CASE REPORT

Double-Chambered Right Ventricle Associated with Coronary-Cameral Fistula

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ABSTRACT

Coronary arteriovenous fistulae are rare but major coronary anomalies. Association with congenital heart defect is still very rare. We present the case of a 40-year-old female with ventricular septal defect and double-chambered right ventricle detected to have coronary-cameral fistula during routine coronary angiography and underwent successful surgical intervention for the same without any post-operative complications.

Key words: Coronary-cameral fistula, coronary steal, double-chambered right ventricle, ventricular septal defect

INTRODUCTION

Coronary arteriovenous fistulae connecting a major coronary artery to one of the cardiac chambers or other vascular structures are a very rare occurrence amounting 0.4% of the cardiac anomalies.[1] This can be either congenial or acquired. A coronary-cameral fistula associated with a congenital anomaly like double-chambered right ventricle is very rare and we could find limited cases in English literature. We describe the case of a 40-year-old female with double-chambered right ventricle and ventricular septal defect (VSD) with associated coronary-cameral fistula.

CASE REPORT

A 40-year-old female presented to our clinic with a history of progressive breathlessness on exertion for the past 5 years. There was no history of cyanosis or spells. Physical examination revealed a regular pulse rate of 86/min, tachypnea on exertion, and room air saturation between 90% and 92%.

Chest X-ray revealed significant scoliosis with rotated heart. Two-dimensional echocardiography showed a large subaortic VSD and double-chambered right ventricle with hypertrophied muscle bundles causing a gradient of 110 mmHg across the right ventricular outflow tract (RVOT). Since she was 40 years of age, we proceeded to a coronary angiography as part of routine evaluation before open-heart surgery. Coronary angiogram revealed a fistulous communication from distal left anterior descending (LAD) as well as RV branch of the right coronary artery (RCA) to the right ventricle. A cardiac computed tomography [Figure 1] confirmed the location of coronary-cameral fistula from the LAD and RCA to the right ventricle.

She underwent median sternotomy. Under standard cardiopulmonary bypass instituted by aortic and bicaval cannulation and root cold blood cardioplegic arrest, the right atrium was opened. The VSD was closed and obstructing RVOT muscle bundles were resected. The LAD coronary artery was opened in the distal portion and multiple fistulous tracts were identified [Figure 2a].

Since the fistulous tracts were nearer to the left ventricular apex, distal LAD was ligated off. The RV branch from the right coronary artery [Figure 2b] was also ligated.

After releasing the aortic cross-clamp, she was weaned off cardiopulmonary bypass support slowly with inotropic

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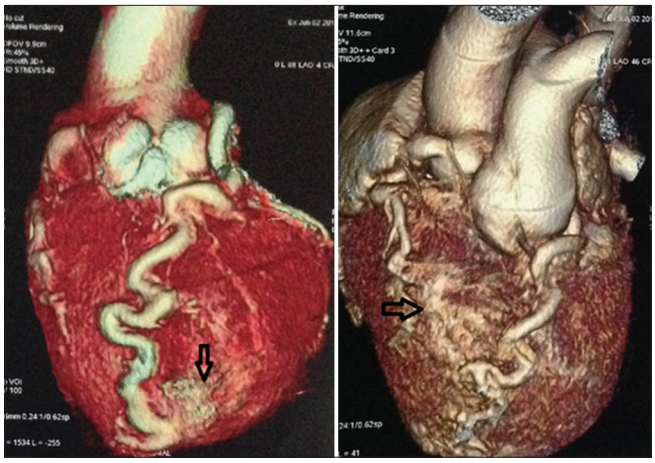

Figure 1: Computed tomography image showing the right ventricle filling from the left anterior descending and right coronary artery (arrows)

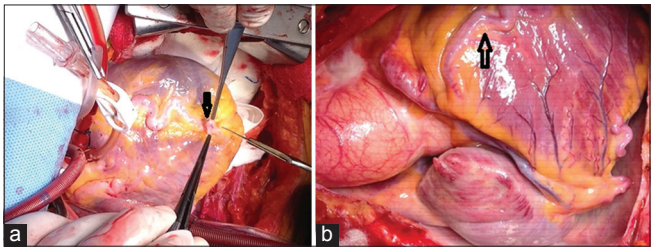

Figure 2: (a) Intraoperative image of open distal left anterior descending (LAD) at the site of fistula (arrow). (b) Ectatic right coronary with the right ventricular branch (arrow)

support. Post-operative echocardiography confirmed the absence of any residual shunts and wide opened RVOT with good biventricular function without any regional wall motion abnormalities (RWMA). She had an uneventful post-operative course and was discharged after 7 days.

At 3-year follow-up, she is asymptomatic with good biventricular function and no residual VSD/RVOT obstruction/RWMA.

DISCUSSION

Association of coronary-cameral fistula with a congenital cardiac anomaly is a rare occurrence. As there was no significant right to left shunt, it was missed in the routine echocardiography. Coronary angiography as well as computed tomography showed ectatic coronaries with filling of the RV chamber.

Since the right and left ventricular chambers were having similar pressures due to large unrestricted VSD and RVOT obstruction, there was no significant flow across the fistula in echocardiography. However, surgical correction of the cardiac anomaly by closing the VSD and relieving the RV outflow obstruction results in a pressure gradient between the two chambers. This can lead to the left to right shunting through the fistulous communications resulting in a coronary steal-like phenomenon producing angina.[2] Furthermore, this abnormal hemodynamics will result in further enlargement of already ectatic coronary arteries, leading to cardiac failure, arrhythmias, and coronary rupture at a later stage. Taking these facts into account, it was decided to address the fistulae also during the surgical correction of the defect.

To the best of our knowledge, we could not find any association with these two disorders after the report of Saxena *et al*.,[3] in 1990. Lee *et al*.[4] described the existence of a multidirectional coronary fistula presenting with symptoms detected 24 years after surgical correction of Tetralogy of Fallot. Hence, it is important to manage them in the initial intervention itself to avoid the possibility of complications in future.

CONCLUSION

Coronary cameral fistula can rarely be associated with tetralogy physiology. Although asymptomatic initially, corrective intracardiac surgery can lead to occurrence of symptoms later on in life. Hence it is important to manage them in the initial intervention itself to avoid the possibility of complications in future.

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How to cite this article: Nair VV, Chooriyil N, Kathayanatt JT, Nair JTK. Double-Chambered Right Ventricle Associated with Coronary-Cameral Fistula. J Clin Cardiol Diagn 2019;2(2):1-2.