

## Evaluating Behçet's Disease Activity Pattern in the Context of Major Organ Involvement

### Abstract

**Objective.** Behçet's disease (BD) is a multi-system and chronic-relapsing vasculitis. Variation in disease presentation and treatment may have a differential effect on disease activity overtime. Despite this, the time varying nature of disease activity has not been well studied in the past. Thus, the aim of this study is to examine the patterns in BD activity over time and the effect of baseline organ involvement and type of medications on disease course.

**Methods.** This is a retrospective cohort study of BD patients who fulfilled the International Criteria for Behçet's disease (ICBD) and has been actively followed in King Saud University Medical City since 2015 to present. A standardized data collected data included demographics, medical histories, clinical manifestations, assessment of disease outcomes and treatment regimens at time of diagnosis and for five consecutive follow-up periods. An overall disease activity index was measured using Behçet's Disease Current Activity Form (BDCAF). A generalized estimated equation model was used to examine factors influencing changes in BDCAF score overtime.

**Results.** A total of 134 BD patients were identified, 77% were male and young. The overall mean BDCAF score was 3.97. The most common presenting complaints were oral aphthosis (55%), ocular lesions (39%) and genital ulcers (25%). Patients who presented with active oral ulcers, were more likely to have high disease activity across time ( $p=0.002$ ) compared to those who presented with other organ manifestations. Conversely, patients who have presented with neurological, fever and/or other vascular complaints, were associated with lower mean disease activity across time ( $p<0.050$ ). The mean BDCAF score was significantly higher among glucocorticoid (GC) users ( $p=0.021$ ). The addition of TNF- $\alpha$  inhibitors (anti-TNF) to GC resulted in a significantly lower mean disease activity on average, ( $p=0.027$ ). There was no similar effect seen with other disease modifying rheumatic drugs (DMARDs) among patients.

**Conclusion.** Generally, BD activity starts high at baseline, then trends downward overtime. This pattern is influenced by baseline organ involvement. Thus, major organ involvement (Vascular and central nervous system) favors this course. A combination of anti-TNF and GC exert the strongest effect on reducing disease activity overtime compared to either one alone, or with other conventional DMARDs (cDMARDs).

**Key words:** Behçet's disease, ICBD, BDCAF, conventional DMARDs, anti-TNF

### Key messages:

- The higher the disease activity at baseline the more severe the disease will be.
- Baseline organ involvement seems to predict the course of disease with oral ulcers being associated with higher disease activity while central nervous system and vascular involvement predict lower disease activity across time.
- GC and anti-TNF have a synergistic effect on reducing disease activity compared to cDMARDs solely.