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Hellp! S.O.S. Call of a Mother

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HELP syndrome is a serious, life-threatening form of pre-eclampsia that is a major cause of maternal and perinatal mortality and morbidity, particularly in developing countries. The syndrome is characterized by a typical laboratory triad: hemolysis, elevated liver enzymes and low platelets (1). It can occur in 0.2-0.6% of all pregnancies and 19-27% of cases may show recurrence (2,3). Cardinal findings of the disease are high blood pressure, proteinuria, and epigastric or right upper abdominal pain. The most common neurological findings in pre-eclampsia are headache, visual alterations, convulsions and stupor or coma.

The etiology of the disease is not clear. It is proposed that endothelial imbalance between vasodilatative and vasoconstrictive substances causes segmental vasospasms, vasoconstriction and a further increase in endothelial dysfunction. This leads to increased platelet aggregation and intravascular coagulation with fibrin deposition in the capillaries and consecutive microcirculatory disorders (4). If not treated disseminated intravascular coagulation may develop within hours.

We describe a clinically and biochemically severe HELLP syndrome case who had no signs or findings of the disease prior to delivery.

Case Report

A 22-year-old primigravid woman was admitted to a local obstetric hospital at due date with regular contractions. At the third stage of labor, the patient had a generalized tonic-clonic seizure. Since she did not regained consciousness and continued to have seizures, she was referred to the neurology department with a diagnosis of status epilepticus. Past medical history was unremarkable; no pathologic finding had been noted on her routine follow-ups. Upon admission blood pressure, heart rate and respiration rate were normal. She had no fever. She was unresponsive to noxious stimuli. Both pupils were equal in size and brainstem reflexes were preserved. Tendon reflexes were diminished and plantar reflexes were indifferent bilaterally. She continued to have tonic-clonic seizures in the intensive care unit.

Laboratory work-up before delivery, including blood chemistry and urinalysis, was unremarkable. However, within the same day, after delivery there was a remarkable increase in hepatic enzyme levels (SGOT: 4202U/l, SGPT: 3949U/l, LDH: 2943U/l, CPK: 1668U/l). The erythrocyte sedimentation rate was 60 mm/h, bilirubin was 8.3 mg/dl and fibrinogen was 3.38 g/dl. Prothrombin time was 18.5 s and partial thromboplastin time was 31 s. CBC was remarkable for low hemoglobin (6.9 mg/dl) and thrombocytopenia (29.000 mm$^3$/µL), and urinalysis for (+++) proteinuria. Antiphospholipid antibodies IgM and IgG and antinuclear antibodies yielded normal levels. There were diffuse bilateral pallidum, thalamus and brainstem hypodensities on cranial tomography. Cranial MRI scanning demonstrated diffuse infarct areas on the medulla, pons and basal ganglia and focal hemorrhagic infarct areas on the right parieto-occipital, left parietal and left temporoooccipital lobes (Figure). A symptomatic therapy
was immediately started including anticonvulsants, anti-
edematous agents, fresh frozen plasma, and erythrocyte
and thrombocyte suspensions. She was also given low
molecular weight heparin. A piece of residual desidua was
removed on control gynecologic examination.

Liver enzyme and bilirubin levels started to decrease
after the second day of treatment. She regained her
consciousness on day 7 and was able to follow
commands. She had slight right hemiparesis on
neurologic examination. She showed progressive general
and neurological improvement. At one and three months,
neurologic examination and laboratory work-up were
normal.

Given the presence of remarkable laboratory findings
the patient was given a diagnosis of Class-I HELLP
syndrome. The patient presented with the most common
and dramatic neurological complications of the disease:
convulsions and coma. However, the peculiarity of this
case was that she had no sign of preeclampsia prior to
labor. This is a rare but not unexpected circumstance; it
has been reported that 6% of cases may have no signs of
preeclampsia before delivery (4). Thus, the possibility of
preeclampsia should not be ruled out in the differential
diagnosis of post-partum neurological complications
based on the absence of previous clinical history.

Differential diagnosis of HELLP syndrome refers
mainly to illnesses not related to obstetrics with
gastrointestinal symptoms, to liver complaints and
thrombotic obstructive microangiopathy (5). Thrombotic
thrombocytopenic purpura, hemolytic uremic syndrome,
systemic lupus erythematosus and antiphospholipid
syndrome should be evaluated in the differential
diagnosis. Our work-up for the existence of any other
disease yielded negative results.

The severity and course of the disease are quite
variable. Clinicians should be alert for a higher risk of
renal failure, consumptive coagulopathy, pulmonary and
cerebral edema, subcapsular liver hematoma and
hypovolemic shock (5,6). In our case, despite a rigorous
onset and rapid deterioration of the disease a good
outcome was observed.

There is no specific treatment for the disease. The
treatment mostly consists of delivery and symptomatic
therapeutical regimens to prevent the possible
complications. Different results have been reported for
the beneficial effect of removal of the placenta (4,7,8).
The use of heparin is controversial, because of a high
possibility of hemorrhage from an open placental vascular
bed, a probable section side or just because of
thrombocytopenia. Use of low molecular weight heparin

Figure. Coronal and axial T1 MR scans showing hemorrhagic infarction on right parietooccipital, left posterior parietal and left temporooccipital
lobes and subacute infarction on right side of pons.
(LMWH) is recommended after the stabilization of coagulation profile. We used LMWH since the early stage of the disease in spite of hemorrhagic infarction and observed no complication.

In this case, the diagnosis was made easily and at an early stage because the remarkable laboratory findings. However, not all patients may have such definite laboratory findings or show such a full-blown and fulminant course. In the differential diagnosis of postpartum complications HELLP syndrome should be kept in mind even if the clinical history is not consistent with pre-eclampsia. Identification of the disease is not only important for decreasing mortality but also for preventing complications that would develop in following pregnancies since the disease may recur in up to one-third of patients.

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