



Comparison of International Study Group Criteria and International Criteria for Behcet's Disease in the Azeri population of Iran

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Article info

Article History:

Received: 23 May 2017

Accepted: 29 June 2017

ePublished: 31 Aug. 2017

Keywords:

Behcet's Disease,
International Study
Group Criteria,
International Criteria for
Behcet's Disease,
Azeri Population

Abstract

Introduction: Behcet's disease (BD) is a chronic systemic inflammatory disease affecting multiple organs and systems such as mucosa, skin, eye, joints and cardiovascular, nervous and gastrointestinal systems. Various criteria are proposed for the assessment of BD. This study was designed to compare performance of two internationally collaborated criteria for diagnosis of BD, namely International Study Group (ISG) criteria and International Criteria for Behcet's Disease (ICBD) in the Azeri population of Iran.

Methods: In a descriptive analytical study, 859 consecutive patients with one of the major clinical manifestations of BD were included. All patients were examined and evaluated by an expert rheumatologist and diagnosis was confirmed clinically. All patients were assessed by ISG and ICBD criteria. Finally, sensitivity, specificity, positive predictive value, negative predictive value, and accuracy of both criteria were calculated.

Results: We included 859 patients in this study. BD was diagnosed in 211 patients. Sensitivity, specificity and accuracy of ISG criteria were 64.9%, 100%, and 91.4%, respectively. Sensitivity, specificity and accuracy of ICBD criteria were 94.7%, 99.6% and 98.5%, respectively.

Conclusion: Based on the finding of the present study, sensitivity and accuracy of ICBD criteria for diagnosis of BD in Azeri population are higher than ISG criteria. So we propose to use ICBD criteria for the evaluation of patients with suspected BD to decrease the rate of missed diagnosis.

Citation: Delnabi L, Gojazadeh M, Hajjalilo M, Ebrahimi A, Khabbazi A. **Comparison of International Study Group Criteria and International Criteria for Behcet's Disease in the Azeri population of Iran.** J Anal Res Clin Med 2017; 5(3): 82-5. Doi: 10.15171/jarcm.2017.016

Introduction

Behcet's disease (BD) is a chronic systemic inflammatory disease affecting multiple organs and systems such as mucosa, skin, eye, joints and cardiovascular, nervous and gastrointestinal systems.¹ The earliest and most common presentation of BD is oral aphthous ulceration. Frequency of oral aphthous ulceration in the Azeri population of Iran is 93.6%.² Other common manifestations of BD are genital ulcers, pseudofolliculitis, erythema nodosum, uveitis and positive pathergy test.² Due to the lack of pathognomonic para-clinical and pathologic findings, diagnosis of BD is

clinical.^{3,4} To date, 17 diagnostic/classification criteria have been developed for diagnosis of BD.

The first set of criteria for diagnosis of BD was described by Curth et al., in 1946 and then other criteria were developed by Hewitt et al. and later, a variety of other criteria from different countries have been established for better diagnosis of BD.^{5,6} But there was no consensus on any of them. To overcome this problem, the International Study Group (ISG) on BD presented the ISG criteria in 1990.^{7,8} ISG criteria had a very high specificity for diagnosis of BD, but the sensitivity of that

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was low.⁹ Davatchi presented Iranian criteria of BD in 1993 to overcome the low sensitivity of the ISG criteria.¹⁰ Finally, the International Criteria for Behcet's disease (ICBD) was developed in 2006.^{11,12} The clinical manifestations of BD differ widely in different ethnic groups.¹ For this reason performance of diagnostic criteria in all of the ethnic groups will not be the same.¹³

This study was designed to compare performance of two internationally collaborated criteria for diagnosis of BD, namely ISG and ICBD in Azeri population.

Methods

In a descriptive analytical study, 859 consecutive patients with one of the major clinical manifestations of BD who referred to the rheumatology clinic of Connective Tissue Diseases Research Center (CTDRC) were included. Study protocol was approved by the ethic committee of Tabriz University of Medical Sciences, Tabriz, Iran. All patients provided written informed consent. All patients were examined and evaluated by an expert rheumatologist and diagnosis was confirmed clinically. Inclusion criteria were having one of the major manifestation of BD i.e. oral aphthous ulceration, genitalia ulcer, uveitis, erythema nodosum, pseudofolliculitis and recurrent arthritis, and being of Azeri ethnicity.

All patients were assessed by ISG and ICBD criteria.^{8,10} With the comparison of final diagnosis with the results of ISG and ICBD criteria, true and false negative and positive patients were defined. Finally, sensitivity, specificity, positive predictive value, negative predictive value, and accuracy of both criteria were calculated.

Statistical analysis was done using SPSS software (version 15, SPSS Inc., Chicago, IL, USA). We calculated sensitivity as the number of patients with BD fulfilled the diagnostic criteria, multiplied by 100, and then divided by the total number of patients.¹⁴ Specificity was calculated as the number of control patients who did not fulfill the diagnostic criteria, multiplied by 100, and then divided by the total

number of controls.¹⁴ Accuracy was calculated as the number of patients with BD who fulfilled the diagnostic criteria, plus number of control patients who did not fulfill the criteria, multiplied by 100, and then divided by the sum of all patients (BD and controls).¹⁴

Data was reported using descriptive methods [frequency, and mean \pm standard deviation (SD)].

Results

We included 859 patients in this study. BD was diagnosed in 211 patients (24.6%) while this diagnosis was ruled out in 648 patients (75.4%). Table 1 shows the final diagnosis in the study patients.

Table 1. Final diagnosis in the patients (n = 859)

Final diagnosis	n (%)
Behcet's disease	211 (24.6)
Rheumatoid arthritis	211 (24.6)
Ankylosing spondylitis	82 (9.5)
Undifferentiated arthritis	49 (5.7)
Other spondyloarthropathies	48 (5.6)
Systemic lupus erythematosus	42 (4.9)
Palindromic rheumatism	33 (3.8)
Recurrent oral aphthous ulceration	23 (2.7)
Psoriatic arthritis	22 (2.6)
Reactive arthritis	18 (2.1)
Familial Mediterranean fever	16 (1.9)
Vasculitis	13 (1.5)
Systemic sclerosis	12 (1.4)
Others	79 (9.1)

Demographic and clinical characteristics of patients with diagnosis of BD are shown in table 2.

Table 2. The demographic and clinical characteristics of patients with diagnosis of BD (Behcet's disease)

Demographic and clinical characteristics of patients (211 patients)	n (%)
Male/female	129/82 (1.57:1)
Age at the disease onset (year) (mean \pm SD)	26.1 \pm 8.4
Oral aphthous ulcer [n (%)]	198 (93.8)
Eye involvement [n (%)]	116(54.9)
Genital aphthous ulcer [n (%)]	102 (48.3)
Positive pathergy test [n (%)]	72 (34.1)
Pseudofolliculitis [n (%)]	54 (26.0)
Erythema nodosum [n (%)]	33 (15.6)
Musculoskeletal [n (%)]	31 (14.6)
Vascular [n (%)]	21 (9.9)
Epididymitis [n (%)]	13 (6.2)

SD: Standard deviation

Table 3. The sensitivity, specificity, positive predictive value, negative predictive value and accuracy ISG (International Study Group) and ICBD (International Criteria for Behcet's Disease) criteria

	ISG criteria (%)	ICBD criteria (%)
Sensitivity	64.9	94.7
Specificity	100	99.6
Positive predictive value	100	99.0
Negative predictive value	89.7	98.3
Accuracy	91.4	98.5

ISG: International Study Group criteria; ICBD: International Criteria for Behcet's Disease

Table 3 shows the sensitivity, specificity, positive predictive value, negative predictive value and accuracy of ISG and ICBD criteria.

Discussion

Up to now, a wide variety of diagnostic criteria has been developed for diagnosis of BD. Currently, the ISG and ICBD criteria are amongst the widely-used diagnostic tools for BD.^{5,15} In this study, 859 patients referred to the rheumatology clinic of CTDR were enrolled and assessed by two rheumatologists. The sensitivity and specificity of ISG criteria to diagnose BD were 64.9% and 100%, respectively while these values were 94.7% and 99.6% for ICBD criteria, respectively. This study finally showed that ICBD criteria had a higher sensitivity and accuracy compared to the ISG criteria in the Azeri population (35.1% vs. 7.1%, respectively).

The findings of our study were in line with previous studies in other ethnic groups. In Davatchi¹⁰ study on 6128 BD patients and 3400 controls, the sensitivity of ISG and ICBD criteria were 78.1% and 98.2%, respectively. Also, they found that the accuracy of these criteria were 85.5% and 97.3%, respectively.¹⁰

The sensitivity and specificity of ISG criteria in China was found to be 65.4% and 87.0%.¹⁶ The sensitivity and specificity of ICBD were 87.0% and 94.1%, respectively.

The sensitivity and specificity of ISG criteria in Germany was 83.7% and 89.5%.¹⁷ The sensitivity and specificity of ICBD were 96.5% and 73.7%, respectively.

In conclusion, based on the findings of the present study and previous literature, both the ISG and ICBD criteria are appropriate diagnostic tools for BD. According to our findings, the sensitivity and accuracy of ICBD were higher than that of ISG criteria.

However, the specificity of ICBD was lower than that of ISG criteria. The observed differences between the findings of the present study with the mentioned literature could be due to the differences in the sample selection and study design. The geographical and genetic differences of the patients also could affect the results of this study.

Conclusion

Sensitivity and accuracy of ICBD criteria for diagnosis of BD in Azeri population are higher than ISG criteria. So we propose to use ICBD criteria for evaluation of patients with suspected BD to decrease the rate of missed diagnosis.

Acknowledgments

We are grateful to Professor Davatchi for helping us in designing of the study.

Authors' Contribution

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Funding

No funding source declared.

Conflict of Interest

Authors have no conflict of interest.

Ethical Approval

Ethic Committee of Tabriz University of

Medical Sciences approved this study (87/23/d/5).

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