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Successful management of a patient with recurrent thrombotic thrombocytopenic purpura (TTP) with rituximab

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Introduction: TTP is a rare life threatening thrombotic microangiopathy due to severe deficiency of the von Willebrand factor cleaving serine metalloprotease, ADAMTS13. Relapse occurs in 20 – 50 % of patients. Plasma exchange and rituximab are the treatment modalities considered in the management of relapsed disease.

Case Report: We report a case of a 26-year-old man who had two relapses over a period of 18 months. At the initial presentation he was admitted to Colombo South Teaching Hospital with a history of fever, multiple purpuric patches and red coloured urine. The laboratory investigations revealed a haemoglobin of 7.2 g/dL, WBC $5.4 \times 10^9/L$ and a platelet count of $58,000/mm^3$. His blood film showed many fragments confirming microangiopathichaemolysis. His coagulation profile was normal. The serum creatinine was 140 mol/L, reticulocyte count was 3% and direct antiglobulin test (DAT) was negative with an indirect hyperbilirubinaemia. Lactate dehydrogenase (LDH) was markedly elevated with a value of 3293 u/L and D-dimer was negative. His ANA was positive. According to the clinical and laboratory parameters he was diagnosed as a case of TTP and managed with cryosupernatant infusions and prednisolone 1 mg/kg/d which was tailed off over several months. First relapse occurred nine months later with a transient ischaemic attack (TIA) and the second relapse occurred 15 months later again with a TIA. On both occasions he was off prednisolone for several weeks. Three months following the second relapse he was treated with four doses of rituximab $375 \text{ mg}/m^2$ weekly and maintained at a low dose of prednisolone for another year. Currently he is off prednisolone for 10 months and in remission without any clinical or laboratory evidence of relapse.

Discussion: We highlight that even though the recurrent TTP is a rare condition it can be successfully managed with immunomodulatory therapy such as rituximab.

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Polycythaemia vera in a patient with congenital heart disease: A rare coincidence

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Introduction: Polycythaemia vera is a clonal myeloproliferative disorder, characterized by autonomous proliferation of red cells due to an acquired somatic mutation of a marrow stem cell. JAK 2 -V617F mutation is present in 97% of patients. Polycythaemia can occur due to secondary causes as well. Tissue hypoxia due to congenital heart disease is one of the well established causes of secondary polycythaemia. Both polycythaemia vera and secondary polycythaemia give similar clinical manifestations due to hyperviscosity.

Case Report: Here we report a case of a 53 year old male who presented with a minor stroke, very high haematocrit and an existing diagnosis of congenital heart disease; an uncorrected atrial septal defect. Two possibilities were considered, one being shunt reversal with Eisenmenger's syndrome causing secondary erythrocytosis and the other being Polycythaemia vera. Together with a positive JAK2