ASS 2022 – OP 06

Demographic and Clinical Characteristics of Behçet's disease among patients attending Rheumatology clinic at National hospital of Sri Lanka - A Cohort Study

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Introduction

Behçet's disease (BD) is a rare vasculitic disorder characterized by recurrent oral aphthae and several systemic manifestations including genital ulcers, skin lesions, ocular disease, vascular disease, arthritis, gastrointestinal disease, or neurologic disease. Demographic and clinical characteristics of Behçet's disease among Sri Lankan patients are not widely available.

Methods

All patients who were diagnosed with BD using the 2006 international criteria for Behçet's disease (ICBD) from January 2018 to August 2022 at Rheumatology clinic – General, National Hospital of Sri Lanka are included in this study.

Results

Total of 9 patients were diagnosed with BD during the study period, of which 5(55.6%) were males and 4(44.4%) were females. Mean age of symptom onset was 22.7 years(+/-10.0). Mean age of BD diagnosis was 29.3 years(+/-12.7). Youngest patient was diagnosed at the age of 16 years and oldest patient at 52 years. Mean time between onset of symptoms and diagnosis was 6.89 years(+/-7.3). While all (100%) patients had recurrent mouth ulcers at some point of the illness, they occurred as the initial symptom in 88.9%. The remainder (11.1%) had genital ulcers as the initial symptom. Seven patients (77.8%) had genital ulcers at some time during the course of illness. Joint symptoms, primarily large joint monoarthritis present in 44.4%. Skin involvement was seen in one-third of patients. Pustular skin lesions, erythema nodosum like lesions, and panniculitis were the skin manifestations encountered.

Eye involvement was seen in 44.4%, with bilateral panuveitis being the manifestation in half of the cases. One patient had bilateral optic neuritis. Vascular involvement was seen in one-third of cases. Brachial artery occlusion was seen in one patient and thrombosis involving the right atrium and inferior vena cava was seen in another patient. Large/medium vessel vasculitis was observed in one patient. None of the patients in this cohort had gastrointestinal involvement.

During the diagnostic workup, 22.2% had a positive pathergy test. Only 55.6% underwent HLA B51 testing of which 40% had positive results. TNF alpha inhibitors (i.e.- Infliximab or adalimumab) were required for treatment in 66.6% of cases. These patients had eye or severe vascular involvement.

Conclusion

Awareness of common manifestations and characteristics of investigation profile of Behçet's disease among Sri Lankan patients will help to identify this rare form of multisystem disease preventing delay in diagnosis. Oral ulcers should not be ignored and other clinical features of Behçet's should be actively sought.