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Left ventricular noncompaction (LVNC) with giant left ventricular aneurysm detected by multi detector computed tomography angiography

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Abstract

Left ventricular noncompaction (LVNC) is a rare form of cardiomyopathy caused by the failure of myocardial compaction during embryogenesis. Clinical features spectrums from are from being totally asymptomatic to systolic dysfunction, heart failure (HF), tachyarrhythmias, and systemic thromboembolism. However, there have been only few reports regarding its coexistence with LV aneurysm. A 34-year-old man with no significant cardiopulmonary history was evaluated for progressive exertional dyspnoea. The chest radiography exhibited cardiomegaly, mild pulmonary congestion and huge calcified shadow at apex. The laboratory data was unremarkable except for elevated brain-type natriuretic peptide. Transrthoracic echocardiography demonstrated global hypokinesia with an ejection fraction of 30%, prominent trabeculation and deep intertrabecular recesses, and calcified aneurysm (12.3 x 7.8 cm), and increased noncompacted (NC) endomyocardial layer depth compared to the compacted (C) epicardial layer (NC/C = 2.4) which was further confirmed on multi detector computed tomography (MDCT). Based on these findings, he was diagnosed as LVNC complicated with a giant calcified aneurysm. He was discharged with carvedilol, ramipril, and frusemide and referred for surgical restoration/resection of aneurysm. Here, to the best of our knowledge, we report the patient who had LVNC with biggest LV aneurysm.

Key Words: Left ventricular noncompaction; Intertrabecular recesses; Left ventricular aneurysm; Multi detector computed tomography

Introduction

Non-compaction of the ventricular myocardium or "spongy myocardium" is a rare, morphologically distinct primary genetic cardiomyopathy characterized by prominent ventricular trabeculations and deep intertrabecular recesses which is believed to represent an arrest in endomyocardial morphogenesis with a reported prevalence of 0.045%[1, 2]. It can be seen as an isolated cardiomyopathy or can be associated with complex congenital heart lesions[1, 4]. These deep recesses are lined by endocardium and are in continuity with the
ventricular endocardium. Its clinical manifestations include systolic heart failure, thromboembolism, tachyarrhythmias, and sudden death resulting into high mortality rate. Although the regional left ventricular wall thinning with myocardial hypo perfusion has been described in a few cases, association with coexisting left ventricular aneurysm is exceedingly rare [5, 6].

Case Report
A 34-year-old male presented with progressive exertional dyspnoea for the past twelve months. Clinical examination revealed blood pressure of 96/80 mm Hg in right upper limb in supine position with pulse rate of 88/min, regular, low volume with no radio-radial or radio-femoral delay and all peripheral pulses were equally palpable. The jugular venous pressure was raised 4cm above the angle of Luis. Bilateral pedal enema was also present. The apical impulse was displaced in the seventh intercostals space, 4 cm outside the midclavicular line, forceful but ill-sustained in character. The S1, S2 was soft and S3 was audible. Auscultation over the lung fields revealed bilateral basal crepitations. The liver was tender, palpable 3.5 cm below the costal margin. Electrocardiogram showed normal sinus rhythm, left atrial enlargement and left ventricular hypertrophy. The laboratory data was unremarkable except for elevated plasma brain-type natriuretic peptide (BNP) which was 1108 pg/ml. The chest X-ray showed cardiomegaly, mild pulmonary venous hypertension and two calcified ring like shadow at the cardiac apex (Fig.1). A two-dimensional (2-D) transthoracic echocardiogram in apical 4-chamber view showed a dilated left ventricle (LV end diastolic diameter- 56 mm) and left atrium (41 mm) with global hypokinesia and an ejection fraction of 30% with mild mitral regurgitation. The epicardium was thin while the endocardium was markedly trabeculated and spongy. Deep intertrabecular spaces communicating with the main left ventricular cavity were evident on both 2-D imaging (Fig.2). The ratio of maximal thickness of the noncompacted to compacted layers in the lateral wall was > 2 at end-systole. The apical 4-chamber and parasternal short axis views defined the left ventricular non-compaction (LVNC) which was seen in all segments of the left ventricle but was more marked in the apical, lateral and inferior segments. The right ventricular endocardium was normal.

Moderate tricuspid regurgitation was present with an estimated right ventricular systolic pressure of 49 mmHg and the inferior vena cava was dilated. There was a calcified aneurysmal sac connecting with LV with a broad neck. As the sac had a broader neck, comprising of all 3 layers and was akinetic, it was diagnosed as true aneurysm. Coronary angiography was done to rule out any associated coronary artery disease, which was normal.
In order to delineate its connection, dimension, and anatomical detail, computed tomography angiography (CT) was performed which revealed calcified aneurysmal sac measuring 12.3x7.8 cm connecting with LV in its posterior wall with a broad neck of 16.3 mm (Fig. 3). CT angiography showed thin epicardium while the endocardium was markedly trabeculated and spongy with deep recesses (Fig.4). The patient was treated with carvedilol-12.5 mg, ramipril- 10 mg and frusemide-40 mg daily and referred to surgery for ventricular restoration following aneurysm resection/repair.

**Discussion**

Isolated left ventricular noncompaction is a congenital cardiomyopathy caused by a defect in endomyocardial morphogenesis [7]. It is usually isolated affecting LV, and rarely right ventricle as well. Coexisting left ventricular aneurysm has rarely been reported. Its diagnosis is multimodality including echocardiography, multi detector CT (MDCT), ventricular angiography, and cardiac magnetic resonance imaging (cMRI), left ventricular angiography (LVG), and gated single photon emission tomography (SPECT). The echocardiographic criteria suggested by Jenni et al.[8] have become widely accepted which includes: (a) thickened myocardium with a two layered structure consisting of a thinner compacted epicardial [C] and a thicker, noncompacted endocardial layer [N] or trabecular meshwork with deep endocardial spaces (N/C ratio > 2.0 at end-systole); (b) predominant location of the pathology in the mid-lateral, mid-inferior, and apical areas; (c) colour Doppler evidence of deep intertrabecular recesses filled with blood from the LV cavity; and (d) absence of coexisting cardiac abnormalities (in isolated LVNC). The echocardiographic characteristics of our patient were consistent with the above criteria.

Although a congenital disorder, noncompaction may present late in adult life which is an outcome of complex interplay of multiple factors including extent of myocardial involvement, progressive myocardial dysfunction caused by subendocardial hypo perfusion and microcirculatory dysfunction. As the patient was symptomatic only for the past one year, we believe that noncompaction of the ventricular myocardium was silent till the progressive hemodynamic burden because giant aneurysm started taking toll along with impaired systolic function. The hallmark signature is echocardiographic appearance of the ventricular myocardium and unlike that seen in a hypertrophied ventricle. Our patient also had a giant LV aneurysm and the clinical presentation is similar to that of a case of isolated noncompaction. The distinctive clinical and echocardiographic features of noncompaction is similar to what have described in one of the largest series on isolated noncompaction of the
left ventricle in adults by Oechslin et al.[1]. It carries a high incidence of heart failure, arrhythmias, thromboembolism and sudden cardiac death.[1] Coexistence of giant aneurysm further adversely alters the prognosis. LVNC coexisting with a left ventricular aneurysm was first reported by Sato Y et al.[5]. Similarly, left ventricular aneurysm and multiple diverticula were described in a patient with normal coronary arteries [9].

Despite availabilities of multiple diagnostic modalities, it can reliably be diagnosed by transrthoracic echocardiogram and computer tomography imaging. The prognosis is mainly related to the presence of LV dysfunction. Treatment of such patients always poses a therapeutic dilemma. Medical stabilization, aneurysm resection and/repair, prophylactic implantable cardioverter-defibrillator (ICD), and ultimately cardiac transplantation are the option for these patients [1].

Conflict of interest
None

References


Figure legends

Figure 1. The chest X-ray postero-anterior view showing cardiomegaly, mild pulmonary venous hypertension and two calcified ring like shadow at the cardiac apex.

Figure 2. Two-dimensional transthoracic echocardiogram in apical 4-chamber and parasternal short axis (PSAX) views in end-diastole showing deep intertrabecular recesses (white arrow) in the apical segments and along the lateral wall.

Figure 3. MDCT showing calcified aneurysmal sac measuring 12.3 x 7.8 cm connecting with LV in its posterior wall with a broad neck of 16.3 mm (A; B; An-aneurysm; LV-left ventricle).

Figure 4. CT angiography showing thick markedly trabeculated and spongy endocardium with deep recesses and relatively thin epicardium (A); Volume rendered CT showing giant LV aneurysm (B).