Case report: non compacted myocardium of the left ventricle in combination with multiple coronary artery aneurysms

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Abstract

We present a clinical case of coexisting left ventricle non compaction and multiple coronary artery aneurysms in 28 years old woman, complicated by myocardial infarction due to thrombosis of aneurysm. A patient presented due to suspected unstable pectoral angina. Cardiac computed tomography study revealed stented right coronary artery, aneurysm of diagonal branch without detectable signs of atherosclerotic changes and a thick layer of trabeculations in inferolateral walls and apex of left ventricle. Magnetic resonance imaging confirmed evidence of non-compaction myocardium with ratio 2.6 with not impaired left ventricular systolic function. Positron emission tomography–computed tomography study rejected active inflammation in coronary arteries.

Keywords: left ventricle non compaction, coronary artery aneurysm, cardiomyopathy
Learning objective

Coexistence of the left ventricle non compaction and coronary artery aneurysms is rare, but should be suspected due to possibility of potentiate cardiovascular complications, such as left ventricle systolic dysfunction, heart insufficiency, arrhythmias, the episodes of arterial embolism and myocardial infarction. LVNC could be in combination with aneurysms as consequence of Kawasaki disease or congenital origin in young age.

Introduction

Left ventricle noncompaction (LVNC) is a rare form of primary genetic cardiomyopathy, which is associated with the altered process of compaction of ventricular myocardium during embryogenesis and leads to formation of trabeculae separated by deep recesses, communicating with cavity of the ventricle [1,2,3]. The main clinical signs are the left ventricle systolic dysfunction, heart insufficiency, arrhythmias (supraventricular and ventricular) and the episodes of arterial embolism [1,3]. Myocardial ischaemia is often observed, but ST segment elevation myocardial infarction is a rare condition, the most commonly associated with coronary artery diseases [3].

Coronary artery aneurysms (CAA) are localized dilatation of coronary artery segment more than 1.5-fold compared with contiguous normal segments [4]. The ethiology of CAA are atherosclerosis, percutaneous interventions, vasculitis or congenital. Kawasaki disease (KD) is the leading cause of aneurysms in children and the second cause of aneurysms in adults [4,5]. To our knowledge, there is only one case report that presents LVNC in combination with CAA as consequence of KD [6].

We present and overview a rare case of coexisting LVNC and multiple CAA in young female patient, complicated by myocardial infarction due to thrombosis of aneurysm.

Case report

A 28 year old woman presented to our clinic due to suspected unstable angina pectoris. A patient’s complaint was chest pain associated with emotional and physical stress. Anamnestic data revealed a previous myocardial infarction with ST elevation, in age of 18, treated with drug eluting stent implantation. Myocardial infarction manifested in postoperative period of cervical neurinoma excision surgery. On the 5th day patient reported intensive chest pain radiating to the left arm, ECG showed ST elevation myocardial infarction. During this episode, a multiple coronary aneurysms were observed by interventional angiography [figure 1]. An aneurysm in RCA was found thrombosed, subsequently stented and named as culprit lesion. During admission to our Hospital blood tests, ultrasound, computed tomography (CT), magnetic resonance imaging (MRI) and positron emission tomography–computed tomography (PET CT) studies were performed. Blood analysis was ordinary – red blood cells 5,64 x 10¹²/l, haematocrit 0,472, C-reactive protein 7,2mg/l, creatinine 79,3 μmol/l, glomerular filtration rate 88ml/min1,73m², Troponine I <0,01 ng/ml (normal 0 – 0,023), cholesterol 3,34 mmol/l, high density lipoprotein 1,24 mmol/l, low density lipoprotein 1,93 mmol/l, triglycerides 1,08 mmol/l, NTproBNP 145. 2D transthoracic Doppler echocardiographic examination was performed. The overall left ventricular (LV) ejection fraction by Simpson's method was decreased to 46%. There were an inferior wall and inferior septum motion abnormalities. There was mild mitral regurgitation and mild tricuspid regurgitation. According to echo findings an left ventricle non compaction was suspected: two-layered structures (recesses and prominent trabeculae) of LV inferior and septal apical segments with an end-systolic ratio greater than two between the non-compacted subendocardial layer and the compacted subepicardial layer was observed [Figure 2]. With color Doppler echocardiography a blood flow was registered into the intertrabecular recesses. There were no other associated cardiac abnormalities. Cardiac CT study revealed stent in proximal part of RCA and aneurysm of diagonal branch, without detectable signs of atherosclerotic changes in coronary arteries [Figure 3]. Moreover, a thick layer of trabeculations in inferolateral walls and apex of left ventricle were observed in this study [Figure 4]. Subsequently performed MRI confirmed evidence of noncompation in referred segments with ratio of non compated to compacted myocardium (2.6), diagnostic for LVNC [Figure 5] without signs of scar on late gadolinium enhancement.

There was no definitive anamnestic data of clinical evidence of KD and PET CT was prescribed to confirm or exclude active vasculitis. PET CT study revealed no signs of active inflammation in
coronary arteries, as in aorta and other middle sized arteries.

Figure 1. Interventional angiography. LAO cranial view of left coronary artery demonstrates aneurysm of first diagonal branch (arrow) and small aneurysm in septal branch (arrowhead).

Figure 2. 2D transthoracic apical: A - four chamber and B - two chamber views show isolated non-compaction of left ventricle with the deepest intertrabecular recesses in inferior wall and interventricular septum.
Figure 3. Cardiac CT. 3D VR reconstruction of coronary artery tree: aneurysm of first diagonal branch (arrow) and stent in proximal part of RCA (arrowhead).

Figure 4. Cardiac CT. 4 chamber view: trabeculated layer in inferolateral wall and apex of left ventricle.

Figure 5. Cardiac MR. 4 chamber view, SSFP sequence demonstrates non compacted and compacted wall layers of left ventricle.
Discussion

We present a rare combination of LVNC and CAA, complicated by myocardial infarction due to thrombosis of aneurysm. The prevalence of isolated LVNC is not accurate because LVNC is relatively unknown among physicians and often undiagnosed, although in observational studies, LVNC has been found in 0.01–0.26% of all adults referred to an echocardiography [7]. Andrews R.E. and co-authors analysed all types of children’s cardiomyopathies in their study and revealed that prevalence of LVNC is 9.2% in children [8]. The cardiac ultrasound is the main method to achieve diagnosis of LVNC, other imaging tests as magnetic resonance imaging were also used to confirm diagnosis. The main lack of echocardiographic diagnostic criteria is a difficulty to derive diagnostic indexes due to small sample of studies [7]. Criteria of Jenni and co-authors are used widely for LVNC [9]. With reference to those criteria, LVNC is characterized as two-layer structure: a thin, normally compacted layer (C) and a markedly thickened non-compacted layer (NC) (with a ratio of NC/C >2), excessively prominent trabeculations, and deep intertrabecular recesses measured at end-systole in the parasternal short-axis views, filled by blood from the ventricular cavity (when other cardiac abnormalities do not exist) [7]. By MRI, the main criteria is NC/C >2,3 at the end of diastole [10]. According to cardiac ultrasound an isolated LV non-compaction cardiomyopathy was suspected and MRI revealed NC/C 2.6 (diagnostic criteria is >2,3) and LVNC was diagnosed. The difference between cardiac ultrasound and magnetic resonance imaging were compared in the study and established that MRI is more accurate to evaluate expansion of LVNC [11]. We assume that ultrasound could be less accurate due to dependance on operator skills. Unclear diagnosis, in condition of poor echo window should be specify by prescribing MRI [11].

The LVNC can be isolated or present with other cardiac and/or systemic illnesses. Clinical manifestations vary from asymptomatic to ventricular arrhythmias, thromboembolic events, heart failure or sudden death [12]. According to Stähli et al., 12% of patients with LVNC had associated various forms of congenital cardiac abnormalities, mostly stenotic defects of the LV outflow tract, Ebstein anomaly, tetralogy of Fallot [13]. To our patient LVNC coexists with CAA. Aneurysms are detected for 0.3%-5% adults after coronary artery angiography [14]. CAA pathogenetically are divided into atherosclerotic, inflammatory and non-inflammatory [6]. The main cause of CAA is atherosclerosis (about 50% of patients), inflammation of coronary artery wall layers is linked to wall weakening [4, 15]. According to young age of patient, the possibility of atherosclerotic aneurysm in our case is negligible, subsequently coronary aneurysms in our case could be post-inflammatory or congenital. Genetic predisposition is associated with developing of CAA. 9p21.3 allele is related to abnormal formation of coronary artery wall development, but there can be synergism with atherosclerotic changes, further studies are required [4]. Percutaneous coronary interventions, connective tissue diseases (Marphan syndrome, Ehlers-Danlos disease), infection, drug use (especially cocaine), other vasculitides (Takayasu arteritis) and congenital aneurysms are other possible causes [4, 5, 14, 16]. Characteristics of congenital CAA are giant aneurysms (>50mm) and coronary artery fistulas [17,18]. Clinical manifestation of congenital CAA can be asymptomatic or can lead to dyspnoe, pectoral angina; imaging tests findings are similar to other ethiology CAA - CT reveals saccular aneurysms with or without thrombosis, MRI – wall motion abnormalities (as consequence of myocardial ischaemia) [5].

CAA are often confirmed as complication of KD [19]. To our knowledge, a combination of aneurysms and LVNC is described only in one case report, where CAA were consequence of confirmed KD [6]. Aneurysms following KD appear for the right coronary artery and the proximal part of the major coronary vessels; 23% of aneurysms are asymptomatic are the main cause of acute coronary syndromes prevalence due to thrombosis in young age (the third decade) [20]. Patients who do not have sufficient principal clinical findings of KD may be considered with incomplete KD.

American Heart Association recommends undergo stress tests regulary (including stress echocardiography), cardiac MRI and PET for all patient with diagnosed medium or large CAA [19]. Literature review and our clinical data suggests that LVNC could be in combination with aneurysms as consequence of KD or congenital.

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Conflict of interest

The Authors declare that there is no conflict of interest.
References


