Karnataka Rheumatology Association (KRA) Bechet's Collaborative Study

Dr Vijay K Rao
Dr Ramya Sri Kodali
Dr Vineeta Shobha

Rao VK, Kodali RS, Patil A, Chebbi P, Subramanian R, Kumar S, Singh YP, Chandrashekara S, Shobha V. Clinical profiling, treatment characteristics and outcome in Behcet's Disease (BD)-A retrospective cohort study from Karnataka Rheumatology Association (KRA). Clin Rheumatol. 2024 Aug 3. doi: 10.1007/s10067-024-07089-x.. PMID: 39096360.

This is a multi-center ambi-directional cohort study conducted across 8 centers in Karnataka. Behcet's is a rare, chronic, and multi-systemic auto inflammatory disease. In India, Behçet's disease is considered rare and its exact prevalence is not well-documented. Most cases reported in India are in small series from dermatologists or Ophthalmologists. Diagnosis is purely clinical.

We, Rheumatologists at KRA wanted to study a rare disease and chose this disease to study by consensus. The study was discussed by the principal investigator Dr Vineeta Shobha in our state chapter academic meeting. A nodal investigating unit of the principal investigator with team of research assistants, statistician and research coordinators prepared the case report proforma based on the parameters from the available validated Bechet's diagnostic criteria. Ethical clearance was obtained at the principal investigator's center and with other independent ethics committees. Periodic updates by the principal investigator in our state chapter WhatsApp group about the study numbers from each center and the closing date accelerated the participation. The final data set was compiled and analyzed at the principal investigator's unit.

This being a referral dependent disease, not all patients met the criteria. Therefore, it was decided to compare the classification criteria in addition to description of clinical phenotype and outcomes. We were happy to report satisfactory therapeutic outcomes.

Challenges: Majority of rheumatologists who contributed data for this study are in the private practice with busy outpatient clinics. There is a lack of research team in private practice, and lesser enthusiasm for clinical research are the possible hurdles for further research options. Better PayScale for teaching institution doctors, research team for private practice and having a research wing of state chapters are the possible solutions to enhance collaborative research.

Way Forwards: Maintenance of rare disease registry, mandatory tracking of patients especially of those with uncommon manifestation of such rare disease especially wrt outcomes can help design dedicated diagnostic and therapeutic protocols and provide guidance towards prognosis and long-term outcomes.

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.