Presentation and response to plasma exchange of thrombotic thrombocytopaenic purpura in a community with high HIV prevalence

Yusuf Moola[1], Zaheera Cassimjee[1,2], Chandni Dayal[1,2], Sheetal Chiba[1,2], Adekunle Ajayi[1,2], Malcolm Davies[1,2]

[1]University of the Witwatersrand, Johannesburg, South Africa
[2]Division of Nephrology at Helen Joseph Hospital, Johannesburg, South Africa

Abstract

Background: Thrombotic thrombocytopaenia purpura (TTP) is a rare disorder which if untreated carries a high mortality. HIV is an important cause of TTP in the local context but remains poorly characterized; increased mortality risk has been suggested by some. Previous South African literature has suggested acceptable rates of remission with plasma infusion therapy, but the efficacy of plasma exchange (PEX) remains largely unreported. We therefore sought to compare the presentation and response to plasma exchange (PEX) between HIV-positive and HIV-negative patients diagnosed with TTP at our institution.

Methods: A retrospective review of 83 patients receiving PEX for TTP between 1/1/2010 – 31/12/2019 was undertaken. Demographics and presenting parameters were compared between HIV-associated TTP and other aetiologies using Mann Whitney U and Kruskal Wallis ANOVA testing, as appropriate. The effect of aetiology and presenting parameters on PEX duration was modelled using Cox proportional hazards; effect of these variables on mortality and residual renal dysfunction in survivors was analysed using stepwise multivariate regression.

Results**:** Uncontrolled HIV infection was the commonest cause of TTP in this series. Thrombocytopaenia was more severe and neurological deficit more frequent in HIV-associated TTP; renal dysfunction was milder in this group. Aetiology did not influence mortality risk. Aetiological category and presenting parameters did not predict PEX duration. Residual renal dysfunction was less frequent in survivors of HIV-associated TTP than in HIV-negative patients.

Conclusion: HIV is an important cause of TTP in the local context. Haematological and neurological involvement are more severe in HIV-associated TTP. Acceptable survival rates are achievable with PEX even in advanced HIV infection; renal sequalae are less common in this group.