Case Report
Dengue fever, a rare cause of thrombotic thrombocytopenic purpura: a case report

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Introduction
Dengue fever (DF) is a vector borne viral disease caused by dengue virus which belongs to the family Flaviviridae and genus Flavivirus [1]. As the incidence of DF is increasing, unusual manifestations are on the rise, although they are under reported due to lack of awareness [2]. Thrombotic thrombocytopenic purpura (TTP) is a rare disorder. It is important to make the diagnosis because the mortality is very high if untreated and can be reduced with the early initiation of plasma exchange [3]. The presence of TTP in dengue viral infection is rare but has been reported in the literature [4,5].

Case presentation
A 53-year-old female presented with an acute febrile illness for two days. She had a medical history of bronchial asthma. On examination, she was afebrile with an altered sensorium and had a Glasgow coma scale (GCS) of 13 along with gum bleeding. Her blood pressure was 80/50 mmHg and she was tachycardic. Respiratory examination was unremarkable. Abdominal examination revealed right hypochondriacal tenderness. No focal neurological signs were found. From the 3rd day of her illness, she started to develop high fever spikes (101°F – 105°F) and gradual deterioration of sensorium (GCS of 8/15).

Her random blood sugar was 133 mg/dL. Complete blood count showed a platelet count of $8 \times 10^3/\mu L$ (150-400× $10^3/\mu L$) and haemoglobin of 10.6g/dL (11–16g/dL). Reticulocyte count was 6.02% and the Direct Coombs test was negative. Her coagulation profile was normal with prothrombin time (PT) of 15.8 seconds, activated partial thromboplastin time (APTT) of 28 seconds and thrombin time (TT) of 18.2 seconds. The thromboelastogram showed normal clotting factors in both the extrinsic and intrinsic pathway and a high fibrinogen reserve without hyperfibrinolysis. Serum creatinine was elevated 165µmol/L (74-110µmol/L) with normal serum electrolytes. Computed tomography of the brain was unremarkable. Ultrasonography of abdomen and chest revealed free fluid in Morrison's pouch. Blood film showed microangiopathic haemolytic
anaemia (MAHA) with some schistocytes and thrombocytopenia. Lactate dehydrogenase (LDH) level was 1988 U/L (140-280). Clinical picture with MAHA, high LDH and normal coagulation profile directed us towards the diagnosis of TTP.

Dengue IgM and IgG serology became positive. Hepatitis B, hepatitis C and retroviral serology and the microscopic agglutination test (MAT) for leptospirosis were negative. Apart from treatment according to the national management guidelines for dengue, she underwent twelve cycles of plasma exchange. Patient was discharged on prednisolone. She had a very protracted course with a prolonged hospital stay of about a month. However, she made a complete recovery. Clinic follow-up was arranged.

**Discussion**

TTP is a rare and life-threatening pathophysiology which occurs as a result of congenital or acquired deficiency ADAMTS13, a metalloprotease that is important to cleave large Von Willibrand factor multimers. This leads to formation of disseminated microvascular thrombi and organ ischaemia. It is characterized by a pentad of thrombocytopenia, microangiopathic haemolytic anaemia (MAHA), fluctuating neurological signs, renal impairment and fever [5]. Our patient manifested the classic pentad, which is observed in less than 10% of patients with TTP [3,5]. The ADAMTS13 assay helps to confirm the diagnosis but it is not essential. We could not perform the ADAMTS13 assay in our patient as it is not available in Sri Lanka. TTP should be treated as a medical emergency [3]. Prompt initiation of plasma exchange saves life.

Dengue haemorrhagic fever (DHF) and dengue shock syndrome (DSS) are severe and life-threatening complications of DF [7]. Our patient presented in a compensated shock state with altered sensorium, gum bleeding and evidence of plasma leakage. Being an endemic country for dengue, the immediate diagnosis was DSS and resuscitation with crystalloid (0.9% NaCl) was done according to the national dengue management guidelines till she was hemodynamically stable [6]. Later she was diagnosed to have serologically confirmed dengue complicated by TTP. She showed a good clinical response to plasma exchange therapy.

**Conclusion**

This case highlights dengue fever as a rare cause of TTP. Early diagnosis and immediate initiation of appropriate treatment carries a good prognosis.

**References**

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