An Atypical Case of Thrombotic Thrombocytopenic Purpura

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Citation

Abstract
Thrombotic thrombocytopenic purpura (TTP) is a syndrome characterised by a classical triad of microangiopathic haemolytic anemia, thrombocytopenia and neurological symptoms. But also, it could be presented with a pentaed by the addition of fever and renal failure (1, 2). There are several reports concerning the aetiology of TTP, which is not always identified (3-5). We report an unusual case of TTP in a patient who has neurological findings and impaired hepatic functions with minimal thrombocytopenia.

CASE REPORT
A 55- year- old female patient admitted to the emergency department with deterioration of her consciousness. She had a history of cerebrovascular accident on right cerebral hemisphere and treated in a neurological clinic of another hospital one week ago. She suffered from hypertension and diabetes mellitus for six years and had been using acetylsalicylate, nitrate, beta-blockers and insulin. She had undergone a cholecystectomy fifteen years ago and thyroidectomy five years ago.

She had been taken to the intensive care unit for further diagnosis and treatment. Physical examination revealed that, body temperature was 38.1°C, arterial blood pressure was 130/70 mmHg, and crackles on basal area of the right lung. She had left hemiparesis. Hepatomegaly was detected on palpation. Initial laboratory tests revealed, AST: 3090 U/L (normal range: 0-35), ALT: 1410 U/L (normal range: 0-40 U/L), GGT: 800 U/L (normal range: 0-50 U/L), LDH: 1175 U/L (normal range: 100-190 U/L), (ALT/LDH: 1.4), total bilirubin: 3.4 mg/dL, direct bilirubin: 2 mg/dL, blood urine nitrogen (BUN): 98 mg/dL (normal range: 7-20), creatinine: 2.5 mg/dL (normal range: 0.6-1.3), potassium: 5.4 mmol/L, haemoglobin: 9.4 g/dL, reticulocytes: 25.7%. Schistocytes, anisopoikilocytosis were seen on peripheral blood smear and megakaryocytes were detected as increased on bone marrow aspiration.

Urinalysis revealed proteinuria, hematuria and density 1015. On chest X-ray, wedge shaped opacity was seen on the right lower zone. Contrast enhanced CT scans of the brain showed subacute infarct of involved areas. Abdominal ultrasound revealed diffuse hepatomegaly, and all markers of viral hepatitis were negative.

She has been diagnosed as TTP with all these clinical and laboratory findings. Fresh frozen plasma, corticosteroid (metyl prednisolon 80 mg/day) and plasmapheresis (twice) administered to the patient. Hepatic and renal functions, and clinical findings revealed completely in one week. She was discharged from the hospital without any sequel.

DISCUSSION
Thrombotic thrombocytopenic purpura is a fulminant, often lethal disorder that may be initiated by endothelial injury and subsequent release of vWF and other procoagulant materials from the endothelial cell. Characteristic findings include the microvascular deposition of hyaline thrombi that stain for fibrin, thrombocytopenia, microangiopathic haemolytic anemia, fever, renal failure, fluctuating levels of consciousness and evanescent focal neurological deficits. The presence of hyaline thrombi in arterioles, capillaries and venules without any inflammatory changes in the vessel wall is diagnostic (, ). The clinical findings of our patient were also similar to these as defined above.

The presence of a severe Coombs negative haemolytic anemia with schistocytes or fragmented red blood cells in the peripheral blood smear, coupled with thrombocytopenia and minimal activation of the coagulation system help to confirm the clinical suspicion of TTP (, ). In our patient,
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severe haemolytic anemia, typical findings in the blood smear and minimal thrombocytopenia were present. Interesting characteristics of our patient were neurological and hepatic findings. She had no thrombocytopenia while having cerebrovascular accident, but then a minimal decrease of platelets was developed. O'Brien et al described a case of TTP with dominant neurological findings as in our case (7) but our patient had also severe liver enzyme increment.

The treatment of acute TTP has changed radically in the past few years. Steroid and heparin or splenectomy, have been abandoned, and the enthusiasm for antiplatelet therapy has diminished. Increasingly, treatment has focused on the use of exchange transfusion or intensive plasmapheresis coupled with infusion of fresh frozen plasma (1, 8, 9). We also gave high dose corticosteroids, fresh frozen plasma and administered plasmapheresis to our patient. Although the prognosis of TTP is usually poor and the response to the therapy is generally not good, in our patient, with early diagnosis and immediate treatment, a dramatic recovery has been achieved in a short period of time.

In conclusion, without severe thrombocytopenia, neurological findings and impaired hepatic functions could be the presenting features of TTP. In such a case as this, clinical suspicion should be thought about TTP. Early diagnosis and treatment would impact on the prognosis.

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