Double Chambered Right Ventricle: A Rare Diagnosis

Nitesh Pansari¹, H Raghavendra¹, Hemant Mahur², Mahesh Dave³

Abstract

A 27 years old female was admitted to our hospital with complaints of swelling of feet and abdomen, pain abdomen and exertional dyspnea from last 1 week. On examination she was found to have congestive heart failure. Chest x-ray revealed mild cardiomegaly with left pleural effusion and electrocardiography showed right axis deviation with right ventricular hypertrophy. By echocardiography she was diagnosed to have double chambered right ventricle without any other congenital heart anomaly. She was started on medical treatment following which she recovered well and she was advised for surgery. This case is unique as usually double chambered right ventricle is associated with other cardiac malformations, common ventricular septal defect, pulmonary stenosis and aortic stenosis but no such association was present in this case.

Introduction

Double-chambered right ventricle is a developmental cardiac anomaly characterized by aberrant muscular bands which obstruct the body of the right ventricle dividing it into a high-pressure proximal chamber and a low-pressure distal chamber.¹ It typically presents in infancy or childhood but has been reported to present rarely in adults as in our case.

Case Summary

A 27 years old female patient admitted with complaints of swelling of feet and abdomen, pain abdomen and exertional dyspnea for past 1 week. She had a full term normal vaginal delivery before 3 months without any adverse event.

On examination she had pallor, raised jugular venous pressure, bilateral pitting pedal edema, ascites and congestive hepatomegaly. Clinical examination of cardiovascular system revealed a palpable P2 with a systolic thrill at pulmonary area with left parasternal heave and a grade IV/VI systolic murmur at mitral and tricuspid area. So a clinical diagnosis of right ventricular failure was established.

We performed a routine blood work up which showed anemia with a hemoglobin level of 7.5 g/dl and raised erythrocyte sedimentation rate with normal liver and kidney function tests. Chest x-ray displayed cardiomegaly with right ventricular dilatation and left pleural effusion (Figure 1). Electrocardiography was suggestive of right axis deviation and right ventricular hypertrophy. Ultrasound of abdomen demonstrated congestive hepatomegaly with mild to moderate ascites and prominent hepatic veins and inferior vena cava indicating a cardiac pathology.

Echocardiography showed dilated right atria and ventricle with prominent muscle bundle in distal right ventricle body near infundibular os (PSG=120 mm of hg between proximal and distal right ventricular chambers) which led to a diagnosis of double chambered right ventricle with right ventricular dysfunction (Figure 2). Medical therapy with diuretics and hematinics helped the patient to get rid herself of symptoms. Then she was advised to consult a cardiac surgeon for further management.

Discussion

The double-chambered right ventricle is a rare congenital heart disorder involving two different right ventricle pressure compartments that is often associated with malalignment ventricular septal defect (VSD). Usually, the obstruction is caused by an anomalous muscle bundle crossing the right ventricle from the interventricular septum to the right ventricle free wall.²

It can be caused by the presence of anomalous muscle tissue, hypertrophy of the endogenous trabecular bands, or an aberrant moderator band; all of which will typically result in progressive obstruction of the outflow tract.³

Frequent associated lesions include ventricular septal defect (VSD), pulmonary valve stenosis, and discrete subaortic stenosis.

As outlined by Restivo et al, several subtypes of divided right ventricle are noted.⁴ These subtypes include anomalous septoparietal band, anomalous apical shelf, hypertrophy of apical trabeculations, anomalous apical shelf with Ebstein malformation, and sequestration of the outlet portion of the ventricle from a circumferential muscular diaphragm in patients with tetralogy of Fallot. Double-chambered right ventricle, the most common form, is noted by the presence of anomalous muscle bundles (AMB) that divide the right ventricle into 2 chambers. However, no uniformity is observed in the position of these anomalous muscle bundles or in the manner in which the right ventricle is divided.

The lesion makes up approximately 0.5-2% of congenital heart disease and occurs in as many as 10% of patients with ventricular septal defect (VSD). Male-to-female ratio is 2:1.

Clinically, patients with double-chambered right ventricle and no ventricular septal defect (VSD) resemble patients with isolated pulmonary valve stenosis. When a ventricular septal defect (VSD) is present, the clinical picture relates to a ventricular septal defect (VSD). Usually, the patient is diagnosed with a ventricular septal defect (VSD) or pulmonary outflow tract obstruction and, subsequently, may show signs of progression of the outflow obstruction, such as cyanosis, fatigue, and decreased exercise tolerance Patients with severe right ventricle (RV) hypertension.

¹Final Year MD Medicine, ²Associate Professor, ³Professor, RNT Medical College, Udaipur, Rajasthan
Received: 18.04.2016; Accepted: 02.01.2017
may present with cyanosis, right ventricle (RV) failure, failure to thrive, and fatigue. Association with other syndromes is well recognized, and double-chambered right ventricle may be found during their workup.

Most patients are nondysmorphic and acyanotic with normal peripheral examination findings. Auscultation reveals a variable intensity of the second heart sound. A holosystolic ejection murmur, which peaks in intensity near mid-systole, with greatest intensity at mid-left and upper-left precordial areas, characterizes double-chambered right ventricle. An right ventricle heave, hepatomegaly, and tachypnea indicate right ventricle (RV) hypertension.

Electrocardiographic findings in double-chambered right ventricle were reviewed in one series of 30 patients. Almost 50% of the patients had evidence of right ventricular hypertrophy (RVH), 40% of them demonstrated an upright T wave in V3R in the absence of other findings of right ventricular hypertrophy (RVH), and the remainder had normal electrocardiographic findings. Similar findings are reported in other series.

Chest radiography may reveal either a left-to-right shunt with increased pulmonary vascular markings or a severe right ventricle (RV) obstruction with diminished pulmonary vascularity. The usual arrangement includes atrial situs solitus, levocardia, and left aortic arch. Cardiomegaly may be seen in some patients.

Echocardiography currently enables diagnosis on a 2-dimensional Doppler echocardiogram; before its advent, diagnosis of double-chambered right ventricle (DCRV) could not be made noninvasively. In infancy, subxiphoid imaging is optimal; parasternal short-axis views may be more useful in older patients. The cardinal feature is demonstration of muscle bundles that traverse the right ventricle (RV) cavity, with an accompanying gradient starting proximal to the infundibulum. Wong et al described a “displacement index,” which is determined by dividing the distance from the pulmonary annulus to the septal insertion of the moderator band by the tricuspid annulus diameter. An index less than 1 may predict that infants with ventricular septal defect (VSD) are at risk of developing an obstruction from a displaced moderator band.

Transesophageal echocardiography has been used to define structures in older patients with poor windows.

Indications for surgery: a simple dual-chamber right ventricle, the patient’s symptoms obvious, right ventricular hypertrophy, evidence electrocardiogram, chest radiograph, right heart pressure chamber systolic blood pressure reached 70mmHg average pressure reaches a pressure gradient of 25mmHg or more, or the high and low pressure chamber exceeds 40mmHg. Merge other the Correction heart malformations.

Conclusion

When a young patient presents with symptoms and signs of right heart failure, a differential diagnosis of Double chambered right ventricle should be considered and patient should be investigated keeping this diagnosis in mind. Double chambered
right ventricle is a potentially treatable condition by surgery, so patient can have a fruitful and healthy life following right diagnosis and proper management. Double chambered right ventricle usually has associated congenital cardiac anomalies but it can also present as a single isolated lesion as in this patient.

References