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The History and Diagnosis of Behçet's Disease

Müzeyyen Gönül, Arzu Kılıç and Bilgen Gençler

Abstract

Behçet's disease (BD) is a multisystemic vasculitis of unknown aetiology, initially reported by Turkish dermatologist Hulusi Behçet in 1937. Hulusi Behçet presented the disease as a triple symptom complex with recurrent aphthosis, genital ulceration and recurrent hypopyon uveitis. But subsequent studies have shown that it can affect many organs with wide clinical spectrum. It is challenging to make a definite diagnosis because there is no pathognomic laboratory test to diagnose Behçet's disease. The diagnosis is based on variable group of clinical manifestations. Many new diagnostic/classification criteria have been developed through the years. International Study Group (ISG) Criteria and the International Criteria for Behçet's Disease (ICBD) are the most commonly acceptable criteria for the diagnosis of BD. However, due to the broad clinical spectrum of Behçet's disease, there will always be Behçet's patients who do not complete the criteria. Therefore, the experience of the physician and evaluation of the findings with a good clinical anamnesis is of great importance in the diagnosis.

Keywords: Hulusi Behçet, Behçet's disease, International Study Group criteria, International Criteria for Behçet's disease

1. Introduction

Behçet's disease (BD) is a multisystemic vasculitis according to the 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides [1]. The autoimmune process, which is triggered by the environmental and infectious factors on the basis of the genetic factors, is held responsible for the formation of the disease [2]. The definition of the disease dates back to ancient times until the time of Hippocrates [3]. However, for the first time in modern medicine in 1937, Prof. Dr. Hulusi Behçet described it as a separate entity consisting of oral aphthosis, genital ulceration, and iridocyclitis with hypopyon [4].

2. Hulusi Behçet: a life dedicated to dermatology (the person behind the eponym)

Hulusi Behçet was born on 20 February 1889 in Istanbul. Hulusi Behçet was raised by his grandmother after his mother's death when he was young and he had a difficult childhood. He settled in Damascus because of his father's work. He studied at a French school and learned French, Latin, and German. In 1906, he started Kuleli Military Medical School when he was only 16 years old and graduated in 1910.

After his specialization training on skin and venereal diseases in Gulhane Military Hospital, he worked in Edirne Military hospital for 4 years. Afterwards, he went to Europe and worked in Budapest and Berlin for a short time and returned to his country. He worked as a freelance doctor for a while, and then he worked at Haskoy Venereal Diseases Hospital and Guraba Hospital after that in 1933; he became the head of the Department of Skin and Venereal Diseases of Istanbul University and continued this position until 1947 [5, 6].

He is interested in many different areas of dermatology such as syphilis, leishmaniasis, dermatitis *Figus carcia*, parasitosis, and mycosis, but he is mostly known for his studies on Behçet's disease, which is named after him. He presented his research in many national and international meetings and congresses and published 126 articles between 1921 and 1940 [5, 6].

Studies on Behçet's disease began with a patient he first saw in 1924. The patient had recurrent hypopyon uveitis accompanied by ulcerations in the mouth and scrotum, painful nodules on the legs, fever, and joint pain and was variously diagnosed in Istanbul and Vienna and was followed by Hulusi Behçet later on for many more years [5–7].

In 1930, he conducted a study on a female patient with recurrent ocular symptoms and oral and genital lesions and in 1936 on a male patient with oral pemphigus-like wounds, acneiform lesions on the back, scrotal ulcer, night fever, abdominal pain, and ocular symptoms [5–7].

As a result of his research on these three patients, Hulusi Behçet first suggested in 1937 that recurrent oral aphthous lesions, genital ulceration, and recurrent hypopyon uveitis were the symptoms of a single disease as a triple symptom complex [4]. He wrote this view in 1937 in the journal *Dermatologische Wochenschrift* and presented it to his colleagues at a congress in France in the same year. At the same congress, it was suggested that viruses may play a role in the etiology of this condition, especially those of dental origin [5, 6, 8] (**Figure 1**).



Figure 1.
Hulusi Behçet MD (1889–1948).

In 1938, *Dermatologische Wochenschrift* published his thoughts in more detail. In the following years, new case reports came from different countries. While ophthalmologists accepted this new disease, dermatologists believed it to be a symptom of another existing skin disease. However, as a result of the new articles published in different parts of the world, the disease was accepted to be a separate entity. At the international medical congress in Geneva, with the suggestion of Prof. Mischner from Zurich Medical Faculty, this newly diagnosed disease was called “Morbus Behçet” [5, 6].

In 1930, Dr. Benedictos Adamantiades (1875–1962), a Greek ophthalmologist, reported a patient with recurrent hypopyonic irritation accompanied by mucocutaneous lesions and arthritis [9]. In later years, he wrote new case reports, but he did not interpret this as a triple complex; focusing only on the eye, he suggested that hypopyonic iridocyclitis was a separate clinical disease by itself [10–12]. Undoubtedly, although his colleagues from many parts of the world contributed to the acceptance of Behçet's disease as a separate clinical entity, the first person to describe the triple triad and to present it to the world through his articles and presentations is our esteemed professor, doctor Hulusi Behçet.

3. The diagnosis and classification criteria of Behçet's disease

Firstly, although Behçet's disease is accepted as a triple symptom complex with recurrent aphthosis, genital ulceration, and recurrent hypopyon uveitis, subsequent studies have shown that it is a multisystemic chronic inflammatory disease that can affect many organs [1, 13].

There is no pathognomic laboratory test to diagnose Behçet's disease. The disease's wide clinical spectrum, its showing ethnical and geographical differences, and the differences that it shows in the time of onset of symptoms and its courses with different findings in each patient are the factors that make it difficult to diagnose. Therefore, the experience of the physician and evaluation of the findings with a good clinical anamnesis is of great importance in the diagnosis [14].

Although it is a relatively young disease, many different diagnostic/classification criteria have been developed. The first one was created by Curth in 1946 [15]. In 1969, Hewitt et al. presented the diagnostic criteria and revised and reformed them in 1971 [16, 17]. Many new diagnostic criteria have been established in the light of the studies that have been carried out until today. The main ones are Mason and Barnes in 1969, Japan criteria in 1972, Hubault and Hamza in 1974, O'Duffy in 1974, Chen in 1980, Dilsen et al. in 1986, Japan revised criteria in 1988, International Study Group (ISG) in 1990, Iran in 1993, Classification Tree in 1993, Dilsen revised in 2000, Korea in 2003, the International Criteria for Behçet's Disease (ICBD) in 2006, and the revised ICBD in 2014 [18–32].

The Japan Research Committee for Behçet's Disease created the Japanese criteria in 1972. There are four major symptoms according to these criteria: oral aphthosis, genital aphthosis, skin lesions, and ocular findings. In the presence of an ocular lesion, one different major finding is sufficient for the diagnosis. If there are no ocular findings, the presence of the other three major findings is essential for the diagnosis. It is called “complete form” if there are four major findings, and it is called “incomplete form” if there are fewer findings [19]. Japan criteria were revised in 1988. Five minor findings were added to the major findings, and two minor findings were suggested to replace one missing major finding. These minor findings include arthritis/arthralgia, gastrointestinal manifestations, vascular thrombosis, neurological manifestations, and epididymitis [24].

In the Fourth International Conference on Behçet’s Disease, which was held in London in 1985, it was planned to establish a diagnostic criterion with high sensitivity and specificity. Therefore, the International Behçet’s Disease Study Group was established [25, 33]. In 7 countries (France, Iran, Japan, Tunisia, Turkey, the UK, and the USA) and in 12 separate institutions, 912 Behçet patients together with 308 control patients were included in a study and followed up. Twenty-eight patients without oral ulceration were excluded from the study [25]. In the light of the data obtained, “International Behçet’s Disease Study Group Classification Criteria” was established and published in 1990. ISG classification criteria consist of five items. The first criterion is recurrent oral ulceration at least three times a year and is a sine qua non. The other four criteria consist of recurrent genital ulceration, eye lesions (anterior uveitis, posterior uveitis, retinal vasculitis), skin lesions (erythema nodosum, pseudofolliculitis, or papulopustular lesions), and positive pathergy test. In addition to the major criterion oral aphthosis, the existence of two of the four findings is enough for the diagnosis of Behçet’s disease [25, 34] (**Table 1**).

The sensitivity and specificity of ISG classification criteria were found to be 92 and 97%, respectively [25]. Although it is one of the most widely used criteria to date, several concerns about its sensitivity have arisen. In fact, many studies have been performed that measure the performance of diagnostic/classification criteria for Behçet’s disease. Because of the high specificity of ISG, the risk of another disease being misdiagnosed and classified as BD is very low. However, many Behçet’s disease patients go undiagnosed due to its low sensitivity [35]. Another controversial issue is that oral aphthosis is an indispensable finding in ISG, but it has been shown that oral aphthosis may not be in 1–10% of BD patients [36]. Behçet patients with severe, specific symptoms such as vascular involvement can also go without diagnosis with these criteria, and thus the concern of delay in treatment has arisen [37, 38].

In 1993, Iran presented the ISG criteria in an international conference by modifying them to overcome the problem of low sensitivity and low accuracy. According to these criteria, it consisted of five items similar to ISG, but oral aphthosis was not mandatory. Oral aphthosis was given 2 points, and other lesions were given 1 point, and 3 points were sufficient to diagnose BD [26].

In 1993, Iran released a new classification system, using the Classification and Regression Tree method, called the classification tree. The presence of one of the five subgroups in the patient alone is sufficient to diagnose BD. These subgroups are as follows: oral aphthosis + genital aphthosis, oral aphthosis + skin lesions + positive

Recurrent oral ulceration	Minor or major aphthous ulceration or herpetiform ulceration observed by physician or patient (at least three occurrences within a 12-month period)
Plus any two of the following criteria	
Recurrent genital ulceration	Aphthous ulceration or scarring observed by the physician or patient
Eye lesions	Anterior uveitis, posterior uveitis, or cells in vitreous on slit-lamp examination or retinal vasculitis detected by an ophthalmologist
Skin lesions	Erythema nodosum observed by physician or patient, pseudofolliculitis, or papulopustular lesions or acneiform nodules observed by the physician in postadolescent patients (not receiving corticosteroid treatment)
Positive pathergy test	Read by physician at 24–48 hours.

**Findings applicable only in the absence of other clinical explanations.*

Table 1.
International study group (ISG) criteria for the diagnosis of Behçet’s disease.

pathergy test, oral aphthosis + ocular findings, genital aphthosis + ocular findings, and positive pathergy test + ocular findings [27].

In 2000, the original criteria were revised by Dilsen, suggesting that three of the six diagnostic criteria were sufficient to diagnose BD. These criteria are oral aphthosis, genital ulceration, skin lesions (pseudofolliculitis, erythema nodosum), ocular findings, thrombophlebitis, and positive pathergy test [28].

In 2003, Korea established its own diagnostic criterion for BD. This criterion contains six items: oral aphthosis, genital ulceration, skin lesions, ocular findings, positive pathergy test, and gastrointestinal involvement. Three points and above is sufficient to diagnose BD with three points for genital ulceration and one point for other findings [29].

In 2003 the First International Workshop of Behçet's Disease was held in Austria. In this workshop, it was decided that a team of eight countries would prepare a proposal to develop the ISG criteria and to establish new criteria for Behçet's disease if necessary [35].

In the 11th International Conference on Behçet's Disease in Antalya (Turkey) in 2004, the International Team for the Revision of the ISG criteria was created which was composed of clinicians from 27 countries. Among these countries, in the first place comes Turkey, and the rest were Austria, Azerbaijan, China, Egypt, France, Germany, Greece, India, Iran, Iraq, Israel, Italy, Japan, Jordan, Libya, Morocco, Pakistan, Portugal, Russia, Saudi Arabia, Singapore, Spain, Taiwan, Thailand, Tunisia, and the USA.

Between January 2005 and June 2006, 2556 BD and 1163 control patients were included in a study who were selected by expert opinion, rather than a classification system. The International Criteria for Behçet's Disease included a new scoring system for the diagnosis of Behçet's disease. It consisted of six items: genital ulceration and ocular findings with 2 points and oral aphthosis, skin lesions, vascular lesions, and pathergy test with 1 point. A score of 3 or more was sufficient for the diagnosis of BD. In 2010, ICBBD criteria were revised and presented at the 14th International Conference on Behçet's Disease in London. In 2014, these criteria were published in the *Journal of the European Academy of Dermatology and Venereology*. The differences from the classical form were the addition of neurological findings as 1 point and the increase of oral aphthosis to 2 points. Pathergy test was optional but 1 point should be added at its presence. Four points and above was sufficient for the diagnosis of Behçet's disease. The sensitivity and specificity of ICBBD was 94.8 and 90.5%, respectively [30–32] (Table 2).

Sign/symptom points	
Oral aphthosis	2
Genital ulceration	2
Ocular lesions	2
Skin lesions	1
Vascular manifestations	1
Neurological manifestations	1
Positive pathergy test*	1

*1-Pathergy test is optional. However, when it is performed, one extra point may be assigned for a positive result.
2-A patient scoring 4 points or above was classified as having Behçet's disease.

Table 2.
The international criteria for Behçet's disease (ICBD)—point-score system.

4. Differential diagnosis of Behçet's disease

Since Behçet's disease has a broad clinical spectrum, many diseases are included in the differential diagnosis. Although many diagnostic criteria have been developed, it can still be difficult to diagnose Behçet's disease, and it may be necessary to exclude diseases that show similar findings in the differential diagnosis.

4.1 Differential diagnosis of mucocutaneous findings

4.1.1 Oral aphthosis

Recurrent aphthous stomatitis (RAS), viral infections such as herpes simplex virus and Coxsackievirus infections, nutritional deficiencies such as vitamin B and iron, recurrent erythema multiform, oral erosive lichen planus, autoimmune bullous diseases, fixed drug eruption, Stevens-Johnson syndrome, toxic epidermal necrolysis, inflammatory bowel diseases (IBD), Reiter syndrome, systemic lupus erythematosus, celiac disease, mouth and genital ulcers with inflamed cartilage (MAGIC) syndrome, must be Periodic Fever, Aphthous Stomatitis, Pharyngitis, Adenitis (PFAPA) syndrome, hematologic malignancies, and trauma [39–42].

4.1.2 Genital ulceration

Herpes simplex infection; erosive lichen planus; autoimmune bullous dermatoses; sexually transmitted infectious diseases, especially syphilis; and fixed drug eruption [41, 43].

4.1.3 Skin lesions

Papulopustular lesions can be confused with acne and bacterial folliculitis [14, 44].

4.2 Differential diagnosis of systemic findings

The diseases of the similar systemic findings with BD are inflammatory bowel diseases, seronegative arthropathies, sarcoidosis, lupus erythematosus and other systemic vasculitis, multiple sclerosis as neurological involvement, and stroke of the young adult [14, 45].

Differential diagnosis of BD from IBD may be challenging for clinicians because findings of both two diseases include gastrointestinal manifestations, fever, oral aphthosis, pyoderma gangrenosum, erythema nodosum, Sweet syndrome like dermatosis, neutrophilic lobular panniculitis, ocular involvement, arthritis, vasculitis, and thrombotic events. Moreover, the manifestations of intestinal BD are similar to IBD. There are no diagnostic laboratory tests or endoscopic findings for the differential diagnosis of both diseases. Endoscopic findings of Behçet's disease demonstrate single or few, large, round, or oval-shaped ulcerations. Longitudinal ulcers in a discontinuous distribution and cobblestone appearance are endoscopic features of IBD. However, neurologic involvement is not observed in IBD [46–50].

Bowel-associated dermatosis-arthritis syndrome (BADAS) is a recurrent neutrophilic dermatosis which is associated with bowel bypass surgery and gastrointestinal disorders like IBD and diverticulitis [51, 52]. The syndrome should be distinguished from BD, due to the similar findings such as oral aphthosis, vesiculopustular dermatosis, erythema nodosum, neutrophilic lobular panniculitis, fever, vasculitis, and arthritis. The other cutaneous findings of BD and ocular and

neurologic involvement are not expected findings for BADAS. The pathergy test is also negative in the BADAS patients [50].

Although many criteria have been developed for the diagnosis of Behçet's disease, there are still a number of difficulties to establish a definitive diagnosis and for making differential diagnosis. Therefore, newly defined molecular markers and universal criteria with high sensitivity and specificity are needed.

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