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Double Chamber Right Ventricle (DCRV) With Ventricular Septal Defect in Adulthood.

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ABSTRACT

Double-chambered right ventricle (DCRV) is a form of right ventricular outflow tract obstruction. It typically presents in childhood or adolescence. Only a handful of previous cases have been described in which DCRV occurred in adulthood. We report here a case of DCRV with ventricular septal (VSD) presenting in adulthood.

Keywords: Congenital Heart Disease, Double Chamber Right Ventricle, Ventricular Septal Defect

INTRODUCTION

Double-chambered right ventricle (DCRV) is a rare form of congenital heart disease characterized by right ventricle outflow tract obstruction by an anomalous muscle band that divides right ventricle (RV) into proximal high pressure and distal low pressure chamber.[1-3] The anomaly is rare, seen in only 0.5–2% of all cases of congenital heart disease[4] It usually presents in childhood and adolescence with most reported cases in patients less than 20 years old. Occasionally, however, patients can present with this condition in adulthood.[5,6] We report here a case of DCRV with ventricular septal defect (VSD) presenting in adulthood.

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CASE REPORT

A 25 Yrs male presented with seven days history of fever with productive cough. There was no h/o PND, Haemoptysis, tuberculosis. At presentation he was febrile, RR-18/mm, BP-110/70 mmhg. General physical examination was normal. Respiratory system showed few crepitations bilaterally. Cardiovascular examination showed normal heart sounds with systolic murmur grade 3/6 in left Intercostal space, best heard in 3rd ICS without any radiation. Nervous system examination was normal. A diagnosis of VSD with LRTI was made. HB, TLC, DLC was normal. ECG and X-Ray chest was normal. On 2D-Echo it showed adult congenital heart disease with small perimembranous VSD with left to right shunt with DCRV with pressure gradient of 80 mmhg between two right ventricular chambers [Figure 1]. Cardiac MRI with contrast showed a thick chordae / muscular band running from the septal wall near valve to the right ventricle free wall suggestive of double right ventricle with lower and upper chambers [Figure 2]. The Pulmonary valve and pulmonary artery were normal. A very thin 2-3 mm subaortic membranous VSD was observed. Final diagnosis was Double chamber RV with subaortic-membranous VSD.

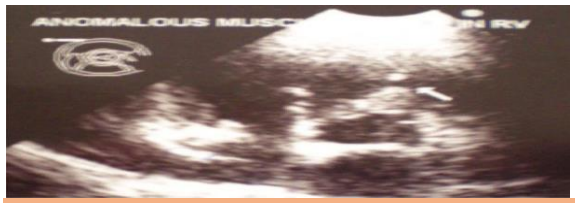

Figure 1: 2D ECHO showing prominent anomalous muscle band in right ventricle cavity.

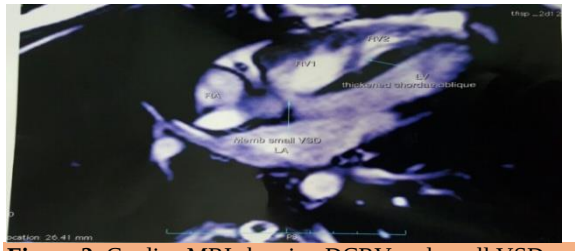

Figure 2: Cardiac MRI showing DCRV and small VSD.

DISCUSSION

DCRV is a rare form of congenital heart disease in which right ventricle is divided into a high pressure inlet portion and a low pressure outlet portion by an anomalous muscle bundle.[2,3] Localisation of the obstructive muscle varies and may be either high (or horizontal), adjacent to the pulmonary valve, or low (or oblique), close to the apex. The anomaly is rare, seen in only 0.5–2% of all cases of congenital heart disease[4] Most cases of DCRV are diagnosed during childhood, and very few reports include cases in which the patients present in adulthood.[5,6] DCRV is exceptionally rare as an isolated anomaly. Most commonly it is associated with a membranous type VSD.[2,3] The correct diagnosis of DCRV is rarely made clinically and confused clinically with tetralogy of Fallot or isolated infundibular stenosis. Because of the unique nature of this anomaly, diagnostic problems are encountered which, if not properly recognized, may lead to inappropriate surgical treatment. Transthoracic echocardiogram although diagnostic sometime fails to demonstrate anomalous muscle band.[5,6] DCRV should be strongly suspected in ECHO if RV hypertrophy is present in absence of infundibular hypertrophy or valvular pulmonary stenosis. MRI or HRCT demonstrates exact anatomy of anomalous muscle bundle and helps in confirmation of diagnosis. Surgical intervention should be considered in all patients who are symptomatic or have peak gradients of ≥50 mm Hg.

CONCLUSION

The surgical approach resects the muscle bundles and repairs any associated lesions. Most patients do well after surgical intervention and lead fairly unrestricted lives.

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