Case Report

Quadricuspid Aortic Valve Associated with Non-Obstructive Sub-Aortic Membrane: A Case Report and Review of the Literature

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Quadricuspid aortic valve (QAV) is a rare congenital cardiac entity. The recognition of QAV has clinical significance because QAV causes aortic valve dysfunction, commonly aortic regurgitation. QAV is often associated with other congenital cardiac abnormalities. Our case represents, to the best of our knowledge, the first reported case of QAV with a non-obstructive sub-aortic membrane.

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Quadricuspid aortic valve is a rare congenital cardiac entity. Recognition of the anomaly has clinical significance because it causes aortic valve dysfunction, commonly aortic regurgitation, and is often associated with other congenital cardiac abnormalities.

Case presentation

A 54-year-old man was referred to our outpatient unit for evaluation of a diastolic heart murmur. He was asymptomatic with no cardiovascular history except for arterial hypertension and he was taking angiotensin-converting enzyme inhibitors.

On physical examination his systemic blood pressure was 120/60 mmHg, his pulse was 73 beats per minute. A 3/6 diastolic murmur was audible over the left sternal border. Chest X-ray was normal and the ECG displayed normal sinus rhythm.

Transthoracic echocardiography showed moderate aortic valve insufficiency and mild mitral regurgitation. The left ventricle had mild eccentric hypertrophy with normal systolic function.

The aortic valve (AV) was quadricuspid and consisted of one large, one small, and two intermediate cusps in both systolic and diastolic frames (type D). The cusps showed a mild degree of thickening and calcification, but well preserved motion. On the short axis view of the AV the commissural lines formed by the adjacent cusps resulted in an "X" shape, whereas the normal tricuspid aortic valve has a "Y" shape.

A mobile, thin, fibrous membrane, which was attached to the basal anterior septum, was fluttering into the left ventricular outflow tract. There was no subvalvular obstruction, as no pressure gradient was identified across the membrane with Doppler echocardiography.

Transoesophageal echocardiography showed in mid-oesophageal short axis view the four cusps of the AV (Figure 1) and confirmed, in mid-oesophageal long axis view, the presence of the sub-aortic non-obstructive membrane (Figures 1 & 2). The smaller accessory cusp was located between the larger non-coronary cusp and the right-sided cusp. The cusps did not meet in the midline. The aortic root and ascending aorta were normal.

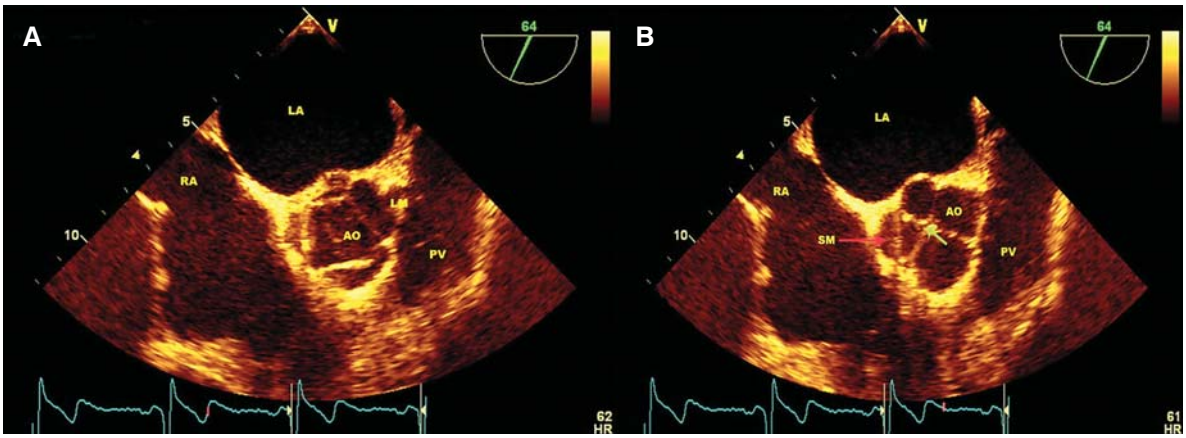

Figure 1. Mid-oesophageal aorta, short axis view, showing the quadricuspid aortic valve, which consisted of one large, two intermediate and one small cusp in both systolic (A) and diastolic (B) frames (type D). The cusps showed a mild degree of thickening and calcification, but well preserved motion. On diastole the commissural lines formed by the adjacent cusps resulted in an "X" shape (B). The smaller accessory cusp was located between the larger non-coronary and the left cusp. The green arrow in B shows the incomplete diastolic coaptation of the aortic cusps. The left main coronary artery (LM in A) was detected in the normal position. B also depicts the sub-aortic membrane (SM, red arrow). AO – aorta; LA – left atrium; PV – pulmonic valve; RA – right atrium.

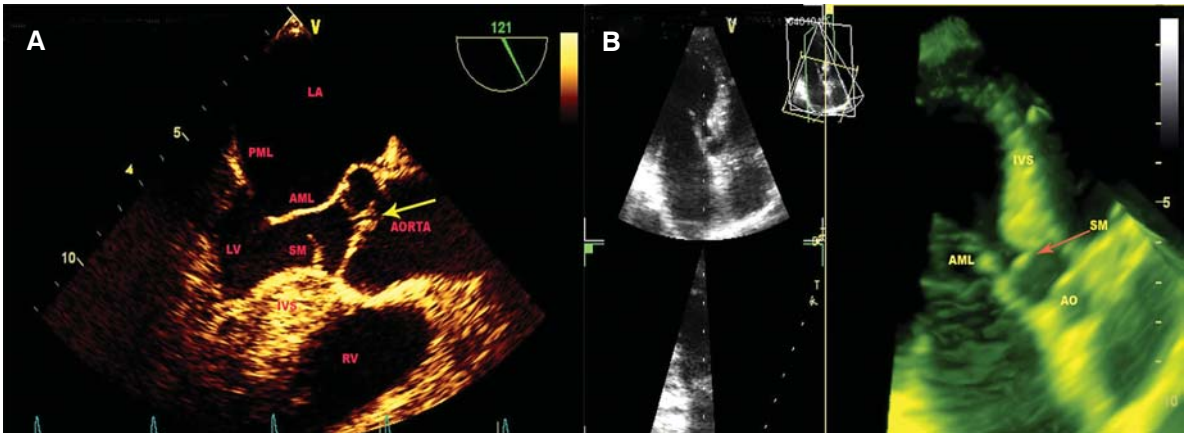

Figure 2. A. Aorta, long-axis, mid-oesophageal view shows a mobile, thin, fibrous membrane (SM) within the left ventricular outflow tract which is attached to the basal anterior septum. The yellow arrow indicates the incomplete diastolic coaptation of the aortic cusps. B. three-dimensional echocardiography showing the sub-aortic membrane (pink arrow, SM) in the left ventricular outflow tract. AML – anterior mitral leaflet AO – aorta; IVS – interventricular septum; LA – left atrium; LV – left ventricle; PML – posterior mitral leaflet; RV – right ventricle.

Moderate aortic regurgitation resulted from incomplete diastolic coaptation of the cusps. The left main coronary artery was detected in a normal position but the right coronary artery was not in the expected position. The patient refused to have a heart computed tomography in order to identify the coronary arterial system.

Having smoking and hypertension as predisposing factors for coronary artery disease, the patient underwent a treadmill test, which was non-diagnostic for ischaemia, and stress echocardiography with dobutamine, which was negative for ischaemia.

The patient was treated medically, remained asymptomatic, and was put on regular clinical and echocardiographic follow up.

Discussion

Quadricuspid aortic valve (QAV) is a rare congenital cardiac entity, with an incidence between 0.008% and 0.033% at autopsy and 1% in patients presenting for aortic valve surgery.[1,2] The first QAV was reported by Balington in 1862. About 200 cases have been published until today. Quadricuspid pneumonic valve is

more common than QAV in a ratio of 9:1. There seems to be a slight male predominance (1.6:1).[1,3]

Most cases of QAV are diagnosed by echocardiography (51%), followed by surgery 22.6%, autopsy 15.6% and aortography 6.5%.[4] Sometimes diagnosis may be missed with transthoracic echocardiography and a transoesophageal approach is needed to detect the morphological and functional status of the valve.[5,6] Improvements in transthoracic and transoesophageal echocardiography, and the increasing use of the latter, may reveal more cases of QAV and alter the incidence rate.

Embryology and classification

The semilunar valves are derived from mesenchymal swellings in the aortic and pulmonary trunk after the *truncus* has been partitioned. The mechanism of formation of a QAV is not well understood. Aberrant fusion of the aorticopulmonary septum or abnormal mesenchymal proliferation in the common trunk may lead to abnormal cusp formation.[7,8] A developmental error during the formation of either the coronary arterial system or the aortic cusps may lead to abnormalities that affect both of them, since septation of the arterial trunk and development of the aortic valve occur immediately after development of the coronary arteries.[9]

Several different anatomical variations of QAV have been described by Hurwitz and Roberts, according to the size of each individual aortic valve cusp.[7] This classification includes 7 different types:

A. four equal-sized cusps (second most common type)
B. three equal-sized cusps and one smaller (most common type)
C. two larger and two smaller cusps
D. one large cusp, 2 intermediate-sized cusps, and 1 smaller cusp.
E. three equal-sized cusps and one larger
F. two equal and two unequal smaller cusps
G. four unequal cusps (least common type).

However, the size of the cusps may be different during surgery compared with echocardiographic findings.[10]

Pathology

The recognition of QAV has clinical significance because a QAV frequently functions abnormally. Incomplete coaptation of the cusp, unequal stress distribution and fibrous thickening may lead to progressive aortic dysfunction.[4]

The predominant valvular abnormality in QAV is aortic regurgitation, with an incidence up to 75% of all cases. Aortic regurgitation tends to be more common in the form of QAV with an additional smaller cusp. The risk of aortic insufficiency is minimal in valves with four symmetric cusps.[4,11]

Cases of QAV stenosis are extremely rare (0.7%),[12] although in some series its prevalence was 7-12%.[13]

Combined QAV stenosis and regurgitation have been reported in 8.4% of all documented cases.[4] Whereas 6% of all QAV had a normal function,[4] surgery was necessary in 45.2% of all patients with a dysfunctional QAV during the fifth or sixth decade.[14] QAV dysfunction is absent or minimal in children and adolescents. The mean age of diagnosis is about 49 years.[4]

QAV is often (18%) associated with other congenital cardiac abnormalities, such as variations in coronary anatomy,[1] ventricular or atrial septal defect,[15] aneurysm of Valsalva sinus (ruptured or not),[16] patent *ductus arteriosus*, Ehlers Danlos syndrome, subaortic fibromuscular stenosis, hypertrophic cardiomyopathy,[17] association with malformation of the mitral valve, dilatation of the ascending aorta (rarely), pulmonary valve stenosis, and supravalvular stenosis with left coronary atresia.[1,4] Most often, it is accompanied by anomalies of the coronary arteries.[4] One case of QAV has been reported with sudden cardiac death due to a dome-like occlusion of the left coronary ostium.[18]

However, attention is needed because some pathological processes, such as bacterial endocarditis or rheumatic valve disease, can mimic the appearance of a QAV.[19]

Our case represents, to the best of our knowledge, the first reported case of QAV with a non-obstructive sub-aortic membrane.

Management

Patients with QAV should be followed closely. There have been some cases of bacterial endocarditis affecting a QAV, although it is not fully established whether QAV is more vulnerable to endocarditis. The risk of endocarditis is probably higher in patients who have valves with unequal cusps.[20] Endocarditis prophylaxis is no longer recommended in the management of patients with QAV.[21]

When there is an indication for surgical intervention, aortic valve replacement is the most acceptable procedure, although successful surgical repair has been reported in few cases.[22] It is very important for the surgeon to know the anatomy of the aortic valve.

If the aortic valve is quadricuspid there is a possibility of abnormal placed coronary ostia; thus, the surgeon must be careful to avoid damaging the left coronary ostium during aortic valve replacement operations.[23]

Conclusion

QAV is an unusual congenital defect, diagnosed mainly in adult life, and may cause aortic valve dysfunction, commonly aortic regurgitation. Sometimes, QAV is associated with other congenital malformations. To our knowledge, this is the first report of a case where QAV was associated with a non-obstructive sub-aortic membrane.

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