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A Case of 'Forme Fruste' Bicuspid Aortic Valve – The Impact On Surgical Strategy

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1. Abstract

We report a case of advanced aortic regurgitation with bicuspid aortic valve (BAV) with very small raphe called as a 'forme fruste' BAV. In this case, surgical strategy should be discussed about the feasibility of repair technique and aortic procedure for aortopathy. We selected BAV repair with the tricuspidization by repositioning of the immature commissure and central plications of the three leaflets. Pathological examination of the aorta showed findings of aortopathy. For potential repairability and proper intervention of aortic complications, preoperative diagnosis of 'forme fruste' BAV is important.

2. Introduction

Recently, BAV with very small raphes (3-6mm) were reported as 'forme fruste' BAV 1,2). In this case, surgical strategy should be discussed about the feasibility of repair technique 3) and aortic procedure for aortopathy 4).

We performed aortic valvuloplasty (AVP) for 'forme fruste' BAV with the tricuspidization by repositioning of the immature commissure and reimplantation technique.

3. Case

The patient was 45-year-old man pointed out left ventricular hypertrophy. Transthoracic echocardiography (TTE) showed severe aortic regurgitation (AR), and the aortic valve showed an apparent tricuspid. The left ventricular (LV) end-diastolic dimension was 67 mm and the LV end-systolic dimension was 47 mm, and LV ejection fraction of 54%. Transesophageal echocardiography (TEE) identified very small commissural fusion of the right coronary cusp (RCC) and left coronary cusp (LCC) (Figure 1a). Prolapse of the fused leaflets was suspected as the cause of regurgitation (Figure 1b). Computed tomography revealed a ventriculo-aortic junction measuring 30×40 mm, sinus of Valsalva measuring 42 mm, and sino-tubular junction measuring 37 mm.

4. Operation

Through mid-sternal incision, standard cardiopulmonary bypass was established. After aortic clamping, aortic valve was observed. The valve had small raphe between RCC and LCC which can be called as 'forme fruste' BAV (Figure 2a). Geometric heights of the RCC, LCC, and non-coronary cusp were 20, 22, and 20 mm, and free margin lengths of these cusps were 35, 26, and 35 mm, respectively. For complete repair, we performed reimplantation technique4) using Gelweave Valsalva graft 26mm (Terumo, Tokyo, Japan). The immature commissure was brought up to the standard commissure position. Central plications of the three leaflets were performed with effective heights ≥9 mm (Figure 2b). After temporary pump-off, TEE showed moderate residual RCC prolapse. Second aortic clamp was applied, fenestration rupture at RCC was recognized. Pericardial patch plasty of RCC around fenestration which did not involve commissure was performed. TEE after pump-off showed trivial central AR. Pathological examination of the aorta showed mild cystic medial necrosis, medionecrosis and elastic fragmentation (Figure 3 a, b).

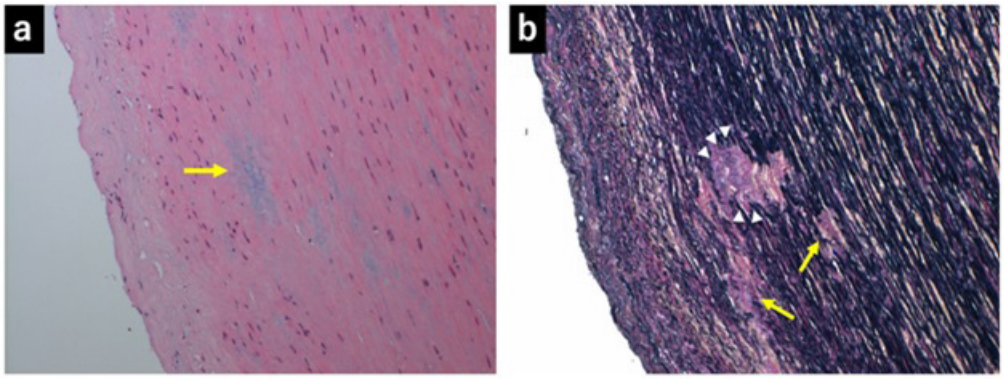

Figure 1: a) Very small raphe commissural fusion of the right and left coronary cusps depicted on 3-dimensional transesophageal echocardiography b) Severe aortic regurgitation

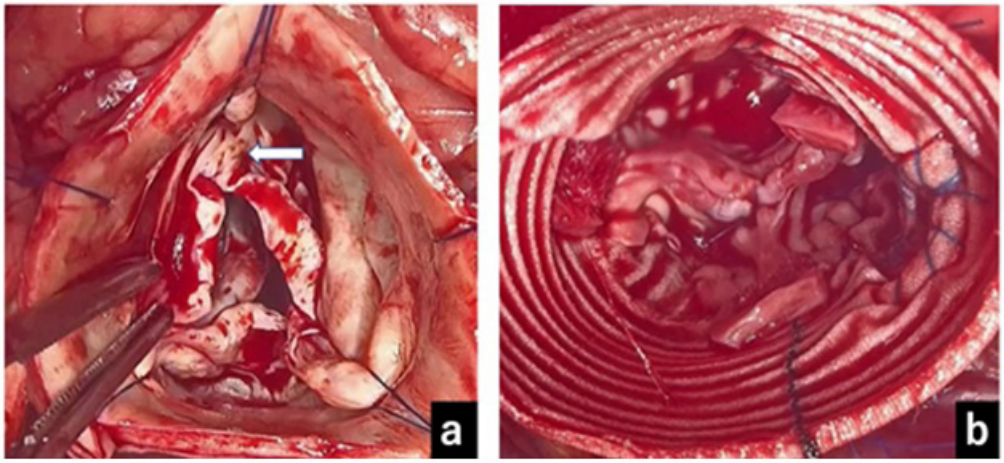

Figure 2: a) Intraoperative findings of 'forme fruste' aortic valve b) Intraoperative view during valve repair and reimplantation

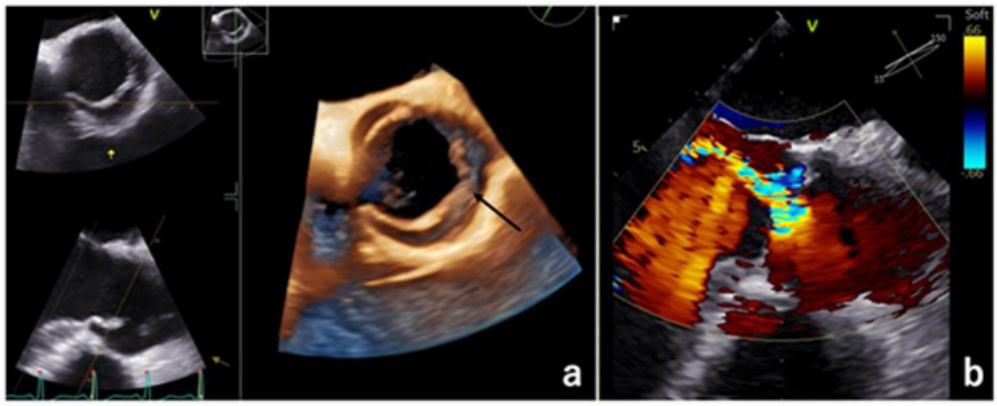

Figure 3: a) Pathological examination of the aorta showed mild cystic medial necrosis (arrow). b) Mild medionecrosis (white arrows) and elastic fragmentation (yellow arrows)

5. Discussion

Recently, Sperling et al. [1] reported that 30% of aortic valves that had been preoperatively diagnosed with tricuspid valves showed an immature, very small raphe classifiable as 'forme fruste' BAV 1). 'Forme fruste' BAV is often recognized as a tricuspid valve preoperatively because it is not easy to detect the very small raphe with routine TTE [2]. There is no scientific basis to support any notion that specific BAV phenotype or raphe length has any impact on the incidence of aortic complications [4,5], and frequency of aortopathy in 'forme fruste' is unknown [3]. However, these aortic valves may potentially exert pathological effects on aortic dilatation as the degenerative changes that we encountered in our case, in which pathological diagnosis showed findings consistent with aortopathy, although the ascending aorta dilation was mild. Therefore, preoperative diagnosis of 'forme fruste' BAV using careful TEE and high resolution CT techniques is meaningful in examining the need for aortic intervention.

A new repair-oriented classification system was recently proposed

by de Kerchove et al. [4] based on those parameters (Type A: symmetrical BAV, Type B: asymmetrical BAV, Type C: very asymmetrical BAV). Commissural orientation correlated positively with the length of raphe fusion and negatively with the height of the non-functional commissure [6]. As an algorithm for repair of BAV under the new classification, they proposed the following techniques: Type A, central plication or direct closure of the non-fused portion of the raphe; Type B, direct closure of the non-fused portion of the raphe ± thinning of the raphe; or Type C, plication of the residual cusp component or creation of a functional commissure at the site of the raphe ± pericardial patch or type B repair. Our case could be classified as their type C, we considered that tricuspidization without need for commissural patch reconstruction was suitable which shows almost the morphology of the tricuspid valve. In fact, the regurgitation could be controlled by central plications of three leaflets basically.

From our experience of 'forme fruste' BAV, we consider that AVP including tricuspidization and central plications might be feasible with reimplantation technique in case of the diameter of ascending aorta <45mm, and that ascending aorta should be replaced in case of its diameter ≥ 45mm when AVR/AVP is indicated following standard indication for BAV.

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