

Coronary Artery Aneurysm and Coronary Fistula in Tetralogy of Fallot: A Case Presentation of A Young Adult Undergoing Total Correction

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ABSTRACT

Congenital presentation of a coronary artery fistula is a rarely observed pathologic finding; especially an association with tetralogy of Fallot is reported in only three cases previously. This is a presentation of a patient with tetralogy of Fallot who had an aneurysmatic fistula communication between the circumflex artery and left ventricle. Preoperative computerized tomography angiographic examination showed a communication between the circumflex artery and left ventricle. 27-year-old female patient underwent an operation for total correction. Intraoperatively, the fistula could not be seen and it had been left unligated. The patient had been discharged from hospital without problems on the tenth postoperative day. Aneurysmatic coronary artery fistula is not found to cause a deleterious effect on the hemodynamics or myocardial performance of the heart after the total correction surgery

Key Words: Tetralogy of Fallot; Coronary artery; aneurysm; coronary fistula.

ÖZET

Fallot Tetralojisinde Koroner Arter Anevrizması ve Koroner Fistul: Tam Düzeltme Yapılan Genç Erişkin Bir Vaka Sunumu

Konjenital koroner arter fistülü oldukça nadir gözlenen bir patolojik bulgudur. Özellikle Fallot tetralojisi ile birlikteliği ise daha önce sadece üç vaka olarak rapor edilmiştir. Bu yazımızda sircumflex arter ile sol ventrikül arasında fistül bulunan fallot tetralojili bir hastanın sunumu yapılmaktadır. Ameliyat öncesi bilgisayarlı tomografik anjiyografide sol ventrikül ile sircumflex arter arasında anevrizmatik fistül ilişkisi gözlemlendi. 27 yaşındaki bayan hasta tam düzeltme ameliyatına alındı. Operasyonda fistül bulunamadı ve bağlanmadan bırakıldı. Problemi olmayan hasta, ameliyat sonrası onuncu günde taburcu edildi. Anevrizmatik koroner arter fistülün tam düzeltme ameliyatı sonrasında miyokardiyal performansda ve hemodinamide bozulma yaratan bir etkisi gözlemlenmedi.

Anahtar Kelimeler: Fallot tetralojisi; koroner arter; anevrizma; koroner fistül.

INTRODUCTION

Congenital presentation of a coronary arteriovenous fistula (CAVF) is a rare abnormal finding and it was reported to present in approximately 1 in every 50.000 in patients with congenital heart disease (1,2). The association of coronary fistula and tetralogy of fallot is reported only in three cases in the literature (3). However, aneurysm in the coronary artery fistula has not been previously reported. The early recognition of coronary arteriovenous fistulas is imperative, as management and treatment could prevent serious complications. In this case report, in the two coronary arteries high degree of fusiform aneurysmatic dilatation and coronary fistula between circumflex artery and left ventricle was detected in the angiographic study preoperatively in a patient with tetralogy of Fallot. The presentation of this case and the surgical intervention and subsequent postoperative course is presented to discuss this unusual case and necessary precautions that is required during management in preoperative, operative and postoperative periods.

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Figure 1: Presentation of aneurysmatic coronary arteries.

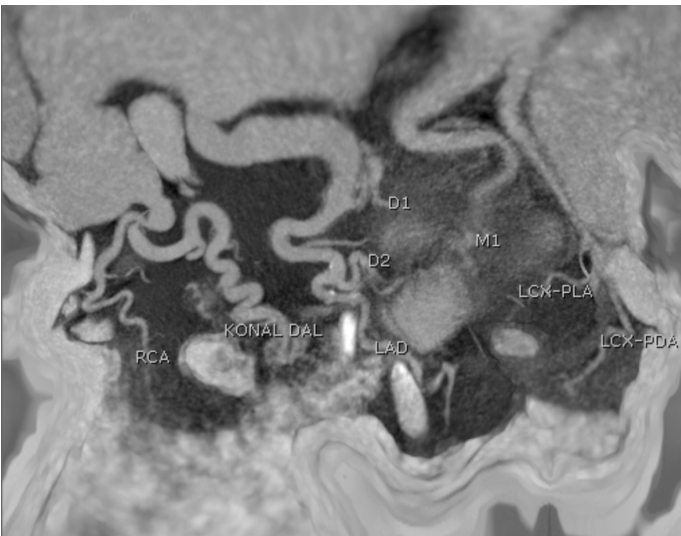


Figure 2: CT showing aneurysmatic coronary arteries and coronary-ventricular fistula.

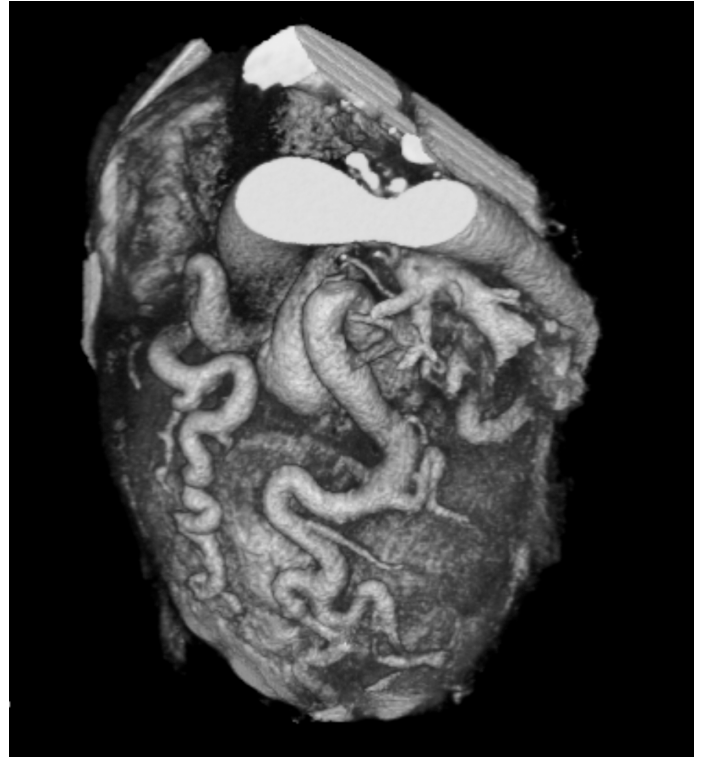


Figure 3: CT showing aneurysmatic coronary arteries.

costal margin and mild pretibial edema is palpated in the lower extremities.

The investigations included; an electrocardiogram which revealed a right ventricular hypertrophy, right axial deviation, atrial fibrillation without any ischemic finding. A chest X-ray showed; cardiomegaly, congestion in basal lung fields bilaterally and biatrial dilatation of the heart. Echocardiographic evaluation revealed; 1) Fallot tetralogy and atrial septal defect (ASD), 2) left ventricular hypertrophy, 3)biatrial dilatation, giant left atrium (right atrium diameter 90 mm). Coronary arteriography by computerized tomographic (CT) study showed; 1) All coronary arteries visualized as having fusiform aneurysmatic presentation and run in an uneven course that especially the diameter of the circumflex artery increases to 2.2 cm at the proximal region and in some regions during its course this diameter is even higher than the diameter of the aorta that has been in a dextraposition. 2) The first obtuse marginal (OM1) branch of the circumflex artery is fistulized into the left ventricle. 3) Left pulmonary artery diameter was 15.3 mm, the right pulmonary artery diameter was 10.2 mm and truncus pulmonalis diameter was 24 mm. 4) There was serious stenosis at the pulmonary infundibulum and the diameter was 11 mm.

The patient was scheduled for total correction and was admitted to surgery electively. The operation was performed under general anesthesia and cardiopulmonary bypass was performed under mild hypothermia (28 °C). Also anterograde blood cardioplegia was used for myocardial protection. During surgical procedure, right atriotomy was performed. An organized thrombus formation is observed at the proximal end of the vena cava superior and a thrombectomy was performed. After

CASE REPORT

A 27 year old woman presented to our clinic with difficulty in breathing, blue discoloration in lips and hands, palpitation. The patient was told to have congenital heart disease but had not received any treatment previously. In the family history, there is no kinship or any sibling with congenital heart disease. She has a healthy brother and a sister who are both alive.

In the physical exam, the blood pressure was 100/60 mmHg, heart rate was 95/minute, respiratory rate was 28/minute, arterial oxygen saturation was 79%. The cyanosis in lips was more prominent than the periphery and there was pertinent respiratory insufficiency and clubbing in hand nails. The auscultation showed a 5/6 systolic murmur which is best heard at the mesocardiac location that is also heard in all other locations as well. Bilateral crackles are heard in lower lobes of lung fields. The palpation of the liver was 2 cm below the

the administration of cardioplegia, the blood was removed from the heart and with appropriate positioning the area to perform right ventriculotomy and pulmonary arteriotomy is visualized between the right coronary artery (RCA) and left anterior descending coronary artery (LAD) which were both appeared as aneurysmatic arteries. ASD was repaired primarily. Ventricular septal defect (VSD) was closed with dacron patch. Right ventricle outflow tract reconstruction was performed with pericardial patch after precautions was taken to protect the posterior cusp of the pulmonary valve. An unsuccessful search was performed to find the first obtuse marginal (OM1) branch of the circumflex artery that was fistulized into the left ventricle. The coronary arteries that were aneurysmatic were left in their places without any further intervention. Patient received inotropic support of dobutamine infusion for a total of 48 hours including the weaning period and was extubated on the postoperative second day. She was discharged to home without any complications on the tenth postoperative day.

DISCUSSION

In tetralogy of Fallot the abnormalities related to the coronary arteries is commonly observed (4). The association of coronary fistula and tetralogy of fallot is reported in only three cases in the literature (3). The pathologic findings related to these abnormalities may vary and the clinical presentation and subsequent treatment modalities change accordingly depending on the clinical symptoms. Some anurysmatic coronary arteries may require medical and surgical treatments whereas others remain asymptomatic (5,6).

In the diagnosis of coronary artery aneurysms, conventional angiography and 64-section multidetector computerized tomography (MDCT) was used extensively as a significant diagnostic tool in several studies (7,8,9). In our case MDCT angiography study was used to evaluate the coronary arteries and revealed aneurysmatic dilatation in two coronary arteries and a fistula between circumflex artery and left ventricle.

Arteriovenous fistulas were reported more commonly between the coronary arteries and the pulmonary arteries. Dabizzi et al. (10) observed fistula between coronary artery and pulmonary artery in 13 patients in a series of 119 patients after preoperative selective coronary angiographic evaluation. Of these 13 cases only 1 patient had fistula between right coronary artery anterior branch and right atrium. Badak et al. (11) have reported a fistula between right coronary artery and right atrium. Saxena et al. (12) have reported a fistula between circumflex artery and coronary sinus. Ito et al. (13) have shown a coronary artery pulmonary artery fistula originating from three major coronary branches associated with exertional chest pain and tachycardia-dependent left bundle branch block

The aneurysm in two coronary arteries especially RCA and LAD and coronary fistula presentation in circumflex artery was significant pathologic findings to hesitate to proceed with total correction procedure in an adult patient with tetralogy of Fallot. We have evaluated the pat-

hologic findings with dual slice CT coronary angiography and were able to make a plan to perform right ventriculotomy without any intervention to the aneurysmatic coronary arteries. Although this surgical plan was successfully performed, the coronary fistula was unable to be detected for further evaluation and for possible repair. Postoperatively no problems related to cardiac performance occurred and this suggests that coronary fistula is a benign presentation that does not require intervention.

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