Epidemiology of Behcet's Disease in Thai Patients

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Objective: *To describe symptoms, signs, laboratory findings and to compare sensitivity of several classification criteria in Thai patients who were diagnosed with Behcet's disease.*
Material and Method: *Using medical records from the rheumatology unit, Siriraj hospital, all cases diagnosed with Behcet's disease by our rheumatology staff were identified and reviewed. Demographic data, clinical presentations, and laboratory data were collected. All cases were also reviewed if they had fulfilled any of the following criteria: Iran classification tree, Japanese, Korean, ISG and O'Duffy's criteria. The sensitivity of each criterion was calculated.*
Results: *Twenty three cases were identified during a 24 year interval (1980-2003). Our population had a mean age of 30.83 years. Common clinical presentations were recurrent oral ulcers 100% (23/23), genital ulcers 69.6% (16/23), eye involvement 52.2% (12/23), skin involvement 60.9% (14/23), GI ulcers 8.7% (2/23), epididymitis 4.3% (1/23), vascular lesions 8.7% (2/23), CNS involvement 8.7% (2/23), fever 60.9% (14/23), and positivity of the pathergy test 33.3% (3/9). The sensitivity of criteria used for diagnosis of our patients with Behcet's disease varied widely. We found that the Iran classification tree criteria had the highest sensitivity followed by those from Japan (82.6%), O'Duffy's(73.9%), ISG (52.2%), and Korean (39.1%) criteria respectively.*
Conclusion: *Behcet's disease is heterogeneous in its manifestations and clinical constellation of the disease varies widely among different parts of the world. This is the first epidemiologic study describing Thai Behcet's patients. We also found the Iran classification tree criteria had the highest sensitivity for diagnosis of Thai patients.*

Keywords: *Epidemiology, Behcet's Disease, Thai Patients*

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Behcet's disease is an uncommon chronic immune-mediated disorder. It can be seen worldwide especially along the Silk Road line. The aetiology is unknown, however genetic background may play roles on the disease pathogenesis. Clinical manifestations can be diverse among different ethnicities, which give rise to important roles of genetic predisposition in the disease pathogenesis[1]. Since there is a wide spectrum of the disease's manifestations[2-13], several authorities have established several different criteria to improve sensitivity and specificity for diagnosing the disease for their population[5,14-21]. This is the first epidemiologic study done on Behcet's disease in Thailand. The objective of this study was to describe the clinical features of Behcet's disease in our patients at Siriraj Hospital, Thailand and to determine an agreed diagnosis of Behcet's disease in our patients using the international diagnostic criteria of Behcet's disease.

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Material and Method

This was a retrospective descriptive study of the patients diagnosed as having Behcet's disease by the rheumatology staff at Siriraj Hospital during the period 1980 to 2003. The information on demographic, clinical features and laboratory test results of the patients with diagnosis of Behcet's disease were collected from their medical records. Several diagnostic criteria for Behcet's disease were reviewed through these records. These established criteria were from ISG[14], research committee of Japan[5,16], O' Duffy[18], classifi-

Table 1. Demographic data and clinical features of Siriraj Hospital patients with Behcet's disease compared with other studies

	Present study	Saudi Arabia[6]	Iraq[7]	Kuwait [8]	Jordan[9]	Turkey[10]		UK[12]	USA[13]
Number of patients	23	119	60	29	20	2313		50	164
M:F ratio	14:9	3.4	3	3.1	2.3	1.03		1.27	N/A
Age	30.8 ± 13.9	29.3	N/A	N/A	N/A	N/A	N/A	30	N/A
Oral ulceration (%)	100 (23/23)	100	97	100	100	Male 100	Female 100	92	98
Genital ulceration (%)	69.6 (16/23)	87	83	93	65	85.6	91	82	80
Ocular symptoms (%)	52.2 (12/23)	56	48	69	65	38.1	19.8	80	Uveitis 51 Retinal vasculitis 20
Skin involvement (%)	60.9 (14/23)	57	75	76	35	EN 45.5 papulo pustular 59.5 thrombo phlebitis 17.5	EN 49.8 papulo pustular 48.3 thrombo phlebitis 3.5	68	63
Joint manifestations (%)	34.8 (8/23)	37	48	69	55	11.3	11.8	62	Synovitis 27 arthralgia13
CNS involvement (%)	8.7 (2/23)	44	0	21	N/A	3.3	1.3	14	23
GI ulceration (%)	8.7 (2/23)	4	0	21	5	1.4	1.4	38	8
Vascular system (%)	8.7 (2/23)	Major venous thrombosis 40.5 Arterial occlusion and/or aneurysm 18.9	17	34	20	11.7	2.1	Peripheral venous occlusion 12 central venous occlusion 10	19
Epididymitis (%)	4.3 (1/23)	4.3	22	N/A	N/A	N/A	N/A	N/A	2
Positive Pathergy test (%)	33.3 (3/9)	17.5	71	34	N/A	N/A	N/A	6	29.6

cation tree (Iran)[20] and (suggested preliminary criteria) Korea[5]. Diagnostic criteria for Behcet's disease from the aforementioned groups were applied to the clinical features of our patients.

Results

Behcet's disease was diagnosed in 23 cases. The demographic, clinical features and laboratory results of the patients are shown in Table 1. Sixty-one percent of the patients were males with an average age of 31 years. The most common symptoms were recurrent oral ulceration, genital ulceration and ophthalmic manifestations such as uveitis and retinal vasculitis. The laboratory results of the patients are shown in Table 2. Anaemia, leucocytosis and thrombocytosis were observed in 52.63% (10/19), 47.37% (9/19) and 23.53% (4/17) respectively. The erythrocyte sedimentation rate was elevated (> 25 mm/hr) in 81% (13/16) of the cases. Most of the patients had normal renal and liver function test results. A pathergy test was done in 9 cases, in which 3 cases were positive. Antinuclear antibody (ANA) was positive in 30% of cases (6/20) and all were speckle type and in low titer ($\leq$ 1:640). Five diagnostic criteria of Behcet's disease, which were from the classification tree from Iran, the research committee of Japan, O'Duffy, the ISG and the suggested preliminary criteria from Korea were applied to our patients. The sensitivity of these criteria was 95.7%, 82.6%, 73.9%, 52.2% and 39.1% respectively (Table 3).

Discussion

The most frequent symptom we found in our patients who had Behcet's disease was oral ulcers which occurred in all of the cases resembling the data from several studies done previously from different parts of the world (Table 1). The other common manifestations were genital ulcerations, cutaneous, ocular and musculoskeletal involvement. Central nervous system, gastro-intestinal and vascular involvement, as well as epididymitis, were not as common. Pathergy tests were not done frequently and showed only about 30% positivity. Because of the wide spectrum of the disease and the difference in prevalence of each symptom in patients from different parts of the world, choosing the best fit diagnostic criteria for each patient population is still problematic. Besides the various diagnostic criteria, clinical judgments of individual cases by a physician should be coordinated to help diagnose this uncommon and multifaceted disease since patients may not have full blown manifestations of the disease in its early course. This study has shown that the classification tree by the Iranian criteria was the most sensitive to the diagnosis of our patients at Siriraj Hospital, Thailand. Surprisingly, the criteria from ISG which has been used worldwide only had a sensitivity of 52.2% in our study, which result resembled several studies done previously[3,16,22] (Table 3). These findings may be due to a low frequency of pathergy test performance,

Table 2. Laboratory results

Test	%	Mean $\pm$ SD
Hemoglobin (<12g/dl)	52.63 (10/19)	11.85 $\pm$ 1.64
WBC count (>12 x 10^3 /mm)	47.37 (9/19)	11.3 $\pm$ 3.5
Platelet count (/mm^3)	23.53 (4/17)	406.470 $\pm$ 130.525
(> 500 x 10^3 /mm^3)		
Creatinine (>1.1 mg/dl)	6.25 (1/16)	0.85 $\pm$ 0.33
SGOT (>45mg/dl)	8.33 (1/12)	22.9 $\pm$ 9.9
SGPT (>45mg/dl)	10 (1/10)	31.9 $\pm$ 17.3
Albumin (g/dl)	N/A	3.8 $\pm$ 0.4
Globulin (g/dl)	N/A	4 $\pm$ 0.6
ESR (>25 mm/hr)	81.25 (13/16)	63 $\pm$ 35
ANA positivity*	30 (6/20)	

* Positive ANA : speckled type 6 cases
1:40 - 2 cases
1:160 - 3 cases
1:640 - 1 case

Table 3. Sensitivity of each individual criteria for diagnosis of Behcet's disease

Diagnostic / Classification criteria	Sensitivity		
	Present study	Calamia, Davatchi[13]	Davatchi et al[20]
1) International Study Group for Behcet's Disease[22]	52.2% (12/23)	75.6 ± 6.6	86.2
2) Research committee of Japan[15,16] (including both complete and incomplete)	82.6% (19/23)	84.8 ± 5.5	93.2 (revised criteria)
3) O'Duffy criteria[18]	73.9% (17/23)	88.4 ± 4.9	80.3
4) Classification tree from Iran[20]	95.7% (22/23)	91.5 ± 4.3	97.1
5) Suggested preliminary criteria from Korea[5]	39.1% (9/23)	N/A	N/A

which may enhance the sensitivity if done routinely. The limitations of this study were the small sample size available for analysis, due to the disease rarity and a single centre study. Since the data collection was done retrospectively, the result may be limited and did not meet the rigid descriptive diagnostic conditions to fulfil the criteria. We felt that there is a need to do a multi-centre prospective study to re-evaluate the sensitivity and specificity of the existing criteria and possibly identify new parameters to facilitate an early and accurate diagnosis.

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ระบาดวิทยาของโรค Behcet ในคนไทย

เอมวลี อารมย์ดี, มณีรัตน์ ธนกิติวิรุณ

วัตถุประสงค์: *เพื่อทราบระบาดวิทยาของอาการ อาการแสดง และผลการตรวจทางห้องปฏิบัติการต่าง ๆ ในผู้ป่วยไทยที่เป็นโรค Behcet รวมทั้งเปรียบเทียบความไวของแต่ละเกณฑ์การวินิจฉัยที่ใช้วินิจฉัยในผู้ป่วยกลุ่มนี้ได้แก่เกณฑ์การวินิจฉัยจากประเทศอิหร่าน, ประเทศญี่ปุ่น, ประเทศเกาหลี, International Study Group (ISG) และ O'Duffy criteria*

วัสดุและวิธีการ: *เก็บข้อมูลจากเวชระเบียนผู้ป่วยของสาขาวิชาโรคข้อและรูมาติสซั่ม ภาควิชาอายุรศาสตร์ คณะแพทยศาสตร์ศิริราชพยาบาล ที่ได้รับการวินิจฉัยว่าเป็นโรค Behcet ตั้งแต่ปี พ.ศ. 2523 - พ.ศ. 2546 และข้อมูลที่มีความจำเป็นต่อการวินิจฉัยโรคโดยอาศัยเกณฑ์การวินิจฉัยต่าง ๆ ข้างต้น และคำนวณความไว ของเกณฑ์การวินิจฉัยแต่ละเกณฑ์*

ผลการศึกษา: *มีผู้ป่วยที่ได้รับการวินิจฉัยว่าเป็นโรค Behcet ในช่วงเวลา 24 ปีนี้ มีจำนวน 23 ราย เป็นเพศชาย 14 ราย (60.9%) เพศหญิง 9 ราย (39.1%) อายุเฉลี่ย 30.83 ปี (ค่าระหว่าง 1-72 ปี) โดยอาการที่พบบ่อยที่สุด ได้แก่ recurrent oral ulceration 100% (23/23), genital ulceration 69.6%(16/23), eye involvement 52.2% (12/23), skin involvement 60.9%(14/23), อาการทางข้อ 34.8% (8/23), GI ulceration 8.7% (2/23), epididymitis 4.3 % (1/23), vascular lesions 8.7%(2/23), CNS involvement 8.7%(2/23), และอาการไข้ 60.9% (14/23) มีผู้ป่วยได้รับการตรวจ pathergy test 9 ราย พบผลการตรวจเป็นบวก 3 ราย (33.3%) ส่วนการเปรียบเทียบความไวของแต่ละเกณฑ์การวินิจฉัยโรค Behcet ในผู้ป่วยไทยพบว่า Classification tree จากประเทศอิหร่าน มีความไวสูงสุด คือ 95.7% รองลงมาคือ เกณฑ์การวินิจฉัยจากประเทศญี่ปุ่น 82.6%, O'Duffy criteria 73.9%, International Study Group (ISG) criteria 52.2% และจากประเทศเกาหลี 39.1%ตามลำดับ*

สรุป: *รายงานนี้เป็นรายงานแรกของระบาดวิทยาของผู้ป่วยโรค Behcet ในประเทศไทย และพบว่า Classification tree จากประเทศอิหร่าน เป็น เกณฑ์การวินิจฉัยที่มีความไวสูงสุดสำหรับการวินิจฉัยผู้ป่วยโรค Behcet ในประเทศไทย*