The HELLP syndrome, originally described by Weinstein in 1982, includes signs of hemolysis (H), elevated liver enzymes (EL), and low platelet count (LP), and is a variant presentation of severe preeclampsia. Ophthalmic manifestations of preeclampsia include retinal arteriolar spasm, optic neuropathy, serous retinal detachment, and cortical blindness. Isolated cortical blindness has been thought to occur in only 1% to 3% of pregnancies complicated by preeclampsia-eclampsia. To date, there are no valid data about the coincidence of HELLP syn drome and cortical blindness. The coincidence of hemolysis, elevated liver enzymes, and low platelet count (HELLP) syndrome and cortical blindness is an uncommon but very dramatic event. We describe a case of HELLP syn drome complicating with acute cortical blindness before delivery. A 27-year-old woman, gravida 1, para 0, with normal medical history, was admitted to a local hospital at the 33th week of gestation due to headache, vomiting, and blurred vision. The ophthalmologic examination showed intact pupillary light reflexes and normal ophthalmoscopic findings, but no light perception in either eye. Brain computed tomography showed no mal findings. HELLP syn drome and preeclampsia was diagnosed based on the findings of hypertension and proteinuria as well as laboratory data. Prompt delivery was performed in order to achieve good maternal and neonatal outcomes.

**Case Report**

A 27-year-old woman, gravida 1, para 0, with normal medical history, was admitted to a local hospital at the 33th week of gestation because of headache, vomiting, and blurred vision. During admission, blood pressure was 134/90 mmHg. Physical and neurologic findings were normal. The ophthalmologic examination showed both pupils were equal in size and briskly reactive to light, but there was no light perception in either eye. Brain computed tomography showed no abnormality. Laboratory data showed...
nor mal liver en zymes, and then the pa tient was trans fer red to our hos pi tal for fur ther eval u a tion and man agement.

In our emer gency de part ment, the pa tient looked well de vel oped, well nour ished, fully alert, and showed no sub cuta ne ous edema. The blood pres sure was 155/97 mmHg and the urine rou tine showed 3+ pro tein. Re ex am i na tion of the eye fundi re vealed nor mal ap pear ance and pupillary func tion. Lab or a tory data on ad mis sion re vealed the fol low ing val u es: white blood cell count 9280/mm$^3$, he mo glo bin 12.7 g%, hemat - ocrit 37.7%, plate let count 66,000/mm$^3$, al bu min 2.9 g/dl, to tal bil i ru bin 2.4 mg/dl, aspartate transaminase (AST) 213 U/l, alanine transaminase (ALT) 224 U/l, lac tate dehydrogenase (LDH) 1052 U/l, creatinine 0.9 mg/dl, prothrombin time 12.0 sec onds (INR 0.95), and urine pro tein 6.5 g/24hrs. In view of these find ings, the di ag no sis of HELLP syn drome and pree clampsia was es tab lished.

The ini tial treat ment in cluded nifedipine 5 mg (s.l., qid prn.) to main tain blood pres sure in the nor mal range and mag ne sium sul fate 1 g/h (i.v.) for sei zure pro phy laxis. Prompt de liv er y by Ce sar ean sec tion was per formed in view of the find ings of HELLP syn drome as so ci ated with the rare com pli ca tion of cor ti cal blind ness. A 1,305 g male in fant was de liv er ed with Apgar scores of 6 and 9 at 1 and 5 min utes re spectively. The post op er a tive course of the pa tient was un event ful, her vi sion im proved grad u a lly about 10 hours af ter the op er a tion, and fi nally re turned to nor mal in 30 hours later. The ma ter nal ab nor mal he ma tologic find ings and liver en zymes re turned to nor mal within 5 days, and she was dis charged from our hos pi tal in good health on the 9th day.

Discussion

HELPP syn drome is a sep a rate en tity from pre eclampsia, and 4% to 12% of hy per ten sive dis eases of preg nancy may com pli cate by HELLP syn drome. In Sibai et al.’s se ries, HELLP syn drome occurred in 20% and 10% in pa tients with se vere pre eclampsia and eclampsia re spectively. Al though vi sual dis tur bances de vel op in per haps 25% of women with eclampsia, blind ness is rare, with a cited in ci dence of 1% to 3%. Nev er the less, Cunningham et al.’s se ries showed blind ness was much more com mon and ac counted for 15% of women with eclampsia. In the past, most cases of blind ness were at trib uted to no r mal ab nor mal i ties that in cluded edema, vas cu lar changes, and detach ment. More re cently, case re ports have placed em pha sis on the cor ti cal blind ness, which is char ac ter ized by intact pupillary light re flexes and nor mal ophthalmoscopic find ings. The most com mon clin ical pre sen ta tions in HELLP syn drome are right up per quad rant ab dom i nal pain or epigastric pain and nau sea or vom iting, from 36% to 65%. A sub set of pa tients ex hibit symp toms of se vere pre eclampsia, in clud ing head a che in 31% of the cases and vi sual dis tur bances in 10%. Preg nancies com pli cated by HELLP syn drome are as so ci ated with poor ma ter nal and fe tal out comes. The ma ter nal mor tal ity rate var ies sig nif i cant ly among the re ported se ries, but it is widely stated to be from 1.0% to 3.5%. The fe tus is sim ilar ly placed at in creased risk for mor tal ity and mor bid ity in the setting of HELLP syndrome, as HELLP syn drome fre quently oc curs be fore term, and prematurity it self sig nif i cant ly con trib utes to per inatal mor tal ity and mor bid ity. The peri natal mor tal ity rate ranges from 56 to 367 per 1,000 births.

There have been a num ber of case re ports of acute cor ti cal blind ness com pli cat ing pre eclampsia-eclampsia and many in cluded neu ro radi o logical as sess ment. Neu ro im aging find ings have ranged from nor mal to wide spread low-den sity ar eas, at trib uted to local ized ar eas of de creased per fu sion as so ci ated with ev i dence of arterial spasm, in farc tion, or ce re bral edema. Le sions of the oc cip i tal lobes are typ i cal and the basal gan glia, brain stem, and cer e bel lum are also com monly af fected. Clin ical re cov er y typ i cally pre ce des the nor mal iza tion of neu ro im aging find ings. These le sions are prob a bly stim u lated by dis par ity in ce re bral re gional blood flow that is char ac ter ized by vasospasm and di min ished flow pri mar ily af fect ing the pos te rior cir cu la tion. Re ports of CT and mag netic re so nance im aging (MRI) in pa tients with HELLP syn drome and pre eclampsia as so ci ated with cor ti cal blind ness range from nor mal to typ i cal find ings are bi lat eral sub cortical and cor ti cal oc cip i tal le sions with

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hypodensity on CT or hyper intensity on T2-weighted MRI. These neuroimaging findings are similar to those of patients with cortical blindness complicating preeclampsia-eclampsia with HELLP syndrome. There is evidence that these brain lesions are induced by vascular changes. Doppler velocimetry and cerebral angiography in patients with HELLP syndrome and preeclampsia associated with cortical blindness demonstrate generalized vasospasm, with a greater sensitivity of the occipital lobes to the deleterious effects of vasospasm and ischemia. Besides, vascular endothelial dysfunction also contributes to the underlying mechanism.

The management of preeclampsia-eclamptic patients, whether they are complicated with cortical blindness or not, includes magnesium sulfate for seizure prophylaxis, control of blood pressure, and fluid restriction to avoid worsening of cerebral edema. Maternal condition as well as the prematurity of delivery are the main contributing factors for termination.

In this case of a primigravida with HELLP syndrome and preeclampsia as so ciated with cortical blindness, prompt delivery by Cesarean section gave a favorable outcome as in previous reports.

References