Anomalous origin of circumflex coronary artery from right pulmonary artery associated with atrial septal defect

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We describe a 4-year-old girl with anomalous origin of the left circumflex coronary artery from the right pulmonary artery and large atrial septal defect. This is the first reported case of such association. Surgical reimplantation of the anomalous left circumflex coronary artery to the aorta and atrial septal defect surgical closure was performed, with no postoperative complications.

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Introduction

Anomalous origin of the left circumflex coronary artery (LCx) from the right pulmonary artery (RPA) is an extremely rare coronary anomaly [1,2]. The clinical course of LCx may not always be favorable, and patients need surgical treatment in early infancy [1,3]. A literature search revealed no cases of anomalous LCx associated with atrial septal defect (ASD) reported to date.

Case report

A 3-year-old girl (weight 15 kg) with large secundum ASD was referred to the Madinah Cardiac Center, Madinah Munawara, Saudi Arabia. Echocardiography showed large ASD with deficient rims, severe dilatation of right ventricle and right atrium, with normal left ventricle size and ejection fraction (62%), and abnormal flow in the RPA presumed to be a small aortopulmonary collateral or small coronary artery fistula.

The ASD was closed surgically at this time. Follow-up echocardiogram after 1 year demonstrated abnormal flow into the RPA with mild dilatation of left ventricle, with suspicion of coronary anomaly (Fig. 1). Diagnostic cardiac catheterization was done (Fig. 2). Aortograms and selective coronary angiograms in different projections showed a normal right and left main coronary arteries arising from the respected coronary sinus.
The left anterior descending coronary artery was communicating with the LCx with multiple significant collaterals, which drained ultimately into the proximal RPA. She underwent reoperation at age 4 years. Operation was performed through median sternotomy using hypothermic (28 °C) cardiopulmonary bypass. The pulmonary artery was dissected, the large LCx originating from the posterior aspect of RPA, to the right of the ascending aorta identified (Fig. 3), and was mobilized almost 2 cm without difficulty. Antegrade Custodiol Cardioplegia (DR FRANZ KOHLER CHIEME GMBH, Bensheim, Germany) cardioplegia was given via the aortic root, RPA and aorta were transected, and a generous button of the orifice of the anomalous coronary artery was mobilized. It was reanastomosed to the ascending aorta above the sinotubular junction posteriorly between the left
coronary and noncoronary commissure after making a j-shaped slit in the wall of the ascending aorta. The RPA defect was patched with an oval patch of bovine pericardium.

The patient was discharged from the hospital 6 days postoperatively without any complications. Eight months after the operation echocardiography showed good left ventricular contractility with an ejection fraction of 65%, good laminar flow to the LCx with no stenosis, and no right pulmonary or ascending aorta stenosis.

Discussion

Anomalous origin of the LCx from the RPA associated with ASD has not been previously reported. Most of the cases reported in the literature are associated with aortic arch anomaly [1,4,5]. The symptoms of the anomalous origin of LCx from the RPA may present in different periods of life or patients may remain asymptomatic with the anomaly being discovered accidentally during diagnostic procedures. There are no standard indications or operative techniques for such patients. Our case was diagnosed at age 3 years to have large secundum ASD not amenable to device closure. Abnormal flow on echocardiography was noticed in the RPA but anomalous connection of the coronary arteries was not suspected at first surgery.

One year after ASD closure, follow-up echocardiogram showed mild left ventricle dilatation, normal left ventricular systolic function, but no evidence of ischemic changes by electrocardiogram. Coronary angiogram was done, which confirmed anomalous origin of LCx from RPA. Most LCx anomalies mentioned in the literature were associated with aortic coarctation and almost all cases were discovered accidentally later on after aortic arch repair [1,4,5]. Chopra et al. [6] reported a 15-year-old female patient who underwent surgical correction of an aortopulmonary window at age 13 months. Fourteen years later, she presented with dyspnea on exertion associated with angina. A coronary artery bypass grafting of the anomalous LCx and ligation of its anomalous origin in the pulmonary artery were performed. Sariglu et al. [4] described a 10-year-old female patient who underwent surgical repair of the aortic coarctation at age 4 years. Six years later, she presented with chest pain during exercise. Cardiac catheterization demonstrated retrograde filling of the LCx from the left anterior descending and right coronary artery, with drainage into the RPA. Alexi-Meskishvili et al. [2] reported two infants with anomalous LCx from PA who had to be operated at ages 40 days and 30 days because of severe myocardial dysfunction. The anomalous LCx was reimplanted to the aorta in the first patient, and simple ligation via left thoracotomy was chosen in the second case.

In conclusion, our patient is the first reported case of anomalous origin of LCx from the RPA associated with ASD. Echocardiography is still the baseline for diagnosis; however, coronary angiogram is required to confirm the diagnosis. Anomalous LCx origin can be silent or can present with ischemic changes, and surgical correction is mandatory even if the patient is asymptomatic.

Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent

Informed consent was obtained from all individual participants included in the study.

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