Abstract

Background: Thrombotic microangiopathies (TMAs) are heterogeneous disorders characterized by widespread occlusive microvascular disease, causing thrombocytopenia, microangiopathic haemolytic anaemia (MAHA) and end-organ dysfunction due to ADAMTS-13 deficiency. TMAs are broadly classified into primary and secondary, with HIV being an increasingly common secondary aetiology, specifically in HIV endemic regions, like South Africa.

Methods: 106 records of patients diagnosed with TMA and treated with plasma exchange (PLEx) from 2010-2019, were assessed at presentation for clinical features of fever, renal and neurological involvement; laboratory parameters including haemoglobin, platelet count, and creatinine levels; treatment required such as blood product support, renal replacement therapy and use of corticosteroids; and patient outcomes such as recurrence, mortality and creatinine at termination of PLEx. Further information was gleaned from the NHLS laboratory database to assess patients' response to PLEx.

Results: HIV was the most common aetiology, accounting for 82.2% of TMA. More than two-thirds of the cohort were females of Black-African ethnicity, and the median age at presentation was 36 years. Patients with HIV-associated TMA had lower platelet counts, and were older at presentation compared to HIV negative patients. The diagnosis of TMA was typically associated with advanced HIV disease (median CD4 count of 147 cells/mm^3). Irrespective of the TMA aetiology, this study found that a median of 10 PLEx sessions was required to induce remission of TMA, with the crude mortality found to be 16.8% in this cohort.

Conclusion: HIV-associated TMA remains an important cause of secondary TMA in South Africa. It presents more often in Black-African females and in those with advanced stages of HIV. Compared to HIV negative individuals, HIV-infected patients present with lower

platelet counts potentially signifying a more severe form of disease. PLEx remains an integral component in the management of TMA and is crucial to improving survival. While this modality is crucial to eliminating the pathogenic ULvWF multimers and auto-antibodies, the important role of combination antiretroviral therapy (cART) in maintaining remission and preventing relapse of TMA should not be underestimated.