RELATO DE CASO

HELLP SYNDROME WITH PLACENTAL ABRUPTION AND FETAL DEATH: A CASE REPORT

SÍNDROME HELLP COM DESCOLAMENTO PLACENTÁRIO E ÓBITO FETAL: RELATO DE CASO

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ABSTRACT

The HELLP syndrome – defined as haemolysis, elevated liver enzymes and thrombocytopenia – consists of an advanced stage of preeclampsia (PE), which can affect 0.6% of pregnancies and 4-12% of patients with severe PE. Being responsible for elevated maternal and perinatal mortality rates, it can present with general malaise, epigastralgia, right hypochondrium pain, nausea and vomits, headache, scotomas, associated with hypertension and proteinuria. Woman, 19 years old, primigest, admitted with 29 weeks of gestational age presenting with arterial hypertension (160/120 mmHg), headache, scotomas, nausea and vomits. She showed regular health status, pallid, closed, anterior and soft cervix, with no fluid losses, and inaudible fetal heart rate on the sonar. She was stabilized with intravenous hydration and antihypertensive drugs, the ultrasonography showed placental abruption and fetal death. An uneventfully caesarean operation was performed under general anaesthesia. Due to the low haemoglobin, haematocrit and platelet levels, she received blood transfusion, progressing with clinical and laboratory improvement. The pressure control was met by the use of methyldopa, nifedipine and pindolol, and she was discharged with a good health status at the 10th day of hospitalization, with scheduled return for reassessment.

Keywords: HELLP syndrome, severe preeclampsia, fetal death, placental abruption.

RESUMO

A síndrome HELLP – definida por hemólise, enzimas hepáticas elevadas e plaquetopenia – consiste em estágio avançado de pré-eclâmpsia (PE), podendo afetar 0,6% das gestações e 4-12% das pacientes com PE grave. Responsável por elevados índices de mortalidade materna e perinatal, pode se apresentar com mal estar geral, epigastralgia, dor em hipocôndrio direito, náuseas ou vômitos, cefaleia, escotomas, assoaciado a quadro hipertensivo e proteinúria. Mulher de 19 anos, primigesta, admitida com 29 semanas de idade gestacional com quadro de hipertensão arterial (160/120 mmHg), cefaleia, escotomas, náuseas e vômitos. Apresentava regular estado geral, hipocorada, colo uterino fechado, anterior, amolecido e sem perdas e batimento cardiofetal inaudível ao sonar. Estabilizada com hidratação venosa e anti-hipertensivos, foi realizada ultrassonografia que evidenciou descolamento prematuro de placenta e óbito fetal. Realizada cesariana sob anestesia geral sem intercorrências. Devido a baixos níveis de hemoglobina, hematócrito e plaquetas, foi hemotransfundida, apresentando melhora clínica e laboratorial. O controle pressórico foi atingido com uso de metildopa, nifedipina e pindolol, recebendo alta em bom estado geral no 10º dia de internação hospitalar com programação de retorno para reavaliação.

Palavras-chave: Síndrome HELLP, Pré-eclâmpsia grave, óbito fetal, descolamento placentário.

ACESSO LIVRE

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INTRODUCTION

Preeclampsia (PE) consists of a multisystem disorder that occurs during pregnancy or puerperium, and it is the main cause of maternal and fetal mortality[1]. It presents after the 20th week of pregnancy with hypertension – systolic arterial pressure (SAP) ≥ 140 mmHg and/or diastolic arterial pressure (DAP) ≥ 90 mmHg – associated with proteinuria (> 300 mg/dL)[2]. The severe PE is defined by SAP ≥ 160 mmHg and/or DAP ≥ 110 mmHg, and it can be followed by proteinuria ≥ 5 g/day, oliguria (< 500 mL/day), elevated serum creatinine, fetal growth restriction, pulmonary edema, neurologic manifestations (headache, visual disturbances, seizure or stroke), hepatic manifestations or HELLP syndrome (Hemolysis Elevated Liver enzymes Low Platelets), condition described by Louis Weinstein in 1982[3].

The HELLP syndrome consists of an advanced staged of PE[3,4] and it can affect 0,6% of gestations and 4-12% of patients with severe PE, developing high maternal (24%) and perinatal (up to 40%) mortality rates[5,6]. It presents clinically with unspecific symptoms, such as general malaise, epigastralgia, right hypochondrium pain, nausea and vomits, headache and scotomas. The thrombocytopenia can lead to the development of mucosal haemorrhage, petechiae or ecchymosis. The arterial hypertension, although frequent, may be absent in 12-18% of pregnant women and the proteinuria may not be detected in 13% of the cases[1].

The diagnosis can be made through the criteria stablished by the American College of Obstetricians and Gynecologists' Task Force on Hypertension in Pregnancy, which are: haemolysis, at least two of the following manifestations: peripheral blood smear with schizocytes or echinocytes, serum bilirubin ≥ 1,2 mg/dL, low serum haptoglobin or severe anemia not related to blood loss; elevated liver enzymes (aspartate aminotransferase – AST – or alanine aminotransferase – ALT – and lactate dehydrogenase – DHL – twice higher than the reference values or more); and thrombocytopenia (< 100.000/mm^3)[1].

Regarding the platelets count, there are three HELLP syndrome categories[4]: class I if the platelets count is lower than 50.000/mm^3; class II if the count is between 50.000/mm^3 and 100.000/mm^3; and class III if it is higher than 100.000/mm^3.

The HELLP syndrome can lead to severe maternal complications, such as disseminated intravascular coagulation, which is the most common, followed by hematoma or hepatic rupture[1,6]. Regardless of gestational age, women with the HELLP syndrome need termination of pregnancy after clinical stabilization or the presence of one or more of the following factors: fetal death, placental abruption (PA), pulmonary edema, hepatic hemorrhage or stroke[7].

CASE REPORT

Nineteen-years-old woman, white, married, residing in Rio dos Bois, state of Tocantins, Brazil. Primigest, admitted to the emergency department of the Dona Regina Siqueira Campos Maternity Hospital (HMDR), Palmas, Tocantins, with 29 weeks of gestational age – based on an ultrasonography (USG) of 15 weeks – referred from the Miracema Regional Hospital showing arterial hypertension, headache, scotomas, nauseas and vomits. She reported at the time of the consultation that she was under treatment for urinary tract infection (UTI). She used hydralazine and methyldopa at the previous hospital. On the physical examination, she presented with hypertension (arterial pressure of 160/120 mmHg), regular health status and pallid. The fetal heart rate was inaudible on the sonar. The vaginal examination revealed a closed, anterior and soft cervix, with no fluid losses.

She was referred to the monitoring room with the diagnostic hypothesis of pregnancy-specific hypertensive disorder (PSHD). An obstetric USG and laboratory tests were requested and intravenous hydration with ringer's lactate solution was performed; hydralazine, magnesium sulphate (MgSO4) and antibiotic therapy with ceftriaxone and metronidazole were administered.

The USG showed a probable PA of eight cm and fetal death (approximately 29 weeks of pregnancy). The initial management was inducing labour with misoprostol (25 μg), strictly monitoring the clinical evolution. After that, the test results became available and demonstrated HELLP syndrome class II (Table 1). The labour induction was then suspended and emergency caesarian was indicated.

Eight hours after admission, the patient was referred to the surgical center, conscious and oriented, to undergo a caesarian section. The extraction of the dead fetus was done under general anesthesia, with a Pfannenstiel incision. The procedure was reportedly uneventful. After the discharge of the post-anesthesia recovery room (PAR), the patient was referred again to the emergency department. Due to the low haemoglobin, haematocrit and platelet levels (Table 1), she received two packed red blood cells transfusions (801mL total) and two fresh frozen plasmas (521 mL total), in addition to one ampoule of dexamethasone (2 mg). The HELLP syndrome severity, defined by the platelets count, decreased from class II to class I. The patient showed clinical and laboratory improvement at the hours following blood transfusion (Figure 1 and 2). After stabilization, she was transferred to gynaecologic ward and a strict pressure control was maintained (Table 2).

At the ward, the patient continued treating with methyldopa (500 mg 8/8h), maintaining hypertensive peaks and reporting nauseas and scotomas, making it necessary the use of hydralazine. After changing the prescription to captopril (50 mg 12/12 h), pindolol (10 mg 12/12 h) and nifedipine (20 mg if AP ≥ 160/110 mmHg), the pressure levels were still not controlled (170x100 mmHg). Nevertheless, there was a clinical improvement and the cardiology team assessment was requested (Day 6 - D6), which prescribed methyldopa (500 mg 8/8h), nifedipine (20 mg 8/8h) and pindolol (10 mg 12/12h). After a new anti-hypertensive regimen, the patient progressed asymptomatic and with improvement of pressure levels and laboratory results (Table 2).

At D9, a swelling at the edge of the surgical wound, with approximately 5 cm in diameter, a softened consistency and no phlogistic signs, and an USG was ordered. The test evidenced a fluid collection with a non-specific aspect, suggestive of hematoma (Figure 4).

At D10, after abdominal wall drainage (Figure 5), the patient was discharge of the hospital asymptomatic, with

Table 1. Laboratory tests according to day and hour of hospitalization.

Days	D1	D2		D3		D4	D5	D6	D7	D9
Hours	2h	15h	26h	45h	53h	69h	94h	118h	146h	194h
Complete blood count (CBC)										
Hb (g/dL)	12,8	7,5	6,5	10,3	11,3	–	10,3	10,7	10,4	11,6
Ht (%)	36,5	21,9	18,2	29,1	32,1	–	28,8	30,8	30,1	33,9
Leuk (cel/mm³)	19.631	–	22.060	21.480	16.980	–	10.860	10.340	11.070	10.220
Plat (cel/mm³)	58.000	–	35.000	31.000	34.000	40.000	65.000	79.000	111.000	204.000
Liver function										
AST (U/L)	56,0	–	–	20,0	–	–	26,0	43,0	–	23
ALT (U/L)	20,0	–	–	14,0	–	–	11,0	20,0	–	18
Bilir										
Dir (mg/dL)	0,34	–	–	0,09	–	–	0,30	–	–	0,18
Ind (mg/dL)	1,38	–	–	0,27	–	–	0,32	–	–	0,35
Renal function										
Ur (mg/dL)	72,0	–	–	60,0	–	–	41,0	36,0	–	17,0
Cr (mg/dL)	1,7	–	–	1,4	–	–	1,2	1,1	–	0,9
Cell lysis markers										
Ác úr (mg/dL)	6,1	–	–	3,1	–	–	–	–	–	2,4
LDH (U/L)	987,0	–	–	1.006	–	–	674,0	702,0	732,0	614,0
Coagulogram										
PT (s)	12,2	–	–	–	12,2	12,2	–	–	–	–
aPTT (s)	30,0	–	–	–	30,0	30,0	–	–	–	–
Analytical urine test and sedimentocopy										
Ptn (mg/dL)	30,0		neg	100			neg	tr	neg	neg
RBC (cell/mL)	180.000	–	71.000	2.500	–	–	20.000	215.000	1.000	2.000
Leuk (cell/mL)	94.000	–	42.000	10.000	–	–	3.000	16.000	3.000	3.000

Captions: D – days; h – hous; Hb – hemoglobin; Ht – hematocrit; Leuk – leukocytes; Plat – platelets; AST – aspartate aminotransferase; ALT – alanine aminotransferas; Bilir – bilirubins; Dir – direct bilirubin; Ind – indirect bilirubin; Ur – urea; Cr – creatinine; Ur ac – uric acid; LDH – lactate dehydrogenase; PT – prothrombin time; aPTT – activated partial thromboplastin time; Ptn – protein; RBC – red blood cells.

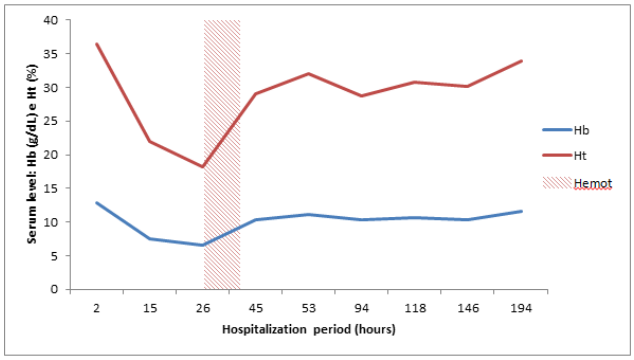
Table 2. Arterial pressure levels (mmHg) per hour and hospitalization period.

Hour	Days of hospitalization										
	D1	D2	D3	D4	D5	D6	D7	D8	D9	D10	D11
06:00	–	100/70	150/90	150/80	170/90	160/100	150/90	130/80	140/100	140/100	140/100
10:00	–	140/80	150/90	170/100	150/90	170/110	130/90	130/80	140/90	140/100	130/80
14:00	160/120	110/70	–	155/90	170/100	150/100	140/90	130/80	160/110	130/90	–
18:00	140/90	–	150/80	140/90	130/70	130/70	140/100	–	140/100	120/80	–
22:00	150/110	–	140/80	150/70	160/100	130/90	120/70	140/90	140/80	110/80	–

Captions: D – days; "systolic arterial pressure/diastolic arterial pressure" (mmHg) data.

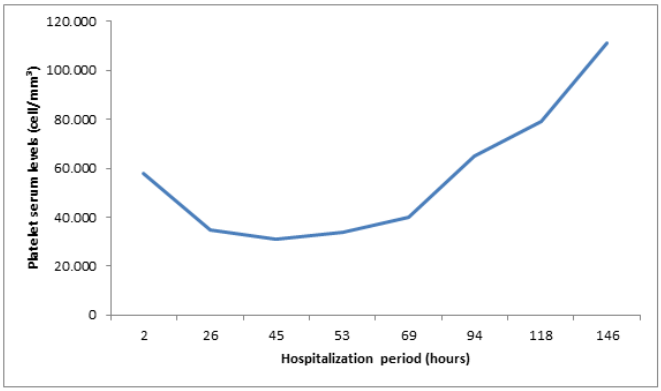
good health status and a significant clinical and laboratory improvement. A return to reassessment was requested, at first weekly, to the emergency department of this same hospital.

Figure 1. Evolution of the hematimetric numbers according to the hospitalization period.



Captions: Hb – hemoglobin; Ht – hematocrit; Hemot – hemotransfusion.

Figure 2. Evolution of the platelet levels according to the hospitalization period.



DISCUSSION

Women at the perimenopause have increased incidence of complications during pregnancy, such as PE and

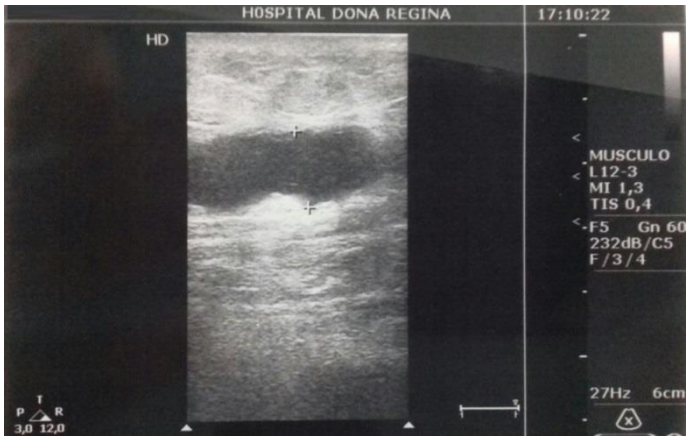


Figure 4. Elongated fluid collection in the anterossuperior abdominal wall, located right above the surgical bed, discreetly heterogeneous, measuring about 18.0 x 6.7 x 0.8 cm (volume = 50 cm^3).

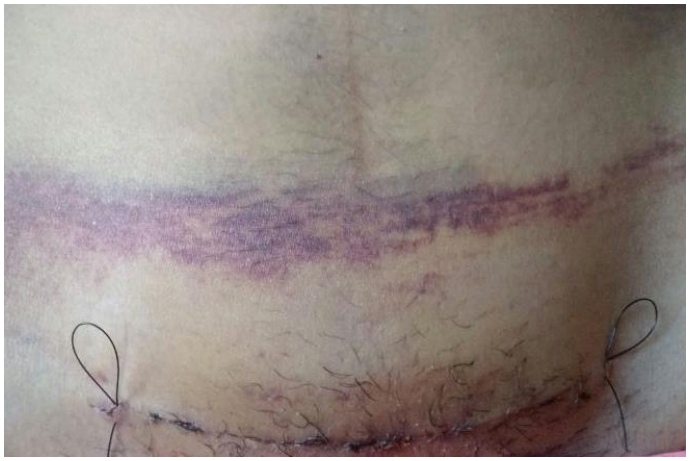


Figure 5. Hematoma in hypogastrium three days after discharge. Image provided by the patient.

HELLP syndrome[8]. This presentation of the severe PE originates from an abnormal placental development, which progresses with the production of factors that lead to endothelial injury through the activation of platelets and/or vasoconstrictor agents[4]. This patient, differently from the usual, was 19 years old.

On the other hand, being primigest is considered a risk factor to the HELLP syndrome[9,10], and the recurrence risk is 14-24%[11]. In face of headache, scotomas, nauseas and vomits, associated with arterial hypertension, the diagnostic hypothesis was set, which was confirmed in a few hours by laboratory tests, leading to the obtained therapeutic success. Literature says that, in light of this condition, the main goal must be the management of the patient's arterial pressure with anti-hypertensive therapy and the seizure control. The administered magnesium sulfate is recommended to these two functions[12]. The maintenance treatment with magnesium sulfate is suggested during the postpartum period because of its vasodilator effect on decreasing the maternal mortality[13].

This syndrome is frequently related to perinatal hypoxia, a severe complication that can lead to impairments throughout life[14]. Therefore, we can consider that the hypoxia, caused by both the syndrome pathogeny and the PA, led to the fetal death. The leukocytosis observed in D1 could be due to the current UTI, associated with the fetal death[15].

Although discrete, the transaminases are related to the hepatic involvement, associated with an impaired renal function and the cell lysis, evidenced by LDH elevation and a severe thrombocytopenia. The endothelial lesion of the liver vessels, followed by platelets activation, aggregation and consumption, resulting in ischemia and hepatocytes death, is the main hypothesis to explain the laboratorial picture of this syndrome[5].

CONCLUSION

The PA, along with the hemolysis characteristic of the HELLP syndrome, is consisted by the probable causes of hematocrit and hemoglobin decrease in the first hours of hospitalization. The blood transfusion enabled a significant improvement in the hematimetric values and the patient stabilization. Thus, due to a prompt medical attention, strict monitoring and adequate treatment, a therapeutic success was reached.

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