

CASE REPORT

Right Ventricular Outflow Tract Obstruction in a Middle Aged Man with Right-Sided Aortic Arch

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ABBREVIATIONS

DCRV = double-chamber right ventricle
MCTA = multidetector computer tomographic angiography
MRI = magnetic resonance imaging
RAoA = right-sided aortic arch
RVOT = right ventricular outflow tract
VSD = ventricular septal defect

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ABSTRACT

Congenital diseases causing an obstruction of the right ventricular outflow tract are difficult to precisely diagnose, especially in elderly patients. We report a case of a 49-year-old man who presented to our hospital with longstanding shortness of breath on exertion. He was finally diagnosed as right ventricular outflow tract obstruction and referred for surgical correction. The patient had a history of descending aorta dissection which was treated by thoracic stent grafting. By that time right-sided aortic arch was diagnosed. The coexistence of right ventricular outflow tract obstruction and right sided aortic arch in the same patient is very rare, to the best of our knowledge.

INTRODUCTION

Congenital diseases causing an obstruction of the right ventricular outflow tract (RVOT) are difficult to precisely diagnose, especially in elderly patients.¹ Obstruction to the RVOT may be the result of abnormalities at the mid-right ventricle, the infundibulum, the pulmonary valve, the supra-ventricular region, or the branch and/or peripheral pulmonary arteries. Previous surgery may be responsible for some obstructive lesions as well.^{1,2} Right-sided aortic arch (RAoA) is an uncommon congenital anomaly with a frequency of approximately 0.1% in the general population.^{3,4} Right-sided aortic arch results from involution of part of left dorsal aorta and persistence of right dorsal aorta. In situs solitus, RAoA is typically associated with cardiac malformations in the outflow tract as it is described in tetralogy of Fallot, pulmonary atresia or common arterial trunk.¹ Herein, we report a case of a 49-year-old man who was found to have RAoA with isolated RVOT obstruction.

CASE REPORT

A 49-year-old man presented to our hospital with longstanding New York Heart

RVOT OBSTRUCTION AND RIGHT-SIDED AORTIC ARCH

Association class II shortness of breath on exertion. His past medical history was significant for hypertension, known for more than 10 years, as well as for dyslipidemia, both controlled with medication. His past surgical history included endovascular stent grafting of the lower part of the descending thoracic aorta for type B aortic dissection 3 years earlier. During that hospitalization a right-sided aortic arch with aberrant left subclavian artery was diagnosed. Besides, in his childhood medical records, a supracristal ventricular septal defect (VSD) was reported, which has not been confirmed. He had a 20-pack-year history of cigarette smoking but stopped smoking 3 years ago. The patient had a family history of coronary artery disease.

On recent admission he was in sinus rhythm and his blood pressure was 120/75 mmHg, on medication. On examination nothing abnormal was detected. A surface electrocardiogram showed negative T-waves in leads II, III, aVF and V1-V5, signs of right ventricular overload, without arrhythmias. A chest x-ray showed the endovascular stent graft in the descending aorta and no sign of pulmonary dilatation. An echocardiographic evaluation, including transthoracic and transesophageal echocardiogram, revealed diffuse right ventricular hypertrophy and a severe obstruction at the right ventricular infundibulum (RVOT Vmax = 6 m/sec) (Fig. 1). There were no signs or findings of right heart failure. The left ventricle was mildly hypertrophied with normal systolic function. The pulmonary valve appeared thickened with mild restriction in opening. Pulmonary artery and branches were normal in size. There was mild tricuspid regurgitation. The false lumen in the descending aorta was thrombosed with no residual flow. Although tetralogy of Fallot was suspected at first, the diagnosis was not confirmed because the ventricular septum was intact, and importantly there was no overriding

aorta. Hence a further work-up was performed.

Contrast right ventriculography with right heart catheterization showed mild pulmonary hypertension with systolic pulmonary artery pressure of 43 mmHg, whereas the right ventricular systolic pressure was markedly elevated at 170 mmHg producing systolic pressure gradient over the obstruction of 127 mmHg. Oxygen saturation indicated no evidence of shunt. The right ventricle appeared significantly hypertrophic. The most severe obstruction of RVOT was located at the infundibulum (Fig. 2). Double cavity right ventricle (DCRV) could not be completely ruled out. The pulmonary valve appeared normal in annular size with thickened cusps and mildly restricted opening.

Left heart catheterization showed a hypertrophic left ventricle, with well preserved systolic function. Left coronary angiography demonstrated filling of two different arteries arising from two separate ostia above the left coronary sinus. The first coronary artery had a course as diagonal and the latter as circumflex artery. Right coronary angiography demonstrated a normal right coronary artery. A branch arose from the proximal part of the right coronary artery and after a course between aortic root and RVOT continued as anterior descending branch. Right-sided aortic arch could be demonstrated



FIGURE 1. Transthoracic echocardiogram and Doppler study demonstrates severe RVOT obstruction (RVOT Vmax = 6 m/sec). RVOT = right ventricular outflow tract.



FIGURE 2. Right ventriculography depicting severe RVOT obstruction. RVOT = right ventricular outflow tract.

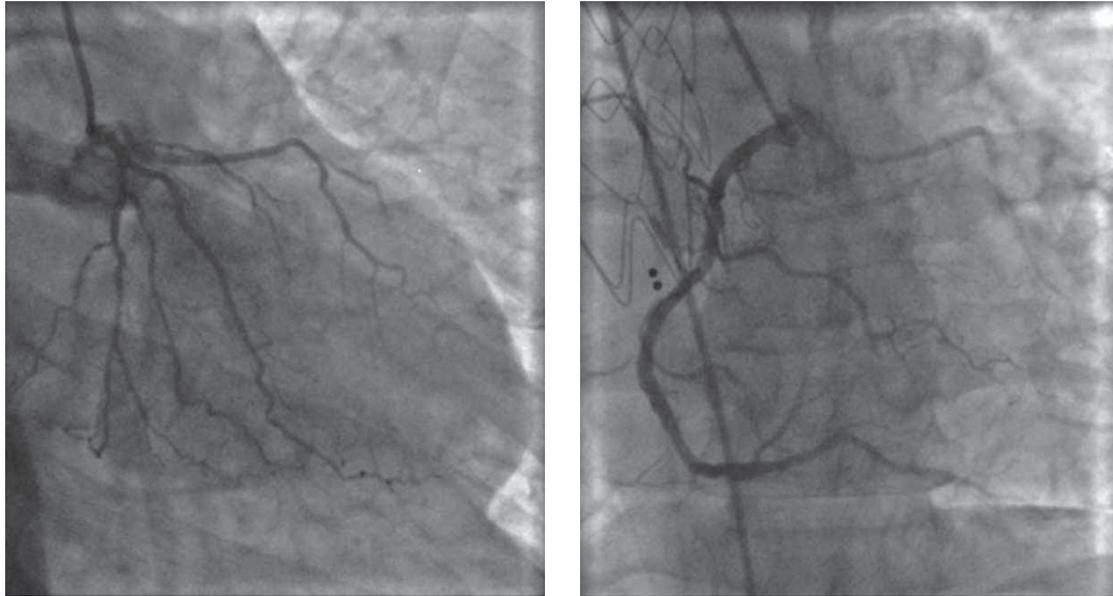


FIGURE 3. Left and right coronary angiography.

(Fig. 3). The above findings were confirmed by multidetector computer tomographic angiography (MCTA) (Fig. 4).

Magnetic resonance imaging (MRI) showed hypertrophic right ventricle with significant trabeculation. The obstruction of the RVOT was located at the infundibulum, with a diameter

less than 1 cm and peak velocity greater than 550 cm/sec at systole. Right and left ventricular volumes were calculated at end-diastole and end-systole. Right ventricular end-diastolic volume was 87 cm³, right ventricular end-systolic volume was 31 cm³; left ventricular end-diastolic volume was 84 cm³ and left ventricular end-systolic volume was 30 cm³. Right and left ventricular ejection fractions were well preserved (Fig. 5).

Cardiopulmonary exercise testing was performed on a bicycle ergometer. Work rate was increased with a ramp protocol. Duration of exercise was 5.6 min. Maximum work achieved was 60 watts. Anaerobic threshold was achieved at 8.7 ml/kg/min (26% of predicted value). Peak O₂ consumption (VO₂ peak) was 1.107 l/min (predicted value=2.709 l/m; VO₂max=14.8 ml/kg/min, 45% of predicted value). There was absence of chest pain or electrocardiographic abnormalities. The above results were consistent with severe cardiac impairment (Weber class C). The patient was referred for RVOT surgical reconstruction.

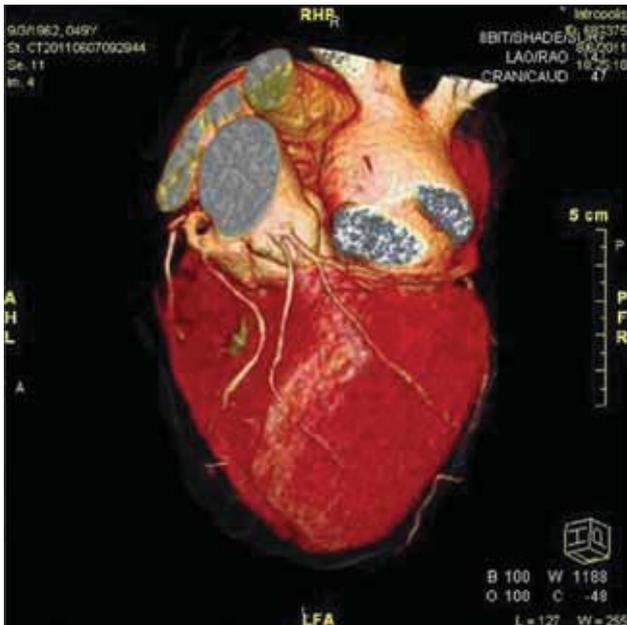


FIGURE 4. Multidetector computer tomographic angiography. Two arteries arose from separate ostia above the left coronary sinus.

DISCUSSION

Right-sided aortic arch (RAoA) is a rare congenital anomaly, presenting in 0.1% of the general population. Embryologically, the aortic arch is formed from the left aortic arch as the right aortic arch is receding. However, if the left aortic arch is degenerated instead of the right aortic arch, the aortic arch will be formed from the right aortic arch.⁴ Right-sided aortic arch can be classified into three types, depending on degenerating pattern of the left aortic arch and branching

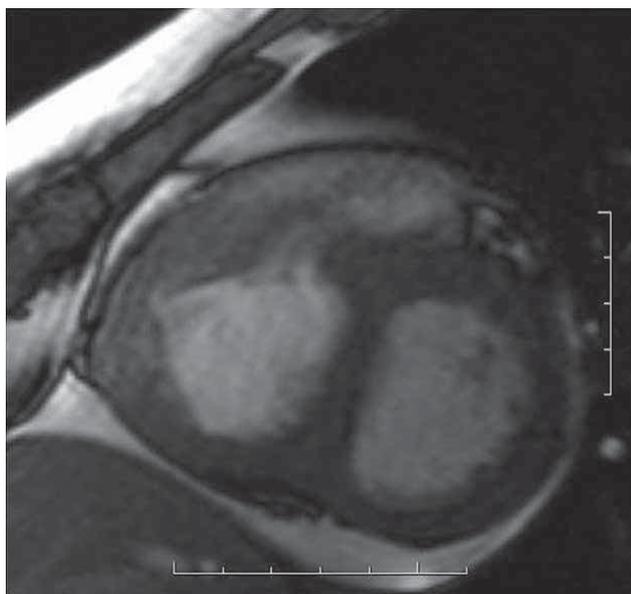


FIGURE 5. Magnetic resonance imaging revealing severe RVOT obstruction. RVOT = right ventricular outflow tract.

pattern of the great vessels. *Type I* (the mirror image type) is when the great vessels originate from the arch in the following order, left innominate artery, right common carotid artery and right subclavian artery. *Type II* (aberrant left subclavian artery), the most common type, is when the great vessels originate from the arch in the following order, left common carotid artery, right common carotid artery, right subclavian artery and left subclavian artery originating from Kommerell's diverticulum. *Type III*, very rare type, is when the left common carotid artery, right common carotid artery and right subclavian artery originate from the right aortic arch orderly and the left subclavian artery is not connected.^{3,4} In the present case, the great vessels originated from the arch in the following order, left common carotid artery, right common carotid artery, right subclavian artery, and aberrant left subclavian artery.

Right-sided aortic arch does not manifest symptoms necessarily and does not need surgical correction itself, except when associated with other congenital diseases including esophageal atresia, tracheoesophageal fistula, vascular ring and aortic arch anomaly around the trachea and esophagus, which can cause clinical symptoms like respiratory obstruction and dysphagia. Type I RAoA is associated with major congenital heart disease in 98% of patients, including tetralogy of Fallot, truncus arteriosus and tricuspid atresia. Type II is less frequently associated with major cardiac anomalies (5-10%).⁴ In the present case, the patient had RAoA associated with RVOT obstruction and there was neither VSD nor overriding aorta.

There are several congenital diseases producing an obstruction of RVOT due to abnormalities at the mid-right ventricle, the infundibulum, the pulmonary valve, the supra-

valvular region, or the branch and/or peripheral pulmonary arteries.² It is sometimes difficult to precisely diagnose such a disease especially in elderly patients because of its rarity and the difficulty in assessing the RVOT.¹

A RAoA, which is of no hemodynamic consequence, is present in one-quarter of patients with tetralogy of Fallot.^{4,6} The diagnostic importance of a right aortic arch accompanying a tetralogy of Fallot complex is of special significance in the so-called acyanotic variety (pink tetralogy of Fallot).⁶ In the present case, although uncorrected tetralogy of Fallot is rare in adults, a misdiagnosed pink tetralogy was considered with the assumption that a small VSD was spontaneously closed.⁷ Additionally to the above, a double-chamber right ventricle (DCRV) is included in the differential diagnosis. Double-chambered right ventricle is a rare form of congenital heart disease in which the right ventricle is divided into a high-pressure inlet portion and a low-pressure outlet portion by anomalous muscle bundles. This form of midcavitary obstruction in most cases is diagnosed and treated during childhood. Adult patients with this condition are often misdiagnosed, and without follow-up or repair, DCRV is a potential cause of morbidity and mortality.⁸ Double chamber right ventricle tended to be misdiagnosed as valvular or infundibular pulmonic stenosis, tetralogy of Fallot, and VSD with or without pulmonic stenosis.¹ The RVOT obstruction is generally believed to be an acquired one and to be progressive over time, although the basic anatomic features are congenital. In 75% of cases, it is associated with a perimembranous VSD that is usually, but not always, below the level of the muscular obstruction.² In the present case, the site of obstruction was at the infundibulum.

Although hypertrophic cardiomyopathy is classically considered a disease of the left ventricle, we found in the literature rare cases of biventricular hypertrophic cardiomyopathy with isolated RVOT obstruction. The mechanism by which obstruction to RVOT occurs in patients with hypertrophic cardiomyopathy results from greatly hypertrophied musculature comprised of crista supraventricularis, moderator band or trabeculae.⁹ Biventricular hypertrophic cardiomyopathy causing right- and/or left-side outflow tract obstruction, as well as valvular pulmonary stenosis, is relatively common in infants with Noonan syndrome. However, this condition without a dysplastic pulmonary valve, or any other valvular dysplasia, is rare in adults. The cardinal features of Noonan syndrome include unusual facies (i.e., hypertelorism, down-slanting eyes, webbed neck), congenital heart disease (in 50%), short stature, and chest deformity. Approximately 25% of individuals with Noonan syndrome have mental retardation.⁹ The above features were not observed in the present case.

In conclusion, we encountered a rare case of a middle-aged patient with right-sided aortic arch associated with isolated RVOT obstruction. For the detailed assessment of RVOT and the precise diagnosis, multiple modalities were used including transthoracic and transesophageal echocardiography, MCTA,

MRI, and right ventriculography with right heart catheterization. Cardiopulmonary exercise testing was used to confirm the cardiac cause of shortness of breath as well as to provide an accurate assessment of the impaired functional capacity. It is important to note that a patient with congenital disease and RVOT obstruction sometimes shows the first symptom at an older age, and surgical treatment is suggested to relieve the symptoms.

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