ORIGINAL ARTICLE

Clinical profiling, treatment characteristics and outcome in Behcet's Disease (BD)—A retrospective cohort study from Karnataka Rheumatology Association (KRA)

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Abstract

Background Behcet's disease (BD) has a heterogeneous and unpredictable phenotype that differs in various geographical areas.

Objective To describe the clinical phenotype & outcome of Behcet's disease(BD) from Karnataka, India and compare them with large cohorts from endemic regions.

Methods Databases of practising rheumatologists from Karnataka were reviewed to retrieve clinical characteristics, course of illness, prescribing information and outcome at last follow-up of patients clinically diagnosed as BD. The classification criteria, namely revised International criteria for Behcet's disease (rICBD) and International study group (ISG) criteria were applied. Outcome was defined as complete or partial remission, persistent disease or relapse.

Results We included 72 patients, equal gender distribution and mean age 37.4 ± 12.8 years from 8 rheumatology centres. Commonest presentations were recurrent oral aphthosis 58(80.6%), genital ulcers 36(50%) and ocular manifestations 40(55.6%). Three-quarters [51/72(70.8%)] fulfilled rICBD criteria whereas only half [36/72(50%)] fulfilled ISG criteria. Apart from glucocorticoids [53/72(73.6%)], frequently prescribed therapies were colchicine 39(54.2%) and azathioprine 35(48.6%). Eleven-patients received biologics(anti-TNF-α) and JAK inhibitors to treat severe organ involvement. HLA-B*51 and pathergy tests were positive in 27/45(60%) and 12/34(35.3%) patients respectively. Outcomes were documented in 94.4%(68/72) patients at median follow-up of 24 (12;36) months. Majority [46/68(67.6%)] had complete remission, 17/68(25%) had partial remission, 4/68(5.9%) had persistent while 1/68(1.5%) had relapsing course.

Conclusion Majority of BD patients had orogenital aphthosis and ocular manifestations and an excellent response to treatment.

Key Points

- *In our region, Behçet's Disease primarily manifests with recurrent oral aphthae and ocular involvement, with comparatively lower incidence of severe genital ulcers and neurological involvement than in endemic regions.*
- *Apart from glucocorticoids, colchicine and azathioprine are the most commonly used agents. Biologics and JAK inhibitors are prescribed infrequently, primarily in cases of severe organ involvement.*
- *A significant proportion of patients achieved either complete or partial remission during follow-up, with no observed mortality suggesting a milder disease course and better outcome compared to endemic regions.*
- *Gender, HLA-B*51 status, and pathergy response did not exert any significant influence on the clinical profile or outcome in BD patients in Karnataka.*

Introduction

Behçet's disease (BD) is a chronic inflammatory disorder that shares the features of both autoimmunity and auto-inflammation. It is characterised by vasculitis affecting

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blood vessels of all sizes and can present with either insidious or acute organ threatening manifestations. It is often associated with a relapsing–remitting course leading to significant morbidity and mortality [1].

The clinical features of Behçet's disease vary across different geographic areas of the world and ethnicities [2–4]. However, orogenital ulcers are usually the most frequent and are considered as a defining feature of BD. The prevalence and severity of uveitis and the neurological manifestations are known to differ across countries and also, several different manifestations can accrue with time.[5] This poses many challenges in diagnosis of BD which remains largely clinical, as there are no specific or defining laboratory abnormalities. Numerous classification criteria have been proposed for Behçet's disease; among them, the International Study Group (ISG) criteria and the revised International Criteria of Behçet's Disease (rICBD) are well-known [5]. The latter is currently preferred due to its improved sensitivity as it was specifically modified to accommodate geographical variation of BD phenotype [6].

The pathergy test has been used in the diagnosis of BD and is included in the revised International Criteria of Behcet's Disease (rICBD) as an optional criterion. HLA-B*51 prevalence is also known to be variable across geographies and influences the clinical phenotype [7]. The treatment for BD requires individualisation and varies based on the severity and extent of the organ involvements. The evidence for treatment is largely driven from the observational studies and expert opinion [8]. Broadly, the therapeutic options include glucocorticoids, colchicine and steroid-sparing immunosuppressive agents including biologic therapies as per the disease extent and or severity [9].

Considering these facts, it is of paramount importance to document region specific clinical phenotype of BD and its outcomes. We undertook the present study to elucidate these characteristics in BD patients in Karnataka state, India and to compare them with findings from larger cohorts in endemic regions.

Materials and methods

This multi-centre retrospective cohort study was conducted across eight rheumatology centres in Karnataka, India. Members of the Karnataka chapter of the Indian Rheumatology Association (KRA) were engaged in searching their databases for patients diagnosed with Behçet's Disease (BD). Their current and prior medical records were meticulously reviewed, and the data was systematically collected using a structured case report form. At the last follow up, currently prescribed medications and the outcome were documented. All centres received ethics approval from their respective local ethics committees.

The variables collected encompassed various aspects, including demographic parameters, the time taken for diagnosis, clinical manifestations, and treatments employed. Patients with incomplete documentation were excluded from the analysis to ensure the reliability and completeness of the dataset. Specifically, efforts were made to gather information necessary for applying diagnostic criteria, viz. rICBD and the ISG criteria. Additionally, laboratory parameters were retrieved in order to assess the severity of inflammation and the extent of organ system involvement. The results of pathergy tests, and the HLA-B*51 was recorded when available. Further, relevant imaging and histopathology information were compiled to provide a comprehensive understanding of the initial presentation and the disease course.

The outcome was documented in a similar manner to those outlined by previous studies, such as Fatma Daoud et al.[10] Complete remission was defined as complete resolution of pre-existing symptoms, without the emergence of any new symptoms at follow-up, while maintaining a stable treatment dose. Partial remission indicated incomplete resolution but significant improvement of pre-existing symptoms, with no new symptoms appearing at follow-up. Persistent disease refers to the stability or persistence of pre-existing symptoms without progression towards improvement or worsening, with a potential risk of relapse observed at follow-up. Relapse was defined as the occurrence of new symptoms following a favourable remission period [10]. These definitions were employed to systematically assess and categorise the outcomes of patients during their follow-up visits.

Further, we assessed potential differences in clinical phenotype and outcome based on gender, pathergy response, and HLA-B*51.

Statistical analysis

The Kolmogorov–Smirnov test was used to test normality. Continuous variables were presented with mean and standard deviation or median and Inter quartile range (IQR) as appropriate. Categorical variables were reported with frequency and percentage. Chi square test/fisher exact test was used to find the association between gender and categorical clinical parameters. Independent-t test/ Mann Whitney U test was used to report the significant difference between continuous variables. The results were considered statistically significant at the level of alpha < 0.05 in all analyses. Statistical analysis was performed using the International Business Machines Statistical package for the social and Sciences (IBM SPSS) Statistics 29.0.

Results

We included 72 patients clinically diagnosed as BD from 8 rheumatology centres across Karnataka, India. The mean age at the disease onset was 37.4 ± 12.8 years and the gender distribution was equal(M:F 1:1). The ISG criteria was met by 36(50%) and the rICBD criteria was fulfilled by 51(70.8%) patients, the median score for the latter being 5(IQR 3;5). However, 20/72 (27.7%) did not fulfil neither ISG nor rICBD criteria.

The demographic profile: clinical manifestations and treatment details are represented in Table 1.

Clinical manifestations Recurrent oral aphthae [$N=58(80.6\%)$] was the most frequent symptom, being major aphthae in 30/58(51.7%), and herpetiform ulcers were noted in only one patient. Half of patients [$n=36(50\%)$], also exhibited genital aphthae which were scarring in 18/36(50%). Ocular involvement was the second most frequent manifestation, observed in 40(55.6%) patients. Remarkably, in 11/25(44%) patients with uveitis, it was the presenting manifestation. Predominant lower limb oligoarthritis was the commonest pattern of arthritis [14/23(60.9%)]. Pseudofolliculitis 13/72(18%) and erythema nodosum 7/72(9.7%) were the commonest cutaneous manifestations. In patients with arthritis, 16/23(69.5%) had concurrent either skin pustules or pseudofolliculitis. Vascular involvement was observed in 10(14.3%) patients, gastrointestinal and neurologic involvement was uncommon and there were no patients with either cardiopulmonary or renal involvement.

Special investigations The Pathergy test was positive in 13/34(38.2%) patients. Histopathology from the skin lesions and mucosal ulcers was available in 7 patients, demonstrating neutrophilic dermatosis. Imaging studies were available for 6 patients, revealing either arterial or venous thrombosis. HLA-B*51 was tested in 45/72 patients out of which 27/45 (62.5%) tested positive.

Treatment details During the course of the illness, 53(73.6%) patients received glucocorticoids (GC) at doses exceeding 7.5 mg/day. Azathioprine [35(48.6%)] and methotrexate [29(40.2%)] were the most commonly used steroid-sparing agents. Additionally, biologics like tumour necrosis factor inhibitors(TNFi) 5(7.1%), and tofacitinib 6(8.6%) were prescribed infrequently, primarily in those with severe organ involvement, particularly refractory uveitis. Colchicine was prescribed in 37(52.9%) patients, largely in those with mucocutaneous manifestations.

Outcomes We assessed outcomes in 68/72(94.4%) patients over a median follow-up period of 24 (IQR 12; 36) months. Among them, 46(67.6%) achieved complete remission, 17(25.4%) were in partial remission. Four (5.9%) patients experienced persistent symptoms; 2 had recurrent orogenital ulcers and another 2 had recurrent uveitis. At the last follow-up, less than half of the patients 35/72(48.6%) were on treatment with glucocorticoids, majority [23/35(65.7%)] being on ≤ 7.5 mg/day.

Neither the gender, the pathergy response, nor the HLA-B*51 demonstrated any influence on the clinical profile or the outcomes in patients with BD.(supplementary Table 1).

Discussion

We present a retrospective cohort study of Behçet's Disease (BD) from Karnataka, India, focusing on the clinical profile, treatment, and outcomes on follow-up with rheumatologists. While BD has been extensively studied in the endemic regions, reports from India are limited. We also compare our findings with few of the largest cohorts from endemic regions to highlight differences and similarities. (Table 2).

Gender differences in BD have been discussed in several reports [11]. Male preponderance was reported in the largest series involving more than seven thousand patients from Iran and several other studies [10, 12–14]. The organ involvement in males is more frequent and severe, particularly, the ocular and vascular manifestations as compared to females in two largest series from Iran and Turkey [12, 15]. However, we did not find any gender disparities in either the clinical profile or disease severity similar to the other Indian study by Pande et.al [14]. The higher proportion of patients meeting the revised ICBD criteria as compared to the ISG criteria in the present study emphasises the already established differences in their diagnostic sensitivity. However, almost a third of our patients were unable to fulfil either of the criteria. Low sensitivity of the criteria poses a challenge when clinicians observe said constellation of manifestations, but are unable to consolidate the diagnosis especially, in the absence of diagnostic or confirmatory tests.

The prevalence of oral ulcers in the current study was comparable to that reported by Davatchi et al. and Gurler et al. [12, 15] However, we observed a lower incidence and less severe genital ulcers in contrast to findings from endemic regions, which have noted a higher prevalence of large, deep perforating genital ulcers [15]. In a study from India by Singhal et al., authors reported a higher frequency of mucocutaneous manifestations (oral ulcers 100%, genital ulcers 100%, skin lesions 93%), possibly due to the inclusion of only patients meeting ISG criteria, wherein the presence of oral ulcers is a mandatory criterion [16]. In the present study, uveitis was noted in approximately two-thirds of BD patients with ocular involvement. A recent study from Tunisia reported a somewhat similar prevalence(77.1%),

Table 1 Clinico-demographic profile of Behcet's disease

Patient characteristics	(N=72)(%)
Mean age at disease onset (years ± SD)	37.4 ± 12.8
ISG criteria	36/72(50%)
Revised ICBD	51/72 (70.8%)
Median Total revised ICBD (IQR)	5(3;5)
Duration behcet's symptoms from onset (in months)(Median IQR)	48 (13;108)
Duration till the establishment of diagnosis (in months)(Median IQR)	18 (3.3; 60)
Duration of follow-up (in months) (Median IQR)	24 (12; 36)
Clinical manifestations	
Oral Aphthosis	58 (80.6%)
Genital Aphthosis	36 (50%)
Ocular manifestations	**40/72(55.6%)**
Uveitis	25/40(62.5%)
Retinal vasculitis /vein occlusion	12/40(30%)
Keratitis	2/40(5%)
Episcleritis	2/40(5%)
Optic neuritis	2/40(5%)
Cutaneous manifestations	**34/72(47.2%)**
Pseudofolliculitis	13/34 (38.2%)
Acne	8/34(23.5%)
Erythema Nodosum	7/34 (20.6%)
Papulopustular lesions	6/34 (17.6%)
Pyoderma gangrenosum	4/34 (11.8%)
Musculoskeletal manifestations	**23/72(31.9%)**
Oligoarthritis	14/23(60.9%)
Polyarthritis	5/23(21.7%)
Arthralgias	3/23(13%)
Sacroiliitis	1/23(4.3%)
Vascular manifestations	**9/72(12.5%)**
Arterial thrombosis	4/9 (44.4%)
Venous thrombosis	1/9 (11.1%)
Superficial thrombophlebitis	2/9 (22.2%)
Arteritis /aortitis	2/9 (22.2%)
Neurological manifestations	**5/72 (6.9%)**
Cerebral Venous thrombosis	2/5(40%)
Headache	2/5(40%)
Demyelination	1/5(20%)
G.I vasculitis	1/72 (1.4%)
Treatment details	
Treatment (anytime)	N=72
Glucocorticoids	
≥7.5 mg	53(73.6%)
<7.5 mg	19(26.4%)
Colchicine	39(54.2%)
Methotrexate	29(40.2%)
Azathioprine	35(48.6%)
Cyclophosphamide	5 (6.9%)
TNF inhibitors (Etanercept)	5 (6.9%)
Tofacitinib	6 (8.3%)
Thalidomide	8 (11.1%)
Treatment (last FU)	
Glucocorticoids (any dose)	35(48.6%)

Table 1 (continued)

Patient characteristics	(N = 72)(%)
≥ 7.5 mg	12(34.3%)
< 7.5 mg	23(65.7%)
Colchicine	24(33.3%)
Methotrexate	25(34.7%)
Azathioprine	23(31.9%)
TNF inhibitors	1(1.4%)
Thalidomide	3(4.1%)

Results are presented as %, Mean ± SD or median with inter quartile range(IQR) as appropriate

ISG: International study group; *ICBD*: International criteria for behcet's disease; *IQR*:Interquartile range; *GI*: Gastrointestinal; *FU*: followup

Table 2 Comparison of characteristics with other BD cohorts

Variable	Present study	Pande et.al [14]	Gurler et.al [15]	Davatchi et.al [12]
Study population (N)	72	58	2147	6075
Ethnicity/region	South India	North India	Turkey	Iran
Mean Age at disease onset (years ± SD)	37.7 ± 12	33.1	38.3	28.3
Gender (M:F) ratio	1:1	1.8:1	1.03:1	1.3:1
Delay in diagnosis of BD(months)	18 (3;60)	55	79	69.6
rICBD criteria fulfilled	51(70.8%)	-	-	(96.9%)
ISG criteria	36(50%)	42(72.4%)	2147(100%)	(77.9%)
Oral ulcers	58(80.6%)	52(89.7%)	1858(86.5%)	5921 (97.7%)
Genital ulcers	36(50%%)	45(77.6%)	1892(88.2%)	3989(65.7%)
Cutaneous	34(47.2%)	37(63.8%)	1162(54.2%)	3927(64.6%)
Ophthalmic	40(55.6%)	25(43%)	621(28.9%)	3528(58.1%)
Vascular	9(12.5%)	20(34.5%)	361(16.8%)	553(9.1%)
CNS/ PNS	5(6.9%)	1(1.7%)	46 (2.2%)	642(10.6%)
Musculoskeletal	23(31.9%)	41(70.7%)	343(16%)	2393(39.4%)
Gastrointestinal	1(1.4%)	3(5.2%)	62(2.8%)	426(7%)
Pulmonary	-	1(1.7%)	20(1%)	59(1%)
Renal	-	2(3.4%)	2(0.1%)	96(1.58%)
CVS	-	-	3(0.1%)	37(0.6%)
Commonest IS used	Azathioprine (48.6%)	Chlorambucil (9%)	-	-
Colchicine	37(54.2%)	47(81%)	-	-
Biologics/JAK inhibitors	11(15.7%)	-	-	-
HLA-B*51	27/45 (60%)	10(30%)	-	888/1815(48.9%
Positive pathergy test	12/34 (35.3%)	5/58(8.6%)	1220/2147 (56.8%)	3122/5969(52.3)

SD: standard deviation; *M*: Males; *F*: Females; *BD*: Behcet's Disease; *ICBD*: International criteria for behcet's disease; *ISG*: International study group; *CNS*: Central nervous system; *PNS*: Peripheral nervous system; *IS*: Immunosuppression; *JAK*: Janus Kinase Inhibitors; *HLA*: Human Leukocyte Antigen

but they noted poor visual outcomes and a higher incidence of blindness (8.3%)[10]. In a study conducted by the ophthalmology group from India, a high prevalence of ocular involvement(92.4%); particularly panuveitis was reported, about 1/3rd of whom had poor visual outcomes[13]. The notable differences in prevalence and outcomes between this study and ours is likely due to the referral bias pertaining to the predominant organ system affected and none of our patients had documented blindness or poor visual outcomes suggesting a milder clinical course. This could also be due to early use of immunosuppressive therapy including TNFi for ocular BD.

Despite BD affecting all types of blood vessels, venous thrombosis is more prevalent than arterial disease. In this

context, superficial thrombophlebitis frequently relapses and presence of aneurysms can be life threatening and challenging to treat [17]. In the present study, the frequency of vascular involvement was similar to that reported by Davatchi et al. and Gurler et al.[12, 15] Pande et.al from India have previously reported a higher prevalence of vascular involvement 20(34.5%), superficial thrombophlebitis being the most frequent, in contrast to the present study [14].

The predominant articular pattern observed in BD is asymmetric large joint mono or oligoarticular of lower limbs, which typically is non-destructive and self-limiting [12, 18]. Previous Indian study reported an equal prevalence of both oligoarticular(34.5%) and polyarticular(32.8%) disease, the majority were symmetrical (86.8%) [14]. In contrast, in the present study, around 60% of patients with arthritis exhibited an asymmetrical oligoarticular pattern. Notably, more than half of our patients with arthritis presented with skin pustules or pseudofolliculitis. This association of skin lesions with arthritis has been well documented in prospective studies from Iran and Turkey [18, 19]. In the present study, one patient was diagnosed with sacroiliitis despite testing negative for HLA-B*27. The prevalence of sacroiliitis in Behçet's disease (BD) typically ranges from 5 to 46% [20, 21]. This association can be attributed to the overlapping features of spondyloarthritis in BD, which is categorised under the umbrella term "MHC-Iopathy" [22].

HLA-B*51 is a split molecule of the HLA-B*5 antigen. In the healthy Indian population, the prevalence of HLA-B*5 antigen is around 30%, similar to the general population in endemic countries [7, 23, 24]. About 60% of patients in the present study were tested positive for HLA-B*51 which falls within the global prevalence range of 50–72% in patients with BD from endemic countries [25]. However, a lower prevalence (30%) of HLA-B*51 in BD patients was reported from North India [13, 14]. Furthermore, a systematic review and meta-analyses have indicated that HLA-B*51 is associated with male predominance, mucocutaneous manifestations, and a lower likelihood of gastrointestinal involvement [26]. We did not notice any significant clinical association with the presence of HLA-B51 in the present study.

The positive pathergy test, prevalent in Japan and Mediterranean countries such as Turkey (60%-90%), is rarely positive in Caucasians [25]. It boasts high specificity (98.4%) and mostly occurs during active disease [27]. Given its geographical variations and lower sensitivity, the pathergy test is primarily employed to diagnose Behçet's disease only in atypical or doubtful cases. We report a prevalence of positive pathergy tests in about one-thirds, similar to findings reported by Davatchi et al. and Singhal et.al [12, 16].

In the current study, 73.6%of patients received glucocorticoids, and azathioprine was the most common immunosuppressant (48.6%) prescribed, similar to other studies[28]. In another Indian study by Pande et al.(1995), chlorambucil was the most common immunosuppressant, and colchicine was used in the majority (84%) of patients. This suggests the evolution of treatment practices for BD overtime. In addition, none of our patients were on anticoagulation for vascular manifestations, in contrast to a study by Fatma Daoud et.al from Tunisia, wherein all patients with venous/arterial thrombosis received anticoagulation.

The majority of patients (93%) in our cohort achieved either complete or partial remission, with no observed mortality during a median follow-up of 2 years. Singhal et.al from India also reported 26/29(89.6%) had complete remission during 1 year follow-up [16]. In contrast, a monocentric study from Tunisia reported complete or partial remission in only about half (48.5%) over a median follow-up of 5.7 years, relapsing disease in 42.3%, predominantly vasculo-BD, and a mortality rate of 1.5% [10]. These differences suggest that Behçet's Disease may exhibit a milder clinical course and better outcomes in India compared to the endemic areas.

However, the clinical profile and the outcomes presented in this study are among patients referred to the rheumatologists and may not be a true representation of all BD patients. Further, this being a retrospective study with its inherent drawbacks, we are limited by a rather short duration of follow up and cannot comment on relapses and flares over a longer period of time. We believe that several BD patients may have remained undiagnosed as our patient population was largely concentrated in the metro area. Despite these limitations, this is the largest real-world description of clinical manifestations of Behcet's from the rheumatologists in Southern India providing insight into varied phenotype and response to therapy.

Conclusion

This is the first real-world data that provides novel insights into the clinical profile, treatment patterns, and outcomes of Behçet's Disease in the Indian population, specifically within Karnataka, India. Oro-genital ulcers followed by uveitis were identified as the most prevalent manifestations. Neurological and gastrointestinal manifestations were less commonly observed. Achieving classification through either rICBD or ISG criteria was challenging in this population. More than two-thirds of patients achieved clinical remission during follow-up. Notably, the gender, HLA-B51 status, and the pathergy response did not exert any influence on either the clinical profile or outcomes. These results significantly contribute to the knowledge of Behçet's Disease phenotype and treatment response in our region.

Declarations

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