Cardiology in the Young

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Acquired multiple coronary artery fistulas: a rare complication of congenital heart surgery in a child diagnosed with right heart valve endocarditis

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Brief Report

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Abstract

Majority of coronary artery fistulas are of the congenital origin, and they have been rarely reported after congenital heart surgery. Here, we present echocardiographic images and prognosis of multiple acquired coronary artery fistulas occurring after surgical myectomy in a child diagnosed with double-chamber right ventricle complicated with infective endocarditis involving the right heart valves.

Introduction

A 12-year-old female patient diagnosed with double-chamber right ventricle and cerebral palsy was admitted for routine control. The patient's history revealed that she had used antibiotics with a complaint of high fever 1 month ago and had intermittent fever and vomiting for the last few days. Physical examination revealed low-grade fever, cardiac murmur, and neuromuscular retardation but no haemodynamic disturbance. Previous transthoracic echocardiography was performed 4 months ago, and it showed severe subvalvular pulmonary stenosis (pulmonary valve velocity: 5 m/s) due to a double-chamber right ventricle. She also had severe degree tricuspid valve insufficiency, mitral valve prolapse with mild degree mitral regurgitation, and a tiny patent ductus arteriosus. At the last admission, we noticed that there were multiple echogenicities compatible with vegetations on the tricuspid (12 × 13 mm) and pulmonary valves (11 × 7 mm) (Figure 1, Video 1). These echocardiographic findings were accepted as major echocardiographic criteria of endocarditis. The acute phase reactants were elevated. The patient was hospitalised with the diagnosis of infective endocarditis. The wide spectrum of antibiotics (ceftriaxone, vancomycin, and amikacin) were started immediately after blood cultures were taken. Six blood cultures were taken from our patient during hospitalisation, and *Enterococcus faecium* growth was detected in only one, so it was accepted as a minor criterion. Congenital cardiac abnormalities, elevated acute phase reactants, and positive blood cultures were accepted as three minor criteria of endocarditis.

Although she has no haemodynamic disturbance in clinical evaluation, we observed the persistence of the vegetation with no change in size and the low grade of fever peaks in spite of 22 days of antibiotics at the follow-up. Then an open-heart surgery was performed. The vegetation on the tricuspid valve was removed with surgery, the tricuspid valve was repaired, the vegetation on the pulmonary valve was cleared, the cleft on the pulmonary valve was repaired, and the ductus was ligated. Muscle band excision was performed through the right ventricle side by deep myectomy. Postoperative transthoracic echocardiogram showed no vegetations on the valves, no obstruction of the right ventricular outflow tract (gradient 15 mmHg), and only mild tricuspid and pulmonary regurgitation. There was a small perimembranous ventricular septal defect by colour and continuous wave Doppler echocardiography that could not be seen in preoperative echocardiography.

Furthermore, there was three colour Doppler left to right jet crossing the interventricular septum resembling ventricular septal defect, but a continuous low velocity flow by continuous wave Doppler was detected on the right ventricle outflow tract on surgical resection site of the muscle bound that was consistent with multiple coronary fistulas (Figure 2, Video 2). Our patient had no signs of volume overload, and there were no findings related to coronary ischaemia by laboratory investigation (normal electrocardiogram and normal troponin level). After 1 month, echocardiography showed only one small size coronary fistula on the interventricular septum and spontaneous closure of the other two fistulas (Figure 2, Video 2). After 4 months, she has no medication and is free of symptoms. At the fourth month follow-up, the last coronary fistula disappeared spontaneously (Video 2).

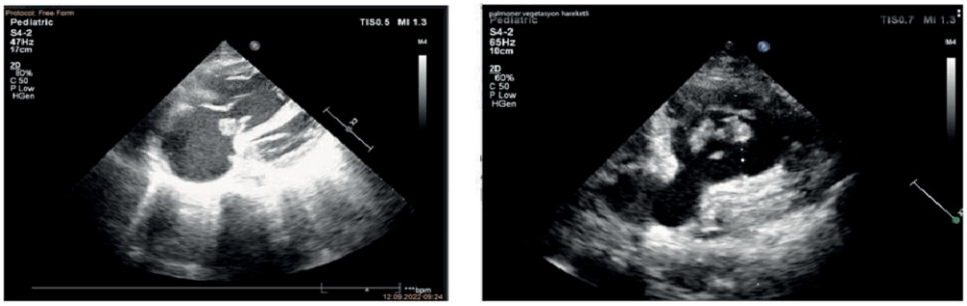

Figure 1. Transthoracic echocardiography showed vegetations in the tricuspid and pulmonary valves.

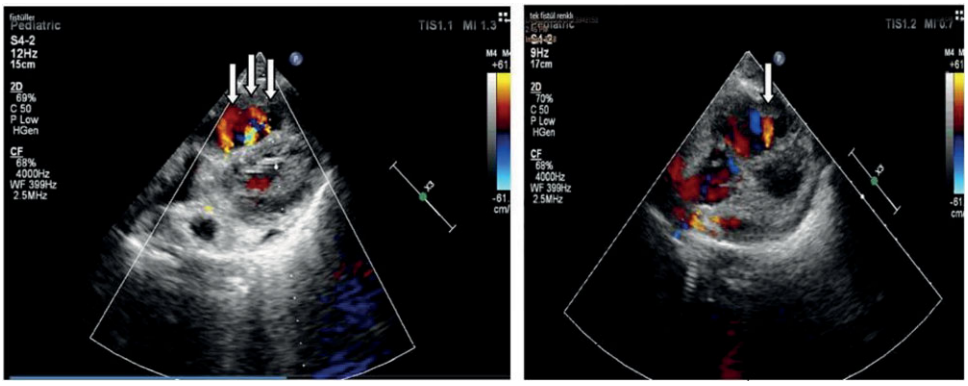

Figure 2. Echocardiography showed three coronary fistulas in the early postoperative period and the single remaining coronary fistula at the first month postoperative visit.

Discussion

The coronary artery fistulas are defined as a direct precapillary connection between a branch of the coronary artery and the lumen of one of the four cardiac chambers or great vessels.[1] The majority of coronary artery fistulas are of the congenital origin. Acquired coronary artery fistula is a rare complication after open-heart surgery. The most common reasons were surgery of tetralogy of Fallot, ventricular septal defect with double-chamber right ventricle, and transposition of the great arteries with ventricular septal defect.[2] Coronary artery fistulas are a well-described phenomenon following surgical myectomy.

The resection of the infundibular muscle or anomalous muscle bundle may damage the precapillary coronary arteriole in the circle of Vieussens and redirect arterial blood flow either from the left or right coronary artery to the right ventricle chamber.[1] However, coronary artery fistulas are commonly located in the left ventricular surface of the septum in patients with hypertrophic cardiomyopathy in the early period after surgical myectomy.[3]

Small-sized fistulas are usually asymptomatic and incidental findings of echocardiography.[4] Echocardiography shows coronary fistulae with continuous low velocity flow in the right ventricle (diastolic > systolic). These lesions can be seen if only the echocardiography performs in the early period after surgery because of their rapid spontaneous closure.[4]

The coronary fistulas were accepted as a complication of deep myectomy in our case. Although the impact of the presence of endocarditis on the likelihood of postoperative coronary fistula is not clear, coronary fistulas can be seen as a complication of infective endocarditis and also occur after endocarditis surgery. Lorena Fernandez-Ruız et al. observed coronary artery fistüla (CAF) from the left coronary artery to the right ventricle in a 6-month-old child with tetralogy of Fallot; the diagnosis was made after immediate postsurgical period and spontaneous closure observed 1 month after the initial diagnosis.[5] Exact mechanisms of spontaneous closure of acquired coronary artery fistula is not known. However, it may resemble the usual mechanism of spontaneous muscular ventricular septal defect closure, muscular encroachment, and superimposed fibrous tissue enclosing the whole defect or hypertrophy of the septal myocardium.

Aurelio Sgalambro et al. defined 40 patients with hypertrophic cardiomyopathy undergoing extended septal myectomy.[3] Preoperatively, none of the patients had evidence of coronary artery fistulas. After surgery, 9 of 40 patients were diagnosed with coronary artery fistula with a prevalence of 23%. At 6 months, coronary artery fistulas could still be detected in two patients but had spontaneously disappeared in the remaining seven patients. Coronary fistula of one patient persisted at follow-up (37 months) without any significant haemodynamic impairment.

Like our case, monitoring may be sufficient in asymptomatic cases. They have excellent prognosis and only need serial echocardiographic examination and clinical follow-up.[4] Treatment options of symptomatic coronary artery fistula consist of trans-catheter or surgical closure.[6] Left ventricular enlargement due to left to right shunt and symptoms of congestive heart failure or ischaemia due to coronary steal are major reasons for surgery.[7]

Supplementary material. The supplementary material for this article can be found at https://doi.org/10.1017/S1047951124026167

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