INTRODUCTION

Behçet's disease (BD) was first described by Hulusi Behçet in 1937.1 In the following years, BD was divided into three subgroups as enteric Behçet with deep ulcers in the ileocecal region, neural Behçet, and vascular Behçet.2 In studies conducted in BD in our country, the rate of vascular involvement is varied between 14% and 49%, and more common in males.3,4 However, mortality due to complications of vascular involvement is very high in the reported study.5 Venous involvement is more common in vascular BD. If the arteries are involved, lesions are mostly seen in the abdominal aorta, subclavian and carotid arteries in the form of aneurysms. BD may involve the aortic root and aortic valve, as well as the ascending aorta. The first clinical finding may be the signs of aortic valve insufficiency. Aortic valve insufficiency occurs as a result of aortic leaflet degeneration, aortic root enlargement or both pathologies. Aortic valve replacement is the only surgical treatment of aortic valve insufficiency due to severe degeneration of aortic leaflets. On the other hand, in cases of aortic insufficiency due to dilatation of aortic root and aortic annulus, the valve-sparing strategy should be planned. In 1992, Tirone David has successfully applied valve-sparing procedure in ascending aortic aneurysm with intact valve leaflet and named after him.6 In later years, David operation in aortic valve insufficiency with ascending aortic aneurysms and aortic dissection has become a major priority because of excellent long-term
result. The study aimed to present 6 years experience of Button Bentall and David operations in BD with ascending aortic involvement.

METHODS

This study was carried out after approval of the Medipol University Ethics Committee. In this study, total of 13 ascending aorta aneurism with BD operated between January 2012 and 2018 were included. All of them were male, mean age was 41±9.5 (ranged 19 to 67 years). Mean follow-up was 51±23 (ranged 23 to 94) months. Eight patients had moderate/severe aortic valve insufficiency, and 3 patients had aortic valve degeneration. The aortic diameter ranged 4.5cm to 7.2cm (Figure 1a and 1b) at root level in echocardiography or CT angiography. Two patients had aneurysm at the level of arcus aorta and type A dissection (Figure 2a and 2b). Coronary angiography was performed in 6 patients due to older age. A 67-year-old patient had RCA stenosis and coronary bypass was performed additionally. All the patients were strictly followed-up by the rheumatology clinic before and after the operation, the steroid treatment was continued until the operation day.

Operative technique

After median sternotomy, arterial cannulation was performed by suturing a dacron graft to the right axillary artery, venous cannulation was performed through the right atrium. The patients were cooled to 28 degrees and aortic clamps were placed. We routinely used cold blood cardioplegia and repeated every 15-20 minutes from coronary artery ostea. Total circulatory arrest was performed in two patients requiring hemiarcus replacement, patients were cooled to 18 degrees, aortic cross clamp was removed, selective partial brain perfusion was provided. After aortotomy, aortic valves were evaluated for leaflet degeneration. Bentall procedure was performed in patients with severe degenerative aortic valve by using the proper size of valvular composite (Figure 1c and 1d).

Statistical analysis

Statistical analysis was performed with SPSS version 24.0 program (SPSS Inc. Chicago IL, USA). Mean and standard deviation values were used to present descriptive analyzes. Percentage was used to evaluate nonparametric variables.

RESULTS

Preoperative patient characteristics and comorbid factors were shown in Table 1.

| Characteristic                       | N (%)       | Mean±SD  |
|-------------------------------------|-------------|----------|
| Age (yrs)                           | 41±9.5      |          |
| Body surface area (m²)              | 1.78±0.23   |          |
| Body mass index (kg/m²)             | 25.8±5.8    |          |
| Diabetes mellitus                   | 2 (15.4)    |          |
| Hypertension                        | 3 (23.1)    |          |
| Hyperlipidemia                      | 5 (38.5)    |          |
| Coronary artery stenosis            | 1 (7.7)     |          |
| Smoking                             | 5 (38.5)    |          |
| Aortic regurgitation                |             |          |
| Normal                              | 4 (30.8)    |          |
| Mild                                | 1 (7.7)     |          |
| Moderate                            | 3 (23.1)    |          |
| Severe                              | 5 (38.5)    |          |
| Aortic valve degeneration           | 3 (23.1)    |          |
| Ejection fraction (%)               | 53±7        |          |
| Aortic root diameter (cm)           | 5.1±0.7     |          |

Seven patients (54%) were performed Bentall procedure and six patients (46%) were David procedure. One patient from each group required hemiarcus replacement, short length of total circulatory arrest was applied. One patient was reexplorated because of massive bleeding five hours after the operation. One patient underwent button Bentall procedure for acute type A dissection died.
at 24th postoperative hour due to multiorgan dysfunction. Nine patients were discharged on the 8th postoperative day without any complications. Renal failure was occurred in two patients on the 29th and 3rd postoperative day, they were recovered without hemodialysis and discharged uneventfully after 11th and 13th postoperative day. One patient who suffered cognitive dysfunction was also discharged on the 10th postoperative day. After 3 weeks, one patient readmitted for dyspnea due to Dressler syndrome, the pericardial effusion was treated by pericardiocentesis by echocardiography guide. One patient died of major gastrointestinal bleeding after three months. Postoperative major complications and mortality were shown in Table 2.

Table 2: Operative and postoperative patient variables.

| Variable                          | N (%) | Mean±SD |
|----------------------------------|-------|---------|
| Bentall procedure                | 7 (54) |         |
| David procedure                  | 6 (46) |         |
| Cross clamp time (min)           | 106±23 |         |
| Cardiopulmonary bypass time (min)| 157±35 |         |
| Total circulatory arrest         | 2 (15.4) |         |
| Postoperative bleeding (ml)      | 765±290 |        |
| Reexploration                    | 1 (7.7) |         |
| Neurological complication        | 1 (7.7) |         |
| Renal failure                    | 2 (15.4) |       |
| Inotrope requirement (more than 6 h) | 9 (69.2) |     |
| Length of hospital stay (day)    | 8.8±1.6 |         |
| Operative mortality              | 1 (7.7) |         |
| Overall mortality                | 2 (15.4) |        |

DISCUSSION

Although the prevalence of rheumatic valve disease has decreased owing to the adequate prophylaxis, valvular studies have been reported that the aortic root dilatation is the most important pathology leading to aortic valve insufficiency. Aortic root dilatation may develop idiopathically in many cases or may develop after Marfan syndrome, aortic dissection or vasculitis such as Behcet. In addition to the ascending aorta, the aortic arch aneurysm is rarely seen. It is reported that the development of true or pseudoaneurysm in the anastomosis regions, the dehiscence of the valve after operation is much more common (40%) than the normal population. The fragile and inflammatory tissue in the Behcet patients was shown to be the main cause of this. To prevent this kind of complication, it is recommended that the surgery should be performed in a silent period of at least 3 months in which the patient is clinically stable and sedimentation tests are normal. In cases with whom the remission cannot be expected, the steroid treatment should be applied both before and after surgery to prevent complications. In study series, the steroid treatment before surgery was strictly followed continued in the postoperative period in a patient who underwent emergency operation because of type A dissection.

It was found in the study that aortic leaflets are mostly preserved in aortic valve insufficiency due to aortic root and valsalva sinuses dilatation except in older (>50 year) patients. Previously, in the pathologies where aortic root dilatation is present but aortic valves are not affected, the patient tissues were completely replaced with graft and prosthetic aortic valve, but in recent years, Yacoub and especially David proposed the preservation of the valve structure. In recent years, valve-sparing aortic root replacements have been discussed in the large aortic root patients with normal aortic valve structure, it has been stood out that valve-sparing root replacement is a preferred choice after strict evaluation in the inflammatory disease group. In present series, it was decided to perform button Bentall or David procedure by considering the degree of aortic valve insufficiency and degeneration. In large aortic root, aortic leaflets are frequently thickened and fenestrated. Since aortic valve should be replaced in these patients in the long-term, we chose to use valvular composite method (Bentall), and in the other patients, valve sparing root replacement was performed. Aortic annuloplasty ring was not preferred to use in practice no new aortic insufficiency other than early postoperative period occurred during follow-up. Initial results of aortic annuloplasty revealed that the aortic root does not dilate over time after remodeling with or without annuloplasty. One of the favorite techniques is that, instead of teflon felt, aortic adventitia tissue strip in anastomosis line was used, the adventitia tissue is a very strong pledget per se was considered, it reduced anastomosis leakage and facilitated easy suture if additional suture needed.

In conclusion, it should be kept in mind that proximal and arcus aortic aneurysms can be seen in BD. Procedures requiring surgical intervention should be performed during the remission period. In cases of emergency where remission cannot be expected, patient should be given steroid treatment before and after the operation, anastomosis lines should be supported with teflon felt or aortic adventitia tissue and secured with tissue adhesives. In patients whose aortic valve structure is considered to be intact, one of the David procedure may be preferred as the valve-sparing surgery. Larger aortic root with degenerative aortic valve may necessitate Bentall procedure. In the postoperative period, patients should be closely monitored by CT angiography and echocardiography, thus, early intervention can be possible to reduced mortality in cases of bleeding, anastomosis leakage, pseudoaneurysm or aortic valve dehiscence.

Limitation

This is a retrospective single-center study, and included a limited number of patients. The results of the study also
showed mid-term results, long-term follow-up should be required.

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