Journal of Clinical & Experimental Cardiology
ISSN: 2155-9880

Pizzino et al., J Clin Exp Cardiolog 2013, 4:5
DOI: 10.4172/2155-9880.1000247

Case Report Open Access

Association between Anomalous Origin of the Left Main and Accessory Mitral Valve Tissue

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Abstract

A 73-year-old man presented to our department for ischemic alterations appeared during a stress electrocardiogram. The diagnostic process including coronary angiography, cardiac computed tomography and echocardiogram revealed an anomalous origin of the left main coronary artery from the commissural region between the left and the non-coronary Valsalva sinus associated with non-obstructive accessory mitral valve tissue. Although these malformations are relatively common the association is not described in literature to date.

Introduction

The congenital coronary artery anomalies are a rare cause of sudden cardiac death and myocardial infarction in children and in adults.

We report the case of a rare congenital malformation of the left coronary artery (LCA) taking off from the posterior non-coronary Valsalva sinus associated with another rare congenital anomaly represented by accessory mitral valve tissue (AMVT). Although the association of several cardiac anomalies is common, the finding of both these malformations in the same subject is not described in literature to date.

Case Description

A 73-year-old male cyclist with hypertension, type 2 diabetes mellitus and dyslipidemia was admitted to our department to further evaluate severe ischemic ST-T changes observed during stress ECG. Past medical history, clinical examination and rest ECG were unremarkable. At trans-thoracic echocardiogram, left ventricle had normal dimensions and thickness and no wall motion abnormalities. The mitral valve was peculiar because of AMVT adherent to the anterior leaflet (Figure 1, panel A, white arrow) not causing left ventricular outflow tract (LVOT) obstruction. At coronary angiography the dominant right coronary artery had non-significant lesions; it was technically impossible to selectively engage the left main artery (LM) because of posterior anomalous origin and aortic wall calcifications close to the ostium. Non-selective injections showed non-significant lesions on both left anterior descending and circumflex arteries. The patient underwent cardiac computed tomography (cCT): the LM originated posteriorly from the left Valsalva sinus (panel B), in the commissural region between the left and the non-coronary sinuses. A huge calcific aortic plaque adjacent, superiorly, to the LM ostium (Figure 1, panel C, white arrow) had an eccentric soft component causing an intermediate stenosis of the LM stem (panel B, white arrow) which, in turn, coursed between the left coronary sinus and the anterior wall of the left atrium (Figure 1, panel B and C). Cardiac CT ruled out any other significant atherosclerotic coronary lesion and further delineated the AMVT adherent to the mitral anterior leaflet (Figure 1, panel D, black arrow). In order to assess the severity of the myocardial ischemia and to establish a correct management, we considered performing additional cardiac stress tests such as myocardial perfusion nuclear magnetic resonance or scintigraphy but the patient refused any other procedure and was discharged uneventfully.

Discussion

The incidence of coronary artery anomalies is reported about 1% but it is estimated about 4-15% in autopsy of young people who experienced sudden death [1].

In a study by Kilner et Al. including 7694 patients with coronary artery anomalies, the authors found that the 95.2% had anomalies of origin and distribution and 4.8% had coronary artery fistulae. The incidence was the highest for the separate origin of the left anterior

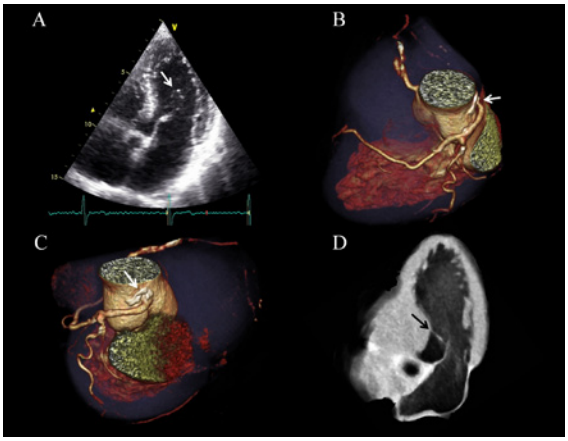

Figure 1: (A) Two-dimensional echocardiography: apical four-chamber view showing accessory mitral valve tissue (white arrow) connected to the anterior leaflet, floating in the ventricular cavity in diastole. (B-C) Cardiac computed tomography (cCT)-3D reconstruction- presenting the anomalous origin of the LCA (white arrow) and the huge atherosclerotic plaque extending from the aorta in the LM (white arrow). (D) 3D reconstruction from cCT: anatomical section showing the accessory mitral valve tissue in diastole (black arrow).

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Received April 11, 2013; **Accepted** May 10, 2013; **Published** May 13, 2013

Citation: Pizzino F, Zito C, Donato R, Carerj S, Andò G (2013) Association between Anomalous Origin of the Left Main and Accessory Mitral Valve Tissue. J Clin Exp Cardiolog 4: 247. doi:10.4172/2155-9880.1000247

descending and circumflex (Cx) from the left sinus of Valsalva (52.4%). Anomalous origin of the left Cx from the right coronary was 8.7% while from the right coronary sinus of Valsalva was 18.4%. Ectopic coronary origin from the opposite aortic sinus (1.9%) and single coronary artery (3.88%) were described but they are rare [2]. The literature is poor of cases describing an anomalous origin of the LCA from the non-coronary sinus of Valsalva [3-6]. Two-dimensional echocardiography, computed tomography angiography and magnetic resonance are the most important diagnostic instruments for the evaluation of the coronary anomalies [7]. The advantage of these techniques is to individuate high risk conditions, or eventual further congenital cardiovascular malformations associated, and to state if the ischemic ECG alterations are related to the anatomical defect or to others coexisting pathological conditions.

Accessory mitral valve tissue consists in the presence of a free floating membrane-like structure contiguous to the ventricular side of the anterior mitral valve leaflet, or as a fixed structure anchored to the interventricular septum by a short chordal apparatus; nevertheless, the literature reports many intermediate and variable configurations. Patients are often asymptomatic; however someone may experience palpitations and fatigue. Although in a restricted number of cases a cerebrovascular embolic event is the first manifestation of the malformation, the benefit of anticoagulant treatment is not yet well determined [8]. A LVOT obstruction is a common finding in patients with AMVT and the presence of other associated malformations such as transposition of the great arteries, coarctation of the aorta, partial atrioventricular canal; double outlet right ventricle or membranous septal aneurysm is frequent [9]. The incidence of this anomaly is unknown. Rovner et al. described 5 cases on about 130.000 echocardiograms performed in their institute, so they assess a single center incidence of about 1 per 26.000 cases [10]. Two-dimensional echocardiography (performed by either transthoracic or transesophageal approach) is the main technique for the evaluation of the AMVT. The most frequent echocardiographic aspect is an irregular parachute or sail-like structure attached to the chordae, anterior MV leaflet, accessory papillary muscle, or the interventricular septum. In some cases it appears as a globular or even cystic mass. The redundant tissue typically prolapsed into LVOT during systole, following the blood flow, and retracting away during diastole. Doppler imaging is useful for evaluation of prognosis: a gradient higher than 50 mmHg in LVOT is correlated to a worse outcome, and in those cases the surgical treatment can be considered [10]. To the best of our knowledge, this is the first demonstration of the coexistence of AMVT and anomalous origin of the LM by means of integrated cardiac imaging.

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