Management of rare case of mifepristone induced thrombotic microangiopathy

BALUSWAMY GAYATHRI BALUSWAMY
Department of Obstetrics and Gynaecology, KILPAUK MEDICAL COLLEGE AND HOSPITAL

ABSTRACT-Thrombotic microangiopathies are a group of disorders characterized by microangiopathic haemolytic anaemia, fragmented RBCs and micro vascular thrombosis. Thrombotic thrombocytopenic Purpura is characterized by pentad of fever, thrombocytopenia, microangiopathic haemolytic anaemia, transient neurologic deficit, and renal failure. Haemolytic Uremic Syndrome is also associated with microangiopathic haemolytic anaemia and thrombocytopenia but is distinguished by absence of neurologic symptoms, prominence of acute renal failure and frequently occurs in children. Mifepristone is an anti progesterone indicated for medical termination of pregnancy along with misoprostol up to 49 days of gestational age. We hereby present a case of mifepristone induced Thrombotic microangiopathy which is a rare complication as such in a patient who took MTP drugs over the counter and underwent plasmapheresis.

INTRODUCTION:
Thrombotic microangiopathies which include both TTP and HUS is characterized by thrombocytopenia, microangiopathic haemolytic anaemia, schistocytes in peripheral smear and micro vascular thrombosis. With out proper treatment the condition is fatal in 90% of the cases. Plasma exchange markedly improves the prognosis of the patient with decrease in mortality from 85-100% to 10-30%.

CASE SCENARIO:
A 23 yr old antenatal patient with obstetric score G3P2L2 and previous two LSCS ,weighing 62kg was admitted in the Labor ward on 14/6/13 at 8.15 PM with 16 weeks of amenorrhoea and history of consumption of over the counter MTP drugs. She was referred from a peripheral hospital with severe lower abdominal pain, bleeding per vaginum and passage of clots with foetal parts. Her menstrual cycles were irregular and LMP was not known. She had history of 2 units of blood transfusion for anaemia in previous pregnancy.

GENERAL EXAMINATION:
Patient was conscious and oriented. She was afebrile, not icteric, not pale and no pedal oedema. All the peripheral pulses were well felt. Her pulse rate was 97/min ,regular and her BP was 120/80 mm Hg in the Right Upper limb in the Supine position. Her oxygen saturation in the room air was 98%

SYSTEM EXAMINATION:
On examination her cardiovascular system, respiratory system and nervous system found to be intact. Her abdominal examination showed uterus 16 weeks size ,scar+, uterus contour intact , uterus not tense and not tender. Her per vaginal examination showed cervix pointing downwards, uterus 16 wks size, os open, bleeding+. She had blood stained urine about 200ml.In view of haematuria , previous two caesarean and attempted MTP, uterine rupture was suspected and emergency laparotomy was done which was found to be negative. Her clinical presentation from day1-day8:
Peripheral smear showed : RBC: normocytic, normochromic, schistocytes +, spherocytes +. WBC: macrocytes+. Platelets: Vesicular megakaryocytes seen with number decreased. Tests were negative for malaria, leptospirosis, dengue, typhoid, SLE. Blood culture, urine culture and vaginal swab showed no growth and direct Coombs for malaria, leptospirosis, dengue, typhoid, SLE. Blood culture, megakaryocytes seen with number decreased. Tests were negative.

3 days. On day 6 since patient developed one episode of anaesthesia opinion obtained and she was on ventilator support for following:
1. Plasmapheresis.

Pathogenesis of TTP: idiopathic TTP is due to deficiency of, or antibodies to, a metalloproteinase that cleaves VWF and ADAMTS13 respectively. VWF is normally secreted as ultralarge VWF molecules which contribute to platelet adhesion and aggregation. Drug related TTP may be secondary to antibody formation. Shiga toxin mediated injury to vascular endothelial cells in the kidney, brain and other organs underlies the pathogenesis caused by enterohemorrhagic ecoli. These potent cytotoxins are released in the gut by bacteria, enter the blood stream, and cause endothelial injury through the binding to the globotriaosylceramide (Gb3) receptor on the plasma membrane of target cells. Atypical HUS due to mutation in the genes encoding complement proteins including C3, factors H, factors B and I and membrane cofactor protein. These mutations result in dysregulation of complement that leads to excess complement activation and endothelial damage. Manifestation of HUS - microangiopathic haemolytic anaemia, thrombocytopenia and acute renal failure in children. CNS involvement in 20-50% of cases. TTP - characterized by more of neurological manifestations, microangiopathic haemolytic anaemia, thrombocytopenia. Confirmation of diagnosis by schistocytes, microspherocytes, reticulocytosis in peripheral smear. Increased LDH, decreased haptoglobin, increased urea and creatinine. Decreased platelet count. Negative Coombs test.

Treatment: