

18 years old patient with Bland White Garland syndrome after non-ST segment elevation myocardial infarction (RCD code: I-1C.3)

Natalia Dłużniewska^{1*}, Jakub Podolec², Magdalena Kostkiewicz¹, Lidia Tomkiewicz-Pająk¹, Maria Olszowska¹, Piotr Podolec¹

¹ Department of Cardiac and Vascular Disease, Jagiellonian University College of Medicine, John Paul II Hospital, Krakow, Poland; ² Department of Interventional Cardiology, Jagiellonian University College of Medicine, John Paul II Hospital, Krakow, Poland

Abstract

49 year-old patient with multiple myeloma diagnosed a year earlier, after a few cycles of chemotherapy was admitted to hospital with heart failure symptoms accompanied by a rapid atrial flutter. Echocardiography revealed eccentric hypertrophy of the left ventricle and reduced ejection fraction approx. 20% and non sustained ventricular tachycardia episodes in the Holter ECG. Due to the gradually deteriorating patient's condition electrical cardioversion was performed restoring sinus rhythm. Applied pharmacological treatment brought relief of symptoms reported at admission. To clarify the etiology of heart failure coronary angiography was performed excluding the presence of coronary artery disease. Because of the frequent occurrence of amyloidosis in patients with multiple myeloma, the amyloid cardiomyopathy was suspected and the investigation in that direction begun. Histology samples abdominal fat and gums mucous in a hematoxylin and eosin and Sirius red staining showed no signs of amyloidosis. However, bone marrow biopsy revealed the presence of protein corresponding to the extracellular amyloid deposits. The patient died a few months later in hematology clinic due to severe multiple organ failure associated with generalized amyloidosis. JRC D 2015; 2 (2): 60–63

Key words: amyloidosis, heart failure, myeloma multiplex

Introduction

According to the literature, coronary anomalies affect ca. 1% of the general population [1] and occur in 1,3% of patients undergoing coronary angiography and in 0,3% of autopsies [2]. 87% of those have anomalous origin or distribution and 13% are described as coronary artery fistulae [3]. Different types of classifications have been published in the literature. Some authors prefer to categorize them as “major,” “hemodynamically significant” anomalies versus “minor” ones. Angelini et al. conclude that a classification of coronary anomalies should consider all possible coronary anatomic variations independently from the clinical and hemodynamic measurements [4]. The majority of the anomalies are considered to be benign and include:

1. separate origin of left anterior descending and left circumflex from the left sinus of Valsalva;

2. ectopic origin of the circumflex from the right sinus of Valsalva;
3. ectopic coronary origin from the posterior sinus of Valsalva;
4. ectopic coronary origin from the posterior sinus of Valsalva;
5. ectopic coronary origin from the ascending aorta;
6. absent circumflex;
7. intercoronary communication;
8. small size coronary artery fistulae [3].

In 19% of those patients anomalies are symptomatic and may present with serious clinical events. The most events occur in patients with:

1. ectopic coronary origin from the pulmonary artery;
2. ectopic coronary origin from the opposite aortic sinus;
3. single coronary artery;
4. large coronary artery fistulae [2, 3].

Bland – White – Garland syndrome is a rare coronary anomaly including anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). It occurs in 0,2–0,5% of all congenital

Conflict of interest: none declared.

* Corresponding author: Department of Cardiac and Vascular Diseases, Institute of Cardiology, Jagiellonian University Medical College, Centre for Rare Cardiovascular Diseases, John Paul II Hospital, Krakow, Poland, Pradnicka str. 80, 31-202 Krakow, Poland; tel. 0048 12 614 22 87, fax 0048 12 423 43 76; email: natalia.dluzniewska@uj.edu.pl

Copyright © 2015 Journal of Rare Cardiovascular Diseases; Fundacja Dla Serca w Krakowie

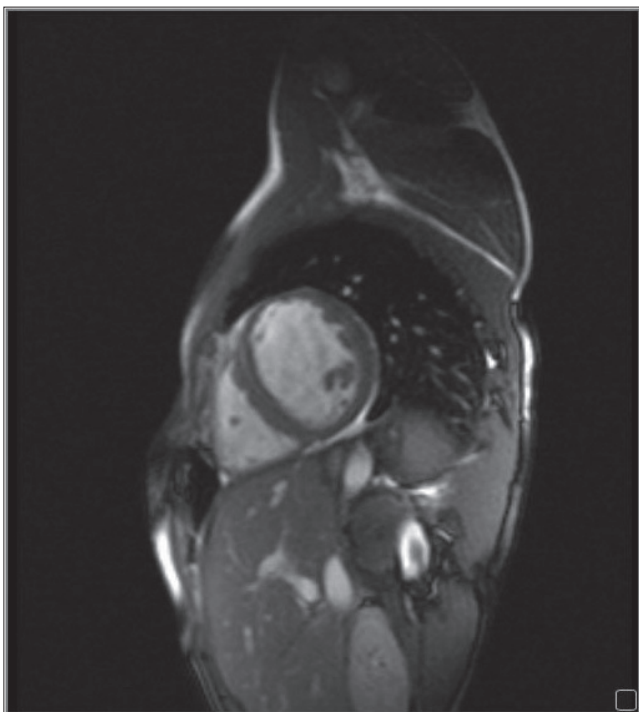


Figure 1. Magnetic Resonance Imaging. Remodeling of the left ventricle in mid and apical segments of anterior wall

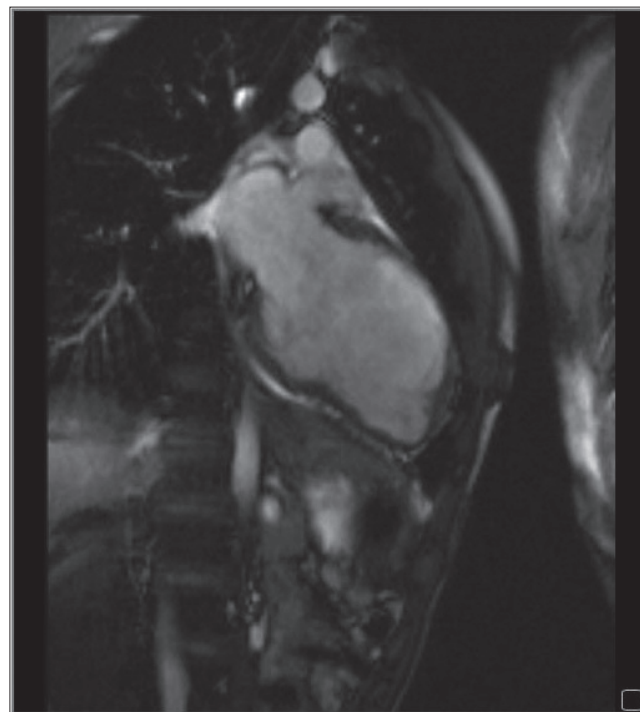


Figure 2. Magnetic Resonance Imaging. Left ventricular wall enlargement

cardiac malformations. Although it has high mortality rate of 90% within the newborn children. Ca. 10–15% of patients reach adulthood. In most cases the existence of extensive coronary collaterals is increasing their chances to survive [5,6,7]. Usually adult patients with ALCAPA are asymptomatic.

Case presentation

We present a case of young 18 years old man suffering an episode of non ST – segment elevation myocardial infarction with Bland White Garland syndrome in march 2006. The patient was admitted to the Department of Cardiovascular Surgery and Transplantation at the Collegium Medicum Jagiellonian University due to cardiac catheterization before the surgical correction of the Bland White Garland syndrome.

No past medical history and no risk factors of cardio-vascular disease were noticed before. The electrocardiogram demonstrated sinus rhythm with heart rate of 70 beats per minute, left axis deviation, pathological Q waves waves in leads V5-V6, negative T wave in aVL, ST segment depression in I, aVL, V4-V6 and left bundle branch block. The cardiac catheterization showed myocardial bridge over the left anterior descending artery (LAD) with 25% lumen reduction during systole, 30% stenosis in the middle part and anomalous artery originating from the pulmonary artery connecting the LAD to the pulmonary artery. Collateral circulation to LAD and left circumflex artery (Cx) is provided form the right coronary artery. Venticulography showed enlargement of the left ventricle and akinesis of the antero-lateral segment. Ejection fraction measured in ventriculography was 40%. Due to ambiguous picture of

the origin of the LAD and Cx computed tomography and magnetic resonance were indicated. Magnetic resonance was done in the University Children Hospital in Cracow and showed remodeling of the left ventricle in mid and apical segments of anterior wall. (Figure 1). Left ventricle wall enlargement was also observed (Figure 2). The parameters measured during magnetic resonance included end-diastolic volume (EDV) of 185 ml end-systolic volume (ESV) 131 ml, systolic volume (SV) 54 ml, and LV mass of 174 gr. The angio-CT showed anomalous origin of the left coronary artery from the pulmonary artery (Figure 3 and 4). Left coronary artery divides into small recessive circumflex branch and strong wide left anterior descending artery, which creates fistulae to the right ventricle in two places (diameter of the fistulae is below 1 mm). Collateral circulation is provided from right coronary artery. Additionally the angio-CT showed remodeling of left ventricle which corresponding to the episodes of ischemia of the anterior wall, aneurism of the apex and middle segment of thin anterior wall (<3 mm, no systolic thickening of wall with dyskinetic move). On the 09th of May 2007 the patient underwent surgery which consisted of re-location of the origin of the left coronary artery from the pulmonary artery to the ascending aorta. Echocardiography performed one week after the procedure showed significant increase of the ejection fraction (EF) to 65%. The patient was asymptomatic and discharged to the Rehabilitation Department in good condition. Currently, after almost 8 years of surgical treatment the patient's general condition is very good. The patient is asymptomatic with good exercise tolerance. Routine echocardiographic and holter EKG monitoring each year shows no abnormalities with preserved EF.

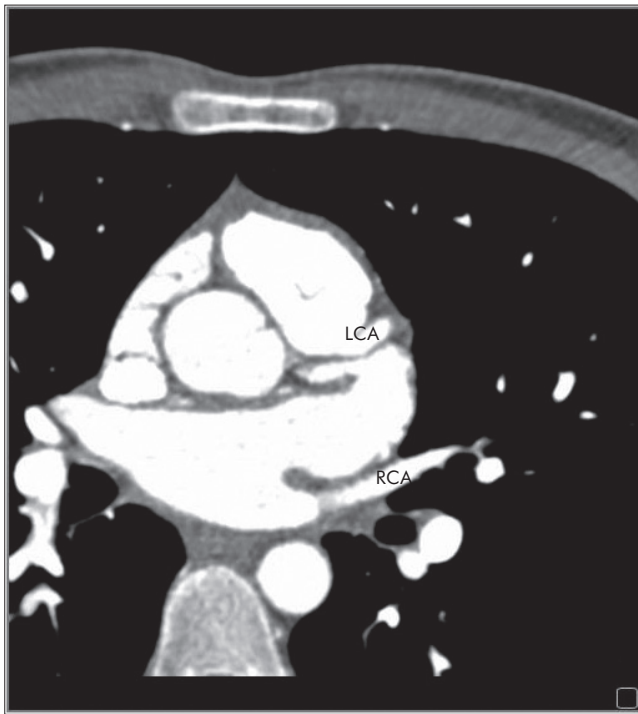


Figure 3. Computed tomography. The anomalous left coronary artery (LCA) with pulmonary artery origin. Right coronary artery (RCA) originates from aorta

Discussion

An anomalous origin of one coronary artery from the pulmonary artery is usually an isolated anomaly, which occurs in 0,4% of patients with congenital heart diseases and is usually discovered during childhood. The most common defect is the anomalous origin of the left coronary artery from the pulmonary artery and in adults without congenital cardiovascular diseases is very rare [8]. The diagnostic process is difficult and often resulting in accidental findings. The minority of those patients suffer serious clinical episodes including myocardial ischemia, infarction, arrhythmias and sudden cardiac death [5, 9]. Symptoms and prognosis are dependent on the extension of collateral circulation [10]. Definite diagnosis is often made during conventional coronary angiography [5], however recent advances of imaging techniques such as computed tomography and magnetic resonance imaging are becoming helpful noninvasive techniques. CT offers better spatial resolution but MRI can potentially provide additional information on myocardial viability and perfusion [11]. However, in case of coronary anomalies, angiography should be performed in order to exclude additional atherosclerosis. Our case report shows that non-invasive examinations as much as invasive procedures provide useful information to complete diagnosis of Bland White Garland [11]. Patients with Bland White Garland syndrome need to be treated surgically [12]. It is recommended in all patients at the time of diagnosis, regardless of age or symptoms [13]. Studies known from literature show immediate clinical improvement after surgery and normalization of left ventricle function should be expected [13,14].

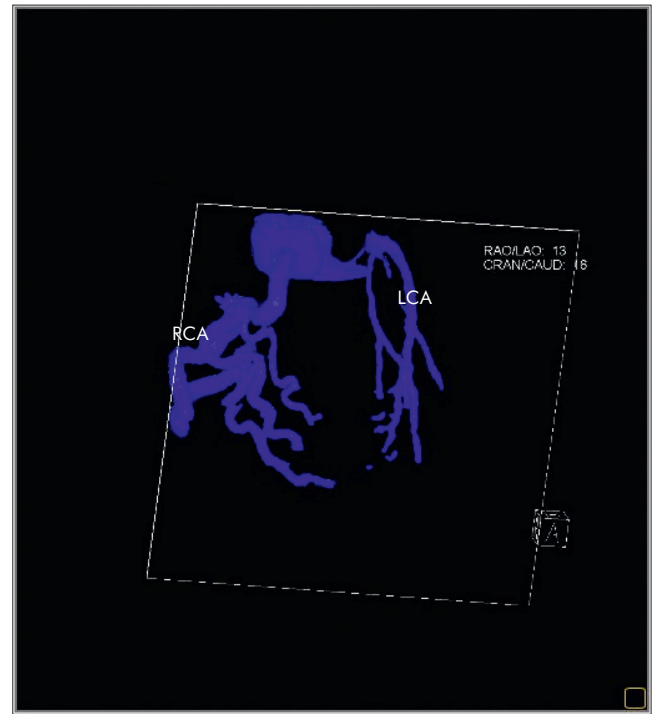


Figure 4. Computed tomography. Reconstruction of the anomalous left coronary artery (LCA) with pulmonary artery origin. Right coronary artery (RCA) originates from aorta

Conclusions

Bland White Garland syndrome is a rare anomaly among the adults and may present with serious clinical presentation. Although the diagnosis is usually made during cardiac catheterization, MRI and CT can provide a comprehensive assessment of the anatomical and functional consequences of coronary artery disease. In most patients surgical intervention should be considered as a safe option, which significantly reduces symptoms and risk of acute coronary episodes re-occurrence.

References

1. Angelini P, Velasco JA, Flamm S. Coronary Anomalies Incidence, Pathophysiology, and Clinical Relevance. *Circulation* 2002; 105: 2449–2454.
2. Zemanek D, Veselka J, Kautznerova D, Tesar D. The anomalous origin of the left coronary artery from the right aortic sinus: is the coronary angiography still a 'gold standard'?. *The International Journal of Cardiovascular Imaging* 2006; 22:1 27–133.
3. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn.* 1990; 21: 28–40.
4. Angelini P. Coronary Artery Anomalies An Entity in Search of an Identity. *Circulation* 2007; 115: 1296–1305.
5. Kim B, Park Y, Hong S et al. Anomalous origin of the left coronary artery from the pulmonary artery initially visualized by echocardiography and multidetector computed tomography coronary angiography. *J Cardiovasc Ultrasound* 2012; 20: 197–200.
6. Kumar RV, Rama Krishna LS, Ramachandran G. Anomalous origin of the left coronary artery arising from the pulmonary artery: Reverse remodeling of coronary arterie following translocation operations. *Indian J Thorac Cardiovasc Surg* 2012; 28: 151–152.

7. Neony P, Beaune J, Champsaur G et al. Anomalous Origin of Left Coronary Artery From the Evolution of Left Ventricular Function and Perfusion 44-Year-Old Man Pulmonary Artery: After Surgery in a 44-Year-Old Man. *Clin. Cardiol.* 1992; 15: 466–468.
8. Bland EF, White PD, Garland J. Congenital anomalies of the coronary arteries: report of unusual case associated with cardiac hypertrophy. *Am Heart Journal* 1933; 8: 787.
9. Acikel S, Dogan M, Turkvatan A et al. Single left coronary artery and coronary sinus opening anomaly in patient presenting with cardiac symptoms. *Canadian Journal of Cardiology* 2012; 28: 612.e1.–612.e3.
10. Blaghith M. Anomalous origin of the right coronary artery from the proximal left anterior descending artery and a single coronary artery anomaly: Three case reports. *Journal Saudi Heart Assoc.* 2013; 25: 43–46.
11. Korosoglou G, Ringwald G, Giannitsis E, Katus H. Anomalous origin of the left circumflex coronary artery from the pulmonary artery. A very rare congenital anomaly in an adult patient diagnosed by cardiovascular magnetic resonance. *Journal of Cardiovascular Magnetic Resonance* 2008; 10: 4.
12. Haponiuk I, Rycaj J, Grzybowski A et al. Leczenie operacyjne zespołu Blanda – White’a – Garlanda u noworodka z rozległym pozawałowym uszkodzeniem lewej komory. Czy korekcja jest bardziej korzystna od transplantacji serca? *Folia Cardiol.* 2004; 11: 687–696.
13. Kuroczynski W, Kampmann C, Kayhan N et al. Anomalous origin of the left coronary artery from the pulmonary artery: mid-term results after surgical correction. *Clin Res Cardiol.* 2008; 97: 266–271.
14. Amaral F, Carvalho JS, Granzotti JA, Shinebourne EA. Anomalous Origin of the Left Coronary Artery from the Pulmonary Trunk. Clinical Features and Midterm Results after Surgical Treatment. *Arq Bras Cardiol.* 1999; 72: 314–320.