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Original Article

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

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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. **(J Rheum Dis 2020;27:100-109)**

Key Words. Behcet syndrome, Takayasu arteritis, Arteritis, Cardiovascular diseases

INTRODUCTION

Takayasu's arteritis (TAK) and Behçet's disease (BD) are systemic vasculitis 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-

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sis is also associated with BD [8].

TAK and BD have distinct clinical characteristics, and the presence of symptoms indicating BD can help distinguish TAK and BD. However, the data comparing clinical, angiographic features and vascular outcome of both diseases are only 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 $\geq$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 basilar 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,

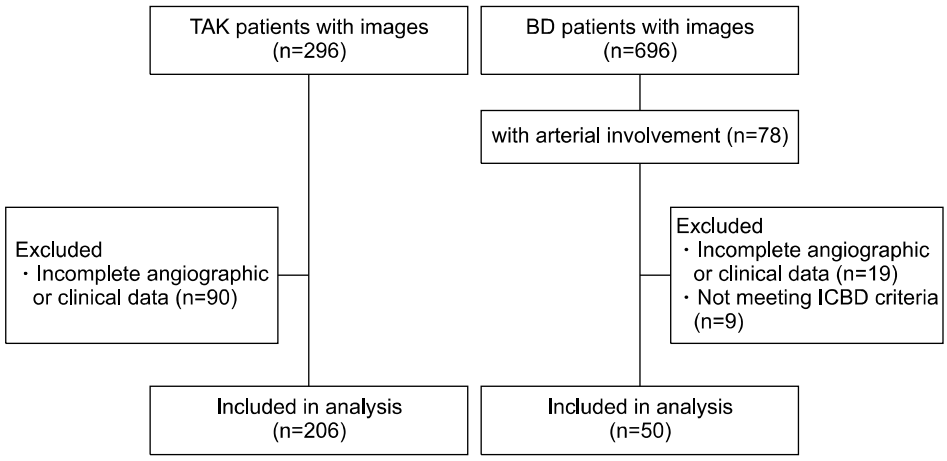

Figure 1. Study participants in Takayasu's arteritis (TAK) and Behçet's disease (BD). ICBD: International Criteria for Behçet's disease.

Table 1. Demographic data and clinical features at diagnosis in Takayasu's arteritis and Behçet's disease

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)	0.572
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.

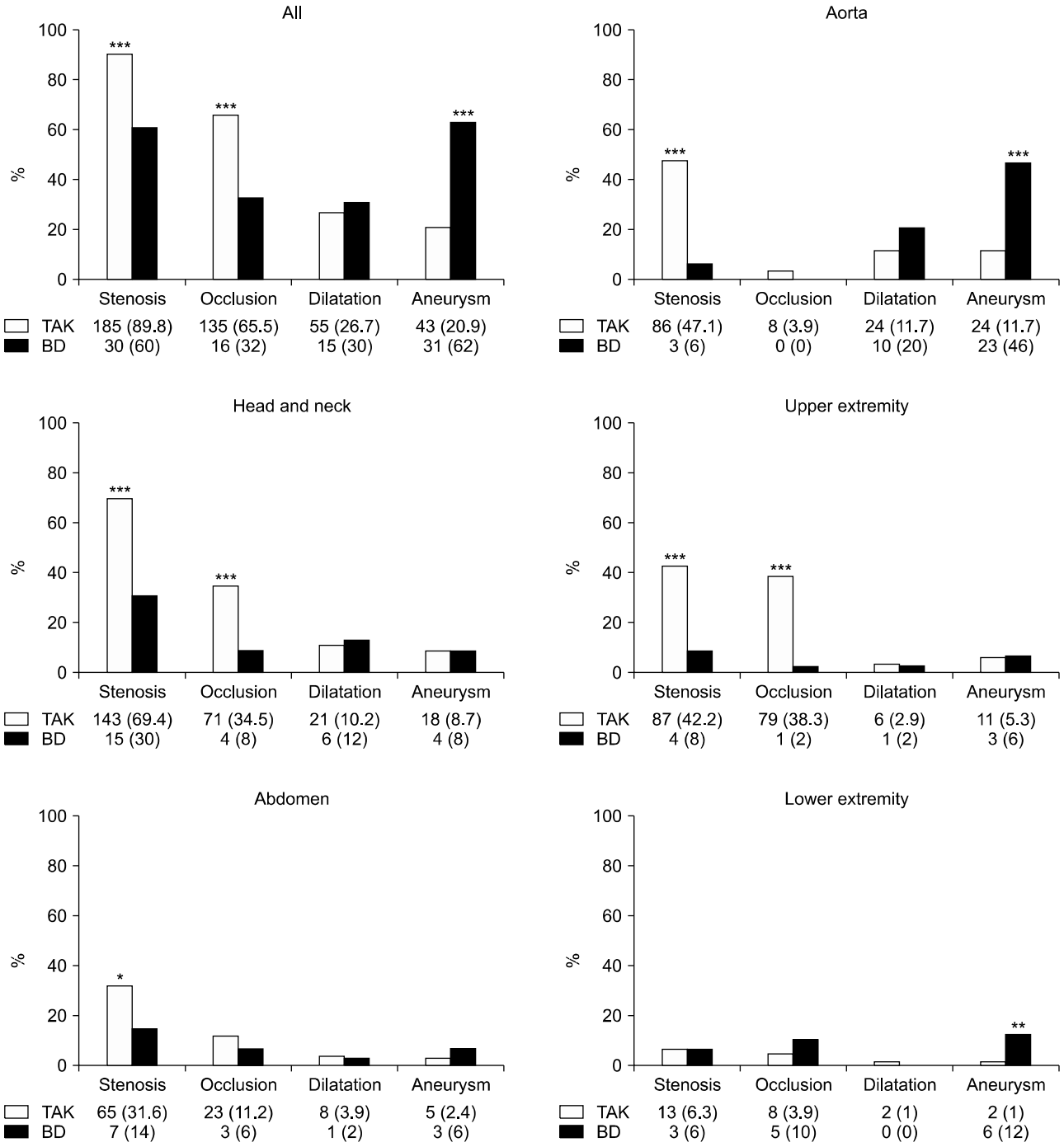

Figure 2. Pattern of angiographic features in Takayasu's arteritis (TAK) and Behçet's disease (BD) according to distribution region. Results presented as number (%). *$p<0.05$, **$p<0.01$, and ***$p<0.001$.

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-

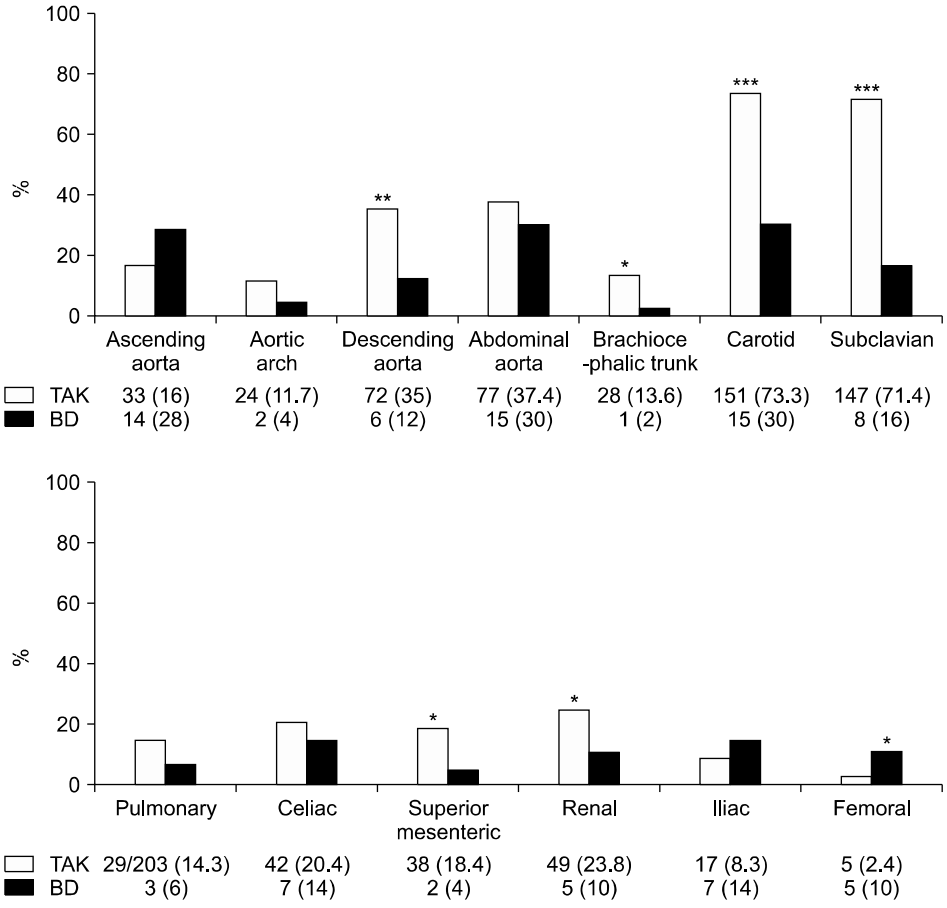

Figure 3. Arterial involvement in Takayasu's arteritis (TAK) and Behçet's disease (BD) according to distribution. Results presented as number (%). *$p<0.05$, **$p<0.01$, and ***$p<0.001$.

Table 2. Angiographic features of arterial involvement in Takayasu's arteritis and Behçet's disease

Artery	Stenosis			Occlusion			Dilatation			Aneurysm		
	TAK (n=206)	BD (n=50)	p-value	TAK (n=206)	BD (n=50)	p-value	TAK (n=206)	BD (n=50)	p-value	TAK (n=206)	BD (n=50)	p-value
Ascending aorta	9 (4.4)	0 (0)	0.213	0 (0)	0 (0)	N/A	12 (5.8)	6 (12)	0.131	13 (6.3)	8 (16)	0.040
Aortic arch	19 (9.2)	0 (0)	0.030	0 (0)	0 (0)	N/A	5 (2.4)	1 (2)	1.000	3 (1.5)	1 (2)	0.583
Descending aorta	60 (29.1)	0 (0)	<0.001	2 (1)	0 (0)	1.000	10 (4.9)	1 (2)	0.697	8 (3.9)	5 (10)	0.141
Abdominal aorta	64 (31.1)	3 (6)	<0.001	7 (3.4)	0 (0)	0.351	5 (2.4)	2 (4)	0.625	9 (4.4)	12 (24)	<0.001
Brachiocephalic	15 (7.3)	0 (0)	0.048	8 (3.9)	1 (2)	1.000	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	5/143 (3.5)	1/29 (3.4)	1.000
Lt. cerebral	14/143 (9.8)	1/29 (3.4)	0.471	3/143 (2.1)	0/29 (0)	1.000	3/143 (2.1)	1/29 (3.4)	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)	0/29 (0)	N/A	1/145 (0.7)	1/29 (3.4)	0.306	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	6 (2.9)	3 (6)	0.384
Lt. carotid	91 (44.2)	9 (18)	0.001	37 (18)	0 (0)	<0.001	11 (5.3)	1 (2)	0.470	4 (1.9)	1 (2)	1.000
Rt. vertebral	28 (13.6)	3 (6)	0.140	13 (6.3)	0 (0)	0.079	3 (1.5)	0 (0)	1.000	1 (0.5)	0 (0)	1.000
Lt. vertebral	34 (16.5)	2 (4)	0.023	15 (7.3)	3 (6)	1.000	1 (0.5)	0 (0)	1.000	0 (0)	0 (0)	N/A
Rt. subclavian	43 (20.9)	2 (4)	0.005	31 (15)	0 (0)	0.003	3 (1.5)	1 (2)	0.583	8 (3.9)	2 (4)	1.000
Lt. subclavian	64 (31.1)	2 (4)	<0.001	67 (32.5)	1 (2)	<0.001	3 (1.5)	1 (2)	0.583	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	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	0 (0)	1 (2)	0.195
Celiac	32 (15.5)	4 (8)	0.169	8 (3.9)	1 (2)	1.000	3 (1.5)	0 (0)	1.000	1 (0.5)	2 (4)	0.098
Superior mesenteric	20 (9.7)	1 (2)	0.088	15 (7.3)	0 (0)	0.048	4 (1.9)	1 (2)	1.000	2 (1)	1 (2)	0.480
Inferior mesenteric	6 (2.9)	0 (0)	0.601	0 (0)	0 (0)	N/A	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	0 (0)	1 (2)	0.195
Lt. renal	30 (14.6)	2 (4)	0.043	4 (1.9)	1 (2)	1.000	3 (1.5)	0 (0)	1.000	2 (1)	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	1 (0.5)	3 (6)	0.024
Lt. iliac	6 (2.9)	1 (2)	1.000	2 (1)	1 (2)	0.480	0 (0)	0 (0)	N/A	2 (1)	2 (4)	0.172
Rt. femoral	3 (1.5)	0 (0)	1.000	1 (0.5)	2 (4)	0.098	0 (0)	0 (0)	N/A	0 (0)	4 (8)	0.001
Lt. femoral	2 (1)	1 (2)	0.480	3 (1.5)	2 (4)	0.252	0 (0)	0 (0)	N/A	0 (0)	1 (2)	0.195
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	5/123 (4.1)	2/25 (8)	0.337
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	2/203 (1)	0 (0)	1.000

Values are presented as number (%). TAK: Takayasu's arteritis, BD: Behçet's disease, N/A: not available.

phalic 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. Drafting the manuscript: S.J.C. Revising the manuscript critically for important intellectual content: S.J.C., H.J.K., D.H.Y., J.W.K., J.S.O., S.C.H., Y.G.K., B.Y., C.K.L.

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