Case Report

Exertional dizziness and syncope caused by anomalous left coronary artery origin from the right sinus of Valsalva

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ABSTRACT

Coronary anomalies are generally rare, but has fascinating congenital cardiac disease entities, recognized in less than 1.3% of all coronary angiograms. Left coronary artery arising from right sinus of valsalva (RSOV) represents an extremely uncommon subtype. Initial presentations include chest pain, myocardial infarction, arrhythmias, sudden death, and rarely exertional syncope. We report a case of exertional dizziness and syncope, diagnosed to have anomalous origin of left main coronary artery from RSOV. Surgical intervention was curative.

Saudi Med J 2004; Vol. 25 (11): 1720-1722

Coronary anomalies are generally rare, carrying an incidence of 0.6-1.3% of angiographic series, and 0.3% of autopsy series.[1] Left coronary artery (LCA) arising from right sinus of valsalva (RSOV) represents an incidence of 0.017-0.03% of angiographic series, and 1-1.3% of all congenital malformations.[2] Initial presentation includes atypical chest pain, angina, myocardial infarction, arrhythmias, sudden death, and rarely exertional syncope. We report a case of exertional dizziness and syncope, diagnosed to have anomalous origin of left main coronary artery from RSOV. Correction by surgery was effective in relieving the patient's symptoms and ischemia.

Case Report. We report here a case of a 46-year-old lady who had a long-standing hypertension, hyperlipidemia, and marked obesity. Patient presented to the clinic with several episodes of dizziness and syncope on mild exertion accompanied by vague left sided chest discomfort. Her examinations revealed a blood pressure of 150/80, heart rate 78, jugular venous pressure was not raised, cardiac auscultation was normal no added sounds or murmurs, chest was clear and the rest were normal. Electrocardiogram showed normal sinus rhythm. Echocardiogram was normal.

Persantine stress thallium showed a reversible ischemic defect in the anterior-apical and lateral walls (**Figure 1**). Cardiac catheterization revealed normal ventriculogram with high-end diastolic pressure; coronary angiogram showed anomalous origin of the left main coronary artery (LMN) from RSOV, passing in between the aorta (AO) and pulmonary artery (**Figures 2 & 3**). No significant lesion in the left anterior descending (LAD) or left circumflex arteries (LCX), the right coronary was dominant and free of disease.

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Received 6th April 2004. Accepted for publication in final form 13th June 2004.

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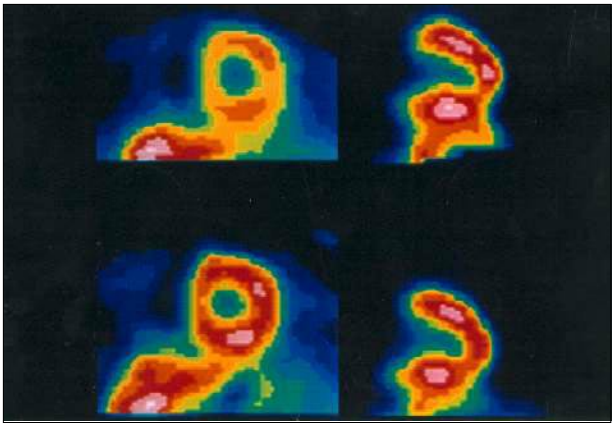

Figure 1 - Myoperfusion scan short and long axis showing anteroseptal, apical and lateral reversible defect during stress (upper panel) and rest redistribution (lower panel).

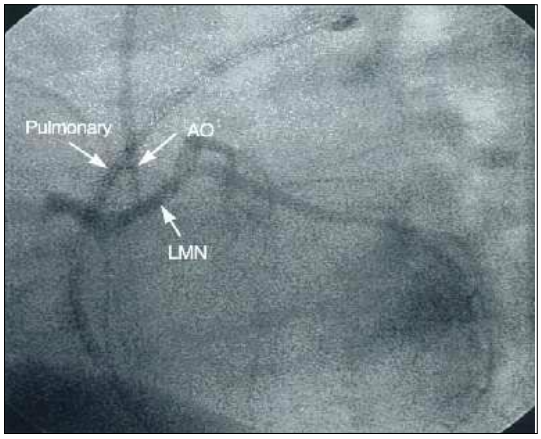

Figure 3 - Coronary angiogram in left anterior oblique 75 degrees showing pulmonary artery anterior, aorta (AO) posterior and left coronary artery passing in between.

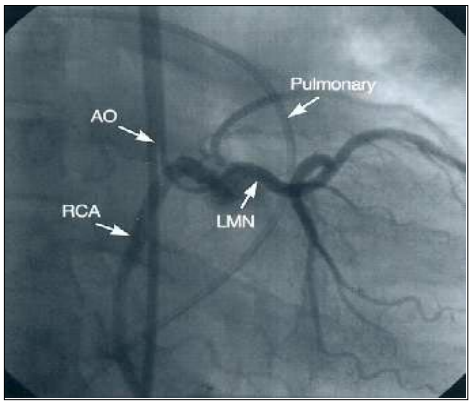

Figure 2 - Coronary angiogram in right anterior oblique 10 degrees showing left coronary artery origin from right sinus and passing between aorta (AO) (posterior) and pulmonary artery (anterior). RCA - right coronary artery, LMN - left main coronary artery

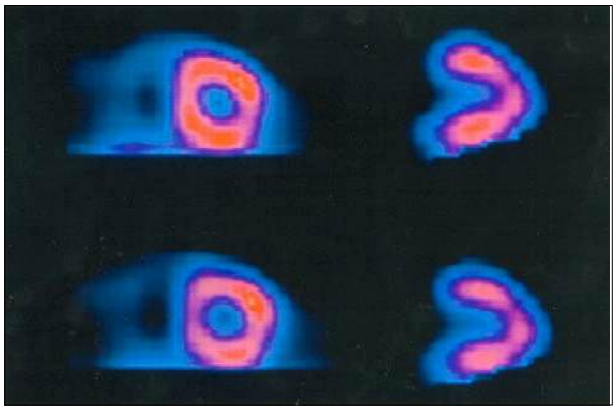

Figure 4 - Myoperfusion scan normalized with no detected reversible defect during stress (upper panel) and rest (lower panel).

The patient had coronary artery bypass surgery for correction of her coronary anomaly (left internal mammary artery to LCX-marginal and saphenous vein graft to LAD). Patient remained asymptomatic one year after her surgery, has no angina or syncope, leading normal life and repeat stress thallium was normal (**Figure 4**).

Discussion. Coronary artery anomalies are some of the most confusing and neglected topics in cardiology. It constitutes around 2.2% of all congenital malformations of the heart.[3] They are usually classified into benign or potentially serious. Left coronary artery arising from RSOV is categorized among the potentially serious anomalies.[1]

Four different anatomic types are identified according to the relation of the anomalous LCA to the AO and pulmonary artery, and classified into LCA passing anterior to the pulmonary artery, retroaortic course, where the LCA passes posterior to the AO, interarterial course between the AO and pulmonary artery, intramyocardial or septal course, along the right ventricular outflow tract. Septal perforator branches from the LMN help differentiate the intramyocardial from the intraarterial form. The most serious type is the intraarterial type, to which the literature has attributed the highest reported sudden cardiac death. Coronary anomalies have been implicated in chest pain, sudden death, cardiomyopathy, dyspnea, ventricular arrhythmia, and myocardial infarction. Quite rarely, they have been related to reproducible effort syncope.[4]

The reason for the sudden fatal event and the mechanism of ischemia are generally unclear. Different proposed theories include compression of

the anomalous LCA between the AO and pulmonary artery during heavy exercise. Other theories propose that ischemia is related to angulation at the origin from the RSOV, spasm or congenital hypoplasia of the anomalous vessel, or that the course of the anomalous artery around the contour of the AO leftward and posteriorly causes flap-like closure of the slit-like orifice as the AO expands during exercise.[1,2]

In the past, coronary angiography was the only tool for the diagnosis of LCA originating from the RSOV. More recently, transesophageal echocardiography as well as magnetic resonance imaging has been used to detect this anomaly. Computed tomography angiography was also reported in confirming the diagnosis.[5]

Patients with the intraarterial form of the anomaly and symptoms, should be considered for surgical therapy. Those with the other types of this anomaly are thought to be at low risk and do not require surgery. However, all patients with LCA arising from the RSOV are cautioned to avoid unmonitored vigorous exercise. All physicians must be aware of exertional dizziness and syncope as a rare presentation of coronary anomaly and myocardial ischemia.

Acknowledgment. The authors thank Dr. Faisal Al-Atwai for his assistance.

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