Clinical case report based study

Biventricular noncompaction presented with symptomatic complete heart block – Report of a case and review of literature

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ABSTRACT

Ventricular noncompaction has been recognized as a distinct form of rare cardiomyopathy characterized by numerous, prominent ventricular trabeculations and deep intertrabecular recesses and is caused by a disorder of endomyocardial morphogenesis. Concomitance of either valvular pathologies or complete atrioventricular block with biventricular noncompaction has rarely been reported. Herein, we present a case of 67 years old male presented with syncopal attack and congestive heart failure due to biventricular noncompaction with significant left ventricular dysfunction associated with complete atrioventricular block. He was formerly diagnosed as dilated cardiomyopathy for last 2 years. Review of literatures of all reported cases has been discussed.

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1. Introduction

Noncompaction of the ventricular myocardium (NCVM) is an unclassified congenital cardiomyopathy resulting from an arrest in normal endomyocardial embryogenesis characterized by an excessively prominent trabecular meshwork with deep intertrabecular recess.¹ It frequently complicated with heart failure, malignant arrhythmias and embolic events.² The clinical manifestations of this entity are variable, from patients who are asymptomatic with a preserved LV ejection fraction to patients that have various degrees of systolic ventricular dysfunction and eventually develop heart failure, systemic embolism and various types of cardiac arrhythmias.³ We are reporting a patient with NCVM who presented with symptomatic complete heart block.

2. Case history

A 67 years old male, non-diabetic, non-hypertensive, non-smoker and had history of dyspnea on exertion of New York Heart Association (NYHA) class II since last two years. He was previously admitted twice due to congestive heart failure (CHF) in the past and was on treatment with oral enalapril, digoxin, diuretics and

carvedilol. This time he admitted in our hospital with a history of syncopal attack and features of CHF. At admission, his heart rate was 20 beats/min (bpm), blood pressure was 130/90 mm oh Hg with engorged neck veins and pedal edema. Systemic examination revealed pansystolic murmur of grade III/VI at apex and left lower parasternal border, early diastolic murmur at neo aortic area along with bilateral basal crepitations and altered sensorium. Electrocardiogram (25 mm/s, 10 mm/mV) was done at admission which showed complete heart block (CHB) with ventricular escape rate of 20 bpm (Fig. 1). Patient was immediately put on bedside temporary pacemaker through internal jugular vein and rate was fixed at 80 bpm. Blood investigation showed renal dysfunction (blood urea-70, serum creatinine-1.6), normal electrolytes and normal complete blood counts. Transthoracic echocardiography revealed non-compaction of ventricular myocardium involving apical portions of both ventricles and lateral & inferio-posterior wall of left ventricle (Fig. 2). The ratio of noncompacted-to-compacted myocardium was >2 in the maximal thickened wall, measured at the end systole. On the color Doppler echocardiography, these recesses were filled with blood from the ventricular cavity. All the four cardiac chambers were dilated with biventricular global hypokinesia was present. The calculated ejection fraction by modified biplane was only 30%. On color Doppler echocardiography, there was presence of aortic, mitral and tricuspid regurgitation of moderate severity. Aortic cusps and mitral valve leaflets were calcified and thickened. Papillary muscles were normal. Doppler echocardiographically

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Fig. 1. ECG showing complete heart block with ventricular escape at 20 bpm.

measured pulmonary artery systolic pressure over tricuspid insufficiency was 64 mm Hg. Patient was on temporary pacemaker along with conservative treatment for CHF. His sensorium and symptoms of cardiac failure improved after four days. Though he was offered dual chamber pacemaker for the management of CHB along with

severe biventricular dysfunction, but due to financial constrain, he was put on VVIR pacemaker and was discharged to home after 10 days of hospital admission. He was put on angiotensin converting enzyme inhibitor (ACEI); loop diuretic, aldactone and acetyl salicylic acid.

Fig. 2. Biventricular noncompaction of apical & lateral wall of LV & apical wall of RV.

3. Discussion

NCVM is classified as an unclassified cardiomyopathy by the World Health Organization in its report on definition and classification of cardiomyopathies. NCVM is increasingly recognized as an important cause of cardiomyopathy and it is the result of the arrest of cardiac lesion densification process and characterized by excessively prominent trabeculations in ventricular wall segments and deep intratrabecular recesses.⁴ It is diagnosed primarily by echocardiography.⁵ The most frequently applied echocardiographic definition of NCVM was established by Jenni et al, defined NVMC as the ratio of the end-systolic thickness of noncompacted/compacted myocardium of greater than 2.0.⁶ Another echocardiographic definition of NCVM which is also frequently applied was given by Stöllberger et al. According to their criteria, NCVM is characterized by more than 4 coarse and prominent trabeculations, which move synchronously with the myocardium and have the same echogenicity and are surrounded by intertrabecular spaces.⁷ Finsterer and Stöllberger proposed a new echocardiographic definition that applies both definitions and categorizes echocardiographic findings into definite, probable and possible NCVM.³ Definite NCVM is present if both definitions are completely fulfilled. Probable NCVM is present if either the criteria of Jenni et al or Stöllberger et al are fulfilled. Possible NCVM is present if the numbers of trabeculations are less than four or if the ratio of noncompacted-to-compacted layers is <2. As in our case, both definitions were completely, so it was a definite case of NCVM. There was a poor correlation between the three echocardiographic definitions: 24% of the study population fulfilled one or more echocardiographic definitions for LVNC, while only 30% fulfilled all three criteria. More surprisingly, 8% of apparently healthy individuals also satisfied one or more diagnostic criteria for LVNC.⁸ New echocardiographic technique, such as tissue Doppler imaging, strain and strain rate, and speckle tracking, may help to evaluate the functional impact of an abnormal myocardial architecture and enable the clinician to distinguish between normally trabeculated myocardium from LVNC.⁸ Left ventricular twist was determined by speckle tracking echocardiography. Rotation was clockwise at the base and counterclockwise at the apex in all controls and patients with dilated cardiomyopathy. In contrast, the LV base and apex rotated in the same direction in all noncompaction patients.⁸ Cardiac magnetic resonance imaging (MRI) has been increasingly used to describe the morphological appearance of the myocardium. Multimodality imaging, including echocardiography and cardiac MRI, is required to confirm or to exclude LVNC.⁸

Patients with NCVM have high incidence of CHF, arrhythmias and thromboembolism.¹ The most common presentation is congestive heart failure. Arrhythmias associated with NCVM include atrial arrhythmias, ventricular tachycardia and sudden cardiac death.² Complete atrioventricular block (AVB) is rarely reported.^{1–5} Complete AVB is a relatively common finding in pediatric series and is rarely reported in adult patients.⁹ The true cause of ventricular conduction abnormalities is still unclear. The fibrotic change in the conduction system develops gradually and this might account for the progressive nature of the atrioventricular block seen in adult patients.² Defects of cytoarchitectural proteins can cause left ventricular noncompaction, which is often associated with conduction system diseases. Recently, a study showed that loss of function of hNav1.5 by a ZASP1 mutation associated with intraventricular conduction disturbances in left ventricular noncompaction.¹⁰

There are five reported cases of CHB associated with ventricular noncompaction reported in the literature so far.^{1–5} Among these cases, only two cases were associated with biventricular

Table 1
Clinical features and management profile of all previously reported cases of NCVM with CHB.

Variables	Age/sex	Clinical presentation	ECG	ECHO	Medical management	Pacemaker	Follow-up
Dagdeviren et al ⁵	18 years/F	Dizziness, presyncope, no CHF	Complete AVB, rate 36/min	LVNC of apical, lateral & posterior wall, LVEF-normal	Oral anticoagulant	TPI followed by DDD pacemaker	Symptoms free
Sajeev et al ²	24 years/M	Recurrent syncope	AF with CHB	LVNC of apex & mid ventricle, LVEF-normal	–	–	–
Yildiz et al ¹	70 years/M	F/S/O CHF & MR, AR, TR a/w palpitation, dizziness for 4 mo	Complete AVB, rate 48/min	BVNC, RWMA, LVEF-53%, PASP-60	ACEI, diuretics, aspirin	–	Symptoms free
Ying et al ³	43 years/M	F/S/O CHF & MR	AF, CHB & RBBB	LVNC of septum & mid ventricle, LVEF-30%	–	VVIR pacemaker	Symptoms free
Nascimento BR et al ⁴	32 years/F	Presyncope, Dyspnea, elevated JVP	Complete AVB, rate 54/min	BVNC of apical area, LVEF-51%	Diuretics & ACEI	Dual chamber pacemaker	Symptoms free
Present case	67 years/M	Syncope, F/S/O CHF, MR, AR, TR	CHB, rate 20/min	BVNC of apex & lateral wall, Dilated chambers, LVEF-30%, PASP-64	ACEI, diuretics, aspirin	VVIR pacemaker	Symptoms free

AF-atrial fibrillation, RBBB-right bundle branch block, LVNC-left ventricular noncompaction, BVNC-biventricular noncompaction, LVEF-left ventricular ejection fraction, RWMA-regional wall motion abnormality.

noncompaction,^{1,4} whereas, other cases were associated with isolated left ventricular noncompaction^{2,3,5} (Table 1).

Our case had biventricular NCVM with dilated cardiomyopathy, concomitant valvular insufficiency and complicated with complete AVB. Valvular insufficiency of aortic and mitral valves might be due to NCVM associated valve dysfunction or age related valvular degeneration. As our patient had complete AVB and was dependent on temporary pacemaker along with severe biventricular dysfunction, therefore, it is an indication for a permanent pacemaker. Pacemaker implantation was thought to be effective to prevent the risk of sudden cardiac death and other complications.⁷ In our case, VVIR pacemaker was implanted and optimum management for CHF was continued.

4. Conclusion

During treatment and follow-up of ventricular noncompaction, not only the heart failure, embolic events or the ventricular arrhythmias but also the valvular pathologies and complete AVB should be kept in mind and treated accordingly.

Conflicts of interest

All authors have none to declare.

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BK, SP, & SKS were involved in the management of this patient. BK & SP reviewed the literature & drafted the manuscript. SKS &

MCN corrected the manuscript. All authors approved the final version of the manuscript.

References

1. Yildiz A, Ozeke O, Akyol S, Demirbag R, Yilmaz R, Gur M. Biventricular myocardial noncompaction presenting with complete atrioventricular block. *Int J Cardiol.* 2009;132:e34–e36.
2. Sajeev CG, Francis J, Shanker V, Vasudev B, Abdul Khader S, Venugopal K. Young male with isolated noncompaction of the ventricular myocardium presenting with atrial fibrillation and complete heart block. *Int J Cardiol.* 2006;107:142–143.
3. Ying ZQ, Xu G. An adult patient with isolated left ventricular noncompaction presented with atrial fibrillation and complete atrioventricular block. *Intern Med.* 2011;50:2597–2599.
4. Nascimento BR, Vidigal DF, Carneiro RD, et al. Complete atrioventricular block as the first manifestation of noncompaction of the ventricular myocardium. *Pacing Clin Electrophysiol.* 2011 Mar 16;36:e107–e110.
5. Dağdeviren B, Eren M, Oguz E. Noncompaction of ventricular myocardium, complete atrioventricular block and minor congenital heart abnormalities: case report of an unusual coexistence. *Acta Cardiol.* 2002;57:221–224.
6. Steffel J, Kobza R, Oechslin E, Jenni R, Duru F. Electrocardiographic characteristics at initial diagnosis in patients with isolated left ventricular noncompaction. *Am J Cardiol.* 2009;104:984–989.
7. Stöllerberger C, Keller H, Finsterer J. Disappearance of left ventricular hypertrabeculation/noncompaction after biventricular pacing in a patient with polyneuropathy. *J Card Fail.* 2007;13:211–214.
8. Oechslin E, Jenni R. Left ventricular non-compaction revisited: a distinct phenotype with genetic heterogeneity? *Eur Heart J.* 2011;32:1446–1456.
9. Ergul Y, Nisli K, Varkal MA, et al. Electrocardiographic findings at initial diagnosis in children with isolated left ventricular noncompaction. *Ann Noninvasive Electrocardiol.* 2011;16:184–191.
10. Xi Y, Ai T, De Lange E, et al. Loss of function of hNav1.5 by a ZASP1 mutation associated with intraventricular conduction disturbances in left ventricular noncompaction. *Circ Arrhythm Electrophysiol.* 2012;5:1017–1026.