Large non restrictive apical ventricular septal defect curtailed by anomalous right ventricular muscle band : An rare variant double chambered right ventricle

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ABSTRACT

Anomalous muscle bands in right ventricle are uncommon findings with variable clinical presentations. We report an interesting case of a 6 year old asymptomatic boy with a large apical non restrictive ventricular septal defect and an anomalous muscle band in right ventricle. The muscle band divided the right ventricle into 2 chambers: smaller apical chamber which was connected to the left ventricle by an large apical non restrictive ventricular septal defect and proximal chamber which included right ventricular inflow and outflow tract. The hemodynamic burden of the ventricular septal defect was curtailed by the restrictive muscle band, hence patient remained asymptomatic. In total the condition behaved like a restrictive ventricular septal defect. Patient was kept under follow up and was not considered for surgical correction in view of negligible hemodynamic disturbances. Previously in literature 1 such case has been reported earlier.

Key words: Double chambered right ventricle, Echocardiography, Right ventricular anomalous muscle band, Restrictive, Ventricular septal defect.

INTRODUCTION

A 6 year old asymptomatic boy came for the evaluation of murmur which was detected on routine clinical examination. He denied history of breathlessness, chest pain, palpitation or any other cardiovascular symptoms. Patient's growth and developmental history were normal. His general physical examination was unremarkable. Cardiovascular examination revealed normal S1 and S2, and grade 3/6 pan systolic murmur at the left lower sternal border. Electrocardiography was done which demonstrated normal QRS axis with no evidence of ventricular hypertrophy. His chest X-rays revealed normal cardiac shadow and pulmonary vasculature. 2D echocardiography showed a large apical muscular ventricular septal defect (17 mm) and an anomalous muscle band across right ventricular (RV) apex (Figure 1). The anomalous muscle bundles divided the RV into 2 parts: proximal chamber, and the distal apical part which is connected to the left ventricle through the large apical ventricular septal defect. Color Doppler showed non restrictive flow across the ventricular septal defect and a restrictive left to right flow across the anomalous muscle band, with a peak systolic Doppler gradient of 100 mm Hg across the muscle band (Figure 2). Echocardiographically there was no evidence ventricular hypertrophy and left to right shunt fraction was insignificant in spite of large ventricular septal defect. Right heart catheterization and angiography was done which revealed muscle band dividing the right ventricle into larger proximal chamber and distal apical chamber. Distal apical chamber could not be visualized in right ventricular injection because of the restrictive muscle band with left to right flow (Figure 3).

Inspite of presence of a large ventricular septal defect, patient was asymptomatic and there were no evidence of hemodynamic consequences because of the restrictive nature of the muscle band. Conservative management with medical follow up was considered as the best treatment option with infective endocarditis prophylaxis.

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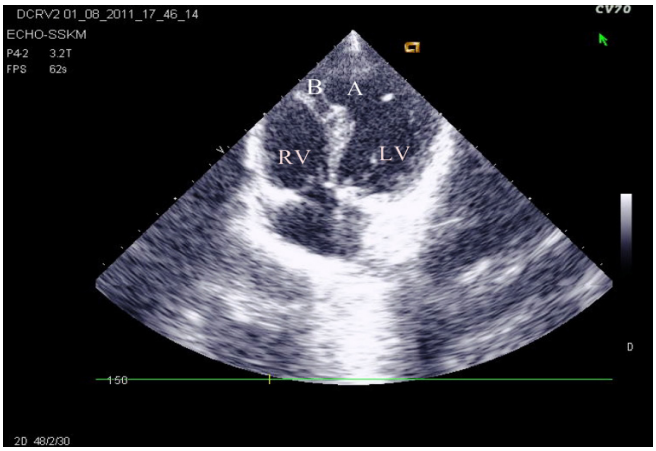

Figure 1: -2 D echocardiography showing a large apical muscular Ventricular septal defect and an anomalous muscle band across RV apex.

(LV-Left ventricle, RV- Right ventricle, A-VSD, B -Muscle band)

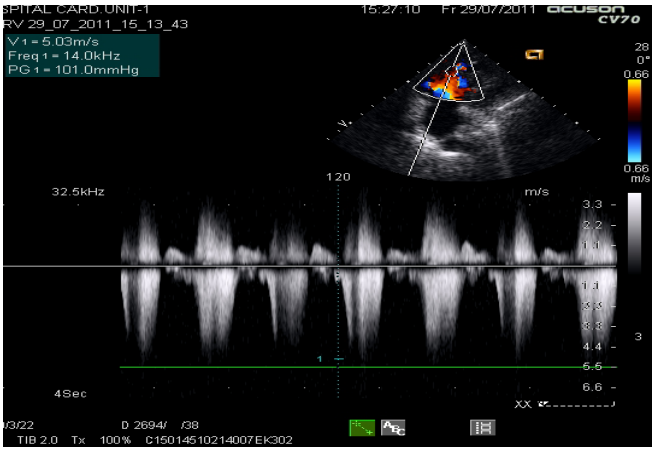

Figure 2: -2 D Color Doppler showing restrictive left to right flow across the anomalous muscle band, with a peak systolic Doppler gradient of 100 mm Hg across the muscle band

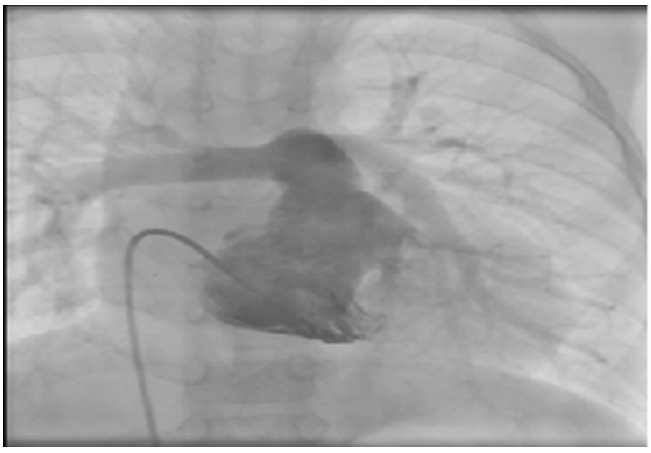

Figure 3: -Right ventricular angiography picture showing muscle band dividing the right ventricle into larger proximal chamber and distal apical chamber which couldn't be Visualized because of restrictive left to right flow across the muscle band.

DISCUSSION

Anomalous muscle bundles when found in the right ventriclular apical region are generally of little functional Significance. On the other hand a muscle bundle situated across the main channel of the right ventricular cavity can cause haemodynamic disturbances, especially when it becomes hypertrophied. Muscle bands across the RV cavity usually divides the RV into 2 chambers, called double chambered right ventricle first reported[1,2] in 1962. Since then a number of cases and case series have been published.[3,4] Usually the anomalous muscle bands divide right ventricle in to 2 chambers with proximal high pressure chamber connected to the inflow and a distal low pressure chamber Connected to the RV outflow.[5] It is commonly associated with anomalies as ventricular septal defect, Right ventricular outlet tract obstruction. Clinically presentation can mimic ventricular septal defect, pulmonary stenosis or tetralogy of fallot.[6]

The index reported case the muscle band in the RV apex divided the right ventricle in to 2 chambers: proximal chamber with both RV inflow and outflow, distal smaller chamber in RV apex which was in communication with the left ventricular cavity via large non restrictive apical muscular ventricular septal defect. The hemodynamic consequences and symptoms of the large non restrictive ventricular septal defect were curtailed by the restrictive muscle band in the right ventricle. Overall clinically, echocardiographically and hemodynamically condition behaved like restrictive ventricular septal defect with left to right shunt.

Small apical muscular ventricular septal defects can close spontaneously but larger defects often persist and needs treatment.[7] In our case it was thought to be unnecessary as there were negligible chances of congestive cardiac failure and pulmonary artery hypertension. Hence it was decided against surgical correction and planned to follow up medically with infective endocarditis prophylaxis. Long term prognosis is expected to be good.

Though number of cases of anomalous muscle band with typical Double chamber right Ventricle has been reported in literature; our case is atypical in nature as discussed earlier. There has been one similar case reported in literature, where an 11 year old asymptomatic girl was found to have an apical muscular ventricular septal defect that was a large defect but behaved like a small defect because of

the restrictive flow across the anomalous muscle bundles in the right ventricular apex, where the patient was kept under medical follow up.[8] On the literature search we also found another anatomically similar case where a in a 48-year old male echocardiography and magnetic resonance imaging revealed a large apical ventricular septal defect with separation of the right ventricular apex from the remaining RV by excessive trabeculations, thereby elimination of any left to right shunt across the Ventricular septal defect. Physiologically there was no hemodynamic disturbance so patient was kept under medical follow up.[9] So this type of apical VSD along with apical muscle bands constitutes a rare and distinct type of morphology and physiology, and treatment needs to be individualized based on the hemodynamic disturbance it causes.

REFERENCES

1. Lucas RV JR, Varco RL, Lillehei CW, *et al*. anomalous muscle bundle in the right ventricle. Hemodynamic consequences and surgical considerations. Circulation 1962; 25(1): 443.
2. Hartmann AF JR, Tsifutis AA, Arvidsson H, *et al*. The two chambered right ventricle: Report of nine cases. Circulation 1962; 26(2): 276.
3. Coates JR, Mcclenathan JE, Scot LP. The double-chambered right ventricle: A diagnostic and operative pitfall. Amer J Cardiol. 1964; 14(4): 561.
4. Hartmann AF JR, Goldring D, Carlson E. Development of right ventricular ostruction by aberrant muscle bands. Circulation 1964; 30(5): 679.
5. 5.Alva C, Ho SY, Lincoln CR, *et al*. The nature of the obstructive muscular bundles in double-chambered right ventricle. J Thorac Cardiovasc Surg. 1999; 117(6): 1180–9.
6. Perloff JK, Ronan JA jir, Deleon AC. Ventricular septal defect with the "two chambered right ventricle. Amer J Cardiol. 1965; 16(6): 894.
7. Shirali GS, Smith EO, Geva T. Quantitation of echocardiographic predictors of outcome in infants with isolated ventricular septal defect. Am Heart J. 1995; 130(6): 1228-35.
8. Khositseth A. Large apical muscular ventricular septal defect: asymptomatic due to anomalous muscle bundles in the right ventricle Congenit Heart Dis. 2007 Jan-Feb; 2(1): 70-3.
9. Grittner F, Daehnert I, Gutberlet M, *et al*. Late diagnosis of a congenital apical ventricular septal defect with complete closure by right ventricular trabeculations. European heart Journal cardiovascular Imaging 2014; 15(2): 230.