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QUADRICUSPID AORTIC VALVE WITH ABERRANT CORONARY OSTIA - A CASE REPORT

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Abstract :
Quadricuspid aortic valve is an uncommon congenital cardiac anomaly often detected incidentally or in association with other congenital cardiac anomaly. The incidence is about 0.003 percent in autopsy studies (1). They may also be associated with coronary anomalies, septal defects, patent ductus arteriosus, mitral or pulmonary valve malformations and conduction defects (2).If undetected in childhood, they may progress to aortic valvular dysfunction later in life, necessitating valve replacement. Associated coronary anomalies, if any, may make prosthetic replacement of the valve challenging. We herein describe a case of a quadricuspid aortic valve who presented with severe aortic regurgitation. She was detected to have low lying right coronary ostia, necessitating precautions during valve replacement.

Keyword : Quadricuspid Aortic valve, low lying coronary ostia

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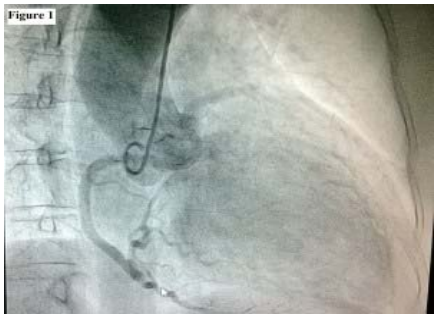
INTRODUCTION:

Quadricuspid aortic valve, first reported by Balington(2) in 1862 is a rare congenital anomaly of the aortic valve, present in about 0.04 % of the population in echo cardiographic studies(2). This is often associated with various congenital cardiac anomalies and syndromes like Ehlers-Danlos and William's syndrome. If undetected in childhood, they often present as valvular dysfunction in adult life, requiring valve replacement. The associated coronary anomalies (10%) may make cardioplegic arrest and prosthetic valve replacement challenging. An exact pre operative diagnosis thus becomes mandatory to surgical planning to avoid surprises on table. We present a case of quadricuspid aortic valve with low right coronary ostia, which required technical adjustments during a prosthetic valve

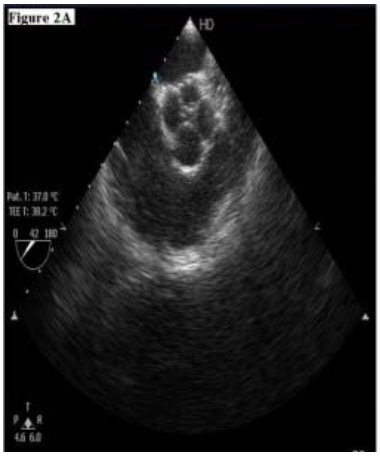
replacement surgery.

CASE REPORT:

A 56 year old lady presented with history of palpitation, exertional breathlessness and occasional chest pain since 4 years. She had no other co morbidities. On examination she was found to have a severe aortic regurgitation. Her chest roentgenogram revealed cardiomegaly and electrocardiogram showed left ventricular hypertrophy. Her transthoracic echocardiogram (TTE) revealed moderate aortic regurgitation with left ventricular systolic dysfunction with an ejection fraction of 40%. A cardiac catheterization demonstrated severe aortic regurgitation, normal coronaries and a low lying right coronary artery ostia **(Figure 1).**

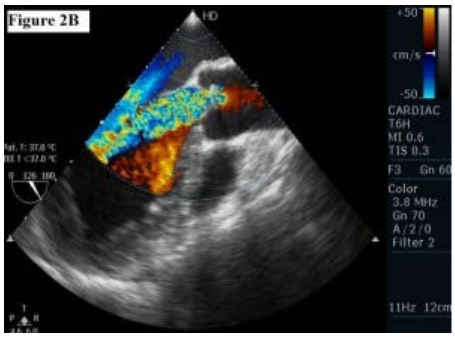


Aortic root injection showing a low lying right coronary artery



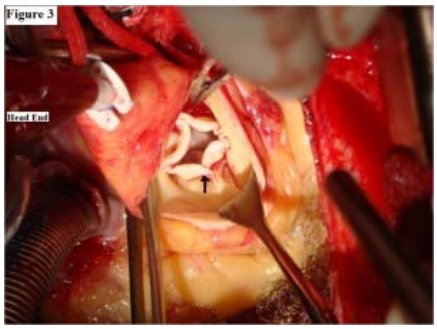
Peroperative transesophageal echo revealed that she had a quadricuspid aortic valve (**Figure 2A**) with low lying right coronary ostia and normally positioned left ostia along with severe valvular regurgitation (**Figure 2B**).

Mid esophageal short axis view of TEE showing the quadricuspid aortic valve.



Mid esophageal long axis view of TEE showing central jet of severe aortic regurgitation.

The valve had two large and two unequal smaller cusps. The right coronary cusp was partitioned to form the extra leaflet (**Figure 3**) leaving the valve quadricuspid.



Peroperative view of the quadricuspid aortic valve. Arrow showing the extra commisure dividing the right coronary cusp.

After excision of the valve leaflets, the low lying right coronary ostia was inspected and the valvesutures were cautiously placed to avoid compromising the same. A prosthetic valve with low profilesewing ring was used. Before closure of the aortotomy, the ostia were inspected again. After thecross clamp was released, an on table Trans Esophageal Echocardiography (TEE) was used toconfirm unobstructed coronary flow, before the termination of cardiopulmonary bypass. Her postoperative recovery was uneventful.

DISCUSSION:

Quadricuspid aortic valve is a fascinating albeit rare entity which is usually associated with othermalformations. There have been only a limited number of such cases described in the literature (2).

Faulty septation of the embryological truncus arteriosus with proliferation of mesenchymal ridges may lead to supernumerary leaflets (3). Associated anomalies include coronary artery anomalies,atrial or ventricular septal defects, mitral valve malformations or hypertrophic cardiomyopathy. Coronary artery anomalies may constitute upto 10% of the associated malformations in such patients.Coronaries arising from a single ostium(4), abnormal origin of the left circumflex artery from the right coronary artery(5) or presence of displaced coronaries have been described. Even minor variations like low lying coronary arteries should caution the presence of associated aortic valvular anomalies.Hurwitz and Roberts have classified this entity into seven types depending on the size of theindividual leaflets (6). The various types are: Type A with four equal cusps, Type B with three equal cusps and one smaller cusp, Type C with two equal larger and two equal smaller cusps, Type D with one large, two intermediate and one small cusp, Type E with three equal cusps and one larger cusp, Type F with two equal larger and two unequal smaller cusps Type G with four unequal cusps.

Our patient had two equal larger sized cusps and two unequal smaller cusps which fall under the type F according to Hurwitz and Roberts. Transthoracic echocardiography may not always detect the anomaly, but a transesophageal echocardiography or a multi detector computed tomography would be of additional benefit in revealing such anomalies (7). Bicuspidizations (8) as well as tricuspidization procedures have been described as satisfactory and durable methods of repair of such valves. Replacement of such valves, if done, must be done with care to prevent damage to the conduction bundle and the abnormally placed coronary arteries. Supra annular sutures may be placed in the region of the supernumerary cusp especially when it lies between the right and noncoronary cusps to safeguard the conduction system (9).

CONCLUSION:

Aortic regurgitation may occasionally be associated with a quadricuspid aortic valve. The management of the associated congenital cardiac defects often takes precedence over the valve defect. However, in the presence of aortic valvular dysfunction, a valve replacement may be required. Diligent study of a coronary anomaly, if any is mandatory. The presence of such coronary anomalies may itself hint towards the presence of an aortic leaflet anomaly. TEE is superior in detecting such anomalies as compared to TTE and conventional angiogram.

Careful excision of the leaflets, judicious placement of the sutures, choosing an appropriate valve, inspecting the patency of the ostia after seating the valve and ensuring unobstructed coronary flow using TEE before terminating cardiopulmonary bypass are essential in such cases.

We present this rare lesion with interesting images to stress upon the necessity to study the associated defects while dealing with quadricuspid aortic valves and to decide upon the appropriate strategy while planning the surgery.

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