A case of anomalous left coronary artery origin from pulmonary artery (ALCAPA) in an asymptomatic adult

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A 36 year old asymptomatic lady presented for the investigation of an incidentally found murmur. Physical examination revealed a flow murmur without any other significant signs. Electrocardiograph (ECG) showed sinus rhythm with no abnormal ST, T or Q wave changes (Figure 1).

Transthoracic 2D echocardiography revealed an increased color flow pattern in the inter-ventricular septum (IVS) (Figure 2). Coronary angiogram showed a dilated right coronary artery (RCA) with numerous and enlarged septal collaterals (Figure 3).

CT coronary angiogram revealed the origin of the left coronary artery from the pulmonary artery (Figure 4) with predominant septal collaterals (Figure 5) confirming the diagnosis of ALCAPA syndrome.

Patient was referred for surgical treatment, which is ligation of the left coronary artery (LCA) at the origin from the pulmonary artery and placing a bypass graft to the LCA [2,3].

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital abnormality. It is usually diagnosed in the childhood and the mortality is 90% during the first year of life [1]. However, some may survive up to adulthood asymptotically as in our patient. In the fetal life, there is an antegrade flow in the LCA as a result of high pulmonary vascular resistance. However, after birth as the pulmonary arterial pressure decreases the blood flow in the anomalous LCA reverses, and finally myocardial ischemia develops, especially in the sub-endocardium. Survival of these patients into adulthood is multifactorial and that may include the development of collaterals from RCA to LCA, having a dominant RCA [3]. Though adult patients may be asymptomatic, they can have subclinical ischemia or sudden cardiac death. In addition, some may present with mitral regurgitation or left ventricular dysfunction as a result of the chronic ongoing subclinical ischemia [3].

Figure 1. ECG of the patient indicating normal sinus rhythm.

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Figure 2. Increased Color Doppler Signal in Inter Ventricular Septum LV: Left Ventricle, IVS: Inter Ventricular Septum.

Figure 3. Invasive coronary angiogram showing large RCA with many septal collaterals.

Figure 4. CT coronary angiogram AO: Aorta, PA: Pulmonary Artery, LMS: Left Main Stem, RA: Right Atrium, LA: Left Atrium.

Figure 5. CT coronary angiogram PA: Pulmonary Artery, LA: Left Atrium, LV: Left Ventricle

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