effectiveness of this approach in relieving ischemia was documented by postoperative nuclear perfusion study, which showed a complete normalization of the perfusion deficit.

We submit that our experience validates the aggressive approach to treatment of patients with aberrant origins of coronary arteries thus far considered benign. The approach avoids the potential of documented ischemia degenerating to SCD or chronic left ventricular failure, as in our case in an asymptomatic patient with a negative ergometric test. As was the case for our patient, although it is very hard to know the sudden death risk, a nuclear perfusion study, combined with exercise stress testing, may potentially assist in stratifying the risk of future cardiac ischemic events in asymptomatic patients with these anomalies.

Authors' Note

Parental informed consent for the patient's treatment and reporting of this case was obtained in accordance with Institutional Review Board guidelines.

Declaration of Conflicting Interests

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