Clinical research

Demographic and clinical characteristics of Behçet's disease in Iraqi patients

Faiq Isho Gorial¹, Mais Ajeel Jabbar²

¹Rheumatology Unit, College of Medicine, University of Baghdad, Baghdad, Iraq

²Rheumatology Unit, Baghdad Teaching Hospital, Baghdad, Iraq

Submitted: 11 December 2020 **Accepted:** 24 January 2021

Arch Med Sci Aging 2021; 4: e1–e5 DOI: https://doi.org/10.5114/amsa.2021.106113 Copyright © 2021 Termedia & Banach

Abstract

Introduction: Behçet's disease (BD) is a chronic autoimmune multisystemic vasculitic disease. This study investigated the demographic and clinical characteristics of BD in Iraqi patients.

Material and methods: This descriptive observational study involved a total of 71 consecutive patients with BD diagnosed according to the International Study Group criteria 1990 for BD at the rheumatology unit. Demographic features, clinical characteristics, and medications used by the patients were recorded. Results: The mean age of patients was 36.0 ±10.8 years with disease duration of 5.1 ±6.0 years. There were 45 (63.4%) males and 26 (36.6%) females, BD activity score was 5.1 ±6.0. Mucosal manifestations were present in 71 (100%) patients, ocular involvement in 62 (87.3%), articular involvement in 51 (71.8%), skin involvement in 48 (67.6%), myalgia in 16 (22.5%), neurological in 9 (12.7%), vascular in 3 (4.2%) patients, and gastrointestinal involvement in 1 (1.4%) patient. The most commonly used treatment in this study was colchicine in 41 (57.7%) followed by infliximab in 34 (47.9%), corticosteroids in 32 (45.1%), azathioprine in 19 (26.8%), adalimumab in 3 (4.2%), cyclosporine in 2 (2.8%), sulphasalazine in 2 (2.8%), and methotrexate in 2 (2.8%). Conclusions: BD showed a higher male predominance in Iraq. Mucosal involvement was the most common clinical feature, followed by ocular involvement and then articular manifestations. The most commonly used treatment was colchicine followed by infliximab and corticosteroids. These findings suggest that early diagnosis and treatment are important to prevent disease complications.

Key words: disease activity, characteristics, Behçet's disease.

Introduction

Behçet's disease (BD) is a multi-systemic disease with a particular geographic distribution [1]. It is characterized by remission and relapse course with highest severity in the first years [2]. Turkey has the highest prevalence worldwide [3]. Iraq has similar prevalence of BD to the nearby countries excluding Turkey [4].

Clinical manifestations of BD are variable [1]. Mucocutaneous manifestations are characteristic and their recognition may permit diagnosis and treatment [5]. Genital ulcers are common [6]. Eye involvement occurs frequently with serious complications typically bilateral non-granulomatous panuveitis [7].

Arthralgia and/or arthritis are often the presenting element before the other manifestations [8]. Vascular symptoms are distinguished by vessel

Corresponding author:

Faiq Isno Gorial Rheumatology Unit College of Medicine University of Baghdad Baghdad, Iraq

E-mail: faiqig@gmail.com

involvement of all types with predominantly venous involvement [9].

Frequency of neuro-Behcet's disease was reported as 3–10% [10, 11]. The ileocecal region is the most involved part of the gastrointestinal tract [12]. This study was designed to assess demographic and clinical characteristics of BD in Iraqi patients.

Material and methods

Study design

An observational descriptive study was performed in the Medical City Complex from July 2017 to January 2018 and informed consent was taken from each participants to conduct the study with ethical approval granted by the ethical committee of the College of Medicine, University of Baghdad.

Participants

Inclusion criteria were: consecutive sample of patients with any age and gender identified as BD according to the International Study Group Criteria 1990 for BD [13]. Criteria for exclusion included overlapping with other inflammatory disease, connective tissue disease, endocrinopathic disease, malignancy, and vasculitis.

Clinical evaluation

Demographic data including age, gender, body mass index (BMI), smoking history, age at onset of disease, disease duration, organ affected, disease

Table I. Baseline characteristics of Behçet's disease patients

Variables (N = 71)	Value
Age [years], mean ± SD	36.0 ±10.8
Gender, n (%):	
Female	26 (36.6)
Male	45 (63.4)
BMI [kg/m²], mean ± SD	28.7 ±6.1
Education level, n (%):	
Primary	36 (50.7)
Secondary	23 (32.4)
University	11 (15.5)
Post-graduate	1 (1.4)
Smokers, n (%)	20 (28.2)
Duration of disease [years] mean ± SD	8.3 ±7.1
Behçet's Disease Current Activity Form, mean ± SD	5.1 ±2.8

N – number, SD – standard deviation.

activity, and medications were recorded. Activity of BD was assessed using the Behçet Disease Current Activity form (BDCAF) [14] which provided a precise history of clinical features that were present throughout the month prior to the measurement date. New clinical characteristics present over the previous 28 days are scored [15]. In 2006, BDCAF was updated and made available at www.behcetdiseasesociety.org/.

Statistical analysis

Quantitative variables were expressed as mean \pm standard deviation and number with percentages for qualitative data; SPSS 20 software package was used to perform the statistical analysis.

Results

Of 71 patients with BD involved in the study, there were 45 males and 26 females with BD. Their mean age was 36.0 \pm 10.8 years. Most of them were males (63.4%), overweight (28.7 \pm 6.1 kg/m²) and had primary education (50.7%). There were 20 (28.2%) smokers among patients. The mean duration of the disease was 5.1 \pm 6.0 years and the mean disease activity score (BDCAF) was 5.1 \pm 2.8 (Table I).

The most common organ involved was mucosal involvement (100%) followed by ocular (87.3%) and then articular (71.8%) involvement. The most common treatment used was colchicine (57.7%), next infliximab (47.9%), and then steroids (45.1%), as illustrated in Table II.

Discussion

BD has significant morbidity and mortality rates with poor quality of life.

The mean age of patients in this study was 36.0 ± 10.8 years. Previous studies from Iraq yielded similar results, a mean age of Iraqi patients of BD was reported to be 37 years [16]. Similarly, the mean age of patients in Turkey was 35.01 years [17].

In the current study, males predominated (45/71, 63.4%) with a ratio of 1.7 : 1, however the ratio is lower than that in the previous findings in Iraq [18]. Similar findings were reported by other studies which observed that there were more males than females. Comparing with other studies, the male-to-female ratio in Saudi Arabia was 3.4, Iraq 3, Turkey 1.03, Iran 2.19, Jordan 2.8, Kuwait 4.9, Korea 0.63, Japan 0.98, the UK 0.36, Sweden 0.67, and the USA 0.38 [18]. In certain Middle Eastern and Mediterranean countries, BD demonstrates male predominance and while female predominance in Japan and Korea [19] The predominance of BD in men in developing countries could be attributable to underdiagnosis of BD in women due to the unwillingness of women of certain ethnic groups to seek medical advice for genital ulcers [18].

The current study showed lower mean age at onset of the disease which was 28.3 ±8.4 years in this study and was similar to the previous study done in Iraq, in which the mean age at onset of the disease was 29.4 years. Other studies also reported comparable low mean age of BD and showed that mean age at onset was in Iran 26.2 years, Saudi Arabia 29.3, Turkey 25.6, Jordan 30.1, Egypt 26.2, Lebanon 26, Tunisia 28.7, the UK (Yorkshire) 24.7, Sweden 33, Ireland 20.8, 31 years in the USA, and 35.7 years in Japan [18].

In the present study, majority of the patients had active disease with the mean BDCAF of 5.1 ± 2.8 , which was in line with another study which reported that most of their patients were active with mean BDCAF of 6.00 ± 3.45 [20].

The most common organ involved was mucosal organ (100%), which was similar to another study which reported that mucosal involvement was the most common organ involved in BD [20], followed by eye involvement (87.3%), which is comparable to ocular involvement in different populations [21]. The higher frequencies of ocular involvement had been also reported in Italy (92%) and Portugal (87%) [18]. Controversial findings were observed in other studies in which eye involvement in BD in Kuwait was 69%, Saudi Arabia 65% and in Egypt 64.6% [5, 22]. However ocular involvement was lower in China (35%) and Korea (55.9%) [22]. The higher level of ocular involvement seen in this study can be explained by some population-specific environmental factors (a lower education level in about half of our enrolled patients) or genetic factors (Human leukocyte antigen (HLA)-B51), which is the strongest genetic predisposition toward BD and this association is more common in the Middle East, Mediterranean, Japan, and some of Western nations other than China and Korea and that may influence the clinical image of the disease [23].

Articular manifestations occurred in 51 (71.8%) patients in the form of arthritis and arthralgia, which is within the detected range of joint involvement in BD (29–76.8%) and was similar to other previous studies [18, 21]. The frequency of articular manifestations was 33% in patients in Iran, 30% in China, 57% in Japan, 38% in Korea, 16% in Turkey, 55% in Tunisia, 53% in Germany, 52% in Morocco, 53% in the USA, and 93% in the UK [18].

Skin involvement was observed in 48 (67.6%), which was comparable to other studies. It was reported in 66% of patients in Iran, 84% in Korea, 69% in China, 87% in Japan, 86% in the UK, 81% in Germany, and 61% in the USA [18].

Myalgia was reported in 16 (22.5%) in the current study which was lower comparing to another

Table II. Organ involvement and medications used in Behçet's disease

Variables	Value
Number	71
Organ involvement, n (%):	
Mucosal	71 (100)
Eye:	62 (87.3)
Bilateral eye	57 (91.9)
Unilateral eye	5 (8.1)
Articular	51 (71.8)
Skin	48 (67.6)
Myalgia	16 (22.5)
Central nervous system	9 (12.7)
Vessels	3 (4.2)
Gastrointestinal tract	1 (1.4)
Medications, n (%):	
Colchicine	41 (57.7)
Infliximab	34 (47.9)
Steroids	32 (45.1)
Azathioprine	19 (26.8)
Adalimumab	3 (4.2)
Cyclosporine	2 (2.8)
Sulphasalazine	2 (2.8)
Methotrexate	2 (2.8)

study that showed that myalgia was reported in 45.9% [20]. This finding may be due to differences in environmental or genetic background that may influence the clinical manifestations of the disease. However, the higher frequency of myalgia in the Italian study could at least be partially biased by a concomitant fibromyalgia in some patients enrolled in the study and a different male-to-female ratio of 0.85: 1.

Neuro-Behçet (NB) was seen in 9 (12.7%) patients in this study, which was similar to other studies which reported that NB was seen in 12% in Tunisia, 11% in Japan, 11% in Germany, 17% in Morocco, 13% in the USA, and 9% of patients in Iran. However, a lower prevalence was reported in China – 6.5%, Korea (4.6%) and Turkey (2.2%) [18].

The prevalence of gastrointestinal tract (GIT) involvement among patients with BD showed a wide variation across geographies, from 1% to 50% [12]. In our patients, gastrointestinal manifestations occurred in 1 (1.4%) patient. Other studies showed that GIT manifestations were numerically higher as seen in 2.8% in Turkey, and

Arch Med Sci Aging 2021 e3

was obviously higher in Iran – 7% of patients, in Korea 7.3%, Japan 15.5%, China 9%, Germany 12%, the USA 29%, and 7% in the UK [18].

Another finding of note was that vascular disease was obviously lower compared to other studies

In the current study, vascular involvement was seen in 3 (4.2%) patients, while in other studies it was observed in up to 40% of BD patients and was within the range of other studies (1.8–40%) [22]. Vascular involvement was seen in 17% in Turkey, 8.9% of cases in Iran, 7.7% in China, 8.9% in Japan, 1.8% in Korea, 13% in Germany, 20% in Morocco, 25% in Tunisia (only venous thrombosis), 10% in the USA, and 32% in the UK [18]. The lower frequency seen in this study may be attributed to the limited sample size and short study length.

The most common treatment in this study was colchicine in 41 (57.7%) patients which was the first line of treatment for mucocutaneous manifestations [23]. A previous study in Saudi Arabia also showed a comparable high frequency (62%) of patients treated with colchicine in BD [21].

The second most used drug in the present study was the anti-TNF monoclonal antibody (infliximab) in 34 (47.9%) patients, the second anti-TNF used in the present study but in lesser frequency (4.2%) was adalimumab. Compared to another study [21] only 8.5% of patients were subjected to anti-TNF therapy, this higher frequency of anti-TNF use in the current study can be explained by the referral of severe and complicated cases that remained unresponsive to conventional therapies to the tertiary care center where the study was done.

Corticosteroid (prednisolone) was used in 32 (45.1%) patients, reflecting the fact that corticosteroids are the first line of treatment during relapses of BD [23]. This finding was lower than in Saudi Arabia (85.15%) and may be explained by differences in disease manifestations and activity.

Azathioprine was used in lower frequency compared to a previous study. It was used in in 19 (26.8%) patients, compared to 62% of patients in another study [21]. This lower frequency of azathioprine use may be explained by severe BD symptoms that were unresponsive to conventional therapies that require the use of anti-TNF. Other medications used in this study were cyclosporine, sulphasalazine, methotrexate, dapsone — each in equal frequency (2.8%), and mycophenolate mofetil, rituximab, anticoagulant with each used in 1 (1.4%) patient.

In conclusion, BD showed a higher male predominance in Iraq with a higher frequency of ocular involvement and lower frequency of vascular disease than the previous studies. Colchicine followed by infliximab then corticosteroids were commonly used treatment.

Acknowledgments

We thank all the participants in the study.

Conflict of interest

The authors declare no conflict of interest.

References

- 1. Yazici H, Seyahi E, Hatemi G, Yazici Y. Behçet syndrome: a contemporary view. Nat Rev Rheumatol 2018; 14: 107-19.
- Kural-Seyahi E, Fresko I, Seyahi N, et al. The long-term mortality and morbidity of Behçet syndrome: a 2-decade outcome survey of 387 patients followed at a dedicated center. Medicine 2003; 82: 60-76.
- Azizlerli G, Köse AA, Sarica R, et al. Prevalence of Behçet's disease in Istanbul, Turkey. Int J Dermatol 2003; 42: 803-6.
- 4. Al-Rawi Z, Al-Rawi ZS, Neda AH. Prevalence of Behçet's disease among Iraqis. Adv Exp Med Biol 2003; 528: 37-41.
- 5. El-Garf A, Abdo M, Alkemary A, Mohamed S. Behçet's disease patterns and subsets in a cohort of Egyptian patients. Egypt Rheumatol 2019; 41: 135-8.
- Scherrer MAR, Rocha VB, Garcia LC. Behçet's disease: review with emphasis on dermatological aspects. An Bras Dermatol 2017; 92: 452-64.
- Mendes D, Correia M, Barbedo M, et al. Behçet's disease: a contemporary review. J Autoimmun 2009; 32: 178-88.
- 8. Saadoun D, Wechsler B. Behçet's disease. Orphanet J Rare Dis 2012; 7: 20.
- 9. Saadoun D, Asli B, Wechsler B, et al. Long-term outcome of arterial lesions in Behçet disease: a series of 101 patients. Medicine 2012; 91: 18-24.
- 10. Akman-Demir G, Serdaroglu P, Tasci B. Clinical patterns of neurological involvement in Behcet's disease: evaluation of 200 patients. The Neuro-Behcet Study Group. Brain 1999; 122: 2171-82.
- 11. Davatchi F, Shahram F, Chams-Davatchi C, et al. Behcet's disease in Iran: analysis of 6500 cases. Int J Rheum Dis 2010; 13: 367-73.
- 12. Yurdakul S, Tüzüner N, Yurdakul I, et al. Gastrointestinal involvement in Behçet's syndrome: a controlled study. Ann Rheum Dis 1996; 55: 208-10.
- 13. International Study Group for Behçet's Disease. Criteria for diagnosis of Behçet's disease. Lancet 1990; 335: 1078-80.
- 14. International Society for Behçet's disease. Behçet's disease current activity form 2006. Available at http://www.behcetdiseasesociety.org/. Accessed in 25 June 2017.
- 15. Baltaci M. A review on disease activity scores in Behçet's disease. Arthritis Res Ther 2003; 5 (Suppl 2): 7.
- 16. Al-Izzi MK, Jabber AS. Fibromyalgia in Iraqi patients with Behcet's disease. J Med Liban 2004; 52: 86-90.
- 17. Melikoglu M, Melikoglu MA. What affects the quality of life in patients with Behçet's disease? Acta Reumatol Port 2014; 39: 46-53.
- 18. Davatchi F, Shahram F, Chams-Davatchi C, et al. Behcet's disease: from East to West. Clin Rheumatol 2010; 29:
- 19. Cho SB, Cho S, Bang D. New insights in the clinical understanding of Behçet's disease. Yonsei Med J 2012; 53: 35-42.
- 20. Fabiani C, Vitale A, Orlando I, et al. Quality of life impairment in Behçet's disease and relationship with disease

- activity: a prospective study. Intern Emerg Med 2017; 12: 947-55.
- 21. El-Najjar AR, Abou El-Soud AM, Amar HA, Al Sayed Diab M. Clinical characteristics and disease activity of Behçet's disease patients in Zagazig, Egypt. Egypt Rheumatol 2015; 37: 191-6.
- 22. Al Zahrani FM, Alsultan B, Alahmari H, et al. Behçet's disease presentations and care outcomes in a tertiary hospital in south-western Saudi Arabia. Int J Rheum Dis 2019; 22: 1466-73.
- Hatemi G, Christensen R, Bang D, et al. 2018 update of the EULAR recommendations for the management of Behçet's syndrome. Ann Rheum Dis 2018; 77: 808-18.

Arch Med Sci Aging 2021 e5