



Cascade Filtration: a successful treatment of Thrombotic Thrombocytopenic Purpura



Abbruzzese L.¹, Caputo M.¹, Mele A.², Vitali R.¹, Carbone F.¹, Pavone V.²

¹Transfusion Medicine Service, "Card. G. Panico" Hospital (Tricase, Italy)

²Hematology Unit, "Card. G. Panico" Hospital (Tricase, Italy)

Background

Thrombotic thrombocytopenic purpura (TTP) is a rare, often fatal blood disorder characterized by thrombocytopenia and microangiopathic haemolytic anaemia. Most patients have immune mediated TTP, that is associated with a deficiency of plasma ADAMTS13 enzyme activity, which is responsible for maintaining normal distribution of von Willebrand factor (vWF) multimers. The mainstay of treatment is therapeutic plasma exchange (TPE) to rapidly remove the causative antibody to von Willebrand factor cleaving metalloprotease (ADAMTS13).

Case Presentation

We describe a case of a 60-year-old male presented to our hospital for stroke suspicion associated to dysesthesia and strength deficits. The encephalic CT scan revealed a small area of hypodensity in the cortico-subcortical area of the right middle frontal gyrus, followed by an MRI scan with a positive result for small ischemic lesions in the acute phase. On the basis of hematochemical results and morphological examination of the peripheral venous blood smear showing the presence of schistocytes, TTP was suspected and confirmed by severe enzymatic deficiency of ADAMTS 13 (3%). Treatment with daily TPE in combination with steroids was started, with improvement of blood crashes and regression of neurological symptoms.

Results

A total of 8 sessions of plasmapheresis were performed and the patient was discharged. Two days after he returned to emergency room following strength deficit with paraesthesia and disturbance in speech production. 3 sessions of TPE were performed, but in the fourth session he developed an allergic reaction, which was thought to be due to caplacizumab, so a fifth session of TPE was done with a new allergic reaction. So, it was decided to continue therapy with Cascade Filtration (CF), that was realized using the automatic system Plasmapher/Apherlungs (Aferetica, Italy), directly connected with Amicus separator system (Fresenius Kabi, Italy). Plasma obtained by centrifugation is conveyed into a fractionator filter, Evaflux 3A20 (Aferetica, Italy), which allows a semi-selective plasma purification and removes autoantibodies responsible for the pathophysiology. After purification the plasma returns to the patient, avoiding plasma donor infusion and consequent possible adverse reactions. A total of 8 sessions of CF were performed with no adverse reactions. Treatment with CF continued on an outpatient basis for another 4 sessions. In figure 1 is showed the PLT Trend.

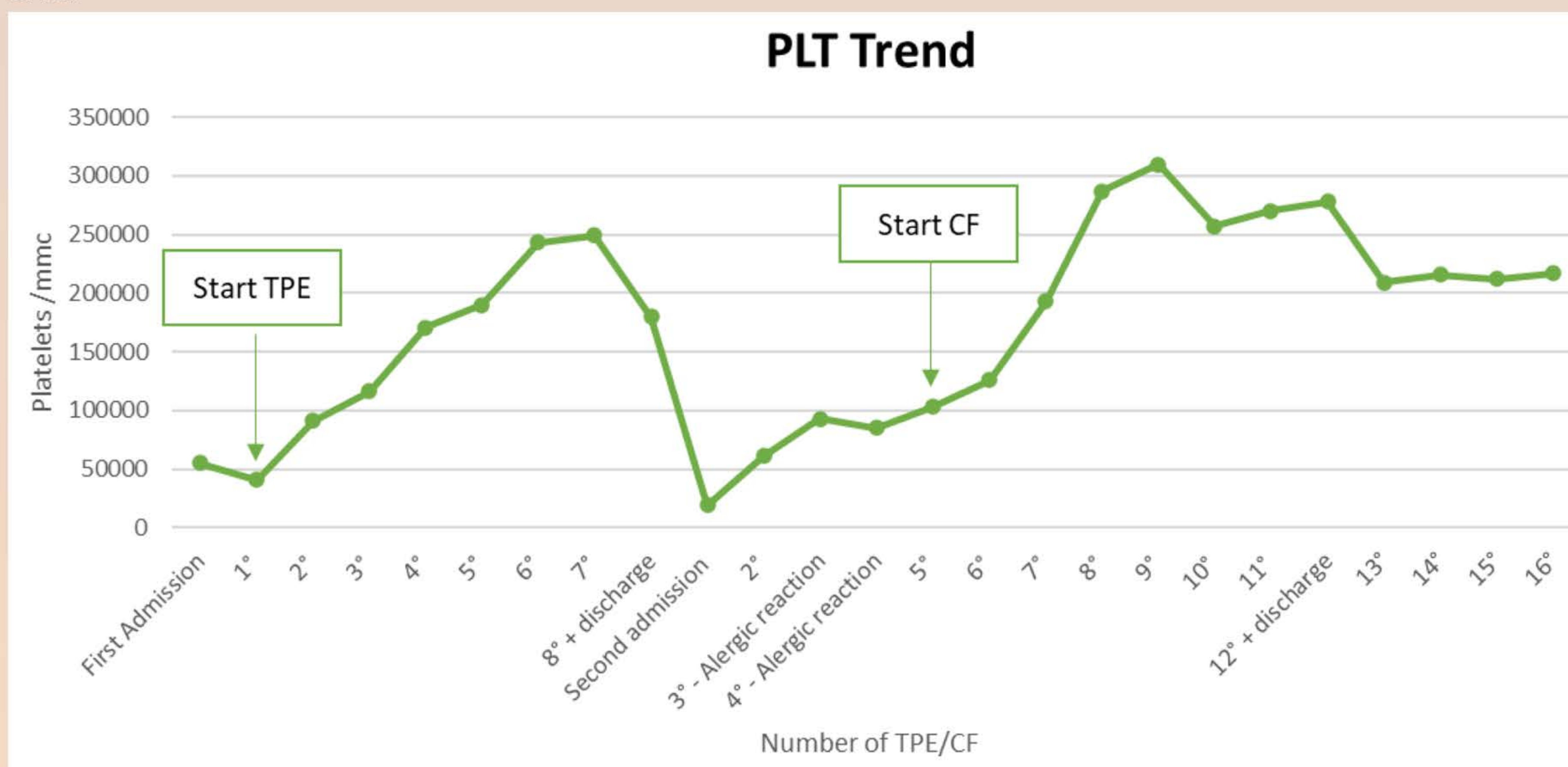


Figure 1: PLT Trend

Conclusions

CF has proven to be effective as TPE, then we conclude that it can be considered as a safe and valid alternative therapy to TPE for the treatment of TTP.