A rare cause of severe aortic regurgitation: quadricuspid aortic valve

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Abstract

Quadricuspid aortic valve (QAV) is a rare cause of valve failure. Often, it is an incidental finding at transesophageal echo (TEE), surgery or post-mortem examination. This anomaly is often associated with abnormally placed left coronary ostium that could be damaged during the operation. We report a case of QAV detected incidentally during perioperative TEE.

Keywords: Aortic valve; Echocardiography; Valve disease

1. Introduction

Quadricuspid aortic valve (QAV) is a very uncommon cause of valve failure and it often is an incidental finding at transesophageal echo (TEE), surgery or post-mortem examination. When valve regurgitation is severe, surgery is indicated in terms of either replacement or plasty, depending on the valve anatomy.

In this article, we report a case of QAV identified only with intraoperative TEE.

2. Case description

A 77-year-old man with a history of severe aortic regurgitation, mild mitral regurgitation and dilated cardiomyopathy with low ejection fraction (0.28), was referred to our department to undergo aortic valve surgery. The intraoperative TEE showed the presence of a QAV (Fig. 1) which was not diagnosed with transthoracic echo (TTE) before. The regurgitation was huge and central, related to incomplete juxtaposition of the four cusps. Moreover, a mild mitral regurgitation was observed due to a reduced motion of the proximal portion of the anterior leaflet of the mitral valve, with an abnormal coaptation of the free margins below mitral valve annulus.

In accordance with the Hurwitz and Roberts classification [1], which describes seven anatomic variants (from type a to type g), this aortic valve is classified as type b, three equal-sized cusps and one smaller cusp. The smaller cusp was located between the non-coronary and left coronary cusps.

Ascending aorta and right atrium were cannulated for cardiopulmonary bypass; heart was arrested with cold blood cardioplegia via ante-retrograde route with warm induction.

The surgical view of aortic valve (Fig. 2a) confirmed the TEE diagnosis, and abnormally placed left coronary ostium was noticed. Some calcifications were identified on the three commissures and retraction of the free margin of the right and left coronary leaflets were observed. Therefore, the anatomy of aortic valve was judged not suitable for aortic plasty and consequently aortic valve replacement was decided.

Aortic valve was removed in toto (Fig. 2b) and it was replaced with a bioprosthesis (St Jude Medical Supra #23, St Jude Medical, St Paul, MN, USA) in supra-annular position. A mitral annuloplasty was performed, implanting a partial semirigid ring (Future Band Ring # 28, Medtronic, Minneapolis, MN, USA). The postoperative course was uneventfully and the patient was discharged home on 6th postoperative day.

The patient was restudied with TTE after 4 months. He was asymptomatic; the TTE showed residual trivial mitral valve regurgitation, an increased ejection fraction (0.38) and reduced left ventricle volumes.

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3. Comment

The QAV is a rare congenital malformation and its mechanism is not fully understood. One of the leading hypotheses is for an abnormal septation of embryological arterial trunk. Normally, the semilunar valves are derived from mesenchimal swellings, which develop into three semilunar cusps. In the setting of QAV, the fourth cusp arises during the early stage of truncal septation, resulting from either different number of primordial aortic leaflets or abnormal cusp proliferation or abnormal aortopulmonary septation [2,3]. In this way, the aortic root deviates from normal configuration with formation of abnormal sinuses, leaflets, and interleaflet triangles.

The first case of QAV was described in 1862. Since then, about 110 cases were described [4]. According to world literature, the incidence of this congenital pathology ranges between 0.003 and 0.043% [5,6] and most of the cases were incidentally recognised at surgery or necroscopy. Nowadays TTE and TEE play an important role in diagnosing this anomaly [7,8]. Often, the TTE maybe suboptimal to recognise this malformation and the origin of the left main, as shown in the present case. In fact, the patient underwent several transthoracic two-dimensional echoes in the past, that showed the aortic regurgitation and not the QAV.

We observed this malformation only during intraoperative TEE before starting with cardiopulmonary bypass. Moreover, the origin of the left main was dislocated leftward and upward to the normal origin site. This is an important standpoint because in these malformations, with abnormal placed left coronary ostium, the probability to damage it, while performing the aortotomy, is very high. Therefore, as a rule, high aortotomies have to be performed not only in these rare cases, but also in all cases of aortic valve replacement. Knowledge of the anatomy of the aortic valve and of the origin site of the coronary ostia could be of great help to the surgeon to prevent ostia obstruction when prosthesis is implanted [7].

The identification of a QAV in case of aortic regurgitation is very important because high is the progressive deterioration of the leaflets due to the asymmetric mechanical stress around the four cusps and consequently high is the risk of endocarditis. For this reason, it is advisable to maintain an echocardiographic follow-up to assess the state of the valve and eventually to start with endocarditis antibiotic prophylaxis [6,9,10].

In conclusion, QAV is a very uncommon congenital malformation noted in adult life, incidentally identified at surgery or necroscopy before echocardiographic era. Nowadays, TEE allows the clinician and the surgeon to reach an appropriate diagnosis. From a surgical point of view, the surgeon is alert of possible abnormal placed coronary ostia, thereby reducing the likelihood of damaging or obstructing the left coronary ostium in aortic valve replacement operations.
References