

THROMBOTIC THROMBOCYTOPENIC PURPURA COEXISTING WITH SYSTEMIC LUPUS ERYTHEMATOSUS: CASE REPORT FROM A RESOURCE POOR SETTING

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ABSTRACT

Thrombotic thrombocytopenic purpura (TTP) co-existing with systemic lupus erythematosus (SLE) is a rare clinical entity with few cases reported in the literature. Good response has been reported with early initiation of plasmapheresis. We report a case of TTP with SLE in a young lady who presented with features of both diseases and highlight the challenges of management in a resource poor setting.

Keywords: *TTP, SLE, challenges, management, resource poor setting*

BACKGROUND

Thrombotic thrombocytopenic purpura (TTP) is a rare blood disorder characterized by clotting in small blood vessels resulting in low platelet count caused by severely reduced activity of vWF-cleaving protease ADAMST13. Systemic Lupus Erythematosus (SLE) is an autoimmune disease characterized by multisystem involvement and production of array of auto antibodies targeted at different body tissues and organs.

SLE can present with thrombotic microangiopathy, differentiating SLE from TTP can be challenging. An association between SLE and TTP has been reported in the literature, with most of the cases occurring after the diagnosis of SLE, but can also occur concurrently. We report a case of TTP occurring in SLE patient and the challenges of management in resource poor setting.

CASE REPORT

A 21yr old female undergraduate presented with 2 months history of recurrent fever, joint pains, oral ulcers, discoid rashes, alopecia and malar rash. Fever

became continuous 5 days before presentation with associated, cough, pleurisy, irrational talk, aggression and marked decline in urine output.

Examination revealed a drowsy but conscious young lady, febrile (T=40.1°C), tachycardic 132bpm, tachypnoeic with respiratory rate of 28cpm, chest expansion was equal, percussion notes were resonant and breath sounds were vesicular. BP was 100/60 mmHg, she had no signs of meningeal irritation, pupils were 3mm bilaterally and reactive, no focal neurological deficit and other systems were unremarkable.

Laboratory investigations showed proteinuria 3+, haematuria 3+, urine and blood cultures were negative as well as HBV, HCB-Ab and HIV serologies. CXR was not remarkable and complete blood count revealed anaemia PCV 12.3%, thrombocytopenia $71 \times 10^9/L$, leucopaenia WBC $2.3 \times 10^9/L$, LDH 8692 u/L, Prothrombin time 14 seconds, APPT 35 seconds, blood smear showed fragmented red cells, INR 0.9, ANA 1/1000 RU/mL granular, anti-RNP 1/1000 RU/mL, anti- DNA (Eia) 8.0iu/ml, Brain CT – features of bifrontal cerebral infarcts and mild cerebral atrophy. Serum urea was 40.5 mg/dL, creatinine 8.6 mg/dL, potassium 4.5mmol/L and bicarbonate 18mmol/L.

An assessment of community acquired pneumonia on background SLE with lupus nephritis and cerebritis was made.

She was commenced on intravenous ceftriazone, azithromycin, IV fluids and antipyretics. Intravenous methyl prednisolone was started at a dose of 500 mg daily for 3 days and thereafter oral prednisolone at a dose of 1 mg/kg body weight to taper. Two days later she developed generalized tonic clonic seizures and lapsed into unconsciousness. Renal

function deteriorated and anuria ensued. Blood was transfused; dialysis initiated and was transferred to the intensive care unit. Renal parameters improved, seizure was controlled with carbamazepine, but clinical condition did not change. Diagnosis of TTP was made based on the pentad of fever, renal impairment, microangiopathic haemolytic anaemia, thrombocytopenia and CNS features.

She was immediately referred to a private facility 700 kilometers away for plasmapheresis. She had 5 sessions of plasma exchanges with 2 doses of rituximab. She regained full consciousness thereafter with complete resolution of all symptoms and normalization of haematological and renal parameters. She is now on mycophenolate mofetil and prednisolone, stable and has completed her university study awaiting national service.

DISCUSSION

Thrombotic microangiopathies are microvascular occlusive disorders that are a common pathway of different pathological processes. Such disorders are characterized by systemic or intrarenal aggregation of platelets and/or fibrin, mechanical injury to red blood cells, and thrombocytopenia [1]. They may be hereditary or acquired. TTP is a complex syndrome and most cases characterized by diminished activity of vWF-cleaving protease, ADAMTS-13. [1, 2] TTP is characterized by the pentad of; fever, thrombocytopenia, microangiopathic haemolytic anaemia, renal failure and neurologic manifestations [1, 2]. The diagnosis of TTP in this patient was made based on this pentad.

In SLE, clinical features are highly variable, ranging from skin and joint involvement to organ-threatening and life-threatening disease [1]. Our patient presented with most of the clinical features of SLE which was later confirmed by laboratory findings.

SLE rarely presents with thrombotic microangiopathy. Our patient met the criteria for the diagnosis of SLE according to the American College of Rheumatology (ACR) criteria for systemic lupus and at the same time fulfilled the pentad for the diagnosis of TTP [1-3].

SLE and TTP occurring simultaneously in an individual patient have rarely been reported in the literature. There are only a few cases reported in the literature to date [1-4].

Our patient had an excellent outcome, similar observations were made by Daniel *et al* [4] and George *et al* [5], in which excellent responses were reported with initiation of plasmapheresis, but another report suggested that TTP in patients with SLE was associated with a higher mortality than idiopathic TTP, even with optimal treatment [3].

The index case presented was diagnosed with SLE and TTP simultaneously and has responded to standard treatment with plasmapheresis, rituximab, mycophenolate and prednisolone.

Major challenges in the care of this patient were significant financial and logistical problems as she had to be transported 700 kilometers to a private facility for this treatment. Inability to assay enzyme activity was a limitation in this setting. In a country where the minimum wage is just about 50 US dollar a month, and cost of care is out of pocket, the strain on the finances of the patient or their family is much.

Although rare, SLE co-existing with TTP can present to our resource constrained practice and good outcome should be expected with standard care.

REFERENCES

1. Muhammed A S, Alamgir S, Fnu Z, Muhammad R, Aisha A, Shante H *et al*. SLE presenting as refractory TTP, A diagnostic and management challenge; *Am J Case Rep* 2016;17: 752- 757
2. Caramaschi P, Riccetti M, Pasini A, Savarin T, Biasi D and Todeschini G. Systemic lupus Erythematosus and thrombotic thrombocytopenic purpura. Report of three cases and review of the literature. *Lupus* 1998; 7(1): 37- 41
3. Hamasaki K, Mimura T, Kanda H, Kubo K, Setoguchi K, Satoh T *et al*. Systemic lupus erythematosus and thrombotic thrombocytopenic purpura: a case report and literature review. *Clin Rheumatol* 2003; 22: 355-358
4. Daniel B and Geoffrey B. Lupus associated TTP- like microangiopathic haemolytic anemia; a case report and literature review. *World J Nephrol* 2015; 6; 4(5): 528-531.
5. George P, Das J, Pawar B and Kakkar N. Thrombotic thrombocytopenic purpura and systemic lupus erythematosus: successful management of a rare presentation. *Indian J Crit Care Med* 2008; 12: 128-131