Left main coronary artery atresia

Atrezja pnia lewej tętnicy wieńcowej

Michał Tomaszewski^{1, 2}, Jarosław Wójcik¹, Andrzej Tomaszewski¹, Elżbieta Czekajska-Chehab³, Andrzej Madejczyk¹, Barbara Zimoń¹

Abstract

We present a report of an extremely rare case of left main coronary artery atresia in a 33 year-old male with severe symptoms of angina pectoris and dyspnoea. During coronary angiography, the left coronary artery (LCA) ostium could not be catheterised. The right coronary artery (RCA) was dilated, and the LCA was filling retrogradely via collateral vessels. The diagnosis was confirmed by multislice computed tomography. Due to severe symptoms and the high risk of sudden cardiac death, the patient was referred for coronary artery revascularisation. The left internal mammary artery was anastomosed to the left anterior descending artery. The operation and subsequent 24 months follow-up were uneventful.

Key words: left main coronary artery atresia, coronary artery anomaly

Kardiol Pol 2012; 70, 3: 275-276

CASE REPORT

A 33 year-old male was admitted with severe symptoms of angina pectoris and dyspnoea. The coronary angiography was performed. The left coronary artery (LCA) ostium could not be catheterised (Fig. 1). The right coronary artery (RCA) was dominant and the LCA was filling retrogradely via collateral vessels (Fig. 2).

The patient was referred for multislice computed tomography (MSCT). The orifice of the left main coronary artery (LMCA) from the left coronary sinus was not visible (Fig. 3). The pulmonary trunk was dilated to 55 mm in the subbulbar segment. The shunt to the pulmonary artery was excluded in MSCT.

Transthoracic and transoesophageal echocardiography was performed. Cardiac chambers were normal in diameter. The ejection fraction was 60%. There was also mild pulmonary artery stenosis with maximal gradient 28 mm Hg and dilation of pulmonary trunk to about 5 cm. Patent foramen ovale with insignificant left to right flow was found. There was hypokinesis of the basal segment of the interventricular septum and hypokinesis of the anterior wall. A dobutamine stress echo showed improved contractility of the above mentioned segments.

Due to severe symptoms and the high risk of sudden cardiac death (SCD), the patient was referred for coronary artery revascularisation. During the operation, the absence of a LMCA orifice was confirmed. The left internal mammary artery was anastomosed to the left anterior descending artery. The operation and the postoperative course were uneventful. The patient was discharged on the seventh postoperative day and sent for early cardiac rehabilitation. The 24 months follow-up was uneventful. The patient remained asymptomatic.

DISCUSSION

The incidence of coronary anomalies is low in the general population, ranging between 0.46% and 1.55% [1]. In a review of 126,595 patients undergoing coronary angiography over a 28 year period, coronary anomalies were found in 1,686 (1.3%) patients. Most of these patients, 1,461 (87%), had anomalies of origin and distribution, and 225 (13%) had coronary artery fistulae. None of the reviewed patients had LMCA atresia [2].

The LMCA atresia is an extremely rare abnormality. Only 26 cases had been reported in the literature by 1997 accor-

Address for correspondence:

Michał Tomaszewski, MD, PhD, Department of Cardiology, Medical University of Lublin, ul. Jaczewskiego 8, 20–954 Lublin, Poland,

tel/fax: +48 81 724 41 51, e-mail: mdtomaszewski@yahoo.com

Received: 13.02.2011 **Accepted:** 27.04.2011 Copyright © Polskie Towarzystwo Kardiologiczne

¹Department of Cardiology, Medical University of Lublin, Poland

²Chair of Internal Medicine and Department of Internal Medicine in Nursing, Medical University of Lublin, Poland

³1st Department of Radiology, Medical University of Lublin, Poland



Figure 1. Well-developed collateral arising from right coronary artery to left coronary artery (RAO 30 projection)

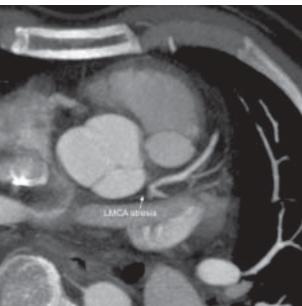


Figure 3. Multislice computed tomography confirms left main coronary artery (LMCA) atresia



Figure 2. Aortography visualises the lack of left main coronary artery orifice

ding to Musiani et al. [1]. To the best of our knowledge, our case is the 64th to be published to date.

Most patients are symptomatic. In paediatric patients, the symptoms are usually syncopal attacks, dyspnoea, tachyarrhythmias, failure to thrive, or myocardial infarction. In adult patients, there are mainly angina symptoms. The SCD may occur in both groups [1].

The most probable mechanism contributing to angina is abnormal timing of blood delivery to the LCA. Relatively long

blood passage may lead to its arrival in systole instead of diastole. The systolic coronary compression, especially during exertion, might contribute to myocardial ischaemia. The development of collateral circulation may explain differences in clinical presentation and variability in survival [3]. The release of angina and improvement of left ventricular function after coronary artery by-pass grafting (CABG) is probably due to the surgical restoration of the two coronary systems with the absence of retrograde flow to the LCA [4, 5].

The LMCA atresia has a poor prognosis and medical therapy is not sufficient. Patients with LMCA atresia can be treated successfully with CABG.

CONCLUSIONS

The LMCA atresia is an extremely rare anomaly. Taking into consideration the severe symptoms, the high risk of SCD, and the good mid-term results of CABG, surgical revascularisation should probably be the treatment of choice.

Conflict of interest: none declared

References

- Musiani A, Cernigliaro C, Sansa M, Maselli D, De Gasperis C. Left main coronary artery atresia: literature review and therapeutical considerations. Eur J Cardiothorac Surg, 1997; 11: 505–514.
- Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. Cathet Cardiovasc Diagn, 1990; 21: 28–40.
- Debich DE, Williams KE, Anderson RH. Congenital atresia of the orifice of the left coronary artery and its main stem. Int J Cardiol, 1989; 22: 398–404.
- Beretta L, Lemma M, Santoli. Isolated atresia of the left main coronary artery in an adult. Eur J Cardiothorac Surg, 1990; 4: 169–170.
- Vidne BA, Nili M, Aygen M, Levy MJ. Congenital atresia of the left main coronary ostium. Scand J Thorac Cardiovasc Surg, 1979; 13: 37–40.