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Association of Significant Mitral Regurgitation and Left Ventricular Dysfunction With ALCAPA Syndrome in a Young Patient

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Abstract

Anomalous origin of the left coronary artery from pulmonary artery (ALCAPA) is not a common anomaly in adulthood. Its early diagnosis requires physician suspicion and the early treatment of disease can prevent its serious side effects. In this article, we presented a young female with pansystolic murmur and heart failure with final diagnosis of ALCAPA syndrome.
Keywords: ALCAPA syndrome, Mitral regurgitation, Large right coronary artery.

Introduction

For first time in 1882, Brooks reported left main coronary artery originating from pulmonary artery (ALCAPA) (1). The clinical characteristics of ALCAPA was described in 1933 by Blandet al (2).

ALCAPA syndrome is not a common disease. The incidence of this rare, life-threatening disease is 1 in 300 000 live births (3-5). Without early diagnosis, most infants die 1 year after their birth because of myocardial infarction and heart failure. Arrhythmia, heart failure, and myocardial ischemia are life-threatening factors; they can result in death in adulthood (6).

Case Report

An 18-year-old female had been admitted to the hospital with progressive exertional dyspnea and palpitation since 2 years ago. She came with dyspnea function class III. On physical examination, a pansystolic murmur of grade III/VI was detected in left sternal border. Her blood pressure was 100/70 and she had sinus tachycardia (heart rate = 110). Transthoracic echocardiography showed moderate to severe mitral regurgitation, mildly reduced left ventricular ejection fraction (LVEF = 45%–50%) and turbulent jet in color doppler study across ventricular septum. Because of low LVEF and dyspnea, aortography was performed on the patient which typically showed a large right coronary artery (RCA) but left coronary ostium was not found in the left aortic sinus. During the late phase of the angiography, left anterior descending (LAD) and left circumflex (LCX) arteries were filled retrograde by collateral circulation from the RCA branches, and in later sequences, retrograde flow from the LAD and LCX arteries opacified the left main coronary artery (LMCA) and the main pulmonary artery (Figure 1). Hence, coronary angiography confirmed the diagnosis of ALCAPA syndrome and the reimplantation of LMCA into the ascending aorta was done for patient. Mitral valve was also repaired. Immediately after operation, the symptoms relieved and after 7 days, coronary angiography confirmed the patency of the left main coronary artery (Figure 2).

Discussion

In ALCAPA, myocardial ischemia and acute myocardial infarction are described as results of coronary blood steal and low oxygenated blood in the left myocardial tissue. The lower blood oxygen level in LCX and LAD enhances collateral vessel formation from RCA to the left system. Increased blood flow leads to the RCA dilatation (Figure 3). Hence, continuous blood flow from RCA into LCA and from LCA into low pressure pulmonary artery results in coronary steal phenomenon. Papillary muscle and LV lateral wall dysfunction induced by chronic ischemia results in mitral regurgitation (4). After birth, continuous drop of pulmonary artery pressure enhances reversed left coronary artery flow into pulmonary artery and this process can lead to severe myocardial ischemia, hemodynamic instability, and sudden cardiac death (5, 6).

In 85% of cases, patients start to have the symptoms of disease around 2 months of age when severe left sided heart insufficiency leads to dyspnea and tachycardia and feeding difficulties (7). However, adequate LCA blood supply from RCA postpones the onset of symptoms. In survived adult patients, the presentation of disease ranges from exertional chest pain and dyspnea to life threatening

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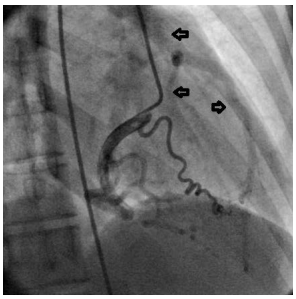

Figure 1. Retrograde flow fills LAD and LCX and subsequently left main coronary artery arises.

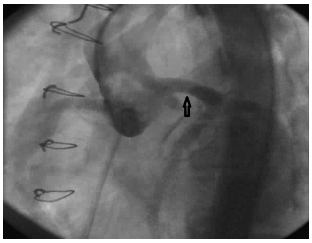

Figure 2. Patient's Left Main Coronary Artery After Surgery.

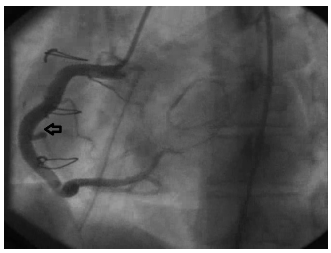

Figure 3. Right Coronary Artery (RCA) Dilatation in ALCAPA Syndrome.

conditions such as sudden cardiac death or malignant ventricular arrhythmias (8). Hence, due to its high mortality rate (about 90% in patients with a mean age of 35 years), surgical correction must be done as soon as possible after the diagnosis of disease (9). When the diagnosis and treatment of disease are performed early after birth, normalization of both ventricular function and mitral valve regurgitation occur in many cases (10). Although we have restricted data about long term prognosis of adult patients after surgical correction, recently it has been shown a 94.8% survival rate for infants in 20 years (11). Generally the prognosis after surgical correction in adults is good. Although in the case described in this study, mitral valve repair was done during surgery, conservative treatment is often preferred when mitral valve function improves after correction of left ventricular circulation and ischemia.

The late outcome after surgery mainly depends on the extent of irreversible myocardial scar tissue.

Conclusion

Heart failure with concomitant mitral regurgitation or arrhythmias in adult patients without ischemic heart disease should be evaluated for signs of ALCAPA syndrome.

Conflict of Interests

Authors declare that they have no conflict of interests.

Ethical Issues

The study protocol was approved by the institutional ethics committee of Tabriz University of medical sciences. The patient provided written informed consent for participation.

References

1. Brooks HS. Two cases of an abnormal coronary artery of the heart arising from the pulmonary artery: with some remarks upon the effect of this anomaly in producing cirsoid dilatation of the vessels. J Anat Physiol. 1885;20(Pt 1):26-29.
2. Bland EF, White PD, Garland J. Congenital anomalies of the coronary arteries: Report of an unusual case associated with cardiac hypertrophy. Am Heart J. 1933;8(6):787-801. doi:10.1016/S0002-8703(33)90140-4.
3. Dodge-Khatami A, Mavroudis C, Backer CL. Anomalous origin of the left coronary artery from the pulmonary artery: collective review of surgical therapy. Ann Thorac Surg. 2002;74(3):946-955.
4. Hauser M. Congenital anomalies of the coronary arteries. Heart. 2005;91(9):1240-1245. doi:10.1136/hrt.2004.057299
5. Peña E, Nguyen ET, Merchant N, Dennie C. ALCAPA Syndrome: Not Just a Pediatric Disease. RadioGraphics. 2009;29(2):553-565. doi:10.1148/rg.292085059.
6. Safaa AM, Du LL, Batra R, Essack N. A rare case of adult type ALCAPA syndrome: presentation, diagnosis and management. Heart Lung Circ. 2013;22(6):444-446. doi:10.1016/j.hlc.2012.10.011
7. Aiyappan SK, Ranga U. Anomalous origin of left coronary artery in an adult. Indian J Med Res. 2013;138(4):566-567.
8. Kristensen T, Kofoed KF, Helqvist S, Helvind M, Sondergaard L. Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) presenting with ventricular fibrillation in an adult: a case report. J Cardiothorac Surg. 2008;3:33. doi:10.1186/1749-8090-3-33
9. Wilson CL, Dlabal PW, Holeyfield RW, Akins CW, Knauf DG. Anomalous origin of left coronary artery from pulmonary artery. Case report and review of literature concerning teen-agers and adults. J Thorac Cardiovasc Surg. 1977;73(6):887-893.
10. Cabrera AG, Chen DW, Pignatelli RH, et al. Outcomes of anomalous left coronary artery from pulmonary artery repair: beyond normal function. Ann Thorac Surg. 2015;99(4):1342-1347. doi:10.1016/j.athoracsur.2014.12.035
11. Lange R, Vogt M, Horer J, et al. Long-term results of repair of anomalous origin of the left coronary artery from the pulmonary artery. Ann Thorac Surg. 2007;83(4):1463-1471. doi:10.1016/j.athoracsur.2006.11.005