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Invited Editorial

Rare but should never be forgotten: HELLP syndrome

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HELLP syndrome (hemolysis, elevated liver enzymes, low platelets) is a rare but life-threatening condition along the spectrum of hypertensive disorders of pregnancy, It was first described as a severe complication of pregnancy in 1982 [1,2].

Hemolysis, elevated liver enzymes, low platelets (HELLP) is seen by many as one of the most severe fulminant consequences of hypertension of pregnancy outside of eclampsia. HELLP syndrome is rare, complicating only 0.5–0.9% of all pregnancies and some 10–20% pregnancies with severe preeclampsia [2]. Clinicians have observed HELLP alone and concomitant with other features of severe preeclampsia. Experts studying this condition have classified HELLP into complete and incomplete and others have developed diagnostic criteria [2]. A pregnant individual may experience symptoms like pain in the right upper quadrant, nausea, and epigastric pain, symptoms that can easily be mistaken as common discomforts of pregnancy. HELLP syndrome also needs to be distinguished from several other rarely seen conditions like acute fatty liver of pregnancy (AFLP), thrombotic thrombocytopenic purpura (TTP)/hemolytic uremic syndrome (HUS), SLE (systemic lupus erythematosus), and antiphospholipid syndrome (APS) [3].

Most cases of HELLP occur in the third trimester (28–40 weeks), but a review of the literature shows several case reports of HELLP prior to the third trimester, occurring as early as 15 weeks [4–6]. A retrospective analysis of HELLP syndrome cases diagnosed before 26 weeks of gestation reported a total of only 6 cases, all of whom had APLS [5]. Another retrospective case series at a large obstetric tertiary-care center found that about half of early-onset (second-trimester) HELLP cases were associated with APLS, with the earliest reported case occurring at 18 weeks [7].

Algorithms are utilized to make problem solving more efficient; they can be used to standardize the care given and to eliminate potential individual biases. While automating processes to make diagnosis more reliable, faster, and potentially easier to perform in medicine, some clinical illnesses are evolving processes. Early-career physicians may never have taken care of a patient with HELLP syndrome. As we move to an era of medicine where algorithms and artificial intelligence are used more to aid in diagnosis and clinical decision making, will the use of algorithms and artificial intelligence (AI) ensure that the rare but indeed deadly conditions are not forgotten? These conditions need to be high on our radar, we need to continue to teach about them, and we need to look for the zebra; we may in fact save a life.

In its most fulminant form, HELLP syndrome may lead to life-threatening consequences to the fetus and the pregnant individual. Without treatment, severe preeclampsia and its severest form, HELLP syndrome, can result in end-organ damage, liver rupture, kidney failure, cerebral hemorrhage, and even death. In the USA after the Dobbs v Jackson US Supreme court decision in 2022, there are now places where if a woman develops HELLP syndrome at an early gestational age healthcare providers may be conflicted about terminating pregnancy, the known treatment for this condition, in fear that this medical decision that may save her life but goes against the laws of the state in which they are practicing, based upon legal interpretations of what constitutes a danger to the mother's life.

The absolute known treatment for HELLP syndrome is delivery. When the gestational age is close to viability, clinicians may offer steroids to benefit the premature fetus and hold off delivery for 24 h for the infant. This is a judgement call and is made very carefully after reviewing the clinical data and trends of the laboratory values and other pertinent data. The condition is like a ticking time bomb and can worsen and lead to life-threatening complications at any time.

The pandemic has changed much in the world and since the COVID-19 pandemic authors have seen a COVID-linked HELLP-like syndrome (CLHLS). Clinical, laboratory, and microvascular pathophysiological similarities between the classic HELLP syndrome and the CLHLS have been observed [8].

It may be too much to expect that all rare conditions and circumstances can be accounted for in algorithms. One of my mentors used to constantly say "The eyes cannot see what the mind does not know." The importance of staying vigilant and on the lookout for these rare but

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deadly conditions of pregnancy is imperative.

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References

[1] L. Weinstein, Syndrome of hemolysis, elevated liver enzymes, and low platelet count: a severe consequence of hypertension in pregnancy, Am. J. Obstet. Gynecol. 142 (2) (1982 Jan 15) 159–167, https://doi.org/10.1016/s0002-9378(16)32330-4. PMID: 7055180.

[2] K. Haram, E. Svendsen, U. Abildgaard, The HELLP syndrome: clinical issues and management. A review, BMC Pregnancy Childbirth (9) (2009 Feb 26) 8, https://doi.org/10.1186/1471-2393-9-8. PMID: 19245695; PMCID: PMC2654858.

[3] B.M. Sibai, Imitators of severe preeclampsia, Obstet. Gynecol. 109 (4) (2007 Apr) 956–966, https://doi.org/10.1097/01.AOG.0000258281.22296.de (PMID: 17400860).

[4] S. Bae, L. Comfort, J. Ng, K. Sarkar, S. Pachtman, Early HELLP syndrome or catastrophic antiphospholipid syndrome? A diagnostic dilemma, Cureus. 15 (4) (2023 Apr 28) e38244, https://doi.org/10.7759/cureus.38244. PMID: 37252601; PMCID: PMC10225157.

[5] M. Pawelec, B. Palczynski, A. Karmowski, HELLP syndrome in pregnancies below 26th week, J. Matern. Fetal Neonatal Med. 25 (5) (2012 May) 467–470, https://doi.org/10.3109/14767058.2011.592875. Epub 2011 Aug 1. PMID: 21801142.

[6] E.L. Berry, S.N. Iqbal, HELLP syndrome at 17 weeks gestation: a rare and catastrophic phenomenon, J. Clin. Gynecol. Obstet. 3 (4) (2014 Dec) 147–150, https://doi.org/10.14740/jcgo297w. PMID: 25806101; PMCID: PMC4369395.

[7] Thuong D. Le Thi, N. Tieulié, N. Costedoat, M.R. Andreu, B. Wechsler, D. Vauthier-Brouzes, O. Aumaître, J.C. Piette, The HELLP syndrome in the antiphospholipid syndrome: retrospective study of 16 cases in 15 women, Ann. Rheum. Dis. 64 (2) (2005 Feb) 273–278, https://doi.org/10.1136/ard.2003.019000. PMID: 15647435; PMCID: PMC1755358.

[8] N.L.E. da Cunha Sobieray, M. Zanela, S.L. Padilha, C.F. Klas, N.S. de Carvalho, HELLP syndrome and COVID-19: A minor revision of a possible new "COVID-19-linked HELLP-like syndrome", Eur. J. Obstet. Gynecol. Reprod. Biol. 283 (2023 Apr) 90–94, https://doi.org/10.1016/j.ejogrb.2023.02.005. Epub 2023 Feb 11. PMID: 36812787; PMCID: PMC9922163.

Damali Campbell Oparaji

E-mail address: campbedm@njms.rutgers.edu.