Case Report / Olgu Sunusu

İntraperikardiyal sol modifiye Blalock-Taussig şant: nadir bir olgu

Intrapericardial left modified Blalock-Taussig shunt : A rare case

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ÖZET

Modifiye Blalock-Taussig şant siyanotik kalp hastalıklarında en sık kullanılan sistemik pulmoner arter şantıdır. Günümüzde yaklaşım palyatif prosedürlerden, erken tüm düzeltmeye kaymıştır. Ancak modifiye Blalock-Taussig şant halen tüm düzeltmeye engel olan koroner arter anomalili ciddi hipoplastik arterli olgularda hayat kurtarıcı bir prosedürdür. Biz, burada Fallot tetralojili, sağ arkus aorta, ayna görüntüsü aortalı ve koroner arter anomalili bir hastayı sunuyoruz. Bu hastaya sternotomi ile nadir bir yaklaşım olan intraperikardiyal sol subklavyen-pulmoner arter şantı yapılmıştır.

Anahtar sözcükler: Blalock-Taussig şant, Fallot tetralojisi, koroner anomali

ABSTRACT

The modified Blalock-Taussig shunt is the most commonly performed systemic to pulmonary artery shunt in children with cyanotic heart disease. Nowadays the approach has shifted from palliative procedures to earlier total correction. But modified Blalock-Taussig shunt is still a lifesaving procedure in cases with severe hypoplastic pulmonary arteries and concomittant coronary artery anomalies which disallow total correction. Here, we present a case with tetralogy of Fallot, mirror image aorta, right aortic arch and coronary artery anomaly. An intrapericardial left subclavian to left pulmonary artery shunt, which is a very rare entity, was performed with sternotomy.

Keywords: Blalock-Taussig shunt, tetralogy of Fallot, coronary anatomy

Introduction

The timing for surgical correction of cyanotic heart disease has shifted towards earlier ages in the modern era. The diminished pulmonary arterial blood flow is the major manifestation, and the presence of severely hypoplastic pulmonary vasculature and concomittant coronary arterial anomalies may lead to a delay in timing of total surgical correction. The modified Blalock-Taussig shunt (MBTS) defined as a systemic to pulmonary artery shunt between ipsilateral subclavian artery and pulmonary artery is the most commonly performed palliative procedure in these cases (1-4). Different approaches are reported for MBTS including thoracotomy and sternotomy (4). Median sternotomy is more commonly preferred due to technical ease and anatomical considerations.

Here we report an intrapericardial left MBTS, which is a rare entity, performed via sternotomy approach in a case with tetralogy of Fallot, mirror-image aortic arch and coexisting coronary arterial anomaly which is a rare presentation.

Case Report

A ten-month old male patient admitted to our clinic with the diagnosis of tetralogy of Fallot and mirror-image aortic arch. He had cyanosis and heart murmur in the newborn period and advanced diagnostic eveluation was made. Physical examination revealed peripheral and perioral cyanosis with clubbing. 4/6° pansystolic murmur was heard on left sternal border. Trancutenous pulse oximetry measured oxygen saturation as 75%. Preoperative echocardiography and catheterization revealed tetralogy of Fallot with mirror image aortic arch. Gradient over pulmonary valve was measured as 75 mm Hg on both echocardiography and angiography with right ventricular hypertrophy. Right ventricular systolic pressure was 90 mm Hg. There was a suspicion for coronary artery anomaly. Right pulmonary artery was measured 8.2 mm and left was 6.9 mm. The aorta was 7.5 mm. McGoon value was calculated as 2.01. With these findings total correction was planned.

Intraoperatively median sternotomy was performed. After the pericardium had been opened an infindibular (or conal) branch arising from right coronary artery crossing the right ventricular outflow tract was observed. Its diameter was same as the right coronary artery. The conal branch divided into two just below the pulmonary annulus. Due to anatomical considerations, we planned a left intrapericardial MBTS. With a 4 mm gore-tex vascular greft a MBTS was performed. Low dose (3µg/kg/min Dopamine) inotropic support was initiated during the procedure and continued postoperatively until the second day. The oxygen saturation increased from 75 to 90% postoperatively. The patient was sedated and paralyzed for 18 hours, and extubated on 27th hour. Heparin infusion was begun intraoperatively and continued for 2 days. Acetylsalicylic acid (10 mg/kg, po) was begun on postoperative first day. The whole postoperative course was uneventful and the patient was discharged on seventh day. To demonstrate the detailed anatomy of the aortic arch, computerized tomography (CT) angiography was performed. Mirror image aorta and right aortic arch, patent left MBTS and high and posterior take off of the right subclavian artery was documented (Figure 1). The patient was lost to follow-up.

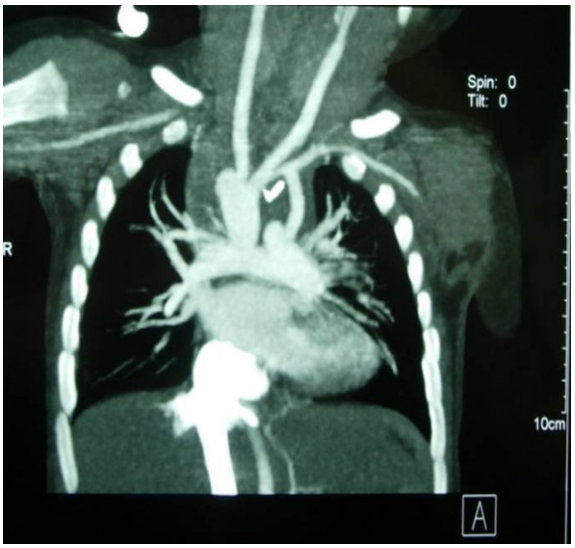

Figure 1. Patent left intrapericardial modified BT shunt

Discussion

Modified Blalock-Taussig shunt is a commonly performed palliative operation in cyanotic congenital heart disease. Sometimes the decision may shift from total correction to palliation intraoperatively due to challenging anatomy most commonly coronary artery anomalies in tetralogy of Fallot. The incidence of a major coronary artery crossing the right ventricular outflow tract in tetralogy of Fallot is 5-12%. These vessels are not always detectable preoperatively and sometimes even intraoperatively (5). There are various surgical techniques to perform total correction in cases with tetralogy of Fallot and concomitant coronary artery

anomalies. Coronary artery may be mobilized, incision may be extended under the coronary artery and by means of a pericardial patch, the right ventricular outflow tract and pulmonary artery may be expanded. Alternatively, a conduit between right ventricle and main pulmonary artery may be anastomozed (6). Transatrial-transpulmonary approach or reverse flap tecniques may also be performed in such cases if the coronary anatomy is suitable. If the coronary artery is very close to the pulmonary annulus, then right ventricular outflow tract reconstruction can not be made adequately (7). If right ventricular outflow tract can not be reconstructed without sacrifying coronary artery or a right ventricle to pulmonary artery conduit can not be placed due to possible sternal compression or proximity to the septum, a systemic to pulmonary artery shunt must be performed to maintain adequate pulmonary blood flow and pulmonary arterial growth. This approach allows time while the child grows as well as pulmonary arterial trunk and the right ventricle. Thoracotomy approach is more commonly employed; when sternotomy is performed, a right sided intrapericardial shunt is preferred due to anatomical ease.

In this case we first planned a right ventricle to main pulmonary artery conduit, but there was no suitable place on the right ventricle for the conduit providing far enough distance from the septum that could avoid sternal pressure. We aimed to preserve the coronary artery and perform total correction by using the reverse flap tecnique or mobilization of coronary artery and use of a pericardial patch, but the coronary artery anatomy prevented adequate repair. In our clinical experience we perform a systemic to pulmonary artery shunt in these patients and delay total correction until suitable anatomy develops. So we decided to perform a MBTS. We usually prefer a right sided subclavian artery to pulmonary artery shunt. The patient had a right aortic arch with left brachiocephalic trunk and right main carotid artery and we could not reach the right subclavian artery from sternotomy due to heavy collaterals. So left subclavian artery and left pulmonary artery were explored. On the left side, the main difference from the right side is that the dissection must be made further to expose the subclavian and pulmonary arteries.

As an alternative, but a very invasive approach to avoid this technical difficulty, left thoracotomy is made following closure of sternotomy and left MBTS is performed still in some centers. In this case we preferred to perform an intrapericardial left MBTS.

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