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● *Original Contribution*

CHARACTERISTIC ECHOCARDIOGRAPHIC MANIFESTATIONS OF BEHÇET'S DISEASE

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Abstract—The objective of this study was to determine the characteristic echocardiographic manifestations of Behçet's disease. Ninety-seven Behçet's patients admitted to our hospital from January 2002 to December 2016 were retrospectively analyzed. There were 63 patients (64.9%) with cardiac involvement; these included 47 men (74.6%) and 16 women (25.4%). Of these 63 patients, most (74.6%) exhibited valvular lesions, especially aortic regurgitation. The primary characteristic manifestations of valves were cusp prolapse with aneurysmal changes, vegetation-like lesions and echo-free spaces within the annulus. Second (30.0%) were aortic lesions, including aortic dilation or aneurysm formation, aortic pseudoaneurysm and coronary sinus aneurysm. Other manifestations, such as cardiac thrombosis, coronary arterial pseudoaneurysm, pulmonary aneurysm and pericardial effusion, were rare. Cardiac involvement is not uncommon in Behçet's patients, especially in males, and it is characterized by valvular disease, especially aortic regurgitation. The diagnosis of Behçet's disease should be considered when evaluating patients with these characteristic echocardiographic manifestations. (E-mail: lrj4020@126.com) **© 2017 World Federation for Ultrasound in Medicine & Biology. All rights reserved.**

Key Words: **Behçet's disease, Echocardiography, Manifestation, Diagnosis, Gender.**

INTRODUCTION

Behçet's disease (BD) is an idiopathic chronic inflammatory disorder characterized by the involvement of large and small arteries and the variable involvement of several organs (Behçet 1937). BD can affect almost all systems, including the mucocutaneous, urogenital, locomotor, ocular, neurologic, gastrointestinal, respiratory and vascular systems. BD involving the cardiovascular system has been reported to have various manifestations, including endomyocardial fibrosis, mural thrombi, active valvulitis and vasculitis involving the pulmonary artery or aorta (Houman et al. 2002; Seyahi et al. 2003). Moreover, patients with valvular lesions caused by BD have a notoriously high incidence of relapse after surgery. Outcomes may be improved by early diagnosis based on the characteristic echocardiographic features and/or pathologic findings, followed by aggressive surgical debridement of the aortic root and post-operative immunosuppressive therapy (Han et al. 2009; Jeong et al. 2009). In this article, we describe and review the spectrum of echocardiographic findings in patients with BD to determine the characteristic echocardiographic manifestations of BD.

METHODS

For this retrospective study, we reviewed a prospective registry of all hospitalized patients with BD by searching the Electronic Medical Record systems at Beijing Anzhen Hospital from January 2002 to December 2016. Because there is no pathognomonic test for the diagnosis for BD, the diagnosis was determined on clinical grounds. A total of 140 patients initially diagnosed with BD were identified from 56,117 hospitalized patients. Of these patients, 34 re-hospitalized cases in this period, 6 patients who did not meet the International Criteria for Behçet's Disease (ICBD) (International Team for the Revision of the International Criteria for Behçet's Disease [ITR-ICBD] 2014) and 3 patients without integrated echocardiographic reports or images were excluded. Our study cohort consisted of 97 patients with BD. We reviewed the echocardiographic reports and images of these 97 patients by searching the ultrasound workstation. Baseline demographic and clinical data were collected from medical records. Data were obtained from standard

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Table 1. Clinical variables and outcomes of patients with cardiac involvement (N = 63)

Age (y)	39.54 ± 10.62
Diagnosis age (y)	37.06 ± 8.59
Males	47 (74.6%)
Females	16 (25.4%)
Cardiac involvement as initial manifestation	38 (60.3%)
Surgical intervention	43 (68.3%)
Multiple surgical interventions	16 (25.4%)
Death or heart transplantation	5 (7.9%)

hospitalization documentation. Our institutional review board approved this retrospective study and waived the requirement for informed consent.

Statistical analysis

Descriptive data were presented as the mean ± standard deviation for normally distributed continuous variables and as the frequency (%) for categorical variables. The significance of group differences in clinical characteristics was evaluated with the independent *t*-test for continuous variables and the χ^2-test for categorical variables. For skewed distributions, the Wilcoxon rank-sum test was used. Statistical analyses were performed using SPSS version 17.0 (IBM, Armonk, NY, USA) and a p value < 0.05 was considered to indicate statistical significance.

RESULTS

Clinical characteristics

Of the 97 patients (age: 18–78 y, mean age: 41.58 ± 12.46 y) diagnosed with BD, 63 patients (64.9%, age: 18–78 y, mean age: 39.54 ± 10.62 y) had cardiac involvement, as detected by trans-thoracic echocardiography. Their clinical and demographic data are summarized in Tables 1–3. Among this group, males (74.6%) were more common, and the initial manifestation of BD was cardiac involvement in 38 patients (60.3%). Forty-three patients (68.3%) underwent surgical intervention, 16 patients (25.4%) experienced complications and underwent re-operation, 2 patients (3.2%) underwent heart transplantation and 3 patients (4.8%) died in the hospital. These patients' erythrocyte sedimentation rates (ESRs) and high-sensitivity C-reactive protein (hsCRP) levels were significantly increased, representing signs of inflammation.

Table 2. Characteristic manifestations of patients with cardiac involvement (N = 63)

Erythrocyte sedimentation rate (mm/1st h)	17.31 ± 18.40
High-sensitivity C-reactive protein (mg/L)	12.59 ± 15.43
Fever	14 (22.2%)
Recurrent oral ulcer	61 (96.8%)
Recurrent genital ulcer	39 (61.9%)
Skin lesions	42 (66.7%)
Ophthalmic involvement	8 (12.7%)

Table 3. Cardiac symptoms of patients with cardiac involvement (N = 63)

Chest distress	36 (57.1%)
Chest pain	10 (15.9%)
Dyspnea	20 (31.7%)
Edema	4 (6.3%)
Fatigue	4 (6.3%)
Hemoptysis	6 (9.5%)

Gender-specific differences

The entire group included 65 male and 32 female patients, and we compared clinical characteristics by gender (Table 4). Age and age at diagnosis did not significantly differ between men and women ($p > 0.05$). Cardiac lesions occurred more frequently in males. Moreover, male patients required more surgical interventions ($p < 0.05$). However, the need for multiple surgical interventions and outcome events such as death and heart transplantation did not significantly differ between the groups ($p > 0.05$).

Echocardiographic findings

The echocardiographic findings in the 63 patients are summarized in Table 5. In our study, the most common cardiac lesion was valvular disease, especially in the aortic valve, followed by the mitral valve, and aortic lesions, such as aortic dilation, aortic pseudoaneurysm and sinus Valsalva aneurysm. Right cardiac thrombosis; coronary artery lesions, such as coronary pseudoaneurysm; pulmonary artery lesions, such as pulmonary aneurysm; and pericardial lesions, such as pericardial effusion, were rare.

Cardiac valve involvement. Cardiac valve involvement was found in 47 patients (74.6%) and manifested mainly as severe valvular regurgitation (Fig. 1A, B), including 41 patients (65.1%) with aortic valvular lesions, 3 patients (4.8%) with mitral valvular lesions and 3 patients (4.8%) with bivalvular lesions. Among these patients, 35 came to our hospital before initial surgery, and the other 12 underwent artificial aortic valve replacement in other hospitals because of aortic valve insufficiency and then came to our hospital for re-operation because of valve dehiscence and paraprosthetic leakage. The initial

Table 4. Comparison of male and female clinical characteristics

Characteristic	Men (N = 65)	Women (N = 32)	*p* value
Age (y)	41.17 ± 12.09	42.41 ± 13.33	0.648
Age at diagnosis (y)	37.93 ± 10.00	36.29 ± 11.36	0.477
Echocardiographic findings	47 (72.3%)	16 (50.0%)	0.03
Cardiac symptoms	52 (80.0%)	14 (43.8%)	<0.001
Surgical intervention	36 (55.4%)	7 (21.9%)	0.002
Multiple surgical interventions	14 (21.5%)	2 (6.3%)	0.08
Death or heart transplantation	3 (4.6%)	2 (6.3%)	0.536