
Case Report

Anomalous Origin Of Left Circumflex Coronary Artery From Non-coronary Aortic Sinus: Classification Of Isolated Anomalies Of Coronary Arteries

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An 18-year-old female suffering from severe mixed rheumatic mitral valve disease was operated on for mitral valve replacement. During the operation, an unusual swelling was noticed near the non-coronary sinus of the aorta, which turned out to be the left circumflex coronary artery. From its origin the artery curved around the right side of the aorta and then traversed in the transverse sinus to follow its normal course. The left anterior descending artery was arising from the left coronary sinus and the right coronary artery was arising from the right coronary sinus. This rare congenital anomaly was not suspected preoperatively as the patient did not have any relevant symptoms. Preoperative transthoracic echocardiography also failed to pick up this anomaly. However, postoperative detailed transthoracic echocardiography provided evidence of an abnormal vessel arising from the non-coronary sinus. (Asia Pacific Heart Journal 1998;7(3):225-227)

Introduction

Isolated congenital anomalies of coronary arteries are not commonly found. The reported incidence in patients undergoing coronary angiography is 0.3-1.3%.¹ Despite this low incidence, they remain unnoticed and mostly present in combination with other serious cardiac diseases. The clinical presentation of an isolated anomaly of coronary artery per se depends on its site of origin and course.

Those cases where an anomalous coronary artery takes origin from the pulmonary trunk or has a sharp angular course may present early with ischaemia or frank cardiac failure. Moreover, these patients have an increased risk of sudden cardiac death at an early age. The diagnosis is difficult with non-invasive diagnostic techniques, and coronary angiography is the best diagnostic tool.²

We report a case of an 18-year-old girl who presented with rheumatic mitral valve disease. During surgery an anomalous origin and course of the left circumflex coronary artery was noticed. The course was such that it did not produce ischaemic effects and therefore did not necessitate surgical intervention.

Case Report

An 18-year-old girl was referred to our unit for mitral valve surgery. She had a 4-year history of rheumatic mitral valve disease. The main presenting complaints were exertional dyspnoea and palpitations. She did not have any complaint which would suggest other cardiac pathology. Her routine blood counts and ECG were

within normal limits. Chest radiograph examination showed cardiomegaly with grossly dilated left atrium and pulmonary artery. Echocardiographic results revealed severe mixed mitral valve disease. The mitral valve area was 1.1 cm² and there was severe regurgitation. The subvalvular apparatus was also diseased. Other valves were normal in function. The left ventricle was volume overloaded and dilated with a systolic dimension of 32 mm and diastolic dimension of 54 mm. Pulmonary artery systolic pressure was estimated to be 90 mmHg.

Surgery was performed through a median sternotomy. While making space around the aorta for aortic crossclamp, we noticed a swelling opposite the non-coronary sinus of aorta. This swelling appeared to be an aberrant blood vessel. A gentle and careful dissection revealed it to be the left circumflex coronary artery which was arising from the non-coronary sinus and curving around the aorta to traverse through the transverse sinus and then follow its normal course. The anterior descending artery was independently arising from the left coronary sinus (Fig. 1).

As there was no evidence of ischaemic attacks in the clinical history of the patient and the coronary arteries appeared normal, this anomaly was left alone. We proceeded to the mitral valve replacement with a Starr Edward (2M) prosthesis. The operation was completed as a routine open heart operation, and the patient had an uneventful recovery. She was discharged from hospital on the fifth postoperative day. Postoperative echocardiography was performed which confirmed 3

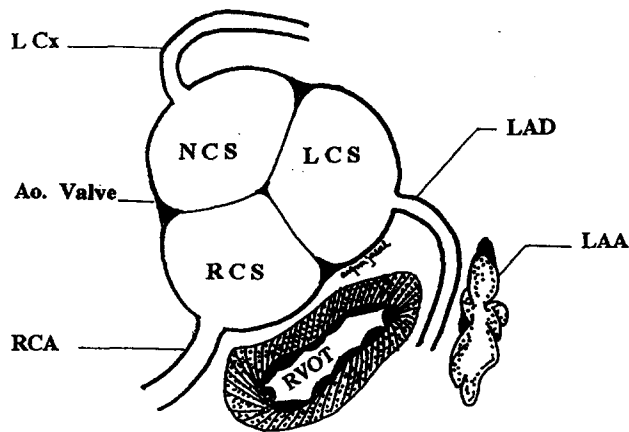


Fig. 1. Diagram shows the origin of the left circumflex coronary artery from non-coronary sinus of valsalva and going retroaortically. The right coronary and left anterior descending arteries are arising normally. Abbreviations: left circumflex coronary artery, LCx; aortic valve, Ao. Valve; right coronary artery, RCA; left anterior descending coronary artery, LAD; left atrial appendage, LAA; non-coronary sinus, NCS; left coronary sinus, LCS; right ventricular outflow tract, RVOT.

vessels arising from the 3 aortic sinuses of valsalva (Fig. 2). The patient's late postoperative period also remained uneventful, and she was found to be in excellent health at a follow-up visit 18 months after the operation.

Discussion

Congenital anomalies of the coronary arteries are found rarely. The preoperative diagnosis is difficult with conventional non-invasive investigations. Echocardiography can identify anomalous origin of a coronary artery from the pulmonary artery, and Doppler colour flow mapping has also been used to detect intercoronary collateral flow as an additional diagnostic sign.³

In our case, the diagnosis was missed on echocardiography. This is not unexpected as, while studying a case of rheumatic mitral valve disease, one usually does not pay much attention to the coronary anatomy. However, when we reviewed the recorded preoperative echocardiograms, there was evidence of an abnormal vessel arising from the non-coronary sinus of valsalva.

Therefore, we did a detailed postoperative echocardiogram which picked up 3 vessels arising from 3 separate sinuses of valsalva. This is an interesting finding because, as far as we know, such an origin of these vessels has never been reported in the literature.

Most cardiology texts have described 5 common forms of coronary artery anomalies. These include origin of the left coronary artery from the pulmonary artery, right coronary artery from the pulmonary artery, left coronary artery from inappropriate aortic sinus, the right coronary artery from the left coronary sinus, the circumflex artery from the right aortic sinus or the right

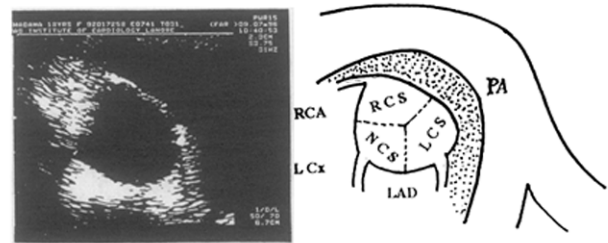


Fig. 2. Postoperative echocardiogram showing 3 coronary arteries arising from 3 separate sinuses of valsalva. Abbreviations: right ventricular outflow tract, RVOT; others as in Fig. 1.

coronary artery and the origin of first septal artery from right coronary sinus or right coronary artery itself.^{1,4}

Moreover, after an abnormal origin, the left coronary artery may follow an abnormal course. Four such abnormal routes have been described in relation to the aorta and pulmonary artery. These are anterior, posterior, interarterial and septal.⁶ The interarterial type poses the most serious risk of ischaemia and sudden death.

In a series of 7,000 coronary arteriograms, Kimbris et al¹ noticed an anomalous aortic origin of coronary arteries in 45 patients (0.64%). The most frequent anomaly was the abnormal origin of the left circumflex coronary artery either from the right coronary sinus or the right coronary artery, which was seen in 26 patients. They noticed associated valvular heart disease in 14 of 45 patients, coronary artery disease in 11 of 45 patients, idiopathic hypertrophic subaortic stenosis in 2 of 45, miscellaneous heart diseases in 8 of 45, and no other heart disease in 11 of 45 patients. They described that the majority of the patients with anomalous coronaries complained of chest pain which was typical angina in 42.2% and atypical chest pain in 28.9%. Other symptoms included syncope (17.7%) and palpitations (46.7%). Their main conclusions were:

- the coronary anomalies were associated with a high incidence of valvular heart diseases and hypertension
- the origin of left main coronary artery from the right sinus of valsalva might, in certain patients, be associated with serious cardiac events as a possible result of impaired myocardial perfusion due to abnormal initial course of the artery.

In another series of 2,996 patients undergoing selective coronary arteriography, Page et al⁶ noted anomalous origin of the left circumflex coronary artery in 20 patients (0.67%). They have described 2 angiographic signs to recognise reliably this anomaly before selective demonstration of the artery itself. One of these signs is the profile view of the artery behind the aortic root during left ventriculography (the "aortic root" sign), and the second is the recognition of absent arterial flow to a significant area of the posterior lateral left ventricle during selective injection of the left main coronary artery (the "sign of non-perfused myocardium").⁶ Interestingly,

Table 1. Classification of isolated anomalies of coronary arteries.

Group I: Origin from the pulmonary artery
a. Of the right coronary artery
b. Of the left coronary artery
Group II: Origin from the aorta
a. From 1 sinus of valsalva
i. From single ostium and then trifurcation
ii. From 2 ostia, i.e. 1 vessel bifurcating
iii. From 3 separate ostia
b. From 2 sinuses of valsalva
i. Each sinus having 1 ostium, i.e. 1 vessel bifurcating after origin
ii. One sinus having single ostium and other with 2 ostia.
c. From 3 sinuses of valsalva
Group III: Miscellaneous
Abnormal origin of first septal artery, and so on

In each subgroup, the following subtypes should be specified according to the course of the anomalously arising artery.

- A. Anterior to aorta and the pulmonary artery
- B. Posterior to aorta and the pulmonary artery
- C. Interarterial
- D. Septal

At the end, the standard abbreviation for the name of the anomalous artery should be given in parentheses. For example, according to this classification, the case reported in this paper will belong to Group II-c-B (LCx).

even in their large series of anomalous origin of left circumflex coronary artery, there was no case where the artery had its origin from the non coronary sinus of valsalva, as noticed in our present report.

In another detailed study of 4,250 patients, Engel et al⁷ reported a 1.2% incidence of anomalous origin of coronary arteries without any congenital heart disease. Their report was based on angiographic observations. They excluded patients with congenital heart disease with a view that they might have a higher incidence than the rest of the population. Their findings included that the majority of variations involved the left coronary artery and that the most common anomaly was origin of the left circumflex from the right sinus of valsalva or the right coronary artery (30 of 4,250 cases).

Despite elaborate descriptions of the pathological anatomy of anomalous coronary arteries and their individual incidences, a comprehensive classification does not exist. Roberts⁸ described the different types of coronary arterial anomalies at length. On the basis of data

then available, they proposed a detailed classification of these anomalies. As a large number of anomalies are related to the right coronary sinus or the right coronary artery, Dollar and Roberts proposed another simple classification applicable to these particular anomalies.⁹

Conclusion

Utilising the knowledge gathered from various published case reports, we have attempted to propose a classification according to the abnormal patterns of origin and course of these anomalous coronary arteries (Table 1). In this classification, Group I comprises patients with serious anomalies which are life threatening. Group II patients need surgical correction if they belong to subtypes C or D, and Group III patients in general do not appear to be at serious risk. We believe that this classification will be helpful for comparison and reference purposes.

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