Gülhane Tıp Dergisi 2008; 50: 125-127

OLGU SUNUMU/CASE REPORT

Anomalous origin of the right coronary artery from the pulmonary artery combined with aortopulmonary window: a case report

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Summary

Aortopulmonary window with anomalous origin of the right coronary artery from the pulmonary artery is a rare but important lesion. We report a 13-day-old patient presenting with anomalous origin of the right coronary artery from the pulmonary artery with aortopulmonary window. We connected the aorta with the anomalous ostium, using closure of aortopulmonary window with the help of autologous pericardial patch.

Key words: Anomalous origin of the right coronary artery from the pulmonary artery, aortopulmonary window, pericardial patch

Özet

Sağ koroner arterin pulmoner arterden orijin aldığı ve aorto-pulmoner pencerenin eşlik ettiği bir olgu

Sağ koroner arterin ana pulmoner arterden orijin almasıyla beraber aortopulmoner pencere bulunması nadir görülen, ancak önemli bir defekttir. Bu tanıyla hastanemize başvuran 13 günlük bir olgu sunulmuştur. Hastada perikardiyal yamadan faydalanılarak aortopulmoner pencerenin kapatılması ile aorta ile anormal açıklığın devamlılığı sağlanmıştır.

Anahtar kelimeler: Sağ koroner arterin pulmoner arterden kaynaklanan çıkış anomalisi, aortopulmoner pencere, perikardiyal yama

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Date submitted: January 22, 2007
Accepted: June 07, 2007

Introduction

Anomalous origin of the right coronary artery from the main pulmonary artery is a rare congenital cardiac malformation. We report a 13-day-old-patient presenting with anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) with aortopulmonary window (APW). Surgical correction was performed and the orifice of the right coronary artery was derivated into the aorta with autologous pericardial patch. The patient's postoperative course was uneventful and he has been asyptomatic in 6 months after the surgery.

Case Report

A 7-day-old male newborn was referred to the division of Pediatric Cardiology of the Baskent University Hospital for the assessment of a cardiac murmur. The infant was 3300 gr. The electrocardiogram showed sinus rhythm and right ventricular hypertrophy. Transthoracic echocardiography revealed good biventricular function, a 3.5-4 mm wide APW and anomalous origin of the right coronary artery from the pulmonary trunk. Cardiac catheterization was performed prior to surgical intervention and demonstrated mildly increased pulmonary pressures. Aortography revealed anomalous origin of the dominant right coronary artery from the pulmonary trunk and a large APW (Figure 1,2).

Surgical correction was performed with the cardiopulmonary bypass support, moderate hypothermia and cold antegrade crystalloid cardioplegic arrest. After longitudinal pulmonary arteriotomy, a 4-mm-wide APW and ARCAPA were identified. The anomalous

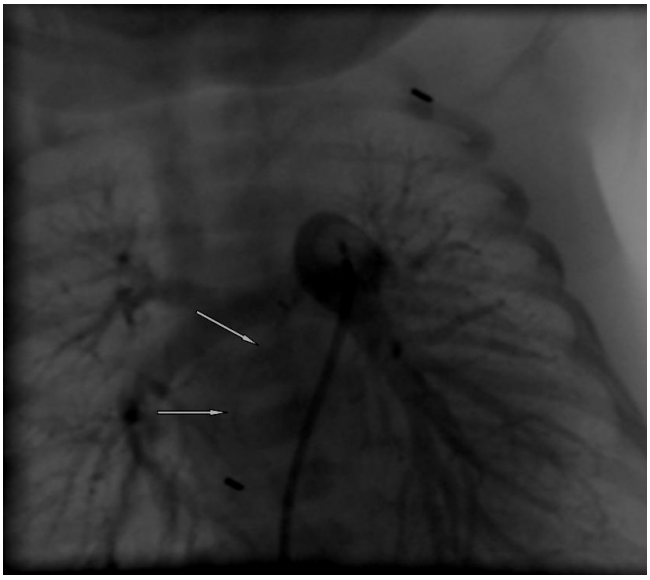

Figure 1. Angiographic view of the right coronary artery originating from the pulmonary artery

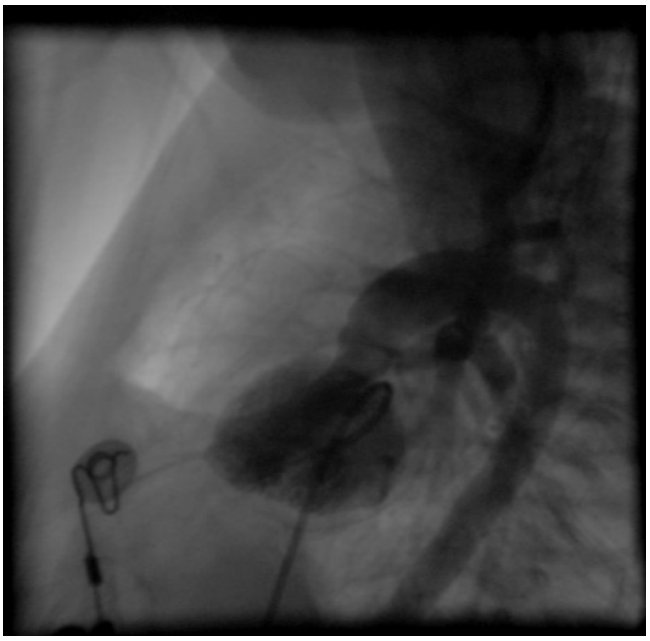

Figure 2. Angiographic view of the aorto-pulmonary window

coronary ostium situated below the APW was derivated to the aorta through intrapulmonary baffle of autologous pericard. The pulmonary artery was closed primarily.

Total cardiopulmonary bypass time was 52 minutes. Weaning from extracorporeal circulation was unproblematic. The postoperative course was uneventful. The patient was discharged in an asymptomatic condition and control echocardiographies showed good right and left ventricular function, no residual shunt and no stenosis of the ascending aorta and pulmonary artery. The right coronary artery blood flow was in normal range in the follow-up period of six months.

Discussion

APW with ARCAPA is a rare but important lesion (1-4). No genetic associations or environmental risk factors are known. The 2 competing embriyologic theories are that APW is part of a spectrum of conotruncal abnormalities, which includes truncus arteriosus at one end of the spectrum, and that APW is unrelated to truncus arteriosus because the lesions associated with each defect are so dissimilar (5,6). Most patients remain asymptomatic because of the presence of well oxygenated blood in the pulmonary trunk owing to the left to right shunt through the APW (4,7). However, there are some cases presenting with angina, myocardial infarction, heart failure and sudden cardiac death (2,3,8). Our case was also asymptomatic and referred for assessment of a cardiac murmur. Because of the simultaneous filling of both great arteries, preoperative angiographic diagnosis could be difficult. Injection of the contrast into both the great arteries is the best way to diagnose APW and to determine the relationship between pulmonary and coronary arteries and the defect (4,8,9).

Echocardiography is often the only imaging obtained prior to the repair of many cardiac lesions. In our case initial diagnosis was also made by transthoracic echocardiography. This demonstrates that coronary artery evaluation should be performed at every new echocardiographic evaluation (7).

In general, a corrective operation is recommended in this malformation even for asymptomatic patients. To correct the aortopulmonary defect through a ligature in association with this coronary anomaly led to intraoperative problems that did not permit the use of this technique. Different techniques were employed for connection of the anomalous coronary ostium with the aorta. One of them is reimplantation of the right coronary artery from the pulmonary artery to the aorta (4). In our case, we connected the aorta with the anomalous ostium, using APW with autologous pericardial patch as reported elsewhere (10,11).

Anomalous origin of the coronary arteries may be seen as isolated or associated with other cardiac defects, in particulary with conotruncal abnormalities. When a conotruncal abnormality is diagnosed, anomalous origin of the coronaries should always be considered. Exact preoperative diagnosis is very important for surgical correction, and it must be done early to prevent the development of irreversible pulmonary vascular disease.

References

1. Stopfkuchen H, Feichter GE, Jungst BK. Anomalous origin of the right coronary artery from the pulmonary artery combined with aortopulmonary window. Z Kardiol 1974; 63: 1086-1098.
2. Luisi SV, Ashraf NH, Gula G, Radley-Smith R, Yacoub M. Anomalous origin of the right coronary with aortopulmonary window: functional and surgical considerations. Thorax 1980; 35: 446-448.
3. Brouwer MH, Beaufort-Krol GC, Talsma MD. Aortopulmonary window associated with an anomalous origin of the right coronary artery. Int J Cardiol 1990; 28: 384-386.
4. Grunenfelder J, Zund G, Vogt PR, Turina MI. Aortopulmonary window with anomalous origin of the right coronary artery. Ann Thorac Surg 1999; 67: 233-235.
5. Backer CL, Mavroudis C. Surgical management of aortopulmonary window: a 40-year experience. Eur J Cardiothorac Surg 2002; 21: 773-779.
6. Berry TE, Bharati S, Muster AJ, et al. Distal aortopulmonary septal defect, aortic origin of the right pulmonary artery, intact ventricular septum, patent ductus arteriosus and hypoplasia of the aortic isthmus: a newly recognized syndrome. Am J Cardiol 1982; 49: 108-116.
7. Greenway SC, Bradley TJ, Caldarone CA, Silverman NH, Hanley FL, Smallhorn JF. Aortopulmonary window in association with anomalous origin of the right coronary artery from the pulmonary artery. Eur J Echocardiography 2006; 7: 379-382.
8. Casillas JA, De Leon JP. Aortopulmonary window with anomalous origin of the right coronary artery from the pulmonary trunk. Texas Heart Inst J 1986; 13: 325-331.
9. Chopra PS, Reed WH, Wilson AD, Rao PS. Delayed presentation of anomalous circumflex coronary artery arising from pulmonary artery following repair of aortopulmonary window in infancy. Chest 1994; 106: 20-22.
10. Hamilton DI, Ghosh PK, Donnelly RJ. An operation for anomalous origin of left coronary artery. Br Heart J 1979; 41: 121-124.
11. Takeuchi S, Imamura H, Katsumoto K, et al. New surgical method for repair of anomalous left coronary artery from pulmonary artery. J Thorac Cardiovasc Surg 1979; 78: 7-11.