Thrombotic thrombocytopenic purpura with terminal pancytopenia

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Summary: A 27 year old housewife developed thrombotic thrombocytopenic purpura during the twelfth week of pregnancy. She had partial response to initial plasma infusion and subsequent plasmapheresis. However, her clinical course was complicated by the development of severe pancytopenia the consequence of a hypocellular marrow. She succumbed to septicemic shock one month after diagnosis. The development of hypocellular marrow in thrombotic thrombocytopenic purpura has not been reported before.

Introduction

Thrombotic thrombocytopenic purpura (TTP) is a rare disorder characterized in its complete form by the pentad of thrombocytopenia, microangiopathic haemolytic anaemia (MAHA), fluctuating neurological abnormalities, renal impairment and fever.¹ We report a patient with TTP in pregnancy who developed terminal pancytopenia secondary to hypocellular marrow.

Case report

A 27 year old housewife gravida 3, para 4 was admitted to a peripheral hospital with a history of fever for a week. She was 3 months pregnant. There were no localizing symptoms or signs and she gave no history suggestive of collagen diseases. Her general condition deteriorated rapidly and 4 days later she lapsed into a coma. Her haemoglobin concentration dropped from 10 g/dl to 4 g/dl. She was transferred to the University Hospital, Kuala Lumpur for further management. On examination, she was in deep coma and responded only to deep pain. She was febrile with marked pallor and a tinge of jaundice. Her blood pressure was 120/80 mmHg. Both liver and spleen were enlarged 4 cm below the subcostal margins. Petechiae were noted over her lower limbs. No fundal haemorrhage or lymphadenopathy was detected. There were no focal neurological signs.

Her full blood count showed a haemoglobin concentration of 4.7 g/dl with a reticulocyte count of 4%. Her white cell count was elevated at 30.8 × 10⁹/l with a slight left shift and the platelet count was 10 × 10⁹/l. The direct Coomb’s test was negative. Peripheral blood film confirmed marked thrombocytopenia and a striking number of fragmented red cells and occasional nucleated red cells were present. Her prothrombin time, partial thromboplastin time, thrombin time and fibrinogen level were within normal limits. Bone marrow examination showed a normocellular marrow with erythroid hyperplasia and presence of normal granulopoiesis and megakaryopoiesis. Urinalysis revealed microscopic haematuria. Her blood urea was elevated (18.9 mmol/l) with serum creatinine of 115 µmol/l. Her liver functions were essentially normal except for elevated serum bilirubin of 50 µmol/l. Computerized axial scan of brain showed no focal lesions and cerebrospinal fluid (CSF) examination was normal. Cultures from blood, CSF and urine showed no growth. Ultrasound of abdomen revealed a viable fetus corresponding to 12 weeks’ gestation. Serological tests for dengue and arbovirus and Widal Weil Felix tests were negative. The diagnosis of thrombotic thrombocytopenic purpura was made on the basis of microangiopathic haemolytic anaemia, profound thrombocytopenia, high fever, and evidence of both central nervous system and renal involvement.

She was started on plasma therapy consisting of daily infusion of 5 units of fresh frozen plasma (FFP). She made a remarkable recovery and regained full consciousness by the day 3 of admission. However, there was persistent MAHA and thrombocytopenia and she required 2 units of packed cell transfusions on alternate days. She was started on plasmapheresis on the day 5 of admission and subsequently six more plasmaphereses
References


