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Postpartum Thrombotic Microangiopathic Syndrome (PTMS): Case Report

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Background:

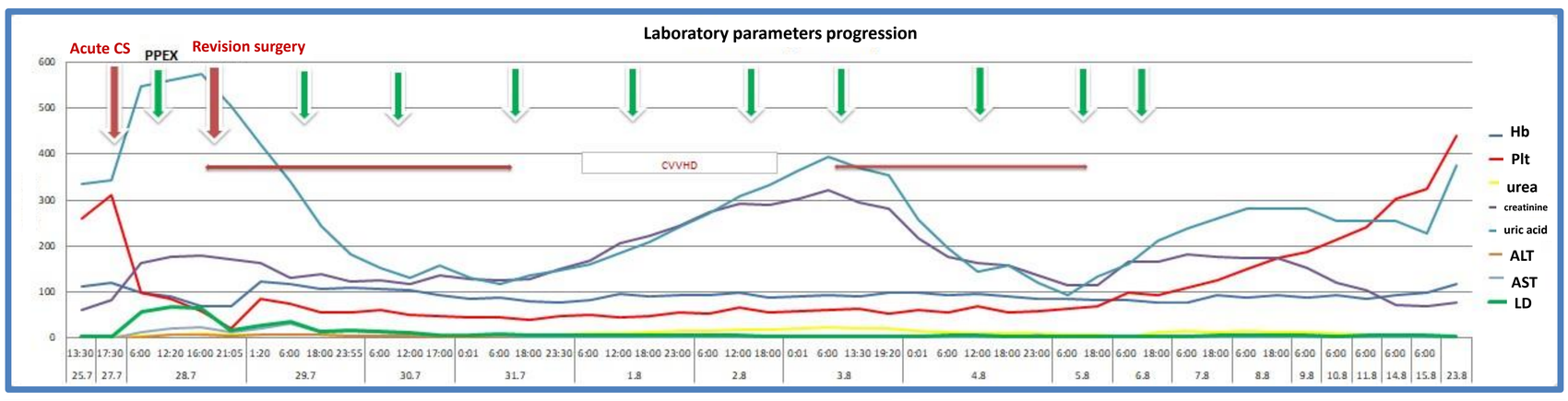
HELLP syndome is a life-threatening condition complicating 0,5-0,9% of all pregnancies. It is characterized by endothelial dysfunction associated with microangiopathy. The syndrome is named after the triad of diagnostic criteria: Hemolysis, Elevated Liver enzyme levels, and Low Platelet levels, and it 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-reacting 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-uraemic syndrome (HUS), Thrombotic thrombocytopenic purpura (TTP), Systemic lupus erythematosus (SLE) and Antiphospholipide 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.Ř., 27 years**
Past medical history:
Family: rather unremarkable,
Personal: breast cysts followed up
Surgeries: umbilical hernia in childhood
laparoscopy for endometrioisis 2015
Medication: Detralex 1-0-1 (venotonics)
Gyn and Ob: endometriosis
nulligravida
Substance abuse: negative

Current pregnancy:
nuliparous
GBS negative
Standard prenatal care



History of present illness, admission to Labour ward

25th July
Grav. hebd. 38+0 referred for hypertension.
PE: blood diastole 85-90, proteinuria +++
Lab.: proteinuria **18,7g**, urine positive for inflammation markers
Recommendation.: urine culture test, check-up 2 days later, 24-hours urine collection.

27th July
Grav. hebd. 38+2
8:30 blood diastole 84-90, proteinuria **11g/24hours, Bishop score 0,** EFW 2870g
Recommendation.: preinduction of labour in the evening

17:13 Brought in by ambulance with epigastric pain, nausea and vomit, headache, blurred vision.
Lab.: normal - Hb 120, platelets 310, ALT 0,32, AST 0,58, LD 4,37, uric acid 343, urea 4, creatinine 82.

18:26 Acute Caesarean section for suspected preeclampsia; During surgery the blood diastole increases over 100, not responding to intravenous Trandate (Labetalol).
Newborn girl 2470g/ 47 cm,
Apgar score 9-10-10
Umbilical cord blood pH 7,33
Patient admitted in the Intensive care unit, intravenously Magnesium sulphate, Trandate (Labetalol).

23:00 Seizures with uncounsciousness - suspected eclamptic seizure, BP 150/120, Apaurin (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, Apaurin (Diazepam) intramuscularly administered; urine output normal.

1:00 Sleeping, BP 154/96; oliguria - Furosemid intravenously administered.

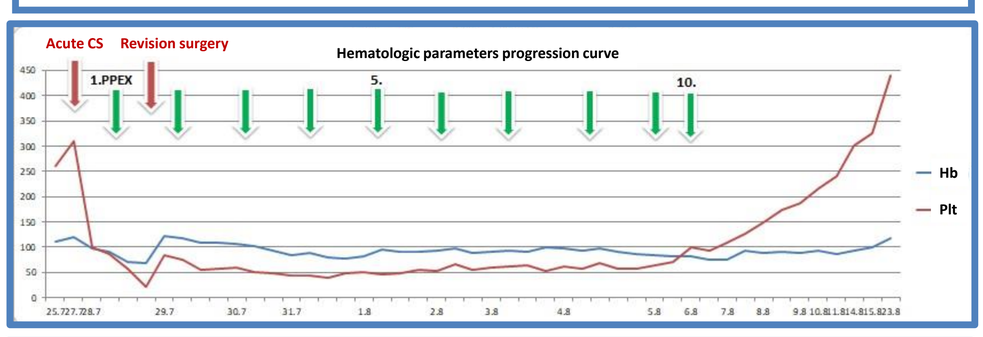
6:00 Hb 98, **Plt 99, LD 56, ALT 3,12, AST 13,7, urea 5,4, creatinine 162**, CRP 16,4 , PU 4g
somnolent, oliguria, blood diastole 95-100,
HELLP syndrom suspected.
Medication: Ursosan (ursodeoxycholic acid), Transmetil (Ademethionin), Mg sulphate, Furosemid, Trandate (Labetalol), Dexamed (Dexamethason).
Tromboelastography (ROTEM) provided.

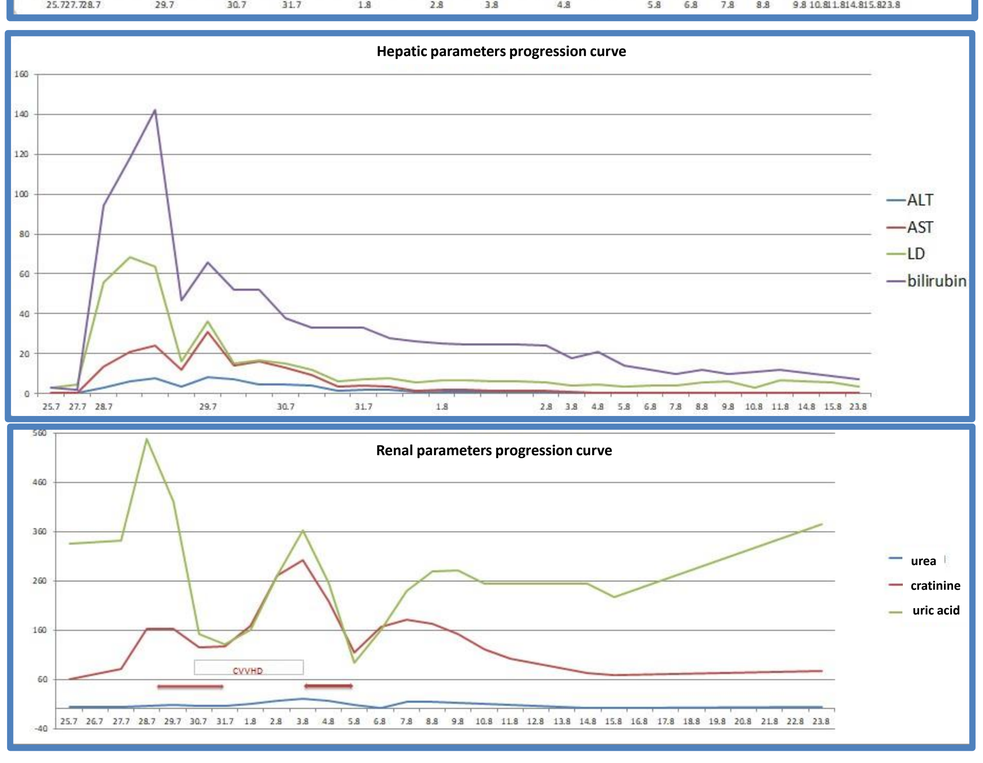
10:00 EEG: abnormal
Neurologist consultant: brain CT scan
11:00 Transfer to Emergency department.
Brain CT scan: hypodense areas in both hemispheras, right thalamus and pons cerebelli - suspect developing ischaemia.

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 hemodialysis (CVVHD) initiated.
Mechanic pulmonary ventilation needed.
Hypertension not responding to medication – Agen (Amlodipine), Ebrantil (Urapidil), Vasocardin (Metoprolol), Zoxon (Doxazosin), Furosemid + intravenous Trandate (Labetalol) and Dihydralazine.
Hemoglobine and platelets decrease. Despite four units of leukocyte-reduced red blood cell transfusion administered, the decrease progressed, two other units of RBT repeated with two units of platelet transfused, 2g of Fibrinogen administered.

22:00 Revision surgery, subcutaneous hematoma, abdominal cavity revision. Repeated transfusions: two units of RBT, one unit of platelet concentrate.
Hematology consultant: suspected **PTMS** (postpartum trombothic microangiopathic syndrome). Plasma exchange initiated (**PPEX**). Eculizumab (Soliris) obtained from Motol University Hospital Prague, however it was not admnistered.

29.7. Persisting MODS signs, coagulation instability
30.7. Stable hemodynamic status, persisting trombocytopaenia, PPEX daily until platelets elevated.
Brain CT scan : partial regression.
31.7. Hematology consultant: **TTP (thrombotic thrombocytopenic purpura) excluded, Adamts13 normal.**
Nefrology consultant: CVVHD discontinued. Atypic hemolytic uremic syndrome (aHUS) unlikel.y
1.8. Brain CT scan: regression. Extubation, somnolence remaining.
3.8. Renal parameters elevation – CVVHD de novo.
Fundoscopic exam normal. Moderate fluidothorax. Cefotaxime discontinued.
4.8. State of conciousness improved.
5.8. CVVHD discontinued.
6.8. PPEX discontined, oral alimentation reestablished.
7.8. 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 (Amlodipine)10mg 1-0-0
Zoxon (Doxazosin)4mg 1-0-0
Hydrochlorthiazide 25mg ½-0-0

Summary:

27-years old pregnant woman, so far healthy. Pregnancy terminated by acute CS in 38 weeks of gestation for suspected preeclampsia with decompensated hypertension, 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 syndrom 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 hematoma. 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 Adamts13 gen 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:

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2. Vlk R, Procházka M. , Měchurová A, Šimetka O, Janků P. Preeklampsie. Maxdorf Jessenius. 2015
3. Owens MY., Martin JN. Jr., Wallace K., et al., Postpartum thrombotic microangiopathic syndrome. Transfus Apher Sci, 2013, 48(1), str. 51-7