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OLGU SUNUMU

CONCOMITANT QUADRICUSPID AORTIC VALVE AND ANEURYSM OF ASCENDING AORTA

KUADRİKÜSPİD AORTİC KAPAK İLE ASENDAN AORT ANEVRİZMASI BİRLİKTELİĞİ

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ABSTRACT: Quadricuspid aortic valve is a congenital malformation which is a rare cause of aortic insufficiency. Although this abnormality usually presents itself as an isolated lesion, it may also be seen simultaneously with other cardiac malformations. In this study, a case with concomitant quadricuspid aortic valve and aneurysm of ascending aorta was presented whose quadricuspid valve was noticed intraoperatively.

Key words: Quadricuspid aortic valve, aortic insufficiency, aneurysm of ascending aorta

ÖZET: Kuadriküspid aortik valv aort yetmezliğinin nadir bir nedeni olarak karşımıza çıkan konjenital bir anomalidir. Bu anomali genellikle izole bir lezyon olarak ortaya çıkmakla birlikte diğer kardiyak anomaliler ile birlikte de görülebilir. Bu çalışmada daha önceden tanılanmamış, aort yetmezliği ve asendan aort anevrizması nedeniyle operasyona alındığında intraoperatif olarak fark edilen kuadriküspid aortik valv olgusu sunuldu.

Anahtar Kelimeler: Kuadriküspid aortik valv, aort yetmezliği, asendan aort anevrizma

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INTRODUCTION

Quadricuspid aortic valve (QAV), as a rare cause of aortic insufficiency, is a congenital abnormality mostly presented as a coincidental finding of echocardiographic, surgical or post-mortem investigations. It may be seen isolated or as an associated lesion to other cardiac abnormalities (1). Concomitant presentation of aneurysm of ascending aorta and QAV is very rare. In this study, a case with concomitant quadricuspid aortic valve and aneurysm of ascending aorta was presented under the light of current literature whose quadricuspid valve was noticed intraoperatively.

CASE REPORT

A 49-year-old male was admitted with a chief complaint of palpitation. His past medical history was significant for hypertension and acute rheumatic fever. Echocardiography revealed severe aortic insufficiency and ascending aortic aneurysm. Contrasted thoracoabdominal computed tomography (CT) and aortography showed that ascending aorta was dilated but sinotubular junction and aortic root were not aneurysmatic (Figure 1). Preoperative investigations led by Department of Cardiology showed no clue about quadrileaflet aortic valve. He was taken to the operating room for aortic valve and supracoronary ascending aortic replacement. Under general anesthesia a median sternotomy was performed.

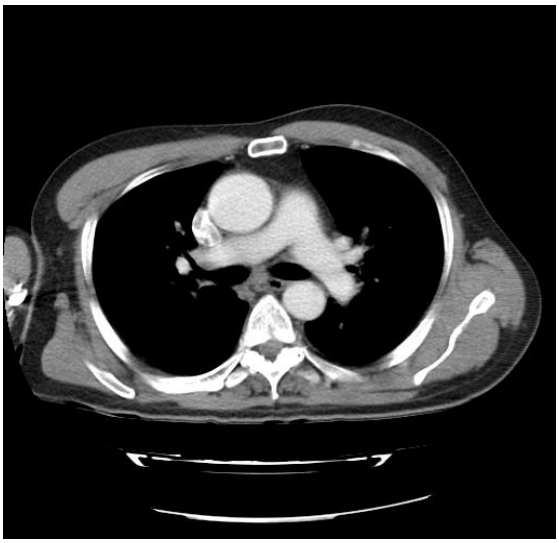

Figure 1. Contrasted CT image showing dilation of ascending aorta.

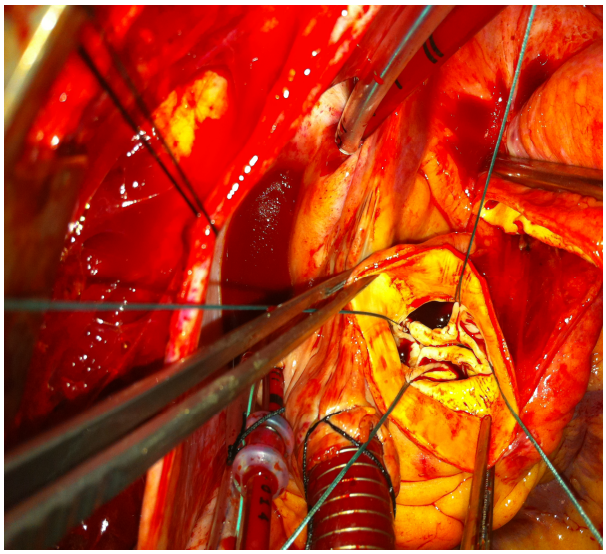

Figure 2. Intraoperative view showing quadricuspid aortic valve.

After longitudinal incision of the pericardium it was observed that whole ascending aorta was aneurysmatic and therefore right femoral artery was cannulated for arterial inflow and the right atrium was cannulated with 2-staged cannula venous outflow.

The patient was cooled down to 28°C. After cross-clamping of aorta an oblique aortotomy was made. For myocardial protection, initially isothermic hyperpotassemic blood cardioplegia was introduced antegradely through coronary orifices and then continuously retrogradely for maintenance. While exploring the aortic valve, 4 leaflets and 4 commissures were observed (Figure2).

It was unsuitable for repair due to extensive fibrotic thickening of aortic leaflets, calcification, lack of pliability and incomplete coaptation. A 23 mm St. Jude mechanical prosthetic valve was implanted. Afterwards, a 26 mm gelatin-coated Dacron graft was implanted with a distal anastomosis carried out under total circulatory arrest. While making the proximal anastomosis, the graft was cross-clamped and circulation was reestablished while warming up the patient. After declamping the aorta, suitable hemodynamic and temperature parameters were achieved and cardiopulmonary bypass was ended. He was transported to the intensive care unit with full monitorization and ventilatory support. After one day of ICU stay he was discharged event-free on 6th postoperative day.

DISCUSSION

Malformations of semilunar valves are the most common congenital defects of the heart and great vessels out of which bicuspid aortic valve is the most common one with a frequency of 2% (1). Quadricuspid aortic valve is much more seldom seen than bicuspid aorta. It is slightly more common in males (male/female ratio 1.6/1). The average age at the time of diagnosis is 50.7 (1,2). Its prevalence is 0.008% in autopsy series but it is seen in 1% of patients referred for aortic valve surgery (3-5). QAV was first defined by Balington in 1862 (2). In literature data, more than 200 cases were reported in echocardiographic and aortographic investigations, intraoperatively or in some autopsy series. Although the frequency of QAV was reported as 1% by some papers including the aortic operations, we did not face with this abnormality in the last decade out of 980 aortic valve explorations.

Although QAV is usually seen alone, there are also some simultaneous congenital heart defects in 18% of cases (1,2,6-8). Some of these abnormalities are coronary artery abnormalities, ventricular septal defect, atrial septal defect, patent ductus arteriosus, pulmonary stenosis, ruptured or unruptured aneurysms of sinus of Valsalva, total AV block, hypertrophic cardiomyopathy, subaortic fibromuscular stenosis and mitral valve abnormalities. Out of these, coronary artery abnormalities are most commonly seen with a ratio of 30%. Concomitance with aneurysm of ascending aorta is extremely rare (6-8). There was no additional abnormality in our case beside aneurysm of ascending aorta. Although the exact embryological origin of this pathology remained unclear, the main pathology was suggested to be either developmental changes of early truncal division or abnormal development or fusion of mesenchymal buds or defect in the number of primordial valves (1,9). Although quadicuspid structure is 9 times more common in pulmonary valve, this defect usually does not cause any insufficiency (9). Nevertheless, quadricuspid structure generally deteriorates the function of aortic valve. Aortic insufficiency is seen in 75% of patients with QAV (1,2). Aortic insufficiency was considered to originate from abnormal commissural fusion or abnormal distribution of the mechanical stress on the leaflets causing structural deterioration or thickening (1,9). Aortic stenosis is seen more seldom than aortic insufficiency.

According to the classification of the quadricuspid valve by Hurwitz and Roberts, there are 7 anatomical types (types A-G) regarding the size of the leaflets. All the leaflets are equal in size in type A. In type B, there are 3 equal leaflets and one small leaflet. In type C, there are 2 equally large and 2 equally small leaflets. In type D, there are 1 large, 2 mid-sized and 1 small leaflets. In type E, there are 3 equal leaflets and 1 large leaflet. In type F, there are 2 equally large and 2 smaller leaflets of different size. In type G, all 4 leaflets are of different size (1,6,10). The most common type is type B, followed by type A (6). In our case, there was a type C quadricuspid valve.

In conclusion, although most of the published data contain the cases with coincidentally diagnosed QAV by echocardiographic studies, as in our case, it may be discovered intraoperatively. In any case that one faces with QAV, associated cardiac

abnormalities, particularly abnormalities of coronary arteries, should be kept in mind and precautions should be taken.

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