Postpartum Thrombotic Microangiopathic Syndrome (PTMs): Case Report

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Background:
HELLP syndrome is a life-threatening condition complicating 0.5-0.9% of all pregnancies. It is characterized by endothelial dysfunction associated with microangiopathy. The syndrome is named after the triad of diagnostic criteria: Hemolysis, Elevated Liver enzyme levels, and Low Platelet levels, and it is mostly classified using The Mississippi-Triple Class System.

The basic treatment usually involves early delivery, antihypertensive drugs, high-dose magnesium sulphate, corticosteroids and close monitoring of fluid balance and coagulation parameters. About 30% of the cases develop after the delivery with a part of patients non-responding to usual therapy. These patients may suffer from PTMs – postpartum thrombotic microangiopathic syndrome. Its symptoms remind of HELLP syndrome by its laboratory parameters and clinical signs, these however do not respond to classic therapy and even progressively deteriorate. The Hemolytic-uricemic syndrome (HUS), Thrombotic thrombocytopenic purpura (TTP), Systemic lupus erythematosus (SLE) and Antiphospholipid syndrome (APS) are some of the diseases included. On the basis of microangiopathy the disseminated intravascular coagulation and multiorgan failure may develop and repeated Postpartum plasma exchange (PPEX) is often necessary.

Case report :
Patient Z.F., 27 years
Past medical history:
Family: rather unremarkable,
Personal: breast cysts followed up,
Surgeries: umbilical hernia in childhood
laparoscopy for endometriosis 2015
Medication:
Dietetics: 1-0-1 (venotocin)

History of present illness, admission to Labour ward
25th July
Grav. hemb. 36+0-referred for hypertension,
PE: blood pressure 84/60, proteinuria +++
Lab.: proteinuria 18.7 g, urine positive for inflammation parameters
Recommendation: urine culture test, check-up 2 days later, 24 hours urine collection.

27th July
Grav. hemb. 36+2
8:00 blood diate 64-69, proteinuria 11 g/24h,
Bishop score 8, EFW 2870
Recommendation: preinduction of labour in the evening

17:13 Brought in by ambulance with epigastric pain, nausea and vomiting,
Lab.: normal – Hb 120, platelets 310, ALT 0.32, AST 0.56, LD 4.37, urea acid 343, urea 4, creatinine 62.

18:26 Acute Caesarean section for suspected preeclampsia: During surgery the blood diastole increases over 100, not responding to intravenous Trandate (Labetalol).

Newborn girl 2470 g, 47 cm,
Apogee score 9-10-10
Umbilical cord pH 7.33
Patient admitted in the intensive care unit, intravenously administration of Trandate (Labetalol).

23:00 Seizures with unconsciousness - suspected eclamptic seizure, BP 150/120, Aparzin (Diazepam) intramuscularly administered, urine output sufficient, trial to transfer the patient to Emergency department, refused due the lack of capacity.

28.7
00:30 Tonic-clonic seizures, BP 173/127, Aparzin (Diazepam) intramuscularly administered, urine output normal

1:00 Sleeping, BP 154/96, oliguria – Furosemide administration

6:00-18:00, PP 99, LD 56, ALT 3.12, AST 13.7, urea 5.4, creatinine 182, CRP 16.4, PUL g somnolent, oliguria, blood diastole 90-100.
HELLP syndrome suspected.

Medication: Urokinase (urothrombolytic acidic), Transatil (Ademetionin), Mg sulphate, Furosemide, Trandate (Labetalol), Dexam (Dexamethason)

Thromboelastography (ROTEM) provided.

10:00 EEG: abnormal – Neurologist consultant: brain CT scan

11:00 Transfer to Emergency department.

Brain CT scan: hypodense areas in both hemispheres, nigh thalamus and pons cerebelli - suspected developing ischaemia.

Hospitalization in Emergency department (28.7-9.8.):
28.7
Cefotaxime administered.
Cardiac markers elevated, echocardiography without kinetic disorder, mitral valve regurgitation.
Signs of renal failure: continuous venovenous hemodiafiltration (CVVHD) initiated.
Mechanic pulmonary ventilation needed.

Hypertension not responding to medication – Agen (Amitidine), Ebenati (Hydralazine), Vasocardin (Metoprolol), Zosan (Doxazosin), Furosemid = intravenous Trandate (Labetalol) and Dihydratrazine.

Hemoglobin and platelets decrease. Despite four units of leucocyt-reduced red blood cell transfusion administered, the decrease progressed, two other units of RBT repeated with two units of platelet transufind, 2g of Fibrogen administered.

22:00 Revision surgery, subcutaneous hemorhoid, abdominal cavity revision. Repeated transfusions: two units of RBT, one unit of platelet concentrate.

Hematology consultant: suspected PTMs (postpartum thrombotic microangiopathic syndrome), Plasma exchange initiated (PPEX).

Exclusmam (Soliris) obtained from Motil University Hospital Prague, however it was not administered.

29.7, Persisting MODS signs, coagulation instability
30.7, Stable hemodynamic status, persisting thrombocytopenia, PPEX daily until platelets elevated.

Brain CT scan: partial regression.

31.7, Hematology consultant: TTP (thrombotic thrombocytopenic purpura) excluded, Adams13 normal.

Nefrology consultant: CVVHD discontinued. Atopic hemolytic uricemic syndrome (aHUS) unlikely.

1.8, Brain CT scan: regression. Estabulation, somnolence remaining.

3.6, Renal parameters elevation – CVVHD de novo.

Fundoscopy exam normal. Moderate fluidothorax. Cefotaxime discontinued.

4.8, State of consciousness improved.

5.6, Persisting thrombocytopenia.

6.6, PPEX discontinued, oral alimentation reestablished.

7.6, Platelet level increased.

Hospitalization in Cardiology department. (9.8-11.8)
Antihypertensive treatment.
Hospitalization in Gynecology department (11.8-16.8)

Patient stable.
Hematologic, renal parameters and liver function tests normalized.
Patient discharged without other difficulties.
Recommended antihypertensive therapy: Vasocardin (Metoprolol) 50mg 1-0-1 Agen (Amitidine)10mg 1-0-1 Zosan (Doxazosin) 4mg 1-0-0
Hydrochlorothiazide 25mg 1-0-0

Summary:
27-year old pregnant woman, so far healthy. Pregnancy terminated by acute CS in 38 weeks of gestation for suspected preeclampsia with decompenasated hypotension, but satisfying blood test values. After the surgery two eclamptic seizures occurred, with developing somnolence and oliguria. 12 hours after the surgery, laboratory signs of HELLP syndrome appeared with no response to usual therapy. Coagulation parameters deteriorated. Ischaemia signs appeared on brain CT scan, EEG abnormal.
Cardiac markers elevated, renal failure signs with necessity of CVVHD, hepatic failure corresponding to multiorgan failure. Revision surgery for abdominal wall hemorhoid. Patient critically ill in the emergency department, pulmonary ventilated, repeatedly transfused with blood components. PTMs suspected, postpartum plasma exchange PPEX initiated (total of 10 plasma exchanges) with gradual improvement. TTP excluded, as Adams13 gene test was negative. Genetic testing for aHUS provided, test results are so far not known.

Conclusion:
Postpartum thrombotic microangiopathic syndrome is a very rare life-threatening condition associated with increased mortality and morbidity. It should be kept in mind when clinical and laboratory signs of presumed HELLP syndrome deteriorate despite adequate treatment.

References: