Case Report

Gross Left Ventricular Voltage with Preexcitation in Isolated Left Ventricular Non-Compaction


Abstract

A 30 years man presented with symptoms of heart failure with prior history of pulmonary tuberculosis. On routine investigation he was found to have gross left ventricular voltage on the electrocardiogram and evidence of ventricular pre-excitation. His echocardiogram confirmed the diagnosis of left ventricular non-compaction. The aetiopathogenesis, clinical features, diagnostic criteria and review of literature of this rare entity is discussed here.

INTRODUCTION

Isolated left ventricular non-compaction (IVNC) was first described just over a decade ago, and is gaining importance as a rare; but important differential diagnosis in patients presenting with cardiac failure. This as yet unclassified cardiomyopathy by the WHO was previously known as “spongy left ventricular myocardium”. Although a variety of electrocardiographic abnormalities have been described in pediatric and adult population, gross left ventricular voltage on ECG may also be associated with IVNC and may mimic hypertrophic cardiomyopathy. The diagnosis of this disorder is mostly missed because of lack of knowledge. Early diagnosis of IVNC and differentiating it from hypertrophic and dilated cardiomyopathy are crucial as clinical manifestation is characterized by important morbidity and mortality caused by early heart failure, life-threatening ventricular arrhythmias, and systemic embolic events. The present patient was diagnosed elsewhere as hypertrophic non-obstructive cardiomyopathy / pericardial effusion. However the recent clear-cut echocardiographic criteria published in literature helped us diagnose the condition accurately. Associated electrocardiographic findings in this patient were gross LV voltage and evidence of atrioventricular bypass tract.

CASE REPORT

A 30 years man presented with history of class III dyspnoea on exertion and intermittent palpitations of three months duration. He was diagnosed to have pulmonary tuberculosis for which he was treated with antituberculous drugs for six months about a year ago. The patient did not have hypertension or diabetes, nor was there a past history of any heart disorder. His haemoglobin was 12 gm%, total count-5800 cells/mm³, ESR-10mm/hr, blood urea-32mg%, Creatinine-0.9mg%, LFT was normal. His chest X-ray showed a thin walled cavity at the apex of right lung. There was cardiomegaly with rounding of the left heart border (Fig. 1). Sputum AFB was negative thrice. His electrocardiogram showed evidence of ventricular preexcitation as evidenced by short PR interval and delta wave; he also had gross LV voltage on the ECG amounting to a total of 100mm of R wave height in lead V2.

Fig. 1 : Chest X-ray showing rounding of cardiac apex along with right upper lobe cavity
His echocardiogram showed that left ventricle was dilated, globally hypokinetic and globular in shape, the end-diastolic dimension was 5.61 cm, end-systolic dimension was 4 cm, posterior wall thickness was 1.9 cm, ventricular septum was 2.4 cm, left atrium was 3.7 cm, aortic root was 2.6 cm and calculated ejection fraction (M mode and area length method) was 45%. There was presence of diastolic dysfunction of left ventricle on pulse wave Doppler. The left ventricle was apparently hypertrophied, but on close examination, revealed prominent trabecular pattern of endocardium in the mid and apical inferior, posterior, lateral and LV apical regions. Prominent deep intertrabecular recesses with color Doppler demonstrating free flow of blood from the LV cavity into the crevices were characteristically demonstrable (Fig. 4). The end-systolic thickness of spongy endocardium was twice the thickness of the compact portion, further confirming the diagnosis (Fig. 3). There was mild mitral regurgitation in addition to the above findings. A diagnosis of isolated non-compaction of left ventricle was made on basis of the above findings.

The patient was put on low dose diuretics and oral anticoagulants to keep an INR between two and three. He was advised regular outpatient follow-up.

**DISCUSSION**

In normal development the myocardium condenses and the intertrabecular recesses are reduced to capillaries, deep intertrabecular recesses communicating with the ventricular endocardium may evolve in some patients because of an arrest of compaction of loose interwoven meshwork of myocardial fibres during intrauterine life. IVNC is characterized by prominent myocardial trabeculations and deep intertrabecular recesses which lie in continuity with the left ventricular cavity. Although prominent trabeculae are seen in the normal right ventricle, the persistence of prominent left ventricular trabeculations is not normally apparent after birth. A similar persistence of non-compacted myocardium is frequently reported in patients with congenital left or right ventricular outflow tract obstruction that generate intraventricular pressure overload and is referred to as “spongy myocardium” or “persisting sinusoids” that communicate with the coronary arteries. Examples include pulmonary atresia with intact ventricular septum or anomalous origin of left coronary artery from the pulmonary trunk. In these hearts, the deep recesses are in continuity with the ventricular cavity and with the coronary arteries and are therefore more accurately described as persistent intramyocardial sinusoids. In contrast, IVNC has no associated cardiac lesions and persistent sinusoids are not seen i.e. the recesses in IVNC have no connection with the coronary circulation. In fact they are covered by endocardial lining continuous with the ventricular cavity, predisposing to local thrombus formation. Isolated IVNC is a distinct entity of cardiomyopathy but is a genetically heterogeneous congenital disorder, both X-linked recessive and an autosomal inheritance is seen. Familial occurrences, relations to genetic disorders or syndromes such as Melnick-needles-syndrome, Roifman syndrome, Leber’s hereditary optic neuropathy and Xq28-linked cardiomyopathy (allelic
with Barth syndrome) have been reported. Since this entity is a congenital disorder, it should be present at birth in all patients, a notion supported by two previous reports. Many of the published reports on this disorder have examined a predominantly pediatric population but now it is apparent that IVNC may present in adulthood, mainly as a result of symptoms of heart failure. The diagnosis of IVNC is mostly missed, because of lack of awareness as was true for almost 90% of the patients with IVNC in a series recently reported by Ichida et al.3 Ritter and colleagues found that the mean time from symptom onset to diagnosis was 3.5 years.4

Prominent left ventricular trabeculations can be found in healthy hearts as well as in hypertrophic cardiomyopathy and in LVH secondary to dilated, valvar, or hypertensive cardiomyopathy. Differentiation between variants and IVNC may be often be challenging. As IVNC may have an impact on morbidity and mortality, early and reliable diagnosis is crucial.2

Non-compaction of the ventricular endocardium primarily affects the left ventricle, but may also involve the right ventricle although distinguishing this from normal anatomy is more difficult. Various forms of semilunar valve obstruction or left ventricular outflow tract obstruction have to be ruled out.1

The thickened left ventricular wall consists of two zones of different structures. The compacted epicardial layer appears as a compact band of uniform tissue while the much thicker endocardial non-compacted layer consists of trabecular meshwork with deep endocardial spaces surrounded by exaggerated hypertrophy of the trabeculae. Strictly speaking a two-layered structure is found only in IVNC, and not in LVH, DCM, or any other condition. It may sometimes appear difficult to define where prominent trabeculations end and non-compaction begins. An end systolic ratio of non-compacted to compacted layers of >2 is diagnostic for IVNC and allows unambiguous differentiation from hypertrophic cardiomyopathy and DCM or LVH.2

The features of IVNC are found predominantly in the apical and lateral, inferior mid ventricular segments of the left ventricle, as confirmed in a large series. These segments are hypokinetic contrary to certain forms of apical cardiomyopathy that can otherwise mimic IVNC. However, hypokinesia may not be confined entirely to these segments but rather extend to morphologically unaffected segments causing decreased global ejection fraction.2

Evidence of direct blood flow from the ventricular cavity into deep intertrabecular recesses by color Doppler echocardiography is one of the hallmarks of the diagnosis of IVNC. This is never observed in other forms of LVH.2

Other modalities such as computed tomography, magnetic resonance imaging, and ultrafast computed tomography may be also helpful, but no diagnostic criteria for these modalities have yet been proposed.2

Accurate diagnosis is important for IVNC as its clinical morbidity includes heart failure caused by progressive ventricular dysfunction both systolic and diastolic, arrhythmias, and systemic and pulmonary embolism.

Mass screening has provided information on the natural history of patients with IVNC who are asymptomatic at presentation. In a Japanese population, it was found that left ventricular dysfunction developed over a longer time course, with restrictive physiology dominating the early stages of the disease. In contrast to the reported cases in symptomatic adults, ventricular arrhythmias and embolic events were rare. Although patients with IVNC who are asymptomatic at presentation have a longer clinical course, the majority of these individuals do go on to develop left ventricular dysfunction. Adult patients are also at risk of ventricular arrhythmias and systemic emboli even during the early stages of the disease, before left ventricular dilatation and systolic dysfunction are apparent. Currently, oral anticoagulation is recommended for all adult patients in whom IVNC is diagnosed, irrespective of left ventricular size and function.1

Electrocardiographic changes in IVNC reported in literature are LBBB, RBBB, LAHB, repolarisation abnormalities, atrial fibrillation and preexcitation.5 In this case we found gross LV voltage with preexcitation, which has infrequently been reported. Although voltage criteria for LVH are unreliable in presence of preexcitation such exceedingly high voltages even in AV-bypass tracts are unusual.

It would have been ideal to do an EP study to substantiate preexcitation and an angiographic study to demonstrate prominent LV trabecular recesses; but since there was no indication, these invasive investigations were not done.

IVNC is associated with poor prognosis, mortality being nearly 40% at five years. Fifty percent of deaths are due to ventricular arrhythmias, heart failure being the second commonest cause. Coronary microcirculatory dysfunction has been shown to be associated with this condition.6

**CONCLUSION**

In conclusion despite a number of conditions mimicking IVNC, awareness of this entity and availability of echocardiographic criterion for diagnosis, coupled with a number of observational studies has led to more correct diagnosis of this particular condition. Although the long-term outlook of IVNC is poor, a correct diagnosis may probably tell us about the actual incidence of this condition in the population, avoiding a mistaken diagnosis of idiopathic dilated cardiomyopathy or hypertrophic non-obstructive cardiomyopathy as was done in our case. Our case also had evidence of AV bypass tract, which has been reported variably in literature from 13% to 15%.3

**REFERENCES**


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