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Chronic Rheumatic Heart Disease with Bivalvular Stenosis in Association with Isolated Left Ventricular Non-Compaction Cardiomyopathy.

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ABSTRACT

We describe a known case of chronic rheumatic heart disease with bivalvular stenosis in a 37year old male. He was incidentally detected to have non-compaction of left ventricle on echocardiography and Doppler examination. He is the first patient to present with bivalvular stenosis in chronic rheumatic heart disease in association with isolated left ventricular non-compaction cardiomyopathy.

Keywords: rheumatic heart disease, stenosis, cardiomyopathy.

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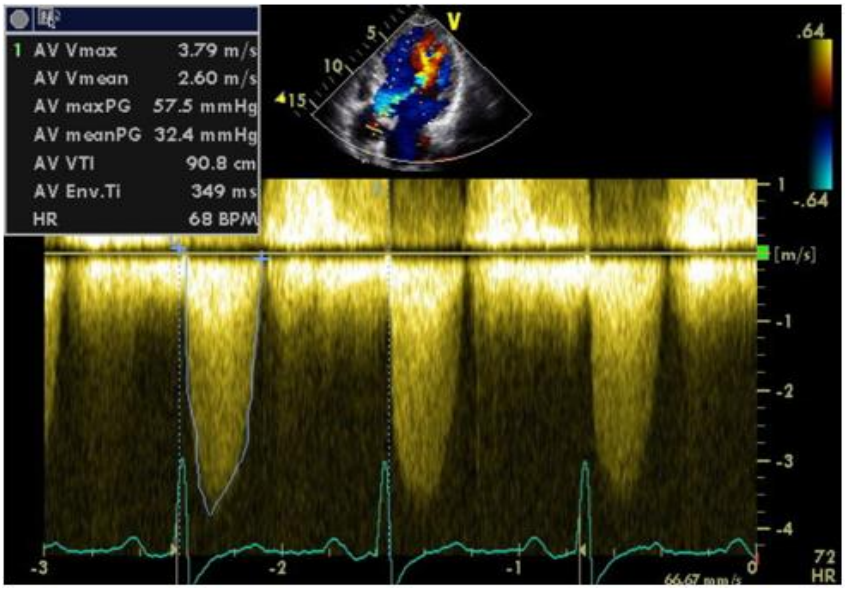
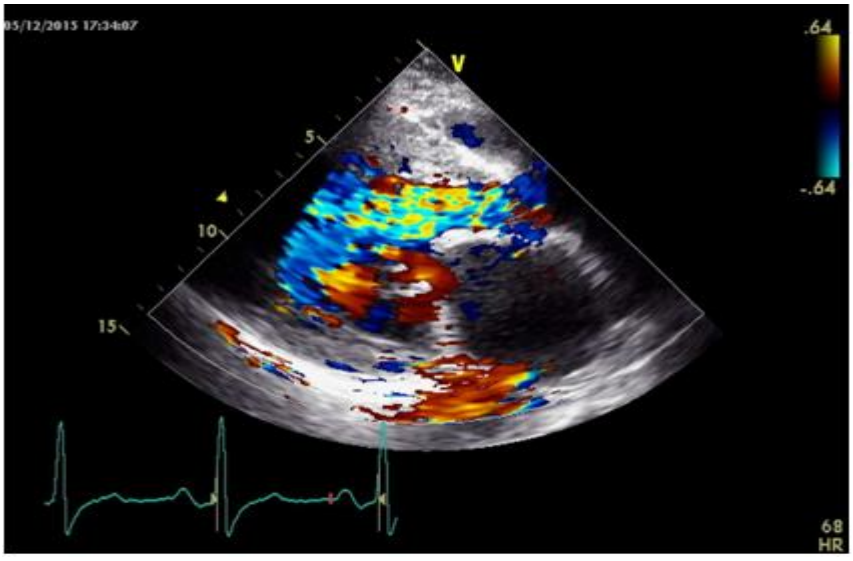
CASE PRESENTATION

A 37 year old male known case of rheumatic heart disease with severe MR and AR presented with complaints of worsening dyspnea NYHA class III, palpitations and fatigue. The frequency of dyspnea had recently increased. A physical examination revealed jugular venous distension, blood pressure of 150/90mmHg, respiratory rate was 20/min and the heart rate was 58 bpm. Cardiac examination revealed regular and rapid rhythm, a grade 3/6 pan-systolic murmur at the cardiac apex, diastolic decrescendo murmur and ESM well heard in the neoaortic area.

INVESTIGATIONS



Continuous wave Doppler showing severe aortic stenosis/regurgitation



PLAX view showing aortic regurgitation

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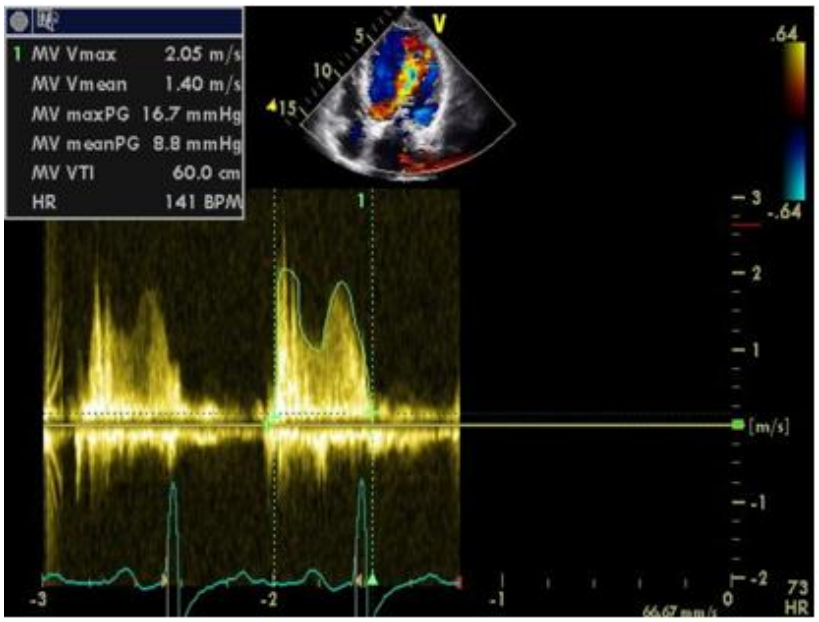
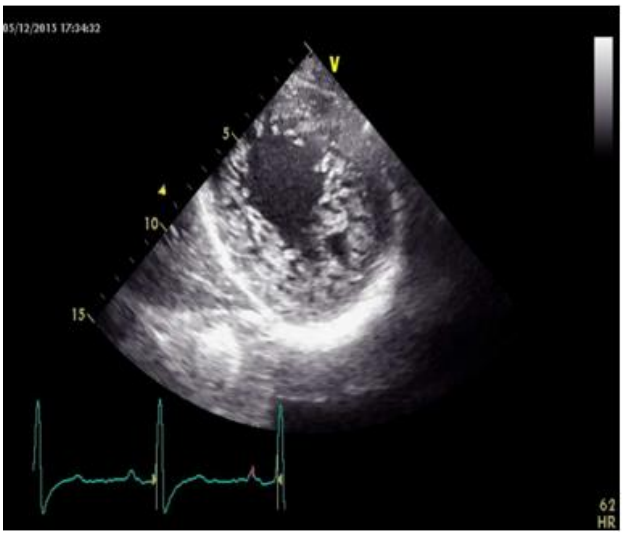
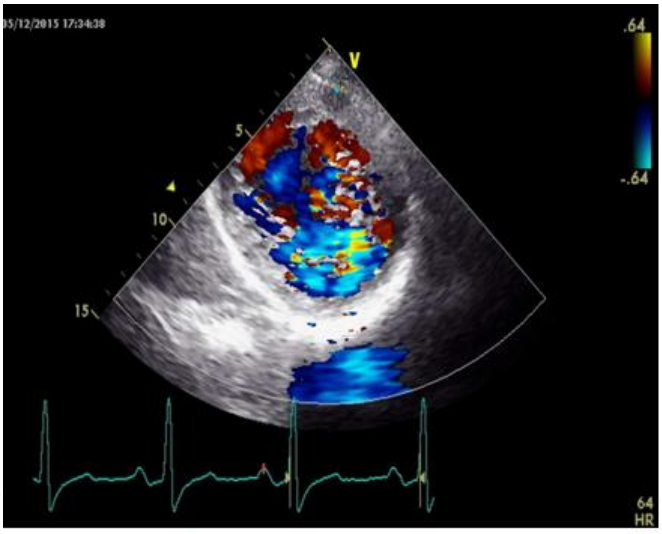


Continuous wave Doppler showing severe mitral stenosis/regurgitation



Left ventricle showing noncompaction



Colour Doppler filling recess in left ventricle

Blood investigations done were essentially normal. The electrocardiography (ECG) revealed bradycardia, P tricuspidale, LVH by voltage criteria, Q waves in V5, V6 suggestive of volume overload without ST changes. ECHO revealed RHD with severe aortic regurgitation with moderate aortic stenosis (figure 1 & 2) and moderate mitral stenosis with mild eccentric mitral regurgitation (figure 3), dilated left atrium, left ventricle and global left ventricular dysfunction. The right atrial and ventricular size and morphology as well as function of pulmonary valves was normal. Holodiastolic flow reversal noted in descending aorta, no clots. On the short axis view, the myocardium of LV had excessively prominent trabeculations and deep intertrabecular recesses at the LV apex (figure 4). The affected segment had a 2 layered structure (a compact epicardial layer with prominent trabecular meshwork and deep intertrabecular spaces. On color Doppler analysis, intertrabecular spaces were filled by direct blood flow from the ventricular cavity throughout cardiac cycle (figure 5).Coronary angiography (CAG) revealed normal coronaries.

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Isolated left ventricular noncompaction cardiomyopathy (IVNC) is a congenital cardiomyopathy caused by a defect in endo-myocardial morphogenesis (1). The major clinical manifestations of IVNC are depressed left ventricular systolic and diastolic function, systemic embolism, ventricular arrhythmias, conduction disorders and neurological manifestations (2, 3). In literature, isolated left ventricular noncompaction cardiomyopathy was associated with severe mitral regurgitation, mitral valve prolapse(4), congenital cleft mitral valve(5), and mitral stenosis in a case of chronic rheumatic heart disease(6). The present case had bivalvular stenosis, chronic rheumatic heart disease accompanied with isolated left ventricular noncompaction cardiomyopathy.

TAKE HOME MESSAGES

Chronic rheumatic heart disease with noncompaction left ventricle may present with bivalvular stenosis, regurgitation.

REFERENCES

[1] Chin TK, Perl off JK, Williams RG, et al.Isolated non-compaction of left ventricular myocardium: a study of eight cases. Circulation 1990; 82:507-13.

[2] Agmon Y, Connolly HM, Olson LJ, et al.,Noncompaction of the ventricular myocardium. J Am Soc Echocar- diogr 1999;12:859-63.

[3] Pignatelli R, McMahon JC, Dreyer WJ, et al. Clinical characteri- zation of left ventricular noncompaction in children: a relatively common form of cardiomyopathy. Circulation 2003; 108:2672-8.

[4] Takashi,Igarashi ,Shinya Takase, et al., Left ventricular noncompaction complicated by mitral valve prolapse: report of a case , Surg Today (2013) 43:818–820.

[5] Amit Malviya , Shillong, Sundeep Mishra, et al., ,Left ventricular (LV) noncompaction associated with isolated congenital cleft mitral valve, Indian Journal of Cardiology, Jul-Dec,2011 Vol.14 No 3 & 4

[6] Rajesh Vijayvergiya, Ashish Jha, Rajasekhar Pal Pandian, et al.,Isolated left ventricular noncompaction in association with rheumatic mitral stenosis, International Journal of Cardiology, Volume 123, Issue 3, 24 January 2008, Pages 54– 56.