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Severe preterm preeclampsia – associated posterior reversible encephalopathy syndrome

Teška preeklampsija u nedonošenoj trudnoći, uz pojavu reverzibilnog encefalopatičnog sindroma

Sandra Benzon, Zdeslav Benzon, Luka Vulić, Marko Vulić*

Summary

Posterior reversible encephalopathy syndrome (PRES) is a clinical syndrome which causes non-specific neurological symptoms such as visual impairment (cortical blindness, diplopia, hemianopia), acute headaches, seizures (focal or general tonic-clonic), vomiting, altered mental status, focal neurologic deficit as a complication of preeclampsia. Preeclampsia is a serious complication specific for pregnancy, characterized by hypertension (systolic > 140 mm Hg and/or diastolic > 90 mm Hg) and proteinuria (> 300 mg u 24-h urine sample , > 1 +) at or after 20th week of gestation. It is one of the leading causes of perinatal morbidity and mortality. We present a 38-year-old pregnant woman, gravida 1 para 0 with PRES which was manifested on the second day after delivery by Cesarean section at 32 weeks of gestation with aphasia and blindness caused by severe preeclampsia. We confirmed the diagnosis by MRI which revealed white matter edema in the posterior cerebral area in a symmetric fashion, hyper intense cortical foci in the right occipital lobe and punctiform lesions in the bilateral occipital lobe. We treated her promptly in the intensive care unit (ICU) with antihypertensive and anticonvulsant therapy after which she fully recovered. PRES should always be kept in mind when a patient in the postpartum period develops one of the above symptoms. If timely recognized and promptly treated, full recover usually follows.

Key words: PRES, severe preeclampsia, early postpartum period, blindness, aphasia

Sažetak

Posteriorni reverzibilni, encefalopatični sindrom (PRES) klinički je sindrom karakteriziran nespecifičnim neurološkim simptomima, kao što su smetnje vida (sljepilo, dvostruki vid, gubitak polovine vidnoga polja), akutna glavobolja, epileptički napadaji (fokalni ili generalizirani toničko-klonički grčevi), povraćanje, izmijenjena stanja svijesti, fokalni neurološki deficiti, u ovom slučaju preeklampsije kao komplikacija. Preeklampsija (PE) je poremećaj specifičan za trudnoću, karakteriziran pojavom hipertenzije (sistolički tlak > 140 mm Hg i/ili dijastolički tlak > 90 mm Hg) udružene s proteinurijom (> 300 mg u 24-h urinu, > 1 +) koji nastaje nakon 20. tjedna gestacije u prethodno normotenzivnih žena, bez proteinurije. Vodeći je uzrok maternalnog i perinatalnog morbiditeta i mortaliteta. U ovom radu prikazujemo slučaj 38-godišnje trudnice, prvorotke, sa simptomima PRES-a koji su se manifestirali gubitkom sposobnosti govora i sljepoćom drugog dana nakon poroda carskim rezom, koji je urađen zbog teške preeklampsije s točno 32 tjedna gestacije. Dijagnozu smo potvrdili magnetskom rezonancom (MR) koja je pokazala simetričan edem bijele tvari u posteriornim područjima mozga, područja hiperintenziteta u desnom okcipitalnom režnju i punktiformne lezije obostrano okcipitalno. Bolesnicu smo odmah prebacili u jedinicu intenzivne njege, gdje je tretirana antihipertenzivnom i antikonvulzivnom terapijom, nakon koje se za nekoliko dana u potpunosti oporavila. Treba uvijek razmišljati o PRES-u kada bolesnica u razdoblju nakon poroda razvije jedno od gore navedenih simptoma. Ukoliko se na vrijeme prepozna i tretira obično uslijedi potpuni oporavak.

Ključne riječi: PRES, teška preeklampsija, rano poslijeporođajno razdoblje, sljepoća, nijemost

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* **Split University Hospital** and **University of Split, Croatia,** Department of Obstetric and Gynecology (Sandra Benzon, MD, PhD; Zdeslav Benzon, MD, prof. Marko Vulić, MD, PhD); **Health Centre Split** (Luka Vulić, MD)

Correspondence address / *Adresa za dopisivanje:* Sandra Benzon, Department of obstetric and gynecology, University hospital Split, Spinčićeva 1, 21 000 Split, Croatia. E-mail: sbenzon68@gmail.com

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Introduction

The posterior reversible encephalopathy syndrome (PRES) is a condition characterized by acutely altered mentation, drowsiness or sometimes stupor, visual impairment (e. g. visual hallucinations, cortical blindness, hemianopia, qudrantanopia, and diplopia), seizures, and sudden or constant, non-localized headaches.[1] The PRES acronyme derives from findings on magnetic resonance imaging (MRI) including white matter edema found in the posterior cerebrum, and the fact that symptoms are reversible, provided that the syndrome is recognized and treated promptly.[2] The pathophysiology of PRES is still not fully understood, although people who are at risk for developing PRES tend to have one or more risk factors, such as: preeclampsia, hypertension, renal failure, liver disease, autoimmune disorders and sepsis.[2,3,4] The most common cause is uncontrolled drug resistant hypertension.[5] Preeclampsia also is acknowledged to be an important cause of PRES.[6] It affects people of all ages, from children to old adults, but most frequently young or middle age females which might be attributable to etiological aspects. Since the syndrome is significantly underdiagnosed, epidemiological data should be interpreted with caution.[1] PRES seems to be linked to failure of cerebral blood flow autoregulation, the brain's ability to maintain constant cerebral blood flow over a range of blood pressure via the constriction or dilatation of the cerebral blood vessels.[2] The second theory regarding the cause of PRES is that the syndrome is triggered by endothelial dysfunction caused by circulating endogenous or exogenous toxins.[1] The third theory include a possible pathological activation of the immune system.[4]

Case report

The patient has provided informed consent for the publication of the case. A previously healthy pregnant 38-year-old woman (gravida 1 para 0) presented to our hospital at 29th week of gestation with preeclampsia. The family history showed that both her mother and father suffered from hypertension. She was treated with methyldopa and amlodipin. Despite the therapy of methyldopa 2g and amlodipin 10 mg per day the blood pressure on 32nd week of gestation was elevated, up to 180/110 mm/Hg with proteinuria 2g/24 hour. A lab panel showed slightly elevated liver enzymes and low proteins (AST 56 U/L, ALT 63 U/L, total protein 50 g/L) while other parameters were normal. Fetal monitoring with cardiotocography showed signs of fetal distress. She received steroids (dexamethason 8 mg per two days) for fetal lung maturation and magnesium (4g intravenous over 30 minutes then 2 g/hour next 12h) for fetal brain neuroprotection. Considering drug resistant hypertension on exactly 32nd week of gestation, she underwent an emergent uncomplicated Cesarean section and she delivered a female baby, 1440 g weight and 36 cm long. Apgar score in the first minute was 5 and the baby was transferred to the Neonatal Ward. Two days after Cesarean section, she experienced a sudden consciousness disturbance which was followed by amaurosis, aphasia, perseveratio, agitatio. The patient's vital signs included high blood pressure, 180/110 mm/Hg although antihypertension therapy was administred the whole time. Heart rate was 120 bpm, respiration rate was12 breaths per minute, SpO_2 was 91%, and body temperature was 37ºC. A lab panel showed slightly elevated liver enzymes and low proteins (AST 70 U/L, ALT 69 U/L, total protein 52 g/L). The diagnosis of PRES was considered. She was transferred to the Intensive Care Unit of our hospital for further management. Immediate rescue measures included: oxygen administration, urapidil 25 mg intravenous bolus, followed by continuous intravenous drip 25 mg/hour, magnesium sulfate 5g intravenous bolus, followed by continuous intravenous drip 1.5 g/hour, dexamethason 8 mg intravenously and levetiracetam 500 mg twice daily. Brain T2-weighted MRI showed hyperintense cortical foci in the right occipital lobe and punctiform lesions in the bilateral occipital lobe (Figure 1). Cardiologic investigations did not show remarkable findings. Her neurological, visual and speech functions gradually improved after 24 hours and the blood pressure stabilized so she continued undergoing antihypertension therapy, methyldopa 2 g and amlodipin 10 mg per day. Twelve days after delivery, she was discharged in good condition with stabile blood pressure and reduced dosage of methyldopa 1 g and amlodipin 5 mg per day.

Discussion

Posterior reversible encephalopathy syndrome is a severe condition that has been underdiagnosed, but owing to the improvement in the quality of imaging, understanding of PRES has improved.[7] In the case described here, the patient showed neurological disorder, second day after undergoing Cesarean section, especially visual impairment and aphasia that lasted for nearly 24 hours, which was connected with preeclampsia. Patients with preeclampsia and PRES may have other abnormalities such as liver enzymes irregularities, decrease of platelets which were not observed in the present case.[2] After diagnostic and therapeutic procedures, the diagnosis of PRES was established. Brain MRI is the most suitable diagnostic tool.[8] It was important that PRES was differentiated

from other diseases such as ischemic stroke and thrombosis of the cerebral venous sinus since it is the most frequent cerebrovascular disorder in the puerperium.[9] The diagnosis has important therapeutic and prognostic implications because the reversibility of the clinical and radiologic abnormalities is contingent on the prompt control of blood pressure and/or discontinuing of the offending drug.[10] Admission to a critical care unit is required in about 40% of patients due to complicating conditions including status epilepticus, cerebral vasoconstriction, ischemia or intracerebral hemorrhage. Prognosis is favorable; in the majority of patients neurological deficits and imaging findings resolve completely.[1] Hypertension has to be treated using antihypertensive drugs and magnesium sulphate should be used to prevent seizures.[7] Delayed treatment might result in permanent brain damage so awareness and education are cornerstones for the proper management of PRES, which should be kept in mind in any patient presented with neurological symptoms in the postpartum period.

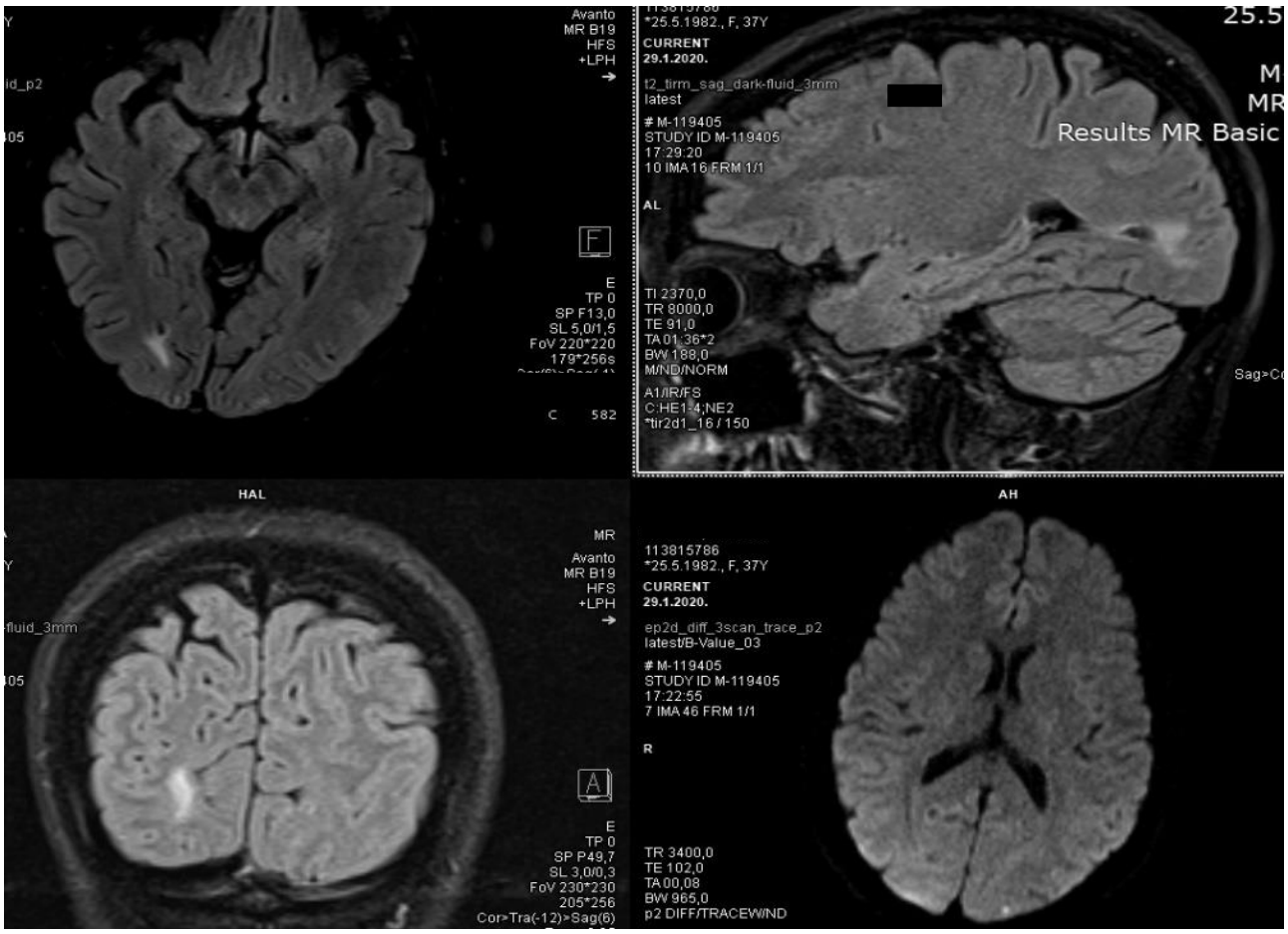

Figure 1 Brain MRI demonstrate hyperintensities in the occipital cortex
MRI – magnetic resonance imaging
Slika 1. MR mozga s hiperintenzitetima u okcipitalnom korteksu
MR – magnetska rezonanca

References

1. Fischer M, Schmutzhard E. Posterior reversible encephalopathy syndrome. J Neurol 2017;264:1608-16.
2. Hobson EV, Craven I, Blnk SC. Posterior reversible encephalopathy syndrome: a truly treatable neurologic illness. Perit Dial Int 2012;32:590-94.
3. Chowdhary M, Kabbani AA, Tobey D, Hope TD. Posterior reversible encephalopathy syndrome in a woman with focal segmental glomerulosclerosis. Neuropsychiatr DIS Treat 2015;11:1111-14.
4. Garner O, Ramirez A, Iardino A. A case of posterior reversible encephalopathy syndrome associated with sepsis. BMJ Case Rep 2018;10:2018.
5. Fugate JE, Claassen DO, Cloft HJ, Kallmes DF, Kozak OS, Rabinstein AA. Posterior reversible encephalopathy syndrome: associated clinical and radiologic findings. Mayo Clin Proc 2010;85:427-32.

6. McDermott M, Miller EC, Rundek T, Hurn PD, Bushnell CD. Preeclampsia: Association with posterior reversible encephalophaty syndrome and stroke. Stroke 2018;49:524-30.
7. Sundin CS, Johnson ML. Posterior Reversible Encephalopathy Syndrome. MCN Am J Matern Child Nurs 2018;43:77-82.
8. Garg RK, Kumar N, Malhotra HS. Posterior reversible encephalopathy syndrome in eclampsia. Neurol India 2018;66:1316-23.
9. Minnerup J, Kleffner I, Wersching H, et al. Late onset postpartum eclampsia: it is really never too late- a case of eclampsia 8 weeks after delivery. Stroke Res Treat. 2010;2010:798616.
10. Lamy C, Oppenheim C, Mas JL. Posterior reversible encephalopathy syndrome. Handb Clin Neurol 2014; 121:1687-701.
11. Engelter ST, Provenzale JM, Petrella JR. Assessment of vasogenic edema in eclampsia using diffusion immaging. Neuroradiology. 2000;42:818-20.