Clinical Case Report

Incidentally detected right aortic arch with mirror image branching in a patient with rheumatic calcific mitral valve disease

KRISHNA KUMAR MOHANAN NAIR, SANJAY GANAPATHI, SONALI INAMDAR, ARUN GOPALAKRISHNAN, TIRUR RAMAN KAPILAMOORTHY, AJITKUMAR VALAPARAMBIL

ABSTRACT

An isolated right-sided aortic arch with no congenital heart disease is extremely rare. We report an adult woman with long-standing rheumatic heart disease causing severe calcific mitral stenosis, moderate mitral regurgitation and moderate pulmonary hypertension, for which she underwent mitral valve replacement and tricuspid annuloplasty. On preoperative work-up, she was detected to have a right-sided aortic arch with mirror image branching along with a ductal dimple. However, there were no associated congenital cardiac defects.

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INTRODUCTION

An incidentally detected isolated right-sided aortic arch (RAA) is usually associated with tetralogy of Fallot or truncus arteriosus. Its occurrence without congenital heart anomalies is rare.

THE CASE

A 50-year-old woman was diagnosed to have rheumatic heart disease with severe calcific mitral stenosis, moderate mitral regurgitation and moderate pulmonary artery hypertension on evaluation for progressively worsening dyspnoea on exertion. She was in atrial fibrillation. In view of her functional class 3 status, she was planned for mitral valve replacement (MVR) and had a diagnostic coronary angiography, which revealed RAA. The aortic root angiogram showed RAA with mirror image branching, proximal right descending thoracic aorta and ductal dimple (Fig. 1a and b). A subsequent cardiac CT scan confirmed the same findings (Fig. 1c and d).

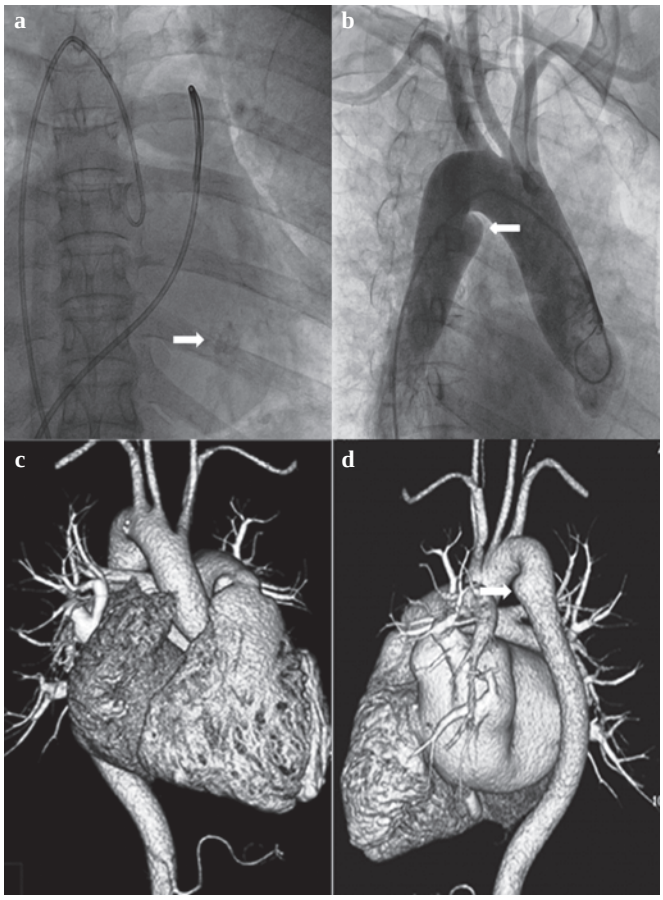

FIG 1. (a) Fluoroscopic image in anteroposterior view showing a pig tail catheter in the right descending thoracic aorta to right aortic arch to ascending aorta. An end-hole catheter is in the main pulmonary artery to left pulmonary artery. The arrow denotes the calcific mitral valve. (b) Fluoroscopic image in the right anterior oblique view of aortic root angiogram showing the right aortic arch with mirror image branching and ductal dimple (arrow). (c, d) CT reconstructed three-dimensional images showing the right aortic arch with mirror image branching and ductal dimple (arrow)

DISCUSSION

From an embryological viewpoint, the development of the aorta and its branches starts between the 4th and 7th weeks of gestation from five pairs of pharyngeal arch arteries in an asymmetrical manner. Evolution of the aortic arch and its branches occurs as a result of asymmetric involution and persistence of specific embryological arch structures, and congenital malformations of the aortic arch arise from disorders in the formation of the primitive double aortic arch.[1] Anomalies of great vessels are incidental findings as they are usually asymptomatic.[2] RAA is a relatively uncommon anomaly, occurring in approximately 0.05% of the population.[3] RAA has been classified into three types according to the branching pattern of the arch vessels: an RAA with an aberrant left subclavian artery (LSA), an RAA with mirror image branching and an RAA with isolation of the LSA. An RAA with mirror image branching is uncommon.[3] The left innominate

Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram 695011, Kerala, India
KRISHNA KUMAR MOHANAN NAIR, SANJAY GANAPATHI, SONALI INAMDAR, ARUN GOPALAKRISHNAN, AJITKUMAR VALAPARAMBIL Department of Cardiology
TIRUR RAMAN KAPILAMOORTHY Department of Imaging Sciences and Interventional Radiology

Correspondence to SANJAY GANAPATHI; *drsanjayganesh@gmail.com, sanjay@sctimst.ac.in*

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artery is the first branch arising from the arch, which is followed by the right carotid artery and right subclavian arteries. This anomaly is usually associated with cyanotic congenital heart disease, especially tetralogy of Fallot and truncus arteriosus.

An RAA associated with vascular rings is invariably completed by a patent ductus arteriosus or ligamentum arteriosus.[4] An RAA with aberrant LSA and a left patent ductus arteriosus is the most common RAA-type vascular ring. When an RAA with mirror image branching forms a vascular ring, it is usually associated with a ductus arteriosus from the descending aorta. If the ductus is closed, it forms the ductal dimple.

Our patient was an adult woman with long-standing rheumatic heart disease causing calcific mitral stenosis and was incidentally detected to have an RAA with mirror image branching and ductal dimple. No associated congenital cardiac defects were found at the time of evaluation. Aortic cannulation during open-heart surgery generally should not cause a problem since the cannulation is done in the ascending aorta. She successfully underwent MVR with partial posterior mitral leaflet preservation with tricuspid annuloplasty without any complications. We believe this is the first report of an adult with rheumatic mitral stenosis and an incidentally detected RAA with mirror image branching and ductal dimple with no congenital cardiac defects.

Conflicts of interest. None declared

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