Postpartum Posterior Reversible Encephalopathy Syndrome (PRES) in a Patient with Severe Preeclampsia: Case Report

Şiddetli Preeklampsi Olgusunda Postpartum Posterior Reversibl Ensefalopati Sendromu (PRES)

Hüseyin Levent KESKİN,[a]
Gamze UYANIK,[a]
Ahmet Akın SİVASLIOĞLU,[a]
Emine ÇELEN,[a]
Ayşe Filiz AVŞAR[a]

[a]Clinic of Obstetrics and Gynecology,
Ankara Atatürk Training and Research Hospital, Ankara

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Yazışma Adresi/*Correspondence:*
Gamze UYANIK
Ankara Atatürk Training and Research Hospital,
Clinic of Obstetrics and Gynecology, Ankara,
TÜRKİYE/TURKEY
gamze_u@hotmail.com

ABSTRACT Posterior reversible encephalopathy syndrome (PRES) is typically characterized by the vasogenic edema in the occipitoparietal area of the brain in clinical and radiological findings. Various factors accounts for the etiology and the symptoms include sudden loss of vision, seizures, headache, and mental status alterations. In this report, we present a 27-year-old primigravida at 34 weeks 6 days of pregnancy who admitted to our hospital with high blood pressure (BP) and headache. The patient was diagnosed preeclampsia with respect to clinical findings and laboratory results, and the baby was delivered by cesarean section because of severe preeclampsia and fetal distress at the 3rd hour of admission to the hospital. In the postoperative period, high BP did not respond to multiple antihypertensive treatment and headache continued. On the 4th day of the follow-up sudden loss of vision developed in the patient, however fundoscopic examination was normal. In the cranial magnetic resonance imaging, some findings interpreting as PRES were detected in bilateral occipital lobes symmetrically, in both of the basal ganglia and left thalamus, and in cortical and subcortical white matter areas. The recovery without any sequelae was occurred with antiedema treatment applying immediately beside antihypertensive therapy. As a result, PRES should be considered in differential diagnosis in preeclamptic cases who has clinical findings supported by radiological methods and the treatment should be immediately administrated.

Key Words: Pre-eclampsia; vision disorders; posterior leukoencephalopathy syndrome

ÖZET Posterior Reversibl Ensefalopati Sendromu (PRES) etiyolojisinde çeşitli faktörler rol alan, ani gelişen görme kaybı, epileptik nöbet, baş ağrısı ve mental durum değişikliği ile birlikte görülebilen, tipik olarak beynin oksipitopariatel bölgesinde vazojenik ödem ile karakterize klinik ve radyolojik bulguları olan bir sendromdur. Bu yazıda, 27 yaşında, primigravid, 34 hafta 6 gün'lük gebeliği mevcut iken baş ağrısı ve kendi ölçtüğü kan basıncı (KB) değerinin yüksek olması nedeniyle hastanemize başvuran olgu sunulmuştur. Yapılan muayene ve tetkikler sonucunda hastaya preeklampsi tanısı konulmuş ve başvurusunun 3. saatinde şiddetli preeklampsi ve fetal distress endikasyonları ile sezaryen ile doğum gerçekleştirilmiştir. Hastanın postoperatif takiplerinde KB yüksekliği ve baş ağrısı devam etmiş çoklu antihipertansif tedaviye yanıt vermemiştir. Takibinin 4. gününde ani görme kaybı gelişen ve fundoskopik muayenesi normal olan hastanın Kranial Manyetik Rezonans görüntülemesinde özellikle bilateral oksipital lobda, her iki bazal ganglionda, talamusta, kortikal ve subkortikal beyaz cevher alanlarında PRES lehine yorumlanan bulgular dikkati çekmiştir. Antihipertansif tedavi yanında, hızla hastaya uygulanan antiödem tedavi ile sekelsiz iyileşme sağlanmıştır. Sonuç olarak, preeklamptik bir olguda klinik bulguların varlığı durumunda ayırıcı tanıda mutlaka PRES'ten şüphelenilmesi, radyolojik yöntemlerle tanının desteklenmesi ve tedavinin hızla uygulanması gerekir.

Anahtar Kelimeler: Pre-eklampsi; görme bozuklukları; posterior lökoensefalopati sendromu

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Posterior reversible encephalopathy syndrome (PRES) is typically characterized by the vasogenic edema in the oksipitopariatel area of the brain in clinical and radiological findings. Various factors involve in the etiology and the symptoms include sudden loss of vision, seizures, headache, and mental status changes.[1] Currently defined etiology of the syndrome include preeclampsia/eclampsia, HELLP syndrome, hypertensive encephalopathy, renal failure, cytotoxic agents, collagen vascular diseases (systemic lupus erythematosus, hemolytic uremic syndrome, thrombotic thrombocytopenic purpura), leukemia and lymphoma.[1-6] In this paper, we present a case of PRES caused by severe preeclampsia present with sudden loss of vision in the postpartum period, and emphasize the importance of early diagnosis and treatment in recovery without permanent damage.

CASE REPORT

A 27 years old, primigravida, with gestation of 34 w 6 d by the date of the last menstruation was admitted to the emergency department with complaints of headache and high blood pressure (BP) according to her own measurement (190/100 mmHg). The patient had no obstetric or maternal complications during routine pregnancy checks in the history. We learned that her BP was measured once 160/100 mm Hg, but later 120/70 mmHg after resting for half an hour in the last antenatal control in the 33rd week of gestation. In the emergency department, the first measured value of BP was recorded 160/100 mm Hg. On physical examination, pretibial edema (++) was detected, whereas there was no neurological deficit. In the obstetric ultrasonography, biometric measurements compatible with 35 weeks of gestation, amniotic fluid index of 120 mm, and single, live fetus were observed.

Electronic fetal electronic monitoring indicated loss of fetal heart rate variability and in spot urine examination of the patient (+++) proteinuria was detected. Laboratory tests were platelet count 257 000/mL, hemoglobin 10.7 g/dL, ALT: 6 U/L, AST: 21 U/L, LDH: 352 U/L respectively, indicating that the LDH value was slightly higher. The patient was given 10 mg of nifedipine sublingually. Because loss of variability continued in the FEM, and BP proceeded > 160/110 mmHg, emergency caesarean section was decided at the 3rd hour of admission with indications of severe preeclampsia and fetal distress. A girl baby, approximately 2300 grams at birth was delivered. Apgar score of 1st minute was 3 and Apgar score of 5th minute was 6 respectively and the infant was hospitalized in the neonatal intensive care service. No additional problems were encountered during caesarean section except enduring high BP. Postpartum BP of the patient was > 180/120 mmHg and alpha-methyl dopa (4x250 mg) and nifedipine (3x10 mg) treatment were started.

On the 2nd day postpartum urine output was > 11 000 cc/24 hour, platelet count, liver and kidney functions were normal and the amount of protein in the urine was 12.2 g/day. The doses of antihypertensive drugs were increased (alpha-methyl dopa 4x 500 mg and 60 mg controlled-release nifedipine) because the BP values endured over 160/110 mm Hg despite the current antihypertensive treatment. Upon the patient's complaints of severe pain in the head and back of neck and enduring high BP, $MgSO_4$ (2 g/h) infusion was started and continued for 24 hours for prophylaxis. Renal artery Doppler examination, echocardiography, fundoscopic fundus examination were performed and a 24-hour urinary catecholamine levels were studied to investigate the etiology of treatment-resistant hypertension. None of these tests revealed pathological findings. On the 4th day postpartum the patient that she had loss of sight when she woke up. Neurological and fundoscopic examinations were normal, natural light reflex was positive bilaterally and she had no change in mental and motor status. In the brain MR imaging of the patient we detected patchy and punctate contrast involvements showing restriction of diffusion in places in bilateral occipital lobes symmetrically, both the basal ganglia and left thalamus, cortical and subcortical white matter areas in the left frontal plane were noted and these findings were interpreted to represent PRES (Figures 1, 2). Treatment of cerebral edema (mannitol, dexamethasone

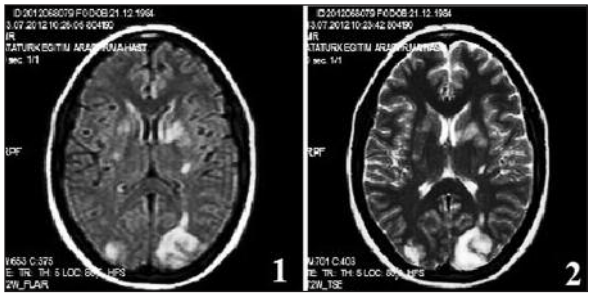

FIGURE 1, 2: In MR-T2A and FLAIR screening; more pronounced in the left occipital lobe of both compatible with vasogenic edema observed hyperintense signal recordings. It also signals the basal ganglia and periventricular white matter are the same.

and low-molecular-weight heparin) started and intravenous glycerol trinitrate (15 mcg/min) infusion started due to BP values enduring higher than 160/100 mmHg. On the 1st day of anti-edema treatment (5th day postpartum) the patient begun to see again, and was able to count fingers at 1 meter even if full clarity. On the 2nd day of the anti-edema treatment (6th day postpartum) the patient begun to see clearer and BP values were controlled (<140/90 mmHg). On the follow-up of no convulsive seizures developed and liver function tests and platelet levels were normal. The antihypertensive treatment with glyceryl trinitrate was completed in 24 hours, whereas the anti-edema treatment was continued for 7 days. On the 12th day postpartum the patient's sense of sight is completely normal and BP was 130/80 mmHg and she was discharged with oral antihypertensive therapy.

DISCUSSION

PRES is a syndrome characterized by headache, mental status changes, epileptic seizures and visual disturbances and was defined by Hincley et al. for the first time in 1996.[1] Pathophysiology of PRES is not known clearly, but two possible hypotheses have been proposed. First hypothesis is that, it may occur as a result of vasospazm due to acute high blood pressure. Ischemia and cytotoxic edema are thought to occur due to sympathetic nerve stimulation and cerebral vasoconstriction after acute episodes of hypertension.[7-9] The second hypothesis is a more widely accepted hyperperfusion theory. According to this theory, the brain tries to maintain a constant cerebral blood flow by autoregulation and the effect of sympathetic system and arteriolar system play significant role.[10] As a result of loss of autoregulation vasoconstriction which occur in order to protect the brain after a rise of systemic arterial blood pressure can not be sustained and arteriolar dilation and endothelial dysfunction develop. In cases of eclampsia-related PRES, maternal endothelial dysfunction, thought to be due to the secretion of trophoblastic cytotoxic factors, plays a role.[11,12]

In this case, deterioration of the blood-brain barrier and endothelial dysfunction allows extravasation of plasma, macromolecules and even erythrocytes and causes vasogenic edema.[4,8] The weaker sympathetic innervation of posterior cerebral arterial circulation is considered to be the reason for lesions to be seen more commonly in the occiptoparietal area.[13]

Radiological methods are quite effective and decisive in the diagnosis of the syndrome. PRES is characterized by the presence of bilateral cortical-subcortical hyperintense lesions in posterior cerebral regions in the Cranial MR imaging.[14] In more severe cases lesions may be seen in the brain stem, cerebellum, basal ganglia and frontal lobes.[15] In our case, the involvement of bilateral occipital lobe, both basal ganglia, periventricular white matter, thalamus, cortical and subcortical white matter areas indicates severe disturbance in cerebral autoregulation. The MRI findings explicitly show that the term "posterior" in the definition of the syndrome is not sufficient, thereby showing lesions which occur in other parts of the brain (white and gray matter-the brain base).[16]

In the differential diagnosis of hypertensive patients who developed PRES encephalopathy, cerebral venous thrombosis, bilateral posterior lobe infarcts, encephalitis, cerebral vasculitic involvement, electrolyte imbalance, and if the patient is pregnant, preeclampsia/HELLP syndrome are of primary significance. Visual disturbances during PRES include a wide range of symptoms varying from loss of sharpness of vision to homonymous hemianopsy and cortical blindness, and the blind-

ness which is characterized by normal pupil reflex and fundoscopic examination turns back generally in 4 hours and 8 days.[7,17] In our case, PRES was present with severe headache and cortical visual impairment on postpartum 4th day after cesarean section. At the 24th hour of the treatment the patient begun to see blurry and her vision improved totally at the 48th hour. The differential diagnosis was performed through the fundoscopic examination, MR imaging and laboratory tests. The diagnosis of PRES was confirmed by differentiating ischemia and vasogenic edema.

The first step in the treatment of PRES is the etiological causes which are the factors triggered the event. In our case; the etiologic factor is severe preeclampsia and hypertension which can not be controlled in postoperative period. In the treatment of hypertension; intravenous and oral antihypertensive agents, diuretics, sedatives and hypnotics can be used. We used alpha-methyl dopa, nifedipine and glyceryl trinitrate as antihypertansive agents. In addition to antihypertensive therapy the anti-edema treatment is started immediately (mannitol, dexamethasone and low-molecular-weight heparin.) In early treatment we used alpha-methyl dopa and nifedipine, but we could not controlled the blood pressure so in hypertensive crisis we used nitroglyserin. The other agents like hydralazin and nitroprusside can also be used but in Turkey, unfortunately, is not present any hydralazin preparation. Half life of sodium nitroprusside is so short but there are some serious side effects like serious hypotension and thiocyanate toxicity. Lowering BP too rapidly may produce cerebral ischemia, stroke or coma. Coronary blood flow, renal perfussion also may deteriorate, resulting in acute renal failure and myocardial infarction.

Because of permanent brain damage and neurological complications such as chronic epilepsy sequelae may result, early detection and treatment is very important. Although remission is possible by providing the autoregulation again; possibility of reversibility could change according to underlying diseases, location and characteristics of the lesions in MRI signal. The typical cortical and subcortical PRES lesions showed reversibility, whereas the brain stem and deep white matter lesions showed less reversibility. PRES due to eclampsia showed maximum reversibility on follow-up imaging compared to PRES with a background of immunosuppressive drugs and hypertension.[18]

PRES may become fatal despite the rapid and effective treatment. It should be considered in the differential diagnosis in presence of severe headache, sudden loss of sight and seizures or altered mental status with high BP particularly in cases with pre-eclampsia, eclampsia and HELLP syndrome and should be noted that rapid diagnosis and treatment of PRES may prevent haemorrhage, infarction, and permanent brain damage.

REFERENCES

1. Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A, et al. A reversibl posterior leukoencephalopathy syndrome. N Engl J Med 1996;334(8):494-500.
2. Bartynski WS. Posterior reversible encephalopathy syndrome, part 1: fundamental imaging and clinical features. AJNR Am J Neuroradiol 2008;29(6):1036-42.
3. Pratap JN, Down JF. Posterior reversible encephalopathy syndrome: a report of a case with atypical features. Anaesthesia 2008; 63(11):1245-8.
4. Ahcar SK, Shetty N, Joseph TT. Posterior reversible encephalopathy sendrome at term pregnancy. Indian J Anaesth 2011;55(4):399-401.
5. Brubaker LM, Smith JK, Lee YZ, Lin W, Castillo M. Hemodynamic and permeability changes in posterior reversible encephalopathy syndrome measured by dynamic susceptibility perfusion-weighted MR imaging. AJNR Am J Neuroradiol 2005;26(4):825-30.
6. Mukherjee P, McKinstry RC. Reversible posterior leukoencephalopathy syndrome: evaluation with diffusion-tensor imaging. Radiology 2001;219(3):756-65.
7. Stott VL, Hurrell MA, Anderson TJ. Reversible posterior leukoencephalopathy syndrome: a misnomer reviewed. Intern Med J 2005; 35(2):83-90.
8. Vaughan CJ, Delanty N. Hypertensive emergencies. Lancet 2000;356(9227):411-7.
9. Rangi PS, Partridge WJ, Newlands ES, Waldman AD. Posterior reversible encephalopathy syndrome: a possible late interaction between cytotoxic agents and general anesthesia. Neuroradiology 2005;47(8):586-90.
10. Akgün N, Karaman M, Başyiğit S, Yılmaz H, Özcan AA. [Posterior reversible encephalopaty sendrome-2 case report]. İstanbul Tıp Dergisi 2010;11(2):82-5.
11. Schwartz RB, Feske SK, Polak JF, DeGirolami U, Iaia A, Beckner KM, et al. Preeclampsia eclampsia: clinical and neuroradiographic correlates and insights into the pathogenesis of hypertensive encephalopathy. Radiology 2000;217(2):371-6.

12. Savvidou MD, Hingorani AD, Tsikas D, Frölich JC, Vallance P, Nicolaides KH. Endothelial dysfunction and raised plasma concentrations of asymmetric dimethyl arginine in pregnant women who subsequently develop pre-eclampsia. Lancet 2003; 361(3968):1511-7.
13. Schwartz RB, Mulkern RV, Gudbjartsson H, Jolesz F. Diffusion-weighted MR imaging in hypertensive encephalopathy: clues to pathogenesis. AJNR Am J Neuroradiol 1998;19(5): 859-62.
14. Demirtaş Ö, Gelal F, Vidinli BD, Demirtaş LO, Uluç E, Baloğlu A. Cranial MR imaging with clinical correlation in preeclampsia and eclampsia. Diagn Interv Radiol 2005;11(4):189-94.
15. Narbone MC, Musolini R, Granata F, Mazzù I, Abbata M, Ferlazzo E. PRES: posterior or potentially reversibl encephalopathy syndrome? Neurol Sci 2006;27(3):187-9.
16. Casey SO, Sampaio RC, Michel E, Truwit CL. Posterior reversible encephalopathy syndrome: utility of fluid-attenuated inversion recovery MR imaging in the detection of cortical and subcortical lesions. AJNR Am J Neuroradiol 2000;21(7):1199-206.
17. Thackeray EM, Tielbor MC. Posterior reversible encephalopathy syndrome in a patient with severe preeklampsia. Anesth Analg 2007;105(1):184-6.
18. Pande AR, Ando K, Ishikura R, Nagami Y, Takada Y, Wada A, et al. Clinicoradiological factors influencing the reversibility of posterior reversible encephalopathy syndrome: a multicenter study. Radiat Med 2006;24(10):659-68.