Elsevier Editorial System(tm) for The Egyptian Rheumatologist

Manuscript Draft

Manuscript Number: ERHE-D-18-00094

Title: Behçet's disease patterns and subsets in a cohort of Egyptian patients

Article Type: Full length research article

Keywords: Behcet's disease, disease relapse, disease patterns, disease subsets.

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Abstract: Aim of the work: To study patterns and disease subsets of Behcet's disease (BD) patients admitted to Cairo University Hospitals and to detect whether relapse of the disease will affect the same system every time or not. Patients and methods: A retrospective study involving 82 BD patients admitted to Cairo University Hospitals, from January 2000 to December 2014. They were reviewed to analyze the frequency of different disease manifestations and to find out disease patterns and subsets. Results: 75 men and 7 women were included in the study, with a mean age of 34.2±9.7 years. Their disease duration ranged from 1 to 34 years with a mean of 9.1±6.9 years. Mucocutaneous manifestations were present in 82 patients (100%), ocular manifestations in 53 patients (64.4%), vascular manifestations in 49 patients (59.8%), and neurological manifestations in 9 patients (11%). Most of our patients, 48 (58.5%) had the same one system pattern throughout the disease course, 25 patients (30.5%) had two systems patterns and 9 patients (11%) had three systems patterns. Conclusion: BD usually affects the same system throughout the disease course whether mucocutaneous, vascular, ocular or neurological and the most common pattern is the one system affection. This will help to predict the system that will be affected in each time the patient presents with a disease relapse. Also, it will help in differentiation between disease relapse and any associated other disease minimizing the need for and the cost of investigations. However, future studies on larger number of patients are recommended.

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Cover Letter

Dear Sir,

I am pleased to submit an original research article entitled "**Behçet's disease patterns and subsets in a cohort of Egyptian patients**" for consideration for publication in The Egyptian Rheumatologist Journal. We aimed to study patterns and disease subsets of Behcet's disease (BD) patients admitted to Cairo University Hospitals and to detect whether relapse of the disease will affect the same system every time or not. The study showed that BD usually affects the same system throughout the disease course whether mucocutaneous, vascular, ocular or neurological and the most common pattern is the one system affection. This will help to predict the system that will be affected in each time the patient presents with a disease relapse. Also, it will help in differentiation between disease relapse and any associated other disease, minimizing the need for and the cost of investigations. We believe that this manuscript is appropriate for publication by The Egyptian Rheumatologist Journal. This manuscript has not been published and is not under consideration for publication elsewhere. We have no conflicts of interest to disclose and all authors have approved the manuscript and agree with its submission. The ethics committee has approved the submission.

Thank you for your consideration.

Sincerely,

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Behçet's disease patterns and subsets in a cohort of Egyptian patients

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Behçet's disease patterns and subsets in a cohort of Egyptian patients

Abstract

Aim of the work: To study patterns and disease subsets of Behcet's disease (BD) patients admitted to Cairo University Hospitals and to detect whether relapse of the disease will affect the same system every time or not. **Patients and methods:** A retrospective study involving 82 BD patients admitted to Cairo University Hospitals, from January 2000 to December 2014. They were reviewed to analyze the frequency of different disease manifestations and to find out disease patterns and subsets. **Results:** 75 men and 7 women were included in the study, with a mean age of 34.2±9.7 years. Their disease duration ranged from 1 to 34 years with a mean of 9.1±6.9 years. Mucocutaneous manifestations were present in 82 patients (100%), ocular manifestations in 53 patients (64.4%), vascular manifestations in 49 patients (59.8%), and neurological manifestations in 9 patients (11%). Most of our patients, 48 (58.5%) had the same one system pattern throughout the disease course, 25 patients (30.5%) had two systems patterns and 9 patients (11%) had three systems patterns. **Conclusion:** BD usually affects the same system throughout the disease course whether mucocutaneous, vascular, ocular or neurological and the most common pattern is the one system affection. This will help to predict the system that will be affected in each time the patient presents with a disease relapse. Also, it will help in differentiation between disease relapse and any associated other disease minimizing the need for and the cost of investigations. However, future studies on larger number of patients are recommended.

Keywords: Behcet's disease, disease relapse, disease patterns, disease subsets.

Introduction:

Behcet's disease (BD) is a relapsing multisystem inflammatory disease [1]. Recurrent oral ulcers are the most common clinical manifestations in BD, present in almost all patients, followed in descending order by genital ulcers, erythema nodosum and papulopustular lesions followed by arthritis, uveitis, thrombophlebitis, gastrointestinal and central nervous system involvement [2]. Most BD patients initially manifest with recurrent oral ulcers with a frequency of 97-100% [3] while genital ulcers vary from 62-100% of patients. Pseudofolliculitis, papulopustular eruption, and erythema nodosum are the most common skin manifestations of BD [4].

In BD, renal involvement is not infrequent being mild in most cases [5]. Cardiovascular involvement occurs in 7-46% in the form of pericarditis, myocarditis, endocarditis, valve diseases, endomyocardial fibrosis, and intracardiac thrombosis. [6]. Pulmonary manifestations are reported in 0.3-18% of BD cases [7] and the patterns are quite heterogeneous and include pleural effusion, pulmonary arteritis or venulitis, emphysema, pneumonia, chronic bronchitis, and fibrosis. Pulmonary artery aneurysm is characteristic of BD and carries a poor prognostic sign [8].

Vasculitis in BD may involve the small, medium and large vessels and can affect both the arterial and venous sides of the circulation [9]. Vascular involvement is seen in 15–38% of BD patients [10]. Prevalence of arterial involvement is 1.5–3% worldwide [11], with an outlier report from Saudi Arabia in 18% of cases [12]. Arterial involvement is mainly in the form of aneurysms and occlusions [12]. Arterial aneurysm frequently occurs in the abdominal aorta [13] however, any artery can be involved and the most common complication is rupture, a main cause of mortality [14]. Venous lesions are the most frequent vascular lesions and include deep venous thrombosis (DVT) and large veins thrombosis (superior and inferior vena cava, mesenteric, portal, hepatic, splenic, iliac, subclavian vein and dural sinus thrombosis) [4].

Ocular manifestations occur within 2–4 years of BD onset [15] in about 70% of patients [16]. Uveitis is the commonest form of ocular involvement and the initial ocular inflammation usually starts anterior and unilateral. Later, inflammation usually affects the posterior segment and becomes bilateral, a vision threatening manifestation [15].

Neurological involvement occurs in 5–30% of BD patients [17] and is the initial presentation of the disease in about 5-23% of patients (neuro-Behçet's disease; NBD) [18]. NBD is divided into parenchymal (more common) and non-parenchymal manifestations [19]. The most frequent neurological manifestations are recurrent meningoencephalitis, cerebral venous thrombosis, cranial nerve palsies and epilepsy [17].

Many studies analyzed clinical characteristics of BD but did not focus on the course of the disease and whether relapse will be involving the same system affected as at the first disease relapse or not. The aim of the present study was to assess this point through studying the frequencies of different disease manifestations, patterns and disease subsets.

Patients and methods:

The medical records of 82 BD patients, who were admitted and followed up in the Rheumatology Department of Cairo University Hospitals from January 2000 to December 2014, were retrospectively reviewed to analyze disease patterns and subsets. Diagnosis of BD was made according to the criteria of the International Study Group for Behçet's disease [20]. The study was approved by the local ethics committee of Cairo University scientific review board, and informed consent was obtained from all subjects according to the 2008 Declaration of Helsinki.

Collected data included age, gender, age at onset and first admission, and disease duration. The main 4 clinical presentations (disease subsets) of BD (mucocutaneous, neurological, ocular or vascular) were considered. Laboratory investigations performed included complete blood picture, erythrocyte sedimentation rate (ESR), liver and kidney function tests. The number and causes of admissions were recorded; causes of first and repeated admissions were assessed. Systems patterns at disease onset and during follow up were analyzed. Radiological investigations to confirm the diagnosis e.g. Doppler, MRI brain, slit lamp and fundus examination were considered. Medications received were also reported.

Statistical Analysis: An IBM compatible PC was used to store and analyze the data and to produce graphic presentation of important results. Calculations were done by means of statistically software package namely "SPSS 13" for Windows (SPSS, Chicago, IL, USA). Results were expressed as mean ± S.D. Chi-square test was used for qualitative data. Values were considered statistically significant if p-value is <0.05.

Results:

Demographic data and laboratory investigations of the patients are shown in table 1 and clinical manifestations in table 2. Frequencies of initial clinical manifestations and causes of first admission are shown in table 3. Number of admissions of BD patients ranged from 1 to 7 (3.02±1.7). The frequency of different causes of admissions is shown in table 4. Disease patterns were observed and the results showed that the majority of patients developed single system pattern from the initial till the last flare (table 5).

Almost all patients (n=80) were on steroids at a mean dose of 22.3±12.7 mg/day. The following medications were received by the patients; cyclophosphamide (n=50; 61%), azathioprine (n=44; 53.7%), cyclosporine A (n=14; 17.1%), colchicine (n=12; 14.6%), biologic therapy (n=9 on infliximab and 1 on adalimumab; 12.2%), methotrexate (n=4; 4.9%) and both sulfasalazine and mycophenolate mofitil by 1 patient each. Oral anticoagulants were received by 27 (32.9%). The number of patterns were comparable between those receiving and those not the different medications ($p>0.05$) and between smokers and non-smokers ($p=0.67$). There was a tendency to an increased number of system patterns in females compared to males ($p=0.22$). The study included 2 juvenile-onset BD cases, one of them had one system pattern which was ocular and the other had 3 systems pattern (ocular, vascular and neurological).

Discussion:

Literature focusing on the course of BD and whether the same system is repeatedly involved is scarse. In this study, disease patterns and subsets of BD patients were analyzed and the cause of repeated admissions determined. The current male to female ratio was 10.7:1. Gender distribution in BD patients differs widely depending on their ethnic origin and country of residence [21]. The frequency of male patients diversly ranged from 27% in USA to 87% in Azerbaijan [22]. A higher frequency of male patients in North African and subSaharan patients was found in comparison to those from Europe. High male to female ratio in developing countries may be due to under diagnosis of BD in females due to reluctance of females to seek medical advice for genital ulcers [21].

The 4 main clinical subsets detected included mucocutaneous (100%), ocular (64.6%), vascular (59.8%) and neurological (11%). In this study, mucocutaneous manifestations represented 75.9% of first presentation of the disease. In agreement, the overall frequency of oral ulcers ranges between 95 and 100% of the BD patients [4]. A similar study reported that oral lesions are the first manifestation perceived in 25 to 75% of cases [23]. Egyptian studies also reported similar results with oral ulcers in 100% of BD patients [24-26], 84.2% [27], 80% [28]. Different results were reported by another Egyptian study with oral ulcers in 50% [29].

Genital ulcers were present in 96.3% of patients. In harmony, genital ulcers are the second most common manifestation present in a large percentage of BD patients [23] and have been reported in different frequencies from 62-100% of patients [22]. Egyptian studies also reported similar results with genital ulcers in 96.8% [26], 95.5% % [24], 85% [25], 78.2% [27], 76% [28].

Erythema nodosum was observed in 23.2% of the patients. Similar results were reported in 23% [30], while others found erythema nodosum in 51% of patients [31].

In the present study, ocular involvement occurred in 64.6% of patients. This was in agreement with another study which reported that eye involvement occurs in 30–70% of BD patients [32]. Egyptian studies showed that ocular involvement was seen in 73.7% [27], 38.9% [29], 63.6% [24], 47.6% [26]. Ocular manifestations occurred as initial presentation in 29% of BD patients and this was in agreement with another study [33].

Vascular involvement was observed in 58.9% of patients and represented 16.8% of first disease presentation. This was higher than that reported in other Arab countries. In Saudi Arabia it was 40% [34]**.** In Asian populations, vascular BD was less common than in Arabs; in Singapore it was 5.4% [35], in Hong Kong 11% [36] and in Korea 1.8% [37]. While in Turkey, Japan and Europe, the frequencies of vascular involvement were 17%, 9% and 10-37% respectively [7]**.** An Egyptian study reported vascular lesions in 57.1% [26].

Neurological (parenchymal) involvement was observed in 11% of patients and represented 1.2% of first disease presentation. Similar results were found in Japan (11%), Germany (11%), Tunisia (12%) and US (13%) [38]. A slightly higher percentage of neurological involvement was reported in Egyptian BD patients (34.9%) [26], 30% [25] and 26,3% [27].

Regarding causes of admissions, ocular disease was the commonest and represented 41.4%, followed by vascular (18.9%), neurological (7.2%) and mucocutaneous (3.7%). In the current work, 58.5% of patients had one system pattern involvment while 30.5% had two system patterns and 11% had three system patterns involved throughout disease course. In patients with one system pattern affection, the most frequently affected was ocular followed by vascular then mucocutanous. There was no isolated neurological affection. The most frequent combination was between ocular and vascular manifestations followed by vascular and neurological then ocular and neurological.

In conclusion, this study suggests that BD usually affects the same system throughout the disease course whether mucocutaneous, vascular, ocular or neurological and the most common pattern is the one system affection. This will help to predict the system that will be affected in each time the patient presents with a disease relapse. Also, it will help in differentiation between disease relapse and any associated other disease, minimizing the need for and the cost of investigations. However, future studies on larger number of patients are recommended.

Conflict of interest: None

Funding: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

References:

1. Bonamigo RR, Razera F, Olm GS. Neutrophilic dermatoses: part I. An Bras Dermatol.2011;86:11-25.

2. Mat C, Yurdakul S, Sevim A, Özyazgan Y, Tüzün Y. Behçet's syndrome: Facts and controversies. Clinics in Dermatology 2013;31:352–61

3. Yurdakul S, Yazici H. Behçet's syndrome. Best Pract Res Clin Rheumatol 2008;22:793-809

4. Davatchi F, Shahram F, Chams-Davatchi C, Shams H, Nadji A, Akhlaghi M, et al Behcet's disease: from East to West. Clin Rheumatol.2010;29:823-33

5. Ardalan MR, Sadreddini S, Noshad H, Ebrahimi A, Molaeefard M, Somi MH, et al. Renal involvement in Behcet's disease. Saudi J Kidney Dis Transpl. 2009;20(4):618-22.

6. Demirelli S, Degirmenci H, Inci S, Arisoy A. Cardiac manifestations in Behcet's disease. Intractable Rare Dis Res 2015;4(2):70-75.

7. Uzun O, Akpolat T, Erkan L. Pulmonary vasculitis in behcet disease: a cumulative analysis. Chest 2005;127:2243-53

8. Ceylan N, Bayraktaroglu S, Erturk SM, Savas R, Alper H. Pulmonary and vascular manifestations of Behçet's disease: imaging findings. AJR Am J Roentgenol 2010;194:W158-64

9. Kurokawa MS, Yoshikawa H, Suzuki N. Behçet's disease. Semin Respir Crit Care Med 2004;25:557-68.

10. Azizlerli G, Köse AA, Sarica R, Gül A, Tutkun IT, Kulaç M et al. Prevalence of Behçet's disease in Istanbul, Turkey. Int J Dermatol.2003;42(10):803–6

11. Kwon TW, Park SJ, Kim HK, Yoon HK, Kim GE, Yu B. Surgical treatment result of abdominal aortic aneurysm in Behçet's disease. Eur J Vasc Endovasc Surg 2008;35(2):173–80.

12. Calamia KT, Schirmer M, Melikoglu M. Major vessel involvement in Behcet's disease: an update. Curr Opin Rheumatol 2011;23:24–31

13. Balcioglu O, Ertugay S, Bozkaya H, Parildar M, Posacioglu H. Endovascular repair and adjunctive immunosuppressive therapy of aortic involvement in Behçet's disease. Eur J Vasc Endovasc Surg.2015;50:593–8.

14. Tüzün H, Beşirli K, Sayin A, Vural FS, Hamuryudan V, Hizli N et al. Management of aneurysms in Behçet's syndrome: an analysis of 24 patients. Surgery.1997;121(2):150–6

15. Paovic J, Paovic P, Sredovic V. Behcet's disease: systemic and ocular manifestations. BioMed Res Int 2013;2013:247345.

16. Kaçmaz RO, Kempen JH, Newcomb C, Gangaputra S, Daniel E, Levy-Clarke GA et al. Ocular inflammation in Behçet's disease: incidence of ocular complications and of loss of visual acuity. Am J Ophthalmol. 2008;146(6):828–36.

17. Siva A, Saip S. The spectrum of nervous system involvement in Behçet's syndrome and its differential diagnosis. J Neurol 2009;256:513-29.

18. Dutra LA, Gonçalves CR, Braga-Neto P, Pedroso JL, Gabbai AA, Barsottini OG et al. Atypical manifestations in Brazilian patients with neuro-Behçet's disease. J Neurol 2012;259:1159-65

19. Lannuzel A, Lamaury I, Charpentier D, Caparros-Lefebvre D. Neurological manifestations of Behçet's disease in a Caribbean population: clinical and imaging findings. J Neurol 2002;249:410-8

20. International Study Group for Behçet's Disease. Criteria for diagnosis of Behçet's disease. Lancet 1990;335:1078–80

21. Savey L, Resche-Rigon M, Wechsler B, Comarmond C, Piette J, Cacoub P et al. Ethnicity and association with disease manifestations and mortality in Behçet's disease. Orphanet J Rare Dis. 2014;9(1):42

22. Davatchi F, Shahram F, Chams-Davatchi C, Sadeghi Abdollahi B, Shams H Nadji A et al. Behcet's disease: is there a gender influence on clinical manifestations? Int J Rheum Dis 2912;15:306–14

24. Hassan SZ, Gheith RE, Baz AA, Zeinab M. Afifi ZM. Evaluation of asymptomatic venous disease by Doppler ultrasonography in *Behçet's* disease patients. The Egyptian Rheumatologist,2017;39(3):165-70

25. El Garf AK, Shahin AA, Shawky SA, Azim MA, Effat DA, Abdelrahman SK. Efficacy of infliximab in refractory posterior uveitis in *Behcet's* disease patients. The Egyptian Rheumatologist, 2018;40(2):93-7

26. El Menyawi MM, Raslan HM, Edrees A. Clinical features of Behcet's disease in Egypt. Rheumatol Int. 2009;29(6):641-6

27. El-Najjar AR, Abou El-Soud AM, Amar HA, Diab MA. Clinical characteristics and disease activity of *Behçet's* disease patients in Zagazig, Egypt. The Egyptian Rheumatologist, 2015; 37(4):191-6

28. Amin AM, Nawito ZO. Preclinical coronary endothelial dysfunction in Egyptian *Behçet's* disease patients; Tc-99m sestamibi pharmacological Gated-SPECT, is it a useful screening tool? The Egyptian Rheumatologist, 2013;35(3):159-66

29. Hammad MA, Sharaf DM, El-Shafey AM, Nasr MM. Peripheral nerve involvement in *Behçet's* disease; an electrophysiological study. The Egyptian Rheumatologist, 2014;36(4):195-9

30. Shafaie N, Shahram F, Nadji A. Iran's aspects of Behçet's disease in children. The 1996 survey. In: Behçet's Disease. Hamsa M (Ed.). Tunis, Pub Adhoua 1997;125- 9.

31. Zouboulis CC, Vaiopoulos G, Marcomichelakis G, Palimeris G, Markidou I, Thouas P, et al. Onset signs, clinical course, prognosis, treatment and outcome of adult patients with Adamantiades-Behçet's disease in Greece. Clin Exp Rheumatol 2003;21 (Suppl.30);S19-S26.

32. Rokutanda R, Kishimoto M, Okada M. Update on the diagnosis and management of Behçet's disease. Open Access Rheumatol. 2014;7:1–8.

33. Alpsoy E, Donmez L, Onder M, Gunasti S, Usta A, Karincaoglu Y et al. Clinical features and natural course of Behcet's disease in 661 cases: A multicenter study. Br J Dermatol 2007;157:901-6.

34. al Dalaan AN, al Balaa SR, el Ramahi K, al-Kawi Z, Bohlega S, Bahabri S, et al. Behcet's disease in Saudi Arabia. J Rheumatol 1994;21:658–61

35. Cheng YK, Thong BY, Chng HH. Behcet's disease: experience in a tertiary rheumatology centre in Singapore and a review of the literature. Ann Acad Med Singapore 2004;33:510-14.

36. Mok CC, Cheung TC, Ho CT, Lee KW, Lau CS, Wong RW. Behcet's disease in southern Chinese patients. J Rheumatol 2002;29:1689–93

37. Bang D, Lee J, Lee E, Lee S, Choi J, Kim Y, et al. Epidemiological and clinical survey of Behcet's disease in Korea: the first multicenter study. J Korean Med Sci 2001;16:615–8

38. Wakefield D, Cuniúngham ET Jr, Tugal-Tutkun I, Khairallah M, Ohno S, Zierhut M. Controversies in Behçet disease. Ocul Immunol Inflamm 2012;20:6-11

Table 1: Demographic data and laboratory investigations of the Egyptian Behçet's disease patients

Demographic data in BD patients (n=82)	
Age (years)	34.2±9.7 (18-57)
Sex (M:F)	75:7 (10.7:1).
Smokers	30 (36.6)
Age at onset (years)	28.5±6.7 (11-47)
Age at first admission (years)	32.1±8.7 (17-53)
Disease duration (years)	9.1±7 (1-34)
Hemoglobin (g/dl)	13.2±1.5
Platelets ($x10^3/mm^3$)	263.3±81.5
TLC ($x10^3/mm^3$)	8.5±2.5
ESR ($mm/1^{st}hr$)	27.2±21.1
AST (U/L)	27.1±15.9
ALT (U/L)	33.5±23.6
Creatinine (mg/dl)	0.83±0.17
Urea (mg/dl)	15.4±6.3

BD: Behçet's disease, TLC: Total leucocytic count, ESR: erythrocyte sedimentation rate, AST: aspartate tranaminase, ALT: alanine transaminase. Values presented as mean±SD (range) or n(%).

Table 2: Different clinical subsets of the Egyptian Behçet's disease patients

Clinical Manifestations n (%)	BD patients (n=82)
Mucocutaneous	**82 (100)**
Oral Ulcers	82 (100)
Genital Ulcers	79 (96.3)
Erythema nodosum	19 (23.2)
Acne	12 (14.6)
Folliculitis	4 (4.9)
Papular rash	2 (2.4)
Others (*pustules, vasculitis, furuncles*)	3 (3.7)
Ocular	**53 (64.6)**
Anterior uveitis	24 (29.3)
Posterior uveitis	25 (30.5)
Panuveitis	9 (10.9)
Retinal vasculitis	16 (19.5)
Vitritis	25 (30.5)
Retinal vein occlusion	8 (9.8)
Retinal artery occlusion	7 (8.5)
Vascular	**49 (59.8)**
DVT	27 (33)
Pulmonary embolism	9 (11)
Aneurysm	8 (9.8)
Stroke	7 (8.5)
Sigmoid sinus thrombosis	2 (2.4)
Transverse sinus thrombosis	2 (2.4)
Superior sagittal sinus thrombosis	1 (1.2)
Brain vasculitis	6 (7.3)
Retinal vein occlusion	8 (9.8)
Retinal artery occlusion	7 (8.5)
Neurological (parenchymal)	**9 (11)**
Cranial nerve palsy	5 (6)
Brain encephalitic changes	1 (1.2)
Brain demyelinating foci	1 (1.2)
Focal encephalomalacia	1 (1.2)
Periventricular leukoencephalopathy	1 (1.2)

BD: Behçet's disease, DVT: deep venous thrombosis.

Table 3: Clinical manifestations (subsets) of the Egyptian Behçet's disease patients at disease onset and at first admission

Manifestations n (%)	BD patients (n=82)	
	At onset	At 1st admission
Mucocutaneous	62 (75.9)	2 (2.4)
Ocular	23 (28.9)	40 (48.2)
Vascular	13 (16.8)	37 (45.8)
Neurological(parenchymal)	1 (1.2)	4 (4.8)

BD: Behçet's disease

Table 4: Clinical presentation causing admissions of the Egyptian Behçet's disease patients

Presentation n (%)	BD patients (n=82)
Mucocutaneous	9 (3.6)
Ocular	103 (41.4)
Vascular	47 (18.9)
Neurological (parenchymal)	18 (7.2)
Others (infection or bleeding)	72 (29)
Total admissions	249 (100)

Table 5: Distribution of systems pattern affection in Egyptian Behçet's disease patients

	System pattern n (%)	BD patients (n=82)
One	Ocular	28 (34.1)
	Vascular	16 (19.5)
	Neurological	0 (0)
	Mucocutanous	4 (4.9)
Two	Ocular + vascular	15 (18.3)
	Vascular + neurological	9 (11)
	Ocular + neurological	1 (1.2)
Three	3 systems involved	9 (11)

BD: Behçet's disease