OLGU SUNUMU CASE REPORT

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A Quadricuspid Aortic Valve that was Misdiagnosed as Rheumatic Carditis: A 13-year-old Case

Yanlış Romatizmal Kardit Tanısı Alan Kuadriküspit Aortik Kapak: 13 Yaşında Bir Olgu

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ABSTRACT Quadricuspid aortic valve anomaly is a very rare congenital heart anomaly. Its incidence is 0.01-0.04 percent. This anomaly is characterized by the presence of four aortic valves of the same or different sizes. This may often lead to progressive aortic regurgitation or rarely aortic valve stenosis. In aortic valve evaluation with transthoracic echocardiographic, if a careful examination is not performed, the diagnosis of quadricuspid aortic valve may be overlooked or may be misdiagnosed as insidious acute rheumatic carditis. Here, we presented a 13-year-old patient with quadricuspid aortic valve who was admitted to another center with chest pain, and was mistakenly diagnosed with insidious rheumatic carditis due to the aortic regurgitation detected on his transthoracic echocardiographic examination. We aimed to draw attention to this rare congenital heart valve disease.

Keywords: Aortic regurgitation; aortic valve; echocardiography

ÖZET Kuadriküspit aort kapak, anomalisi çok nadir görülen bir konjenital kalp anomalisidir. Bu anomalinin insidansı yüzde 0,01-0,04'tür. Bu anomali, aynı veya farklı boyutlarda dört aort kapağının varlığı ile karakterizedir. Bu durum, progresif aort yetersizliğine veya nadiren aort kapak darlığına yol açabilir. Transtorasik ekokardiyografi ile yapılan aort kapak değerlendirmesinde, dikkatli bir inceleme yapılmadığında, kuadriküspit aort kapak tanısı gözden kaçabilir veya sinsi akut romatizmal kardit olarak yanlış teşhis edilebilir. Bu yazıda, farklı bir merkeze göğüs ağrısı şikâyeti ile başvuran ve transtorasik ekokardiyografik incelemesinde saptanan aort yetersizliği nedeniyle yanlışlıkla sinsi romatizmal kardit tanısı alan 13 yaşında bir kuadriküspit aort kapak hastası sunduk. Ve bu nadir konjenital kalp kapakçık hastalığına dikkat çekmeyi amaçladık.

Anahtar Kelimeler: Aort yetersizliği; aort kapak; ekokardiyografi

Quadricuspid aortic valve (QAV) is a quite rare congenital cardiac anormaly, 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 undergo surgery.[2] Here, we presented a 13-year-old patient with QAV who was admitted to another center with chest pain, and was mistakenly diagnosed with insidious rheumatic carditis due to the aortic regurgitation detected on his transthoracic echocardiographic examination. We aimed to draw your attention to this rare congenital heart valve disease.

CASE REPORT

A 13-year-old male patient, applied to pediatric cardiology at another center with chest pain 3 months ago. On transthoracic echocardiography (TTE) performed there, mild 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

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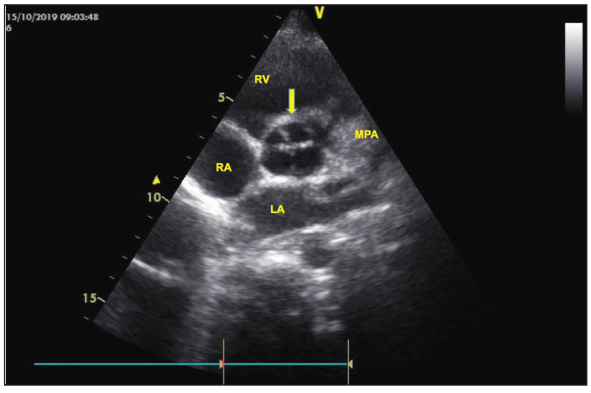

FIGURE 1: Transthoracic parasternal short-axis view showing quadricuspid aortic valve (arrow). LA: Left atrium; RA: Right atrium; RV: Right ventricle; MPA: Main pulmonary artery.

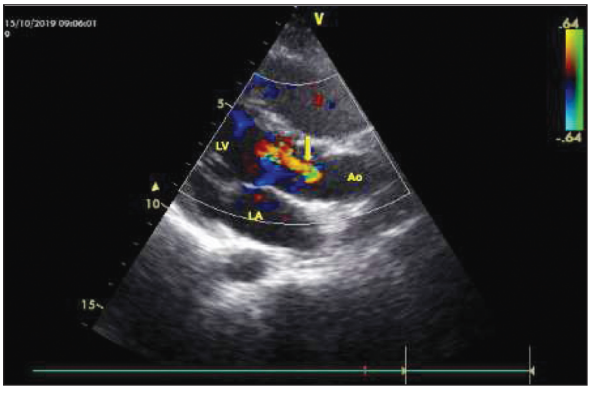

FIGURE 2: Transthoracic parasternal long-axis view showing aort regurgitation (arrow). LA: Left atrium; LV: Left ventricle; Ao: Aorta.

O titre were normal. The 12-lead electrocardiogram (ECG) was normal. On TTE, the aortic valve was quadriquspid (Figure 1) and there was mild aortic regurgitation (Figure 2). The patient's mitral valve was normal. The patient was diagnosed with quatricuspid aortic valve and it was thought that aortic regurgitation was due to valve anomaly. Secondary penicillin prophylaxis was discontinued. The patient was followed up at regular intervals.

Written informed consent was obtained from the parents.

DISCUSSION

QAV anomaly is a very rare congenital heart anomaly. Its incidence is 0.01-0.04 percent.[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 phase 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, coronary artery anomaly, patent ductus arteriosus (PDA), atrial and ventricular septal defect, Tetralogy of Fallot, sinus of valsalva fistula, mitral valve abnormalities, subaortic discrete membrane, and great artery transposition 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 stenosis may occur. 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 a different center, and the TTE performed there revealed mild 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 TTE, we noticed that the patient's aortic valve was

quadricuspid and mild 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. Similar to our case, Kosecik et al. described a seven-year-old patient with quadricuspid aortic valve who was false diagnosed with rheumatic carditis.[9]

With a careful TTE, almost all patients with QAV may be diagnosed. If a valve anomaly is suspected and a definitive diagnosis cannot be made with TTE, transesophageal echocardiography or cardiac computed tomography may be required for a definitive diagnosis.[8,10] Approximately 25-50% of patients with QAV need surgical intervention in older ages. Surgical options for QAV include aortic valve repair and aortic valve replacement. The technique applied in QAV repair is the tricuspidation technique.[2,11]

As a result; QAV is a very rare condition among congenital cardiac anomalies. Whether or not clinical signs or supportive laboratory findings of ARF are present, the patient should also be evaluated for monocuspid, bicuspid or quadricuspid aortic valve when evaluating for rheumatic carditis. It should be kept in mind that, as in our patient, if the patient with this valve anomaly is mistakenly diagnosed with rheumatic carditis, s/he will unnecessarily have to an intramuscular penicilin G benzathine every 21 days until at least 40 years of age or lifelong, which is a very painful procedure and carries the risk of anaphylaxis.

Source of Finance

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Mecnun Çetin;* ***Design:*** *Mecnun Çetin;* ***Control/Supervision:*** *Murat Başaranoğlu, Serap Karaman;* ***Data Collection and/or Processing:*** *Murat Başaranoğlu;* ***Analysis and/or Interpretation:*** *Eser Doğan, Serap Karaman;* ***Literature Review:*** *Mecnun Çetin, Eser Doğan;* ***Writing the Article:*** *Mecnun Çetin, Eser Doğan;* ***Critical Review:*** *Mecnun Çetin, Murat Başaranoğlu;* ***References and Fundings:*** *Serap Karaman;* ***Materials:*** *Eser Doğan.*

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