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Dilated Cardiomyopathy Caused by Isolated Ventricular Non Compaction in Young Adult

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Abstract **Case Report**

Isolated ventricular non-compaction (IVNC) is often described in the context of dilated cardiomyopathy; still; it is unclear whether this genetic disorder is a distinct cardiomyopathy or an acquired condition co existing with other forms of myocardial diseases. To help distinguishing IVNC from idiopathic dilated cardiomyopathy; echocardiographic features of the myocardial wall need to be carefully defined. We describe a case of dilated cardiomyopathy caused by IVNC in severe ventricular dysfunction and cardiac thrombosis in a young adult

Keywords: Dilated cardiomyopathy, isolated ventricular non compaction.

INTRODUCTION

Isolated ventricular non-compaction (IVNC) is a morphogenetic abnormality characterized by a spongy appearance of the left ventricle (LV) due to the presence of an extensive non-compacted myocardial layer lining the ventricle cavity. It can potentially leads to malignant arrhythmias, thromboembolism, and cardiac failure [1].

The phenomenon has an embryological origin caused by intrauterine interruption of the normal myocardial compaction process still it is often diagnosed in adults and children.

IVNC could be considered a subtype of dilated cardiomyopathy since the frequent association with varying degrees of LV dilation in most patients [2].

We report the incidental discovery of IVNC in a young patient at the stage of dilated cardiomyopathy with severe cardiac failure.

CASE REPORT

A 20-year-old man with no medical history was admitted for global heart failure. On physical examination, the patient had conjunctival jaundice, his heart rate was 100 bpm and blood pressure was 85/57 mmHg. The cardiac examination revealed right congestive signs. The electrocardiogram (ECG) demonstrated left ventricular hypertrophy and right branch block. Chest radiography showed cardiomegaly and bilateral hilar overload. Transthoracique echocardiography (TTE) showed a dilated left ventricle with global hyponikésia and severe dysfunction (EF=20%). Deep trabeculated recesses with free flow of blood from the LV cavity on color Doppler study were seen in apical and medioventricular segments of the lower and lateral wall. A mobile apical thrombus was present. The right ventricle was also dilated and dysfunctional. Coronary angiography was normal.

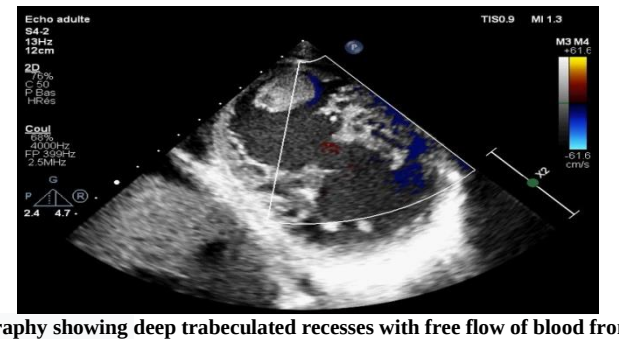

Fig-1: Transthoracic echocardiography showing deep trabeculated recesses with free flow of blood from the left ventricle cavity on color Doppler

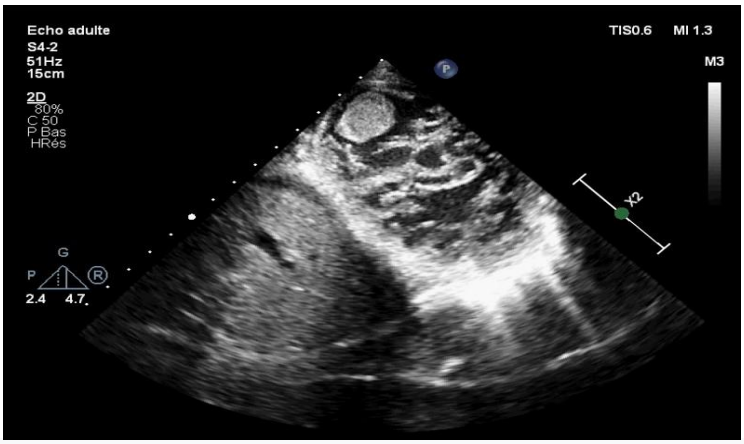

Fig-2: Transthoracic echocardiography showing apical thrombus and multiple trabeculations in the left ventricle

Magnetic resonance imaging (MRI) confirmed the diagnosis of IVNC by the presence of a structured myocardium in two layers with a myocardial ratio between the compacted and uncompacted surface> 2 associated with multiples perfusion defects. The patient was treated with diuretics, oral anticoagulation and beta-blockers.

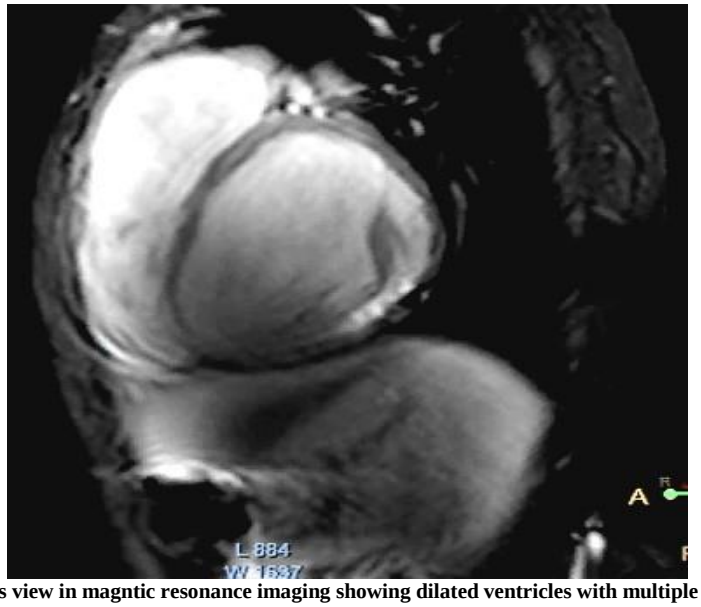

Fig-3: A small axis view in magntic resonance imaging showing dilated ventricles with multiple perfusion defects

DISCUSSION

Engberding and Bender first described isolated ventricular non-compaction in 1984, since then it is recognized as an important differential diagnosis in patients with heart failure [3].

Due to incomplete myocardial morphogenesis, leading to persistence of the embryonic myocardium [4], the actual prevalence of IVNC is uncertain because only data based on case reports are available. Even though, it appears to be the third most commonly diagnosed cardiomyopathy

Discriminating IVNC from other causes of heart failure is difficult, recent studies have shown that symptoms may present late in adulthood as a result of progressive myocardial dysfunction [5].

The most common criteria for diagnosis uses a ratio of the thickness of the noncompacted layer to the thickness of the compacted layer (T/C) with a ratio >2 at end-diastole in with color Doppler evidence of perfused intratrabecular [6].

It is unclear whether IVNC is a distinct cardiomyopathy, a subtype of dilated cardiomyopathy, a morphogenetic disorder, or an acquired condition [7] but recent studies showed that association with dilated cardiomyopathy is very frequent [8].

Trying to elucidate the cause of systolic dysfunction in patients with IVNC, Chin et al suggest that an abnormal endocardial perfusion despite the absence of epicardial coronary artery disease can lead to heart failure [9].

The diagnosis of IVNC relies on noninvasive imaging studies, most typically transthoracic echocardiography (TTE) and cardiac magnetic resonance imaging (CMR) by determining the ratio of thickness of the trabecular and compact layers, the trabeculations being more easily identified by CMR in the relaxed heart. The diastolic ratio of >2.3 showed high diagnostic accuracy for distinguishing pathological LVNC from the degrees of non-compaction observed in healthy, dilated, and hypertrophied hearts [10].

Increasing awareness of the disease helped decreasing mortality rate, Oeschlin et al report a 35% mortality rate after a follow-up of 44 months, and they have described some prognostic factors: higher left ventricular end diastolic diameter at baseline, NYHA Class III or IV presentation, persistent or permanent atrial fibrillation, branch block [5].

The treatment is identical to that of other cardiomyopathies. It is based on the management of heart failure rhythm disorders and anticoagulation to prevent systemic embolism.

CONCLUSION

IVNC is a complex congenital disorder considered rare but more and more described. The key of the diagnosis is echocardiography, which highlights the numerous trabeculations. The poor prognosis along with absence of specific treatment should lead to early detection among first-degree relatives.

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