

Abstract

Thrombotic Thrombocytopenic Purpura (TTP) is characterized by thrombocytopenia and micro-angiopathic haemolytic anaemia (MAHA) on blood picture although fever, acute kidney injury and neurological manifestations also occur in many patients. It is a condition carrying significant mortality. It can be inherited or acquired usually following drugs, infections, pregnancy, organ transplantation and tumours. It can also occur secondary to connective tissue disorders. TTP following Systemic Lupus Erythematosus (SLE) is considered to be a rare occurrence in literature.

We present a case of a 35 year old lady who had been investigated for inflammatory arthritis 6 years ago, now presenting with shortness of breath for one week, bilateral leg swelling for 4 months as well as pain and swelling of bilateral knees, shoulders, elbows for one week. She also had a painless oral ulcer, alopecia and frothy urine. She was found to have a moderate left side pleural effusion, small pericardial effusion and lupus nephritis type IV on renal biopsy.

She was managed as SLE with Lupus Nephritis with IV Methyl Prednisolone 1g daily for 3 days which was converted to oral prednisone 1mg/kg/daily. A regime of IV Cyclophosphamide 500mg monthly for 6 months was started. Enalapril 2.5mg 12 hourly was started to reduce proteinuria but later with-held due to acute kidney injury.

While in the ward she developed headache with mild confusion, acute kidney injury, fever, worsening anaemia and thrombocytopenia. These were suggestive of TTP which were supported by blood picture revealing Microangiopathic Haemolytic Anaemia (MAHA) features. She underwent 6 cycles of plasma exchange under the guidance of Transfusion Medicine consultant and recovered from TTP.