Comparison of Clinical, Angiographic Features and Outcome in Takayasu’s Arteritis and Behçet’s Disease With Arterial Involvement

Su Jin Choi, M.D.¹, Hyun Jung Koo, M.D., Ph.D.², Dong Hyun Yang, M.D., Ph.D.², Joon-Won Kang, M.D., Ph.D.², Ji Seon Oh, M.D., Ph.D.¹, Seokchan Hong, M.D., Ph.D.¹, Yong-Gil Kim, M.D., Ph.D.¹, Bin Yoo, M.D., Ph.D.¹, Chang-Keun Lee, M.D., Ph.D.¹

¹Division of Rheumatology, Department of Internal Medicine, ²Department of Radiology and Research Institute of Radiology, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea

OBJECTIVE. Takayasu’s arteritis (TAK) is a vasculitis that primarily involves the aorta and its branches. In Behçet’s disease (BD), systemic vasculitis is one of major manifestations. We aimed to compare clinical and angiographic features and outcome between TAK and BD with arterial involvement. METHODS. We retrospectively reviewed medical records of 206 TAK patients and 50 BD patients between 1995 and 2015. Angiographic lesions were evaluated via computed tomography, magnetic resonance imaging, and/or conventional angiography. RESULTS. Fever (30% vs. 9.2%, p < 0.001) and arthralgia (36% vs. 7.3%, p < 0.001) were more common in BD. C-reactive protein was higher in BD compared with TAK (5.85 mg/dL vs. 2.08 mg/dL, p < 0.001). Stenosis (89.8% vs. 60%, p < 0.001) and occlusion (65.5% vs. 32%, p < 0.001) were more observed in TAK. In contrast, aneurysm was common in BD (62% vs. 20.9%, p < 0.001). The carotid artery (73.3% vs. 30%, p < 0.001), subclavian artery (71.4% vs. 16%, p < 0.001), descending aorta (35% vs. 12%, p = 0.002), renal artery (23.8% vs. 10%, p = 0.032), superior mesenteric artery (18.4% vs. 4%, p = 0.012), and brachiocephalic trunk (13.6% vs. 2%, p = 0.020) were more commonly involved in TAK, whereas the femoral artery (10% vs. 2.4%, p = 0.027) was more frequently involved in BD. During follow-up, arterial dissection (10% vs. 1.9%, p = 0.016), rupture (12% vs. 0.5%, p < 0.001), and arterial replacement/resection (66% vs. 9.7%, p < 0.001) were more observed in BD. CONCLUSION. TAK differs from BD regarding clinical features and vascular involvement patterns. BD exhibits a higher rate of vascular complications.

KEY WORDS. Behçet syndrome, Takayasu arteritis, Arteritis, Cardiovascular diseases

INTRODUCTION

Takayasu’s arteritis (TAK) and Behçet’s disease (BD) are systemic vasculitides that may affect the large arteries. They are usually characterized by clinical and angiographic features. TAK is a chronic granulomatous vasculitis that affects the aorta and its branches [1]. One of its major clinical manifestations includes vascular symptoms caused by ischemia and constitutional symptoms. Stenosis in the aorta and the cervicobrachial area is a common angiographic pattern of its arterial involvement. Cardiovascular complications are associated with TAK, which generates higher mortality rates than the general population [2,3]. BD is a systemic disease characterized by mucocutaneous, ocular, neurologic, and gastrointestinal involvement [4]. Vascular manifestation is one of BD’s typical clinical features, which involves veins and arteries of all sizes. Arterial involvement occurs in 1% ~ 18% of BD patients [4-6] and can be fatal, with cardiovascular complications including arterial occlusion and aneurysm rupture [7]. BD mainly presents as an aneurysm and occlusion in the aorta and lower extremities. Arterial thrombo-
Takayasu's arteritis and Behçet's disease are both chronic inflammatory vasculitides. The presence of symptoms indicating Behçet's disease can help distinguish Takayasu's arteritis (TAK) and Behçet's disease (BD). However, the data comparing clinical, angiographic features, and vascular outcome of both diseases are limited. This comparison may advance the understanding of the angiographic characteristics and outcomes between two diseases as parts of large vessel vasculitis. In this context, we investigated to compare the clinical and angiographic features as well as outcomes between TAK and BD with arterial involvement.

**MATERIALS AND METHODS**

**Study population**

We retrospectively evaluated 206 patients with TAK and 50 patients with BD who had arterial involvement and were treated at a tertiary referral hospital in Seoul, South Korea between January 1995 and December 2015 (Figure 1). The diagnoses were confirmed according to the classification of the American College of Rheumatology [9] for TAK, and the International Criteria [10] for BD. All patients underwent computed tomography (CT), magnetic resonance imaging, and/or conventional angiography to evaluate the entire aorta and its branches. We collected patient information as follows: demographic data such as age, sex, comorbidities; clinical symptoms and laboratory findings such as erythrocyte sedimentation rate and C-reactive protein (CRP); angiographic findings; complications; interventions (endovascular or surgical); and death. The study was approved by the Institutional Review Board of the Asan Medical Center at Seoul, Korea (protocol number: 2017-1157). The requirement for informed consent was waived because of the retrospective design.

**Clinical, angiographic features, and outcome**

Clinical manifestations of both diseases included constitutional, vascular, cardiopulmonary, neurologic, mucocutaneous, and gastrointestinal symptoms at diagnosis. Patterns of angiographic lesions were categorized into four types: stenosis, occlusion, dilatation, and aneurysm. Stenosis was a narrowing of the vessels compared to the normal upper or lower portions. Occlusion was defined as the case where the contrast material did not pass through the vessel in the affected segment. An aneurysm was defined as a dilated artery which was more than 50% of normally expected arterial diameter compared to upper or lower normal sites from the lesion. In large vessels, ascending aorta aneurysm was considered when the maximum transverse diameter was the same or larger than 50 mm [11]. Descending thoracic aorta aneurysm was diagnosed when the aorta reached a diameter of 40 mm [12]. In the abdominal aorta, aortic diameter ≥ 30 mm considered as aneurysm [13]. If a dilated lesion did not meet the criteria of an aneurysm, it was defined as dilatation. Regions of the arterial lesions were classified on the basis of anatomical location as follows: head and neck (brachiocephalic, carotid, vertebral, cephalic, and basilary artery), upper extremity (subclavian, axillary, and distal upper extremity artery), abdomen (celiac, superior mesenteric, inferior mesenteric, and renal artery), as well as the pelvis and lower extremity (iliac, femoral, and distal lower extremity artery). We also reviewed arterial thrombosis.

To evaluate the outcomes, we obtained information about transient ischemic attack (TIA), stroke, angina, heart failure, aortic valve regurgitation, arterial dissection, and rupture. Endovascular intervention included percutaneous transluminal angioplasty and percutaneous coronary intervention. Operations included bypass surgery,
| Variable                                | TAK (n = 206) | BD (n = 50) | p-value |
|-----------------------------------------|---------------|-------------|---------|
| **Demographic characteristics**         |               |             |         |
| Sex, female                             | 172 (83.5)    | 20 (40)     | <0.001  |
| Age at diagnosis (yr)                   | 43.2 ± 13.8   | 46.5 ± 14.0 | 0.207   |
| Age at clinical onset (yr)              | 39.8 ± 13.6   | 42.9 ± 13.8 | 0.225   |
| Diagnosis delay (mo)                    | 41.8 ± 71.5   | 25.2 ± 40.5 | 0.180   |
| Follow-up duration (mo)                 | 79.8 ± 66.9   | 96.6 ± 67.4 | 0.083   |
| Diabetes mellitus                       | 21 (10.2)     | 7 (14)      | 0.439   |
| Hypertension                            | 85 (41.3)     | 12 (24)     | 0.024   |
| Hyperlipidemia                          | 97 (47.1)     | 25 (50)     | 0.711   |
| Chronic kidney disease                  | 8 (3.9)       | 3 (6)       | 0.453   |
| Smoking                                 | 35/191 (18.3) | 23/49 (46.9)| <0.001  |
| ESR (mm/hr)                             | 40.85 ± 34.83 | 49.70 ± 32.45 | 0.123  |
| CRP (mg/dL)                             | 2.08 ± 4.17   | 5.85 ± 6.43 | <0.001  |
| Anemia                                  | 82 (39.8)     | 35 (70)     | <0.001  |
| Arterial thrombosis                     | 13 (6.3)      | 10 (20)     | 0.005   |
| **Constitutional symptoms**             |               |             |         |
| Fever                                   | 19 (9.2)      | 15 (30)     | <0.001  |
| Malaise                                 | 62 (30.1)     | 13 (26)     | 0.568   |
| Arthralgia                              | 15 (7.3)      | 18 (36)     | <0.001  |
| Night sweat                             | 5 (2.4)       | 1 (2)       | 1.000   |
| Weight loss                             | 29 (14.1)     | 9 (18)      | 0.484   |
| **Vascular symptoms**                   |               |             |         |
| Vascular bruit                          | 69 (33.5)     | 2 (4)       | <0.001  |
| Decreased arterial pulse                | 61 (29.6)     | 3 (6)       | 0.001   |
| Upper limb claudication                 | 22 (10.7)     | 0 (0)       | 0.010   |
| Lower limb claudication                 | 16 (7.8)      | 5 (10)      | 0.572   |
| Systolic blood pressure difference      | 142 (68.9)    | 2 (4)       | <0.001  |
| Carotidynia                             | 13 (6.3)      | 0 (0)       | 0.068   |
| Syncope                                 | 23 (11.2)     | 3 (6)       | 0.278   |
| **Cardiopulmonary symptoms**            |               |             |         |
| Dyspnea                                 | 66 (32)       | 17 (34)     | 0.790   |
| Palpitation                             | 22 (10.7)     | 5 (10)      | 0.888   |
| Anginal chest pain                      | 16 (7.8)      | 5 (10)      | 0.372   |
| Hemoptysis                              | 3 (1.5)       | 1 (2)       | 0.583   |
| **Neurologic symptoms**                 |               |             |         |
| Headache                                | 53 (25.7)     | 8 (16)      | 0.148   |
| Dizziness                               | 44 (21.4)     | 6 (12)      | 0.134   |
| Motor weakness                          | 36 (17.5)     | 5 (10)      | 0.196   |
| Sensory change                          | 45 (21.8)     | 9 (18)      | 0.550   |
| **Mucocutaneous/gastrointestinal symptoms** |           |             |         |
| Erythema nodosum                        | 0 (0)         | 11 (22)     | <0.001  |
| Oral ulcer                              | 20 (9.7)      | 48 (96)     | <0.001  |
| Genital ulcer                           | 7 (3.4)       | 30 (60)     | <0.001  |
| Abdominal pain                          | 5 (2.4)       | 13 (26)     | <0.001  |
| Diarrhea                                | 4 (1.9)       | 0 (0)       | 1.000   |
| Hematochezia/melena                     | 0 (0)         | 2 (4)       | 0.038   |

Values are presented as number (%) or mean±standard deviation. TAK: Takayasu’s arteritis, BD: Behçet’s disease, ESR: erythrocyte sedimentation rate, CRP: C-reactive protein.
arterial replacement and resection, aortic valve replacement, and coronary artery bypass surgery.

**Statistical analysis**

All statistical analyzes were performed using SPSS version 24.0 software (IBM, Armonk, NY, USA). Continuous data were expressed as means±standard deviations and compared using the Student’s t-test or Mann–Whitney test. Categorized data were expressed as percentiles and compared by the chi-square or Fisher’s exact test. A p-value less than 0.05 was considered statistically significant.

![Pattern of angiographic features in Takayasu’s arteritis (TAK) and Behçet’s disease (BD) according to distribution region. Results presented as number (%). *p < 0.05, **p < 0.01, and ***p < 0.001.](image-url)
RESULTS

Differences in clinical features in TAK and BD
A comparison of demographic data and clinical aspects of TAK and BD at diagnosis are shown in Table 1. There was no significant difference in age at clinical onset, diagnosis, and diagnosis delay. Arterial involvement was observed at diagnosis in 60% of BD patients. Mean follow-up duration was 79.8 months in TAK and 96.6 months in BD. Females were more prevalent in TAK than in BD (83.5% vs. 40%, \( p < 0.001 \)). Hypertension was more common in TAK than in BD (41.3% vs. 24%, \( p = 0.024 \)). In patients with hypertension, renal artery stenosis or occlusion was found in 28 (32.9%) of 85 TAK patients and only 1 (8.3%) of 12 BD patients. CRP was higher in BD than in TAK (5.85 mg/dL vs. 2.08 mg/dL, \( p < 0.001 \)), and anemia was also more observed in BD than in TAK (70% vs. 39.8%, \( p < 0.001 \)). Arterial thrombosis (20% vs. 6.3%, \( p = 0.005 \)) was likewise more common in BD than in TAK. 5 patients with TAK had arterial thrombosis of the left subclavian artery, 3 of the aorta, 2 of the left iliac artery and lower extremity, 1 of the coronary artery, 1 of the left renal artery, and 1 of the left carotid artery. Fever (30% vs. 9.2%, \( p < 0.001 \)) and arthralgia (36% vs. 7.3%, \( p < 0.001 \)) were more frequently presented in BD than in TAK. Abdominal pain (26% vs. 2.4%, \( p < 0.001 \)) was also more common in BD than in TAK. Abdominal pain in BD was caused by abdominal aneurysm in 9 patients, gastrointestinal involvement in 2, a duodenal delayed passage in 1, and from an unknown cause in 1 patient.

Comparison of angiographic features in TAK and BD
Stenosis (89.8% vs. 60%, \( p < 0.001 \)) and occlusion (65.5% vs. 32%, \( p < 0.001 \)) were more observed in TAK, whereas aneurysm (62% vs. 20.9%, \( p < 0.001 \)) was more frequent in BD (Figure 2).

When compared according to their distributions (Figure 3), the following were more involved in TAK than in BD: carotid (73.3% vs. 30%, \( p < 0.001 \)), subclavian (71.4% vs. 16%, \( p < 0.001 \)), descending aorta (35% vs. 12%, \( p = 0.002 \)), renal (23.8% vs. 10%, \( p = 0.032 \)), superior mesenteric artery (18.4% vs. 4%, \( p = 0.012 \)), and brachioce-
Table 2. Angiographic features of arterial involvement in Takayasu’s arteritis and Behçet’s disease

| Artery            | Stenosis (n=206) | Occlusion (n=206) | Dilatation (n=206) | Aneurysm (n=206) |
|-------------------|------------------|-------------------|--------------------|-------------------|
|                   | TAK              | BD                | p-value            | TAK              | BD                | p-value            | TAK              | BD                | p-value            |
| Ascending aorta   | 9 (4.4)          | 0 (0)             | 0.213              | 12 (5.8)         | 6 (12)            | 0.131              | 13 (6.3)         | 8 (16)            | 0.040              |
| Aortic arch       | 19 (9.2)         | 0 (0)             | 0.030              | 5 (2.4)          | 1 (2)             | 1.000              | 3 (1.5)          | 1 (2)             | 0.583              |
| Descending aorta  | 60 (29.1)        | 0 (0)             | <0.001             | 10 (4.9)         | 1 (2)             | 0.697              | 8 (3.9)          | 5 (10)            | 0.141              |
| Abdominal aorta   | 64 (31.1)        | 3 (6)             | 0.001              | 5 (2.4)          | 2 (4)             | 0.625              | 9 (4.4)          | 12 (24)           | <0.001             |
| Brachiocephalic   | 15 (7.3)         | 0 (0)             | 0.048              | 4 (1.9)          | 0 (0)             | 1.000              | 2 (1)            | 0 (0)             | 1.000              |
| Rt. cerebral      | 13/143 (9.1)     | 2/29 (6.9)        | 1.000              | 6/143 (4.2)      | 0/29 (0)          | 0.591              | 3/143 (2.1)      | 2/29 (6.9)        | 0.198              |
| Lt. cerebral      | 14/143 (9.8)     | 1/29 (3.4)        | 0.471              | 3/143 (2.1)      | 0/29 (0)          | 1.000              | 7/143 (4.9)      | 1/29 (3.4)        | 1.000              |
| Basilar           | 5/145 (3.4)      | 0/29 (0)          | 0.592              | 0/145 (0.7)      | 0/29 (0)          | N/A                | 1/145 (0.7)      | 0/29 (0)          | 1.000              |
| Rt. carotid       | 79 (38.3)        | 9 (18)            | 0.008              | 32 (15.5)        | 0 (0)             | 0.001              | 6 (2.9)          | 1 (2)             | 1.000              |
| Lt. carotid       | 91 (44.2)        | 9 (18)            | 0.001              | 37 (18)          | 0 (0)             | <0.001             | 11 (5.3)         | 1 (2)             | 0.470              |
| Rt. vertebral     | 28 (13.6)        | 3 (6)             | 0.140              | 13 (6.3)         | 0 (0)             | 0.079              | 3 (1.5)          | 0 (0)             | 1.000              |
| Lt. vertebral     | 34 (16.3)        | 2 (4)             | 0.023              | 15 (7.3)         | 3 (6)             | 1.000              | 1 (0.5)          | 0 (0)             | 0.000              |
| Rt. subclavian    | 43 (20.9)        | 2 (4)             | 0.005              | 31 (15)          | 0 (0)             | 0.003              | 3 (1.5)          | 1 (2)             | 0.583              |
| Lt. subclavian    | 64 (31.1)        | 2 (4)             | <0.001             | 67 (32.5)        | 1 (2)             | <0.001             | 3 (1.5)          | 1 (2)             | 0.583              |
| Rt. axillary      | 2 (1)            | 0 (0)             | 1.000              | 1 (0.5)          | 0 (0)             | 1.000              | 0 (0)            | 0 (0)             | N/A                |
| Lt. axillary      | 6 (2.9)          | 0 (0)             | 0.601              | 5 (2.4)          | 0 (0)             | 0.586              | 0 (0)            | 0 (0)             | N/A                |
| Celiac            | 32 (15.3)        | 4 (8)             | 0.169              | 8 (3.9)          | 1 (2)             | 1.000              | 3 (1.5)          | 0 (0)             | 1.000              |
| Superior mesenteric| 20 (9.7)         | 1 (2)             | 0.088              | 15 (7.3)         | 0 (0)             | 0.048              | 4 (1.9)          | 1 (2)             | 1.000              |
| Inferior mesenteric| 6 (2.9)          | 0 (0)             | 0.601              | 0 (0)            | 0 (0)             | N/A                | 0 (0)            | 0 (0)             | N/A                |
| Rt. renal         | 35 (17)          | 1 (2)             | 0.006              | 3 (1.5)          | 1 (2)             | 0.583              | 3 (1.5)          | 0 (0)             | 1.000              |
| Lt. renal         | 30 (14.6)        | 2 (4)             | 0.043              | 4 (1.9)          | 1 (2)             | 1.000              | 3 (1.5)          | 0 (0)             | 1.000              |
| Rt. iliac         | 11 (5.3)         | 2 (4)             | 1.000              | 2 (1)            | 1 (2)             | 0.480              | 2 (1)            | 0 (0)             | 1.000              |
| Lt. iliac         | 6 (2.9)          | 1 (2)             | 1.000              | 2 (1)            | 1 (2)             | 0.480              | 0 (0)            | 0 (0)             | N/A                |
| Rt. femoral       | 3 (1.5)          | 0 (0)             | 1.000              | 1 (0.5)          | 2 (4)             | 0.098              | 0 (0)            | 0 (0)             | N/A                |
| Lt. femoral       | 2 (1)            | 1 (2)             | 0.480              | 3 (1.5)          | 2 (4)             | 0.252              | 0 (0)            | 0 (0)             | N/A                |
| Coronary          | 37/123 (30.1)    | 9/25 (36)         | 0.560              | 14/123 (11.4)    | 5/25 (20)         | 0.321              | 1/123 (0.8)      | 0/25 (0)          | 1.000              |
| Pulmonary         | 19/203 (9.4)     | 1 (2)             | 0.138              | 5/203 (2.5)      | 1 (2)             | 1.000              | 11/203 (5.4)     | 1 (2)             | 0.470              |

Values are presented as number (%). TAK: Takayasu’s arteritis, BD: Behçet’s disease, N/A: not available.
phal trunk (13.6% vs. 2%, p=0.020). On the other hand, the femoral artery was more involved in BD than in TAK (10% vs. 2.4%, p=0.027).

In the aorta, stenosis (47.1% vs. 6%, p<0.001) was more frequent in TAK, whereas aneurysm (46% vs. 11.7%, p<0.001) was more presented in BD. In the head and neck region, stenosis (69.4% vs. 30%, p<0.001) and occlusion (34.5% vs. 8%, p<0.001) were more presented in TAK than in BD. These patterns were similar in the arteries of the upper extremity and the abdomen region. In the lower extremity region, aneurysm was more observed in BD than in TAK (12% vs. 1%, p=0.001). The difference between TAK and BD sorted by the distribution and pattern of each artery is shown in Table 2. Patients who had multiple lesions were 98.1% in TAK and 82% in BD (p<0.001).

Comparison of outcome in TAK and BD

There were no significant differences in TIA, stroke, angina, and heart failure between the two groups, as detailed in Table 3. However, the p-value for differences in aortic valve regurgitation was right at 0.05. Aortic valve regurgitation above grade 3 was more frequent in BD than in TAK (38% vs. 21.4%, p=0.014). Arterial dissection (10% vs. 1.9%, p=0.016) and rupture (12% vs. 0.5%, p<0.001) occurred more frequently in BD than in TAK. Arterial replacement/resection (66% vs. 9.7%, p<0.001), aortic valve replacement (38% vs. 13.1%, p<0.001), and bypass surgery (22% vs. 10.7%, p=0.032) were performed more often in BD than in TAK. Death occurred in 7 TAK patients and in 6 BD patients. Cause of death in the TAK cases was post-intervention bleeding, sepsis (2 patients each), stroke, and ischemic colitis (1 patient each). In the BD cases, death occurred from sepsis (2 patients), sudden cardiac arrest, as well as myocardial infarction and post-intervention bleeding (1 patient each). Cause remained unknown in 1 TAK patient and 1 BD patient.

DISCUSSION

This study analyzed and presented the differences in clinical, angiographic features, and outcome between TAK and BD. TAK is mainly associated with stenosis of the head and neck, upper extremity, aorta, and abdomen region. In BD, a common manifestation is aneurysm in the aorta and lower extremity region. Vascular complications were more observed in BD than in TAK. To the best of our knowledge, this is the first study to compare characteristics between TAK and BD.

Fever, arthralgia, high CRP, and anemia were more frequent in BD than in TAK. Constitutional symptoms are relatively common manifestations in both TAK and BD. In TAK, acute inflammation leads to constitutional symptoms such as fever, arthralgia, malaise, and night sweats. The incidence of fever and arthralgia has been relatively varied in earlier studies, with fever occurring in 3.4%~29% and arthralgia in 1.8%~39% [2,3,14]. In BD, vas-

Table 3. Complications and interventions in Takayasu’s arteritis and Behçet’s disease

| Variable                        | TAK (n = 206) | BD (n = 50) | p-value |
|---------------------------------|---------------|-------------|---------|
| Complication                    |               |             |         |
| Transient ischemic attack       | 9 (4.4)       | 2 (4)       | 1.000   |
| Ischemic stroke                 | 25 (12.1)     | 7 (14)      | 0.721   |
| Hemorrhagic stroke              | 5 (2.4)       | 3 (6)       | 0.190   |
| Angina                          | 21 (10.2)     | 9 (18)      | 0.124   |
| Heart failure                   | 38 (18.4)     | 15 (30)     | 0.071   |
| Aortic valve regurgitation      | 68 (33.2)     | 24 (48)     | 0.050   |
| Arterial dissection             | 4 (1.9)       | 5 (10)      | 0.016   |
| Arterial rupture                | 1 (0.5)       | 6 (12)      | <0.001  |
| Intervention                    |               |             |         |
| Bypass surgery                  | 22 (10.7)     | 11 (22)     | 0.032   |
| Percutaneous transluminal angioplasty | 37 (18) | 4 (8) | 0.085 |
| Arterial replacement/resection  | 20 (9.7)      | 33 (66)     | <0.001  |
| Aortic valve replacement        | 27 (13.1)     | 19 (38)     | <0.001  |
| Coronary artery bypass surgery  | 11 (5.3)      | 4 (8)       | 0.502   |
| Percutaneous coronary intervention | 12 (5.8) | 5 (10) | 0.339 |

Values are presented as number (%). TAK: Takayasu’s arteritis, BD: Behçet’s disease.
cular involvement tends to be associated with constitutional symptoms and a high acute phase response [15,16]. In a previous study of BD with arterial involvement, fever and arthralgia at onset were present in 29.7% and 38.6% of the cases, respectively [17]. From our study’s results, it appears that inflammatory symptoms at diagnosis may be closer to characteristics of BD than TAK.

Arterial thrombosis was more frequent in BD than in TAK. Although there are a few reports of thrombosis related to TAK [18,19], such an outcome is very rare. In contrast, cases of arterial thrombosis are often observed in BD. The mechanism of thrombosis in BD is not yet fully understood, but may be associated with endothelial damage and activation, abnormal fibrinolysis, and altered platelet function [8,20]. Several studies suggest that BD is more likely to be associated with arterial thrombosis than TAK.

Lower limb claudication was more frequent in BD than in TAK, although this difference was not statistically significant. This might be because BD had more occlusion in the lower extremities. Abdominal pain was more frequent in BD than in TAK, and its major cause in BD was arterial rather than gastrointestinal involvement. This outcome is consistent with previous results, in which abdominal aorta aneurysm was relatively common and the incidence of gastrointestinal lesions was as low as 3.6% in BD with arterial involvement [21]. Gastrointestinal symptoms were rarely seen in TAK, as also noted in a previous study [22], although mesenteric stenosis is relatively common in TAK.

The pattern and distribution of angiographic lesions was different in TAK compared with BD, in that there was more stenosis and occlusion in TAK, and more aneurysm in BD. In TAK, dendritic cells, T lymphocytes, and macrophages infiltrate media and adventitia with granulomatous inflammation and progress to intima. This course leads to intima proliferation, medial necrosis, and adventitial fibrosis. In its late phase, marked fibrosis causes stenosis or occlusion [23]. BD is characterized by neutrophilic vasculitis around the vasa vasorum. Infiltration of neutrophils and lymphocytes exhibits in media and adventitia. Inflammation contributes to the fragmentation of elastic fibers in media with weakening of the arterial wall, resulting in aneurysmal change [24].

However, fibrous thickening of the intima can also occur in BD [25]. In our study, BD cases had a relatively large number of stenoses and occlusions, and the major location of this development was the carotid artery. It might be because we included the lesions evaluated by the carotid Doppler, which had higher sensitivity than CT that was usually used for evaluation of cardiovascular lesion [26-28]. The occlusion of lower extremity region in BD was also relatively common. This result was also shown in a previous study of BD, which demonstrated that stenosis and occlusion were common patterns, occurring in 13.5% and 36.5% of cases, respectively [17]. In TAK, the ascending aorta had more aneurysms than stenoses, as shown in previous studies [2,14]. This observation was unexpected, since TAK is usually known to have stenosis. This might be due to hemodynamic mechanisms and the anatomical structure of the ascending aorta, including the abundance of elastin fiber, collagen, and smooth muscle cells [29]. Therefore, diagnosis based only on the pattern of arterial involvement may require attention.

Considerable research has shown that TAK and BD are both associated with cardiovascular complications including angina, aortic valve regurgitation, heart failure, and stroke [2,4,14,22,30]. In our study, most complications and interventions showed no significant differences between the two groups. However, BD was associated with more severe aortic valve regurgitation and more frequent aortic valve replacements, perhaps because BD involved more aneurysmal changes in the ascending aorta. Vascular complications, including arterial dissection and rupture, and the related intervention procedures were more common in BD. TAK and BD are considered to be causes of arterial dissections and ruptures. However, TAK generates relatively low risk compared with BD, because TAK involves intimal fibrosis in the chronic phase [31]. In BD, destruction of the elastic cells in the media leads to arterial aneurysm or dissection with the corresponding risk of arterial rupture [7,32]. We found that BD had higher rates of vascular complication than TAK.

Our study had several limitations. First, it was retrospective in design and based on data from a single medical center, so there might be a bias in our particular clinical data. Second, patients who were not fully evaluated for major vascular lesions were excluded. However, this study showed mostly similar results to previous studies on the distribution and pattern of arterial involvement in TAK and BD. Third, this study only included Korean patients, and the numerous manifestations of arterial involvement with these two diseases could be different across a variety of ethnic populations.
CONCLUSION

In this study, TAK and BD are different in their clinical presentations, angiographic features, and outcome. TAK produces stenosis and occlusion in the head and neck, upper extremity, aorta, and abdomen region. BD presents symptoms of inflammation, such as fever and high CRP, and causes aneurysms in the aorta and lower extremities. The rates of vascular complications are higher in BD than in TAK.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

AUTHOR CONTRIBUTIONS

Conception and design of study: S.J.C., J.S.O., S.C.H., Y.G.K., B.Y., C.K.L. Acquisition of data: S.J.C., H.J.K. Analysis and interpretation of data: S.J.C., S.C.H., Y.G.K., B.Y., C.K.L. Acquisition of data: S.J.C., H.J.K. Drafting the manuscript: S.J.C. Revising the manuscript critically for important intellectual content: S.J.C., H.J.K., S.C.H., J.W.K., J.S.O., S.C.H., Y.G.K., B.Y., C.K.L.

REFERENCES

1. Kerr GS, Hallahan CW, Giordano J, Leavirt RY, Fauci AS, Rottem M, et al. Takayasu arteritis. Ann Intern Med 1994;120:919-29.
2. Schmidt J, Kermani TA, Bacani AK, Crowson CS, Cooper LT, Matteson EL, et al. Diagnostic features, treatment, and outcomes of Takayasu arteritis in a US cohort of 126 patients. Mayo Clin Proc 2013;88:822-30.
3. Lee GY, Jang SY, Ko SM, Kim EK, Lee SH, Han H, et al. Cardiovascular manifestations of Takayasu arteritis and their relationship to the disease activity: analysis of 204 Korean patients at a single center. Int J Cardiol 2012;159:14-20.
4. Sakane T, Takeno M, Suzuki N, Inaba G. Behçet’s disease. N Engl J Med 1999;341:1284-91.
5. Sarica-Kucukoglu R, Akdag-Kose A, Kayabali M, Yazganoglu KD, Disci R, Erzenjin D, et al. Vascular involvement in Behçet’s disease: a retrospective analysis of 2319 cases. Int J Dermatol 2006;45:919-21.
6. al-Dalaan AN, al Balaa SR, el Ramahi K, al-Kawi Z, Bohlega S, Bahabri S, et al. Behçet’s disease in Saudi Arabia. J Rheumatol 1994;21:658-61.
7. Tüzün H, Beşirli K, Sayın A, Vural FS, Hamuryuden V, Hızlı N, et al. Management of aneurysms in Behçet’s syndrome: an analysis of 24 patients. Surgery 1997;121:150-6.
8. Ames PR, Steuer A, Pap A, Denman AM. Thrombosis in Behçet’s disease: a retrospective survey from a single UK centre. Rheumatology (Oxford) 2001;40:652-5.
9. Arend WP, Michel BA, Bloch DA, Hunder GG, Calabrese LH, Edworthy SM, et al. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. Arthritis Rheum 1990;33:1129-34.
10. International Team for the Revision of the International Criteria for Behçet’s Disease (ITR-ICBD). The International Criteria for Behçet’s Disease (ICBD): a collaborative study of 27 countries on the sensitivity and specificity of the new criteria. J Eur Acad Dermatol Venereol 2014;28:338-47.
11. Saliba E, Sia Y. The ascending aortic aneurysm: when to intervene? Int J Cardiol Heart Vasc 2015;6:91-100.
12. Gagné-Loranger M, Dumont É, Voisine P, Mohammadi S, Dagenais F. Natural history of 40-50 mm root/ascending aortic aneurysms in the current era of dedicated thoracic aortic clinics. Eur J Cardiothorac Surg 2016;50:562-6.
13. Forsdahl SH, Solberg S, Singh K, Jacobsen BK. Abdominal aortic aneurysms, or a relatively large diameter of non-aneurysmal aortas, increase total and cardiovascular mortality: the Tromsø study. Int J Epidemiol 2010;39:225-32.
14. Bicakcigil M, Aksu K, Kamali S, Ozbalkan Z, Ates A, Karadag O, et al. Takayasu’s arteritis in Turkey - clinical and angiographic features of 248 patients. Clin Exp Rheumatol 2009;27(1 Suppl 52):559-64.
15. Seyahi E, Karaaslan H, Ugurlu S, Yazici H. Fever in Behçet’s syndrome. Clin Exp Rheumatol 2013;31(3 Suppl 77):64-7.
16. Müftüoğlu AU, Yazici H, Yurdakul S, Tüüzün Y, Pazarli H, Güngen G, et al. Behçet’s disease. Relation of serum C-reactive protein and erythrocyte sedimentation rates to disease activity. Int J Dermatol 1986;25:235-9.
17. Saadoun D, Asli B, Wechsler B, Houman H, Geri G, Desseaux K, et al. Long-term outcome of arterial lesions in Behçet disease: a series of 101 patients. Medicine (Baltimore) 2012;91:18-24.
18. Ostertag-Hill CA, Abdo AK, Alexander JQ, Skeit N. Unique case of Takayasu arteritis with severe distal aortic stenosis and iliac thrombosis. Ann Vasc Surg 2016;32:128-e7-13.
19. Purkayastha S, Jayadevan ER, Kapilamoorthy TR, Gupta AK. Suction thrombectomy of thrombotic occlusion of the subclavian artery in a case of Takayasu’s arteritis. Cardiovasc Intervent Radiol 2006;29:289-93.
20. Wu X, Li G, Huang X, Wang L, Liu W, Zhao Y, et al. Behçet’s disease complicated with thrombosis: a report of 93 Chinese cases. Medicine (Baltimore) 2014;93:e263.
21. Yang SS, Park KM, Park YJ, Kim YW, Do YS, Park HS, et al. Peripheral arterial involvement in Behçet’s disease: an analysis of the results from a Korean referral center. Rheumatol Int 2013;33:2101-8.
22. Park YB, Hong SK, Choi KJ, Sohn DW, Oh BH, Lee MM, et al. Takayasu arteritis in Korea: clinical and angiographic features. Heart Vessels Suppl 1992;7:55-9.
23. Vaidheeswar P, Deshpande JR. Pathology of Takayasu arteritis: a brief review. Ann Pediatr Cardiol 2013;6:62-58.
24. Kobayashi M, Ito M, Nakagawa M, Nishikimi N, Sakurai T, et al. Neutrophil and endothelial cell activation in the vasa vasorum in vasculo-Behçet disease. Histopathology 2000;36:362-71.
25. Matsumoto T, Uekusa T, Fukuda Y. Vasculo-Behçet’s disease: a pathologic study of eight cases. Hum Pathol 1991;22:45-51.
26. Suwanwela NC, Suwanwela N. Takayasu arteritis: ultra-
sonographic evaluation of the cervico-cerebral arteries. Int J Cardiol 1998;66 Suppl 1:S163-73.
27. Koo HJ, Yang DH, Kang JW, Lee JY, Kim DH, Song JM, et al. Demonstration of infective endocarditis by cardiac CT and transoesophageal echocardiography: comparison with intra-operative findings. Eur Heart J Cardiovasc Imaging 2018;19:199-207.
28. Koo HJ, Lee JY, Kim GH, Kang JW, Kim YH, Kim DH, et al. Paravalvular leakage in patients with prosthetic heart valves: cardiac computed tomography findings and clinical features. Eur Heart J Cardiovasc Imaging 2018;19:1419-27.
29. Yang KQ, Meng X, Zhang Y, Fan P, Wang LP, Zhang HM, et al. Aortic aneurysm in Takayasu arteritis. Am J Med Sci 2017;354:539-47.
30. Tsui KL, Lee KW, Chan WK, Chan HK, Hon SF, Leung TC, et al. Behçet’s aortitis and aortic regurgitation: a report of two cases. J Am Soc Echocardiogr 2004;17:83-6.
31. Wu XP, Zhu P. Clinical features of aortic dissection associated with Takayasu’s arteritis. J Geriatr Cardiol 2017;14:485-7.
32. Hosaka A, Miyata T, Shigematsu H, Shigematsu K, Okamoto H, Ishii S, et al. Long-term outcome after surgical treatment of arterial lesions in Behçet disease. J Vasc Surg 2005;42:116-21.