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To cite this article: D Elsandabesee, R Hamzeh & A Pozyczka (2004) Hemiparesis as an unusual presentation of haemolysis, elevated liver enzymes, low platelet count (HELLP) syndrome, *Journal of Obstetrics and Gynaecology*, 24:8, 926-927

To link to this article: <http://dx.doi.org/10.1080/01443610400019179>



Published online: 02 Jul 2009.



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Theoretically, the enhanced thrombogenesis can lead to a hypercoagulable state that could potentially worsen DIC; however, studies in animals suggest no increase in risk of DIC (Bregengaard *et al.*, 1993). Thrombotic events such as myocardial infarction, cerebrovascular ischaemia, deep vein thrombosis and pulmonary embolism have been reported, but appear to be related to risk factors for these conditions and the severity of the condition for which NovoSeven is used other than to use of NovoSeven or dosage (Roberts, 1998). The major drawback is that the product is expensive.

Alternative treatments such as hysterectomy lead to loss of fertility, an issue for many woman. Uterine artery embolisation (Drife, 1997) had also been used in limited cases but is not available in many units and will probably never be used outside specialist centres. In addition, it is an invasive procedure with potential risks of infection, vascular injury and radiation exposure.

In summary, we have demonstrated a new therapy for the treatment of postpartum bleeding in association with a traumatised placental bed. It provides an easy therapy to add to the therapeutic options for this difficult problem.

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DOI: 10.1080/01443610400019120

Hemiparesis as an unusual presentation of haemolysis, elevated liver enzymes, low platelet count (HELLP) syndrome

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Case report

A 35-year-old woman in her second pregnancy had a normal delivery followed by manual removal of the placenta. The pregnancy was uneventful and there was no medical or surgical history of relevance. Five hours after delivery, the woman complained of headache and muscle weakness on the left side followed by three episodes of tonic and clonic seizures. The blood pressure was 124/74 mmHg and remained normal. There was no proteinuria. Flaccid paralysis was demonstrated on the left side with extensor plantar response. An average score of 14 was given on the Glasgow Coma Scale. The provisional diagnosis was that of cerebrovascular stroke; however, the woman was treated as a case of eclampsia until the diagnosis of stroke could be confirmed.

Initial blood results revealed marked rise in liver enzymes, bilirubin and uric acid (alanine transaminase 550 IU/l, alkaline phosphatase 314 IU/l, bilirubin 45 $\mu\text{mol/l}$, urate 0.6 $\mu\text{mol/l}$). Platelet count was found to drop progressively to reach $66 \times 10^9/l$ (220×10^9 prior to manual removal of placenta). Coagulation screen was normal, apart from raised D-dimer (8174 ng/ml). Peripheral blood film showed no evidence of haemolysis. The diagnosis of haemolysis, elevated liver enzymes, low platelet count (HELLP) or more strictly ELLP syndrome was made. CT scan revealed three areas of intracerebral haemorrhage (ICH) involving the right frontal, temporal and parietal lobes with midline shift.

The patient was transferred to a tertiary referral Neuroscience Critical Care Unit for further management. Conservative management with physiotherapy as well as haematological and neurological monitoring was adopted. MRI excluded lateral and sagittal sinus thrombosis. Platelet count and liver function tests improved gradually and the woman was transferred back to our hospital to be managed with physiotherapy for left hemiparesis. She showed

progressive improvement and was discharged 23 days after the incident.

Discussion

The importance of not missing the diagnosis of HELLP syndrome lies in the potentially disastrous consequences of the condition. Right upper quadrant pain is the presenting symptom in 86–92% of cases. The majority of patients (90%) will give a history of malaise while 45–86% complain of nausea and/or vomiting (Rath *et al.*, 2000).

A striking feature in this case report is the unusual presentation as hemiparesis. ICH is a recognised complication and has been identified as the most common cause of death in patients with HELLP syndrome (Hashiguchi *et al.*, 2001). We claim, however, that no cases published in the literature presented with hemiparesis. We conducted a MEDLINE search for the English-language articles dealing with HELLP syndrome and intracranial haemorrhage or stroke published from 1983 to 2003. None of the three cases reports identified (Gliemroth *et al.*, 2000; Hashiguchi *et al.*, 2001; Ezri *et al.*, 2002) presented in this way.

Another unusual feature is the occurrence of ICH in the presence of normal blood pressure. Although up to 20% of patients present no hypertension (Rath *et al.*, 2000), there have been no reports of ICH in normotensive patients with HELLP. Using strict diagnostic criteria, this case would be considered partial, as opposed to complete HELLP syndrome. It should be borne in mind, however, that haemolysis is the most difficult feature to capture in a laboratory process (Rath *et al.*, 2000). Although a higher incidence of maternal complications has been highlighted with complete HELLP syndrome, this case report demonstrates that there is no

place for complacency in the management of partial HELLP syndrome.

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DOI: 10.1080/01443610400019179

Methylmalonic acidaemia: a rare metabolic disorder in pregnancy

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Case report

A 23-year-old primigravid woman was booked for antenatal care at 9 weeks' gestation in a district general hospital. As a neonate she had been diagnosed to be suffering from a rare metabolic disorder: methylmalonic acidaemia due to deficiency of the methylmalonyl CoA mutase enzyme (a vitamin B₁₂-dependent form of the disease). She developed epilepsy at the age of 7 years, possibly secondary to MMA (methylmalonic acidaemia)-related central nervous system effects in early childhood. Convulsions, which had the features of both absence and generalised seizures, were controlled with carbamazepine.

She was on a high-dose combined pill for contraception as she was on antiepileptic drugs, but missed a pill and became pregnant. Her diet was unremarkable. She smoked 10–15 cigarettes a day. She had one sister who does not have MMA. She had not suffered any metabolic decompensation for many years.

She weighed 60.3 kg with a body mass index of 23. Her general and systemic examination was unremarkable. She was booked for antenatal care at 9 weeks' gestation. Booking bloods, dating and anomaly scans were normal. Downs and neural tube defect screening tests were normal. The partner's carrier status was unknown. The patient was advised that the neonate would be tested for methylmalonic acidaemia by blood spots and urine examination.

She was seen in combined obstetric/medical clinic on a fortnightly basis. Specialist advice was sought from a consultant in adult inborn errors of metabolism at a tertiary centre. Dietary protein was restricted with adequate carbohydrate supplementation. Oral cyanocobalamin therapy 500 µg twice a day was continued throughout pregnancy. Oral L-carnitine was supplemented at 1 µmol/10 ml per day. Because essential fatty acid deficiency is common in MMA, she was prescribed Maxepa capsules. Seizure control was achieved with carbamazepine 800 mg twice a day, the dose was being adjusted in relation to serum levels. Steroid inhalers were continued for asthma.

Full blood count, urea and electrolytes and liver function tests were within the normal range throughout pregnancy. The MMA/creatinine ratio was raised throughout pregnancy. Plasma total and free carnitine were decreased, suggesting relative carnitine deficiency. Propionyl carnitine/palmitoyl carnitine ratio was 5.8.

She presented at 20 weeks' gestation with vaginal discharge and a high vaginal swab did not reveal any pathological organisms. At 25 weeks' gestation she was admitted with abdominal pain. An

endocervical swab confirmed a chlamydial infection, which was treated adequately with clarithromycin. She was followed-up in the genitourinary clinic. She was admitted at 27 weeks' gestation with vomiting and backache, which settled with intravenous fluids and anti-emetics. At 32 weeks' gestation she was admitted with preterm prelabour rupture of membranes and was managed conservatively. Dexamethasone was administered, and regular temperature monitoring, twice-weekly full blood count and daily cardiotocogram were carried out. At 34 weeks' gestation, induction of labour was commenced in view of suspected chorioamnionitis on clinical grounds, although the white cell count was within normal range. She underwent emergency caesarean section for failure to progress in labour and delivered a live female baby weighing 1.9 kg with an apgar score of 9 and 10 at 1 and 5 minutes, respectively. Umbilical cord pH and base excess were within normal range. The baby was admitted to neonatal unit for observation. The baby's urine showed no evidence of methylmalonic acid excretion. The baby was discharged home with multivitamins iron and folic acid. The patient had an uneventful postpartum recovery.

At 6 weeks' follow up the baby was making appropriate developmental progress. At 5 months' follow up the baby weighed 6.15 kg, which was around the 50th centile for her corrected age. Her examination was unremarkable with a satisfactory developmental progress.

Discussion

Methylmalonic acidaemia is a rare genetic metabolic autosomal recessive disorder. Methylmalonic acid is derived from propionic acid as a part of the catabolic pathway of isoleucine, valine, threonine and methionine. A racemase enzyme converts the D-isomer of MMA to L-isomer and the latter is converted to succinic acid by a mutase, which required adenosyl cobalamin as the co-enzyme. Our patient had a deficiency of MMA CoA mutase, which is the most common variant of the disease.

Accumulation of methylmalonic acid in the body leads to metabolic acidosis, ketosis, hyperammonaemia and relative carnitine deficiency (as it is excreted as methyl carnitine). Neurological manifestations include seizures, encephalopathy and stroke (Behrman *et al.*, 1996).

Rational treatment of this condition is by a low-protein diet that is deficient in MMA precursors. Large doses of vitamin B₁₂ are needed in the B₁₂-responsive form of the disease. Acidosis may need