Sir,

Behçet’s disease (BD) is a multi-systemic disease of unknown aetiology characterized by chronic relapsing orogenital ulcers, uveitis and systemic involvement including articular, gastrointestinal, cardiopulmonary, neurological and vascular pathologies (1). The incidence and nature of the cardiovascular involvement in BD is not yet clearly documented. The aim of this study was to evaluate the cardiovascular findings in patients with BD.

MATERIALS AND METHODS

This study was performed at the Outpatient Clinic for Behçet’s Disease, Numune Education and Research Hospital, Ankara, Turkey in 30 consecutive patients with BD. The patients were diagnosed according to the International Criteria for Behçet’s Disease (2). The patients’ medical histories were obtained. The cardiovascular disorders were diagnosed by clinical and radiological examinations, including echocardiography, electrocardiography and lower extremity Doppler ultrasonography. For statistical analysis, $\chi^2$ and Mann-Whitney $U$ tests were used.

RESULTS

Thirty patients with Behçet’s disease, (9 men and 21 women, age range 19–49 years, mean age 34 years) were enrolled in the study. Eighteen of the patients had been taking therapies for BD, including colchicine, oral steroids and cyclosporine. The mean duration of the disease was 6.8 years. Sixteen (53%) of the patients had at least one skin lesion during the examinations, including oral and genital aphthae, papulopustular lesions, erythema nodosum like lesions and trombophlebitis. None of the patients had clinically apparent cardiovascular disease on physical examination or had a history of cardiovascular disease.

Ten (33%) patients had findings in echocardiography including minimal tricuspid valve insufficiency (8 pat.), minimal mitral valve insufficiency (6 pat.), mitral valve prolapse (2 pat.), minimal pulmonary valve insufficiency (1 pat.) and mild pulmonary hypertension (1 pat.). Three (10%) of those patients were male and 7 (23%) were female. There was no significant difference between the number of males and females who had echocardiographic findings ($p > 0.05$, $\chi^2$ test). While the mean duration of the disease in patients with echocardiographic findings was 6.8 years, the mean duration of the disease in patients without echocardiographic findings was 6.2 years. No significant relation between the duration of Behçet’s disease and the presence of echocardiographic findings was found ($p > 0.05$, Mann-Whitney $U$ test).

Thirteen (43%) patients had findings in venous Doppler ultrasonography. Twelve (40%) of them had venous insufficiency and one (3%) had chronic deep vein thrombosis. Five (38%) of the patients who had venous disorder were male and 8 (62%) were female. There was no significant difference between the number of males and females who had venous disorder ($p > 0.05$, $\chi^2$ test). While the mean duration of the disease in patients with venous disease was 6.8 years, the mean duration of the disease in patients without venous disease was 6.7 years. No significant correlation between the duration of Behçet’s disease and the presence of venous disorder was found ($p > 0.05$, Mann-Whitney $U$ test).

DISCUSSION

BD is a multi-systemic disorder characterized by vasculitis which may lead to functional disturbances in highly vascularized organs (3). Resected vascular specimens have been shown to have fragmentation and splitting of the elastic fibres in media with perivascular mononuclear cell infiltration (4).

Cardiovascular involvement has been reported in BD (1, 3–12). Vascular involvement in BD has been reported in 7–40% of patients in the literature. As nearly 8% of these will have serious complications during follow-up, identification of the vascular lesions is important. Venous occlusions, variceal development, and arterial occlusion might be seen in patients with BD (5, 6). Although any large or small arteries or veins may be involved in BD, venous lesions are the predominant vascular lesions (5). The incidence of venous lesions were reported to be between 2% and 46% (7). So far, little information is available on the extent of vascular involvement in patients with BD who are free of vascular symptoms. Kuzu et al. (8) evaluated peripheral vascular system disorders in 34 individuals with BD who did not have any vascular symptoms. They detected venous insufficiency in 32% patients and reported that vascular symptom-free patients with BD had a high incidence of peripheral vascular involvement compared with healthy controls.

We have found the incidence of venous insufficiency to be 43% in patients with BD. This high frequency may explain the frequent development of venous occlusion in the lower extremities of patients with BD (6).
Various cardiac abnormalities have been described in patients with BD (1, 3, 4, 9–11). The number of reports remains small, but increasing awareness has widened the spectrum of manifestations. The incidence and nature of cardiac involvement in BD are not yet clearly documented. Gurgun et al. (9) used trans-oesophageal echocardiography and electrocardiography to define cardiac involvement in patients with BD. Higher incidences of inter-atrial septum aneurysm, mitral valve prolapse, mitral regurgitation and aneurysmal dilatations of the sinus of Valsalva and ascending aorta were observed in the patients with BD. In addition, increased dispersion of ventricular repolarisation and positive late potentials were detected with electrocardiography. Giordano et al. (10) evaluated cardiac involvement in patients with BD and reported mitral valve prolapse and dilatative cardiomyopathy. Gullu et al. (3) reported silent myocardial ischaemia incidence significantly higher in BD compared with the control group and suggested that impaired endothelial cell function may be the underlying cause in the pathogenesis of BD or of its vascular complications. Also, cases of cardiac involvement have been reported in the literature, including intracardiac thrombosis, myocardial infarction and left ventricular dysfunction due to coronary arteritis and myocarditis (3, 11).

In our study we detected minimal mitral and tricuspid valve insufficiencies, mitral valve prolapse and mild pulmonary hypertension in 33% of patients. As streptococcal infections were proposed in both the pathogenesis of the valve diseases and BD, this might be the reason for the increased incidence of valvular heart diseases in patients with BD (12, 13).

In conclusion, cardiovascular findings are not uncommon in BD. Assessment of the cardiovascular system is advised in patients with BD even if there is no clinically apparent cardiovascular disease.

REFERENCES