A quadricuspid aortic valve that was mistakenly diagnosed as insidious rheumatic carditis: A 13-year-old case.

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Abstract

Quaricuspid aortic valve (QAV) is a very rare congenital heart defect characterized by the presence of four valves of the same or different sizes. This anomaly may cause progressive aortic regurgitation or rarely valve stenosis. The diagnosis of QAV may be difficult, misdiagnosed or overlooked if careful evaluation is not made. Here, a 13-year-old patient with QAV who presented to the outer center with a complaint of chest weight, who was mistakenly diagnosed with insidious rheumatic carditis due to aortic regurgitation, was presented, and and it was aimed to draw attention to this rare congenital heart valve disease.

INTRODUCTION

Quadricuspid aortic valve (QAV) is a very rare congenital cardiac abnormally, far less common than unicuspid or bicuspid aortic valve.[1] Aortic dilatation and other structural cardiac abnormalities in patients with QAV are relatively common. Aortic valve regurgitation is the main hemodynamic abnormality and the indication for aortic valve surgery in the majority of patients who undergoing surgery.[2] Here, a 13-year-old patient with QAV who presented to the outer center with a complaint of chest weight, who was mistakenly diagnosed with insidious rheumatic carditis due to aortic regurgitation, was presented, and and it was aimed to draw attention to this rare congenital heart valve disease.

CASE REPORT

A 13-year-old male patient, applied to pediatric cardiology at the outer center with chest pain 3 months ago. In transthoracic echocardiography performed there, 1st degree aortic regurgitation was detected, and secondary penicillin G prophylaxis was initiated considering the insidious rheumatic carditis. Laboratory tests performed in the outer center at that time were normal. The patient applied to our outpatient clinic for his second control, and he did not have any complaints. An anamnesis taken by us revealed that there was no history of acute rheumatic fever (ARF) in any period. The patient's physical examination was completely normal. In laboratory examination, hemogram, biochemical parameters, acute phase reactants and anti-streptolysin O titre were normal. 12-lead electrocardiogram (ECG) was normal. On transthoracic echocardiography, the aortic valve was quadriquspid (Figure 1) and there was a 1st degree aortic regurgitation (Figure 2). The patient was diagnosed with quatricuspid aortic valve and it was thought that aortic regurgitation was due to valve anomaly. Secondary penicillin prophylaxis, which was started for rheumatic carditis in the outer center, was stopped. The patient was followed up at regular intervals.

DISCUSSION

Quadricuspid aortic valve (QAV) is a very rare congenital cardiac anomaly with an incidence of 0.01–0.04%.[3] Aortic valve anomaly may be anatomically unicuspid, bicuspid or quadricuspid as in our case. The most common of these variants is the bicuspid valve, followed by the unicuspid valve.[2,4] The mechanisms of QAV development is not exactly known. Embryological truncus arteriosus is thought to develop as a result of

abnormal decomposition. In general, after septation of the arterial trunk, three mesenchymal swellings develop into semilunar leaflets of the aortic and pulmonary trunks. However, in QAV, the fourth cusp emerges during the early stage of truncal septation, resulting in either a different number of primordial aortic leaflets or abnormal cusp proliferation.[3]

QAV is often in the form of an isolated anomaly. However, anomaly such as coronary artery anomaly, atrial septal defect, ventricular septal defect, patent ductus arteriosus, fallot tetralogy, sinus valsalva fistula, subaortic fibromuscular stenosis, mitral valve regurgitation, mitral valve prolapse, asymmetric septal hypertrophy, and transposition of great arteries may be accompanied.[1,5,6] In our case, there was no other cardiac anomaly accompanying QAV, that is, it was in the form of isolated QAV. In our case, there was no aortic dilation that could be seen in some patients.

In patients with isolated QAV, clinical signs are generally not seen in childhood, the symptoms mostly appear after the age of 40. Significant valvular disorder often occurs after the 5th - 6th decade. Clinical symptoms of patients with QAV often depend on the functional status of QAV and associated disorders. There is often regurgitation in the valve, but rarely may be stenosis. Clinical findings mostly develop as a result of valve regurgitation. As valve regurgitation increases, there may be symptoms of heart failure such as palpitations, chest pain, shortness of breath, fatigue and syncope.[7,8]

When patients are evaluated quickly or carelessly, the diagnosis of QAV may be overlooked or the patient may be misdiagnosed. Our case was also evaluated by a pediatric cardiologist at the outer center, and the transthoracic echocardiography performed there revealed a 1st degree of regurgitation in the aortic valve. Meanwhile, although all other laboratory evaluations were normal and the patient had no history of ARF, the patient was taken to secondary penicillin prophylaxis, considering the insidious rheumatic carditis. However, when we carefully evaluated the patient with transthoracic echocardiography (TTE), we noticed that the patient's aortic valve was quadricuspid and 1st degree valve regurgitation developed as a result of QAV. Therefore, we thought that aortic regurgitation was not related to rheumatic carditis. After the diagnosis of QAV, secondary prophylaxis applied to the patient every 21 days for rheumatic carditis was stopped.

With a careful TTE, almost all patients with QAV may be diagnosed. In cases where valve anomaly is suspected and definitive diagnosis cannot be made with TTE, Transesophageal echocardiography or Cardiak CT may be needed for a definitive diagnosis.[8,9] Approximately 25-50% of patients with QAV have to need surgical intervention in older ages. Surgical options for QAV include aortic valve repair and aortic valve replacement. Tricuspidalization is a preferred technique for QAV repair.[2,10]

As a result; QAV is a very rare congenital cardiac anomaly. When evaluating patients in terms of rheumatic fever, patients should be evaluated in detail for monocuspid, bicuspid or quadricuspid aortic valve in case of valve regurgitation, which is thought to develop as a result of rheumatic carditis, with or without a patient's history, active complaint, or supportive laboratory finding. It should not be forgotten that, as in our patient, if the patient with this valve anomaly is mistakenly diagnosed with rheumatic carditis, this patient will unnecessarily have to have an intramuscular depot penicillin every 21 days until at least 40 years of age or for life, which is a very painful procedure for this patient and carries the risk of anaphylaxis.

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Figure legends.

Figure 1. Transthoracic parasternal short- axis view showing quadricuspid aortic valve (arrow). LA = left atrium; RA = right atrium; RV = right ventricle; MPA = main pulmonary artery

Figure 2. Transthoracic parasternal longt- axis view showing aort regurgitation (arrow). LA = left atrium; LV = left ventricle; Ao = aorta