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Behcet's Disease in Bahrain, Clinical and HLA Findings

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Objective: To study the clinical features and the HLA findings of Behcet's disease in Bahrain.

Method: A retrospective study of nine patients with Behcet's disease from the Salmaniya Medical Complex who were treated over a 15-year period are analysed.

Results: The male to female ratio was 3.5:1. Oral ulcers were present in all patients, genital ulcers in 88.9%, skin lesions in 66.7%, ocular involvement in 44.4%, arthritis in 44.4%, epididymitis in 57% of the males, and DVT in 22.2%. HLA B5 was positive in 66.7% of the patients.

Conclusion: 66.7% of the cases had the HLA B5 allele. Their clinical manifestations are similar to those reported in the other Gulf countries.

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Behcet's disease (BD) is a multisystem disorder which was first described by Hypocrites as a triad of oral ulcers, genital ulcers, and uveitis[1]. The disease received the name after its description by Turkish dermatologist Hulusi Behcet in 1937[2]. The disease is common among people over the old Silk Road extending from China in the Far East to Turkey[3]. The prevalence varies from 8/10000 in Turkey[4] to 1/10000 population in Hokkaido[5] to 6.4/10000 in Yorkshire, UK[6].

Studies from the neighbouring countries showed a prevalence rate of 2.1/100000 in Kuwait[7] and 1.67/10000 in Iran. We present the first study of Behcet's disease in Bahrain. The aim of this study is to describe the clinical manifestations and HLA type in Bahraini patients suffering from Behcet's disease.

METHODS

All Bahraini patients diagnosed to have Behcet's disease in Salmaniya Medical Complex over a 15-year period from 1986 to 1999 were included in the study.

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The diagnosis of BD was made using the international criteria for diagnosis (classification) of BD proposed by the international study group (ISO) for BD[8] (Table 1).

Table 1. **International criteria for the diagnosis (classification) of Behcet's disease.**

Recurrent oral ulceration	major, minor, or herpetiform observed by physician or patient recurrence at lease thrice in any 12 month period.
PLUS TWO OF:	
Recurrent genital ulceration	aphthous or scarring observed by physician or patient
Eye lesions	anterior uveitis posterior uveitis cells in vitreous on slit lamp examination, OR retinal vasculitis observed by ophthalmologist.
Skin lesions	erythema nodosum observed by physician or patient pseudofolliculitis or papuloputular lesions, OR, acneform nodules observed by physician in post-adolescent patients not on corticosteroid treatment.
Positive pathergy test	- read by physician at 24-48 hours.

The clinical information was obtained from the patients' records and by clinical examination of the patients. HLA studies were performed on all patients using the microlymphotoxicity method of Terasaki[9]. The relative risk was estimated using the method described by Dahl[10].

RESULTS

Table 2 shows the frequency of the clinical features. A total of 9 patients 7 males and 2 females were diagnosed to have BD in the mentioned period. The mean age was 35.88 ± 8.36 years and the M: F ratio was 3.5:1. The disease prevalence is therefore 2.23 per 100,000.

Table 2. **Frequency of the clinical features**

Clinical feature	**Number of patients**	**%**
Recurrent oral ulcers	**9**	**100**

Genital ulcers	**8**	**88.9**
Skin lesions Folliculitis Erythema nodosum Erythema multiforme Dermatitis herpetiformis	**6** 3 2 1 1	**66.7**
Eye lesions Anterior uveitis Posterior uveitis Choreoretinitis	**4** 2 1 1	**44.4**
Arthritis	**4**	**44.4**
DVT	**2**	**22.2**
Epididymitis	**4**	**57% of males**
HLA B5 +ve	**6**	**66.7**

Clinical manifestations

Orogenital Ulcers

Painful Oral ulcers (Figure 1) were present in all the patients. They all had recurrent painful oral ulcers both on the buccal mucosa as well as on the tongue.

Figure 1. Picture showing an oral ulcer in one of our patients with Behcet disease.

Painful Genital ulcers (Figure 2) were found in 88.9% of our patients. In males they were all painful scrotal ulcers. In the female subjects they were both vulvar as well as vaginal ulcers.

Figure 2. Picture showing a genital ulcer in one of our patients with Behcet disease.

Eye lesions

Eye involvement was found in 44.4% of the patients. These manifestations ranged from anterior uveitis to choreoretinitis. The most common was the anterior uveitis.

Skin manifestations occurred in 66.7% of the patients. These manifestations included folliculitis (2 patients), erythema nodosum (1 patient), erythema multiforme (1 patient), and dermatitis herpetiformis (1 patient).

Arthritis was diagnosed in 44.4% of the patients. The arthritis involved mainly the large joints. It was recurrent but with full remission between attacks.

Epidydimitis/orchitis was diagnosed in 57% of the male patients.

HLA tissue typing
HLA B5 was +ve in 66.7% of the patients.

DISCUSSION

Only Bahraini patients are included in our study. It shows that the disease is more frequent in men (men to women ratio 3.5:1). The findings of this male to female ratio as well as the incidence of clinical manifestations are very similar to that found in Saudi Arabia[11]. Considering the small sample we are not able to compare the incidence of manifestations between males and females. Salmaniya Medical Complex, being a referral hospital makes the true incidence in Bahrain difficult to assess. The prevalence of Behcet's disease in the Bahraini population is approximately 2.23/100000.

The manifestations of Behcet's disease in Bahrain are similar to those reported by other studies in the region[10,11,13]. Oral ulcers are present in 100% of the patients, and genital ulcers in 88.9%. The incidence reported in Saudi Arabia[10,11], Kuwait, Jordan, Egypt, Turkey, and Greece[11] is comparable.

It is worth noting that none of our patients had CNS symptoms while the incidence reported has been 44% of the patients in Saudi Arabia, which is a neighboring country, and 12% in Kuwait, another Gulf State[11]. This could be partly explained by the small sample but keeping in mind the reported prevalence of 44% we would expect to have around 4 patients with some form of CNS manifestation of the disease.
There is a significantly higher prevalence of the HLA B5 allele amongst patients with Behcet's disease – 66.7% compared to a prevalence of 23% in the general population in Bahrain[14]. The incidence of HLA B5 in the population was estimated in a total of 425 healthy Bahraini kidney transplant donors. It was found in 23% of the sample. The relative risk for developing Behcet's disease in those who are HLA B5 positive compared to the general population is therefore 6.35.

CONCLUSION

There is a high prevalence of HLA B5 amongst patients with BD in Bahrain 66.7% compared to 23% in the general population. There is a significant relative risk (6.35) amongst HLA B5 positive individuals to develop Behcet's disease

compared to the general population. No CNS manifestations of Behcet's disease in our Bahraini patients are demonstrable.

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