Anomalous Left Coronary Artery from Pulmonary Artery presenting associated with Isolated Cleft Palate

Asadollah Tanasan[1], Ahmad Jamei Khosroshaei[2], Farshad rostampour[3], Poya Tanasan[4]

1-Department of Pediatric, Faculty of Medicine, Hamadan University of Medical Sciences and Health Services, Hamadan, Iran
2-Department of Pediatric, Faculty of Medicine, Tabriz University of Medical Sciences and Health Services, Tabriz, Iran
3-Student Research Committee (SRC), Faculty of Medicine, Hamadan University of Medical Sciences and Health Services, Hamadan, Iran
4-Medical Student, Faculty of Medicine, Tehran University of Medical Sciences and Health Services, Tehran, Iran
Corresponding author
Farshad Rostampour**,** Student Research Committee (SRC), Faculty of Medicine, Hamadan University of Medical Sciences and Health Services, Hamadan, Iran. farshad.rostampour@yahoo.com

Abstract: Anomalous Left Coronary Artery from the pulmonary artery (ALCAPA) is a rare congenital cardiac anomaly accounting for approximately 0.25%-0.5% of all congenital heart diseases. This anomaly is usually isolated, but has been associated with patent ductus arteriosus (PDA), ventricular septal defect (VSD), tetralogy of fallot (TOF) or coarctation of the aorta (COA). This anomaly hasn't been associated with extracardiac anomaly.Here we reported a 2-year of old male with cleft palate and the ALCAPA, who presented with cardiomegaly and patent ductus arteriosus. As the early diagnosis of ALCAPA is important, we suggest this association consider in the cleft palate population with congenital heart disease. AS the delay of ALCAPA diagnosis may be associated to elevated mortality and morbidity of cleft palate repair, we suggest ALCAPA association was carefully searched in the cleft palate population

Key words: *Anomalous left coronary artery, pulmonary artery, patent ducts arteriosus, cleft palate.*

Introduction

Cleft lip and cleft palate are common congenital defects, occurring alone or in combination with other malformations in approximately 1.36 per 1000 live birth (1). Some studies was done on the prevalence of congenital heart disease in patients with cleft lips (1, 2). The prevalence of congenital heart disease in these patients was 6.9 % in one study (1). Anomalous Left Coronary Artery from the pulmonary artery (ALCAPA) is a rare congenital cardiac anomaly accounting for approximately 0.25%-0.5% of all congenital heart diseases (3). The mortality of untreated ALCAPA has been estimated to be more than 90% during first year of life (4). Also ALCAPA may be an important cause of malignant ventricular arrhythmias and sudden cardiac arrest in teenagers and adults (5). This anomaly is usually isolated, but has been associated with patent ductus arteriosus (PDA), ventricular septal defect (VSD), tetralogy of fallot (TOF) or coarctation of the aorta (COA) (6). This anomaly hasn't been associated with extracardiac anomaly. We want to report a case of anomalous left coronary artery from pulmonary artery presenting associated with isolated cleft palate that has been not reported until now.

Case presentation

The patient is a 2-year-old male infant referred to Hamadan Besat hospital with cardiac murmur. He had holosystolic grade 2/6 murmur at the apex radiating to the axillary area and continuous murmur 3/6 at left sternal border (LSB). He had cleft palate (Fig1) and his growth indices indicated no failure to thrive (FTT). Electrocardiogram (ECG) showed abnormal Q wave in lead I, aVL and inverted T wave in precordial leads V4 to V6. Mild cardiomegaly without passive congestion were evident in chest x – ray (CXR). Echocardiography showed LA and LV enlargement, mild to moderate mitral regurgitation and small patent ductus arteriosus and abnormal ascending flow were seen within the ventricular septum in four chamber and five chamber views. The left ventricular ejection fraction (LVEF) was 50%. Cardiac angiography was done and revealed patent ductus arteriosus and anomalous origin of the left coronary from the main pulmonary artery (ALCAPA) associated to multiple collaterals between the right and left coronary artery (Fig 2). The patient was referred for re implantation of left coronary artery to the aorta. The surgical re implantation was done

successfully and the patient was candidate for cleft palate repair in the future.

Figure1: Cleft palate shown in this figure.

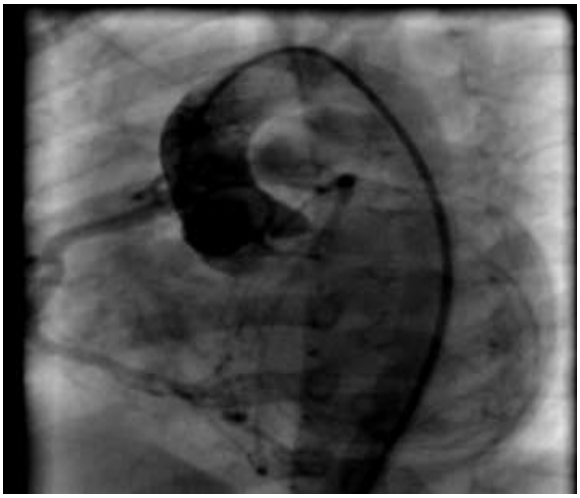

Figure2: In angiography, patent ductus arteriosus and anomalous origin of the left coronary from the main pulmonary artery (ALCAPA) associated to multiple collaterals between the right and left coronary artery revealed.

DISCUSSION

The ALCAPA is a rare congenital cardiac anomaly, accounting for approximately 0.25%-0.5% of all congenital heart diseases (1). This anomaly is usually isolated, but has been associated with patent ductus arteriosus, ventricular septal defect, tetralogy of fallot, or coarctation of the aorta (6). Immediately after birth, as the pulmonary arterial pressure decreases, the perfusion pressure to the left coronary artery gradually falls and myocardial ischemia occurs. On the other hand, as the left-to-right shunt from the left coronary circulation to pulmonary artery increases, Left ventricular myocardial perfusion becomes dependent on the intercoronary collaterals from the right coronary artery (7, 8). Coronary circulation in ALCAPA is two type: infantile and adult type. In the infantile type, the collateral circulation is poorly developed; therefore, presentation of ischemia or infarction and mitral regurgitation is early. In adult type, the collateral circulation is better and the patient presents later in life. 80-90% incidence of sudden death at the mean age of 35 years in this group, estimated (9). The gold standard diagnostic method is cardiac angiography. ALCAPA doesn't associated with extracardiac anomaly (10). Siegel reported 11 patients with congenital heart disease among 110 patients (11%) with facial cleft (11), but in one study in the Tufts-New England Medical center, the overall prevalence rate of CHD was 6.7% among facial clefts and the prevalence of CHD ranged from 0% in the patients with isolated cleft lip to 16.7% in the patients with cleft palate and syndrome diagnosis (1). There is no report of ALCAPA among the facial cleft population. This idea that between ALCAPA and facial cleft anomaly may be relationship, needs more search. Although the timing and type of surgical management for the ALCAPA has remained controversial, the most now agree that the surgical re implantation of left coronary artery is preferable (12-13). AS the delay of ALCAPA diagnosis may be associated to elevated mortality and morbidity of cleft palate repair, we suggest ALCAPA association was carefully searched in the cleft palate population.

References:

1. Geis N, Seto B, Bartoshesky L, Lewis M, Pashayan H. The prevalence of congenital heart disease among the population Metropolitan cleft lip and palate clinic. Cleft palate Journal 1981; 18(1): 19-23.
2. Cohn M. Syndromes with cleft lip and cleft palate. Cleft palate Journal 1978; 15(4): 306-328.
3. Keith JD. The anomalous origin of the left coronary artery from the pulmonary artery. Br Heart J. 1959; 21:149–161.
4. Wesselhoeft H, Fawcett JS, Johnson AL. Anomalous origin of the left coronary from the pulmo-nary trunk. Its clinical spectrum, pathology, and patho-physiology, based on a review of 140 cases with seven further cases. Circulation 1968; 38: 403-425
5. Orem C, Kiriş A, Korkmaz L, Oztürk S, Kahraman N, Koşucu P, Karaci AR, Celik, S. Adult-type anomalous origin of the left coronary artery from the main pulmonary artery: One case report. Echocardiogra-phy 2009; 26: 1232-1235. doi:10.1111/j.1540-8175.2009.00977.x
6. Moss and Adams, Heart Disease in Infants, Children, and Adolescents; 7th edition, page: 731-49.
7. Khatami AD, Mavroudis C, Backer CL. Anomalous origin of left coronary artery from pulmonary artery: collective review of surgical therapy, Ann Thorac surg 2002 ; 74 : 946-55
8. Raghuram AR , Krishnan R , Kumar S , Balamurugan K , Anand ND. Anomalous left Coronary artery from pulmonary artery (ALCAPA) in an adult. IJTCVS 2004; 20:213 -215
9. Benjamin W. Eidem, Frank Cetta, Patrick W. Oleary, Lippincott Wiliams & Wilkins, a wolters Kluwer business, Philadelphia .Echocardiography in Pediatric and Adult Congenital Heart Disease 2010, Michele A.Frommelt. Peter C .Frommelt, vascular abnormality page 345-346.
10. Dodege-Khatami A, Mavoroudis C, Backer C. Anomalous Left Coronary Artery from the pulmonary artery collective review of surgical therapy . Ann Thorac surg 2002; 74: 946-955.
11. Siegel B. A racial comparison of cleft patients in a clinic population: Associated anomalies and recurrence rates, Cleft palate Journal,April 1979 ;16(2),193-197.
12. Arciniegas E,Farooki ZQ, Hakimi M, Green EW: Management of anomalous left coronary artery from the pulmonary artery .Circulation 1980;62(1):180-189.
13. Paridon SM, Farooki ZQ, Kuhns LR, Arciniegas E, Pinsky WW. Exercise performance after repair of anomalous origin of the left coronary artery from the pulmonary artery .Circulation 1990; 81:1287-1292.