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CASE REPORT

A RARE COMBINATION: BIVENTRICULAR NON-COMPACTION, EBSTEIN'S ANOMALY, WOLF PARKINSON WHITE SYNDROME AND MITRAL VALVE PROLAPSUS

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ABSTRACT

Ventricular non-compaction and Ebstein's anomaly are 2 rare congenital abnormalities. In some cases both abnormalities share common genetic predisposition. Both abnormalities could be associated. We report a case of 25-year old male patient with biventricular non-compaction, Ebstein's anomaly, mitral valve prolapsus and Wolf-Parkinson-White syndrome. The combination of these congenital abnormalities could infer a common genetic predisposition with different presentation of myocardium and leaflets.

Keywords: Biventricular non-compaction; Ebstein's Anomaly; Wolf Parkinson White Syndrome; Mitral Valve Prolapsus

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INTRODUCTION

Ventricular non-compaction is a rare congenital disorder characterized with excessive trabeculations and recesses in ventricules [1,2]. Even though most often left ventricule myocardium affected, right ventricule also could be affected. As being a congenital malformation ventricular non compaction could present with various congenital abnormalities. Ebstein's anomaly is also a rare congenital abnormalities, characterized with apical displacement of tricuspid valve leaflets leading to the atrialization of the some part of right ventricule [1,2]. Apical displacement of valves lead to insufficiency that is the main causes of symptoms. Ebstein's anomaly is not just a right heart disorder. Rather it seen with various cardiac abnormalities like non-compaction, wolf-parkinson white syndrome (WPW), bicuspid aortic valve, ventricular and atrial septal defect [3-5].

In the present case, we reported a rare combination of biventricular non-compaction, Ebstein's anomaly, WPW syndrome and mitral valve prolapsus.

CASE REPORT

A 25-year old male patient present to outpatient clinic with atypical chest pain.

The surface electrocardiogram demonstrated normal sinus rhythm with Wolf-Parkinson-White syndrome. (Figure 1) Transthoracic echocardiography showed left ventricular non-compaction. (Figure 2a) Left ventricle apical and lateral parts showed extensive trabeculation and thickness with a 2.6 ratio between trabeculated portion and left ventricle lateral wall. Color Doppler demonstrated intratrabecular recesses flow. Left ventricular global systolic function was preserved. At the parasternal long axis view mitral anterior leaflet was prolapsing to the left atrium with mild mitral regurgitation demonstrating mitral valve prolapses (Figure 2b).

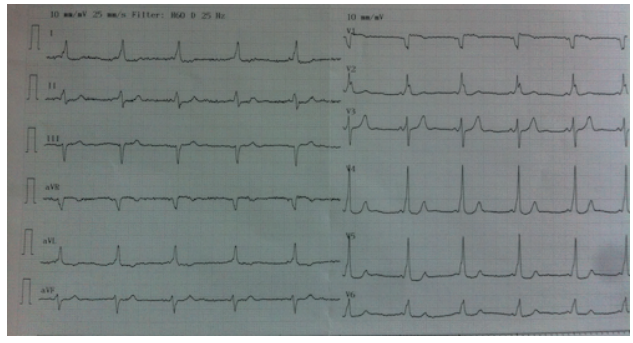

Figure 1: The surface electrocardiogram demonstrated normal sinus rhythm with Wolf-Parkinson-White syndrome

The tricuspid valve showed Ebstein's anomaly physiology; septal leaflet was located apically with atrialization of right ventricle along with mild tricuspid regurgitation on Doppler color flow (Figure 2c). Pulmonary artery systolic pressure was 30 mmHg. Right ventricular systolic function was normal. Due to patient choice and payment strategies of the health care institution a cardiac magnetic resonance imaging was not undertaken.The present report demonstrated a combination of biventricular non-compaction, Ebstein's anomaly, mitral valve prolapses and type B Wolf Parkinson White syndrome. There were few cases presenting non-compaction with different anomalies. To our search this is the first case presenting this combination.

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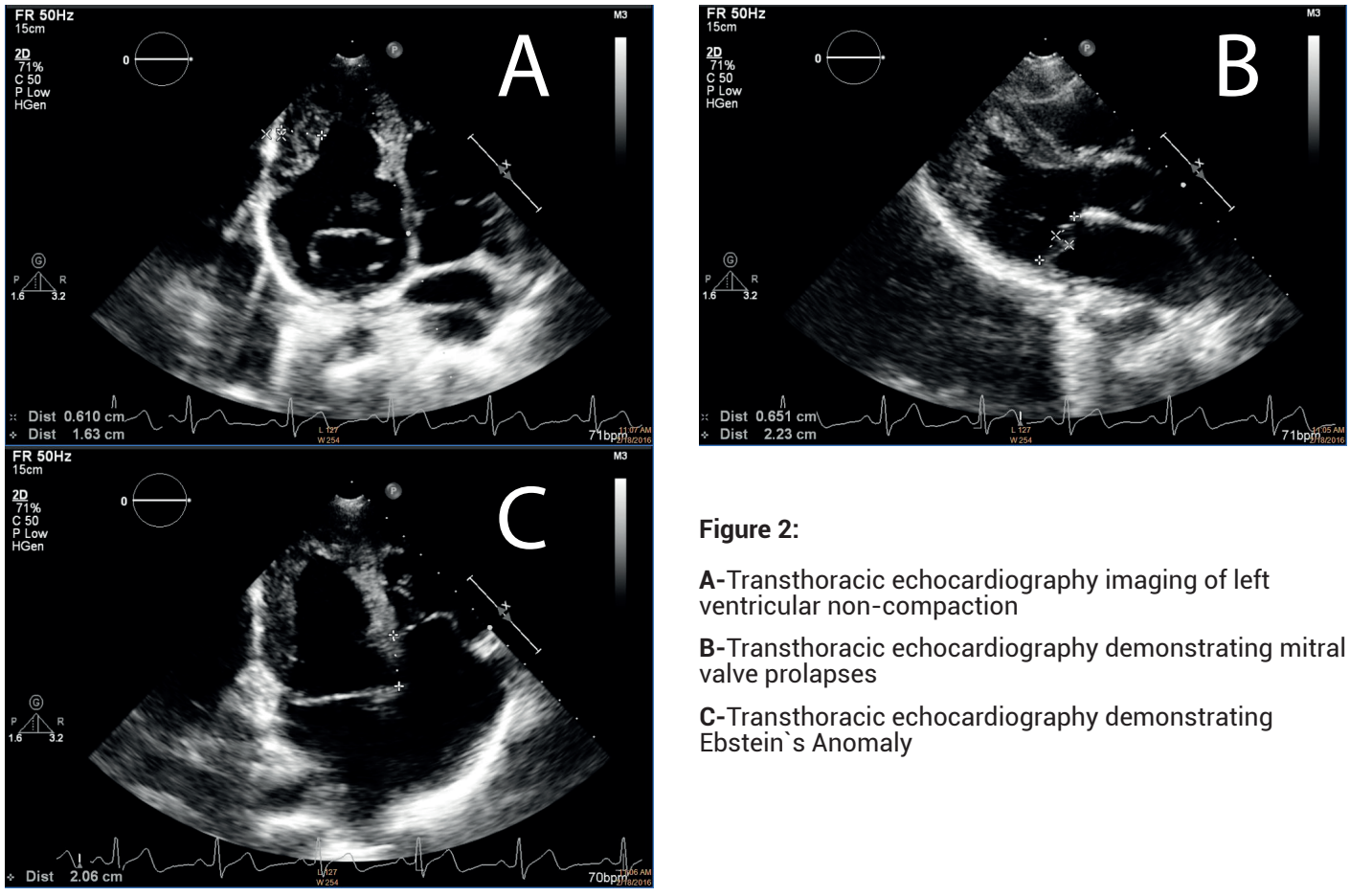

Figure 2:

A-Transthoracic echocardiography imaging of left ventricular non-compaction

B-Transthoracic echocardiography demonstrating mitral valve prolapses

C-Transthoracic echocardiography demonstrating Ebstein`s Anomaly

DISCUSSION

Ventricular non-compaction is a congenital cardiomyopathy that arises from endomyocardial developmental arrest and results in deep recesses and hypertrabeculation in myocardium[1]. Hypertrabeculation of ventricular myocardium can cause ventricular insufficiency, arrhythmia and even sudden cardiac death[6,7]. Ebstein's anomaly is a rare congenital anomaly caused due to tricuspid leaflets incomplete delamination from right ventricule endocardium and displaced apically. Apically displaced tricuspid valves lead to right ventricule volume overload which is the main cause of symptoms. It is proposed that ventricular non-compaction and Ebstein's anomaly could share same genetic cause [8]. It has shown that among ventricular non-compaction patients, the incidence of Ebstein's anomaly is 3%. On the other hand in patients with Ebstein's anomaly ventricular non-compaction found in 18% [8]. The beta-myosin heavy chain encoding MYH7 mutation is the most known genetic cause of ventricular non-compaction and Ebstein's anomaly [1]. Also other more rare mutations have been reported [9]. It has been proposed that, ventricular non-compaction and Ebstein's anomaly could be cause of a common genetic predisposition that end up with different results in ventricular myocardium and atrioventricular valves [9]. Previously it has indicated that Ebstein's anomaly is not just a disorder of right heart [3]. Rather in 39% of Ebstein's anomaly patients, left sided heart abnormalities observed. These are atrial septal defect, patent foramen ovale, ventricular non-compaction, bicuspid aortic valve, mitral valve dysplasia, ventricular septal defect and Wolf-Parkinson-White syndrome[3].

In the present case we reported a combination of mitral valve prolapsus, WPW syndrome, biventricular non-compaction and Ebstein's anomaly. In the literature the co-incidence of ventricular non-compaction and Ebstein's anomaly has been reported. As it has been proposed this could be a syndrome or syndrome-like condition presenting with valvular (mitral valve prolapsus, apical displacement of tricuspid valve and bicuspit aortic valve) and myocardial (biventricular non-compaction) abnormalities.

Patients diagnosed with either congenital abnormalities should be searched for other concomitant abnormalities. A simple surface ECG should be handled for arryhtmia and pre-excitation.

Conflict of interest: none declared

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REFERENCES

1- Budde BS, Binner P, Waldmuller S, et al. Noncompaction of the ventricular myocardium is associated with a de novo mutation in the beta-myosin heavy chain gene. PLoS ONE 2007 2:e1362.
2- Sinkovec M, Kozelj M, Podnar T. Familial biventricular myocardial non-compaction associated with Ebstein's malformation. Int J Cardiol 2005;102:297-302.
3- Attenhofer Jost CH, Connolly HM, O'Leary PW, et al. Left heart lesions in patients with Ebstein's anomaly. Mayo Clin Proc 2005;80:361-368.
4- Saltik L, Eroglu E, Bayrak F, et al. Left ventricular non-compaction associated with Ebstein's anomaly. Turk Kardiyol Dern Ars 2009;37:269-272.
5- Baysan O, Yokusoglu M, Bugan B, et al. E-page original images. A case of Ebstein's anomaly and biventricular non-compaction. Anadolu Kardiyol Derg 2010;10:E26.
6- Captur G, Nihoyannopoulos P. Left ventricular non-compaction: Genetic heterogeneity, diagnosis and clinical course. Int J Cardiol 2010;140:145–153.
7- Towbin JA, Lorts A, Jefferies JL. Left ventricular non-compaction cardiomyopathy. Lancet 2015;386:813–825.
8- Stahli BE, Gebhard C, Biaggi P, et al. Left ventricular non-compaction: Prevalence in congenital heart disease. Int J Cardiol 2013;167:2477–2481.
9- Kelle AM, Bentley SJ, Rohena LO, et al. Ebstein anomaly, left ventricular non-compaction, and early onset heart failure associated with a de novo α-tropomyosin gene mutation. Am J Med Genet A. 2016;170:2186-2190.

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