

Congenital Absence of Left Circumflex Coronary Artery

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Abstract

Congenital absence of left circumflex artery is a rare congenital anomaly of the coronary arteries. The prevalence of the anomaly in different studies ranges from 0.6% to 1.3%. Of these, 80% are benign and asymptomatic and 20% are clinically important. We report a 56-year-old man presented with acute resting chest pain who was diagnosed as having acute anterolateral infarction accompanied by electrocardiographic changes and elevated cardiac enzymes. Coronary angiography in different views was conducted, however, no left circumflex artery was found. The territory supplied by the artery had been perfused by the super dominant right coronary artery. There was no left circumflex coronary artery with anomalous origin. Severe stenosis of left anterior ascending artery superimposed to the absent left circumflex artery was presented as acute anterolateral infarction. Although absence of the artery is mostly considered as a benign condition, atherosclerotic lesions may be more important in such cases because of diminished compensating mechanisms.

Iran J Med Sci 2009; 34(3): 217-219.

Keywords • Circumflex artery • coronary anomaly • congenital anomaly

Introduction

Congenital coronary artery anomalies, infrequently seen during coronary angiographic studies, are mostly diagnosed incidentally. The prevalence in different studies ranges from 0.6% to 1.3%.¹ Of these, 80% are benign and asymptomatic and 20% are clinically important.² Congenital absence of the left circumflex coronary artery (LCX) is an extremely rare vascular anomaly in which the artery fails to develop in the left atrioventricular groove. The prevalence of absent LCX has been shown 0.003% in other studies.³ In previous case reports of absent LCX, there has been an association with systolic click syndrome,⁴ and ischemic changes in the zone of hypoperfusion, which cause chest pain and a poor prognosis related to dilated cardiomyopathy.⁵ Here we described a case of absent LCX with acute myocardial infarction.

Case Report

A 56-year-old man was presented with acute typical resting chest pain for the first time. In medical history, he had hyperlipidemia for 6 years and has been receiving lipid-lowering treatment.

In first physical examination at emergency room his blood pressure was normal and no arrhythmia was seen. Cardiac

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Received: 11 November 2008

Revised: 17 March 2009

Accepted: 23 April 2009

and lung auscultation were normal as well. The initial electrocardiogram showed ST segment elevation in anterior leads (V2-V5) (figure 1). Laboratory evaluation showed serial rise of CPK-MB (51 IU/l – 177 IU/l – 308 IU/l). Transthoracic echocardiography showed mild cardiomegaly with ejection fraction about 35%.

The patient was diagnosed as having acute anterolateral infarction. After initial medical treatment, he underwent coronary angiography using standard right femoral Judkins technique.⁶ The angiogram showed one artery arose from the left sinus of Valsalva as left anterior descending artery (LAD) with severe stenosis of LAD- diagonal bifurcation, and no obvious LCX was demonstrated even after taking several different angiogram views (figure 2). In right coronary artery

(RCA) injection, it was normally originated from the right sinus of Valsalva. It was a super dominant RCA ascended the posterior atrioventricular groove beyond the crux (figure 2). Aortic root angiogram showed no evidence of anomalous origin of the left circumflex coronary artery (figure 2).

According to angiographic data, non-surgical management was primarily considered for the patient. Percutaneous coronary intervention on LAD and diagonal was done with mini crush technique.⁷ Stent Taxus-liberte 3-14 (Boston Scientific, USA) for diagonal and stent 3-18 for LAD were implanted and after final kissing balloon inflation, the LAD lesion was dilated and re-perfusion was achieved. The distal LAD was subsequently visualized but the LCX was not seen again.

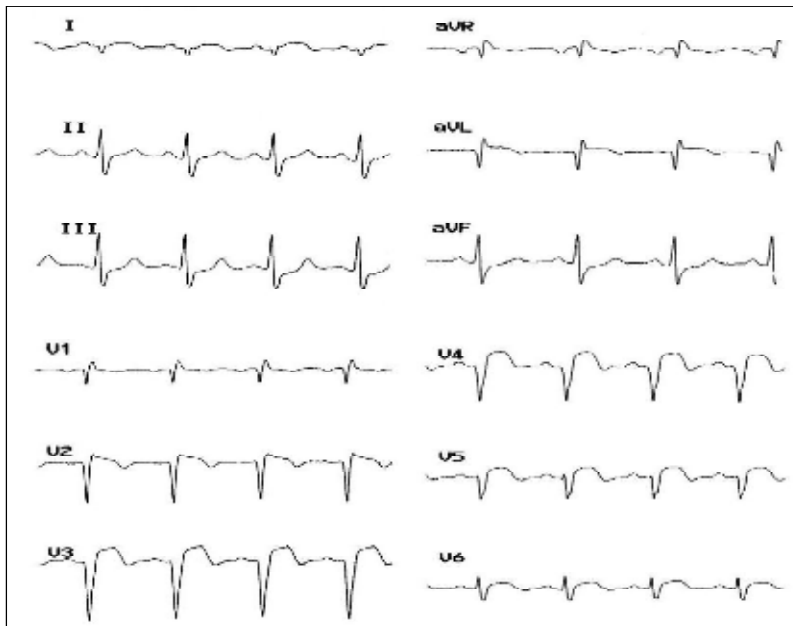


Figure 1: Electrocardiogram on admission showed ST segment elevation in precordial leads (V₂-V₅)

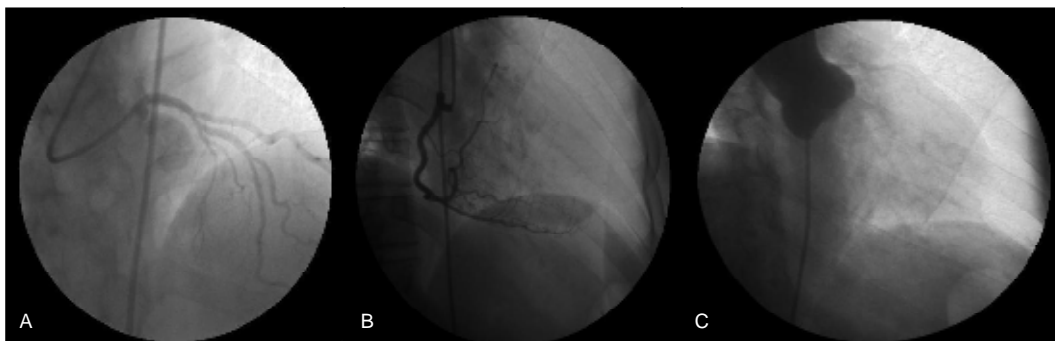


Figure 2: A) Left coronary angiogram in right anterior caudal view. No circumflex artery is demonstrated. B) Right coronary angiogram in right anterior oblique view. The right coronary artery is a super dominant vessel with large branches. Its markedly developed branches are going up to the LCX territories. C) Aortic root angiogram in right anterior oblique view shows no evidence of the left circumflex artery.

Discussion

Coronary artery anomalies are a group of diseases with various severities ranging from mild to life threatening conditions.⁸ Separate origins of the LAD and LCX arteries from the left sinus of Valsalva are the most common anomaly. Absence of LCX is rare,¹ and lack of a coronary artery in the left atrioventricular groove confirms the diagnosis of absent LCX.

Normally, the LCX and RCA run around the atrioventricular groove and form a circle,³ therefore, the absence of LCX is usually compensated with the large super-dominant RCA, crossing the crus and perfusing the LCX territories,⁹ as it was presented in a double LAD pattern in our patient. Because of this anatomical compensating mechanism, the condition is generally thought to have no symptoms with a benign outcome unless it is superimposed by atherosclerotic coronary artery disease.^{8,10}

Mievis has reported myocardial infarction in a 31-year-old man with no obvious coronary artery narrowing because of absent LCX.¹¹ Likewise, severe stenosis of LAD often leads to a fatal outcome, although in the present patient well-developed and super dominant RCA branches that perfused a zone extending to the left ventricular inferior and posterior wall, preserved cardiac function despite lethal stenosis of LAD.

In a case of absent LCX, poorly-developed RCA considered as a subtype of coronary artery hypoplasia syndrome.³

The absence of LCX can be considered mostly as a benign condition. Detailed angiographic evaluation of coronary anomalies may be useful for understanding the clinical course such as chronic stable angina, variant angina, myocardial infarction, and sudden cardiac death. It can also help for selection of the best treatment option. Additionally, atherosclerotic lesions may be more important in such patients because of diminished compensating mechanisms.

Conflict of Interest: None declared

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