Posterior Reversible Encephalopathy Syndrome in a young woman:A rare case report

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Abstract: Posterior reversible encephalopathy syndrome**(**PRES**)** is a clinicoradiologic entity that characterized by variable associations of seizure activity ,consciousness impairment, headache, nausea/vomiting and visual abnormalities**.**It has certain characteristic radiological features, which allow diagnosis by the clinical findings .PRES has been associated with chronic renal disease,preeclampsia,eclampsia,SLE and administration of immunosuppressive agents.
Case Report: A 25-year-old woman was hospitalized due to headache, nausea/vomiting, acute visual disturbance and consciousness impairment (lethargy).She was developed generalize tonic clonic seizure. She had been admitted several times over a 5 month period with kidney stones and renal failure, so she was undergoing dialysis program because of ESRD. Her initial blood pressure was 150/90 mmHg. Bilateral pupils mydriasis with weak light reflex and bilatral papilledema were detected. In motor examination, left limbs movements were slower than right limbs and Bilateral up plantar **r**eflexes were detected .Admission Bun:54 and Cr:7.8 but the other laboratory data were in normal ranges **.**MRI findings demonstrated bilateral and symmetrical high signal intensity change in white matter of occipital and parietal lobes.
Conclusion: The pathogenesis of PRES is based on theory that the elevated blood pressure above patient`s baseline ,exceeds the cerebral vascular auto regulatory abilities, results in vasogenic edema in brain parenchyma and the posterior sub cortical white matter is commonly involved. In many cases also in our case, clinical and radiological recovery of PRES seems to be occure within a period of days to weeks, after removal of the inciting factor and control of blood pressure .

Key words: *PRES, ESRD, Seizure*

Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinical radiographic syndrome of heterogeneous etiologies that increasingly recognized neurological disorder with characteristic MRI findings. The global incidence of PRES is unknown .It has been reported in patients aged 2 to 90 years, although most cases occur in young to middle-aged adults and there is a marked female predominance.[1, 2]

PRES is characterized by headache, nausea, vomiting, seizure ,visual disturbances and confusion.[3, 4]

PRES is commonly associated with a sudden increase in blood pressure (BP).[4] elevated pressure exceeds the cerebral vascular autoregulatory abilities ,resulting in breakthrough vasodilatation of the cerebral arterioles. This results in vasogenic edema in the brain parenchyma. The posterior cerebral arteries, being sparsely innervated by sympathetic fibers, are more susceptible to blood pressure elevation and the occipital subcortical white matter is commonly involved. Adjacent gray matter may be involved, and extension into the frontal lobes is also not uncommon. Isolated frontal lobe involvement is rare.[3]

several disease conditions associated with PRES include hypertension(malignant), chronic renal failure(uremia),preeclampsia, eclampsia, transplant patients on immunosuppressant therapy such as cyclosporine and tacrolimus,patient receiving cisplatin,hemolytic-uremic syndrome, thrombotic

thrombocytopenic purpura, systemic lupus erythematosus.[3]
Clinical and radiologic aspects of this syndrome is typically reversible after removal of the inciting factor and control of the blood pressure.[2]
The clinical findings are not specific forRPES, which can resemble other neurologic conditions, such as stroke, venous thrombosis, toxic or metabolic encephalopathy, demyelinating disorders, vasculitis, or encephalitis among others.[2]

Case presentation

A 25- year- old , married, woman presented to our hospital with a sudden bilateral visual impairment which has begun from 2 hours ago and she had a severe headache, nausea and vomiting that has started from 2 days ago. she had a generalized tonic clonic seizure that last for about 2-3 minutes,some minutes after admission.Her initial vital signs were: PR: 88/min, BP: 150/90mmHg, RR: 19/min Toral:37 ^{0}c. In neurological examination the patient was lethargic and she obeyed the commands. She had bilateral mydriatic pupils with a weak light reflex and a bilateral papilledema. Bilateral planter reflexes were up. Deep tendon reflexes and other examinations were normal. During two days patient deteriorated and connected to ventilator.

Past Medical history : 5 months ago, when the patient was in the eighth week of gestation,she was admitted because of fever, chills and flank pain and ultrasonography had reported hydronephrosis and several stones in both kidneys so she had medical treatment and different urological surgeries like :TUL,embedding DJ and nephrostomy,within 5 last months. However, 3 weeks ago due to a uremia (Cr: 6.5 Urea: 67) and diagnosis of ESRD and according to the nephrologist's recommendation a dialysis program was started for her. But over a week of performing dialysis due to uncontrollable uremia (Cr: 5.2 Urea: 52), the command of pregnancy termination was given by nephrologists and perinatologists. The patient was undergoing induction of labor and the baby was born at 28th week. The patient was discharged 4 days after delivery with Cr: 6, Urea: 53. The patient had no history of other diseases such as diabetes melitus,HTN and collagen vascular disease.

Below are the initial laboratory data of the patient.

A cranial computed tomography (CT) scan demonstrated bilateral hypodense region with dominanttinvolvement of white matter of occipital lobe with extension to the parietal lobe. Based on the patient's symptoms and CT scan findings , diagnosis of PRES syndrome was proposed for the patient. To confirm the diagnosis and rule out other possible diagnoses, MRI was done and it reveal bilateral symmetrical high signal intensity change in white matter of parieto-occipital lobes which are highly According to radiologic findings of PRES syndrome(Figure 1,2).

BS	79 mg/dl
Urea	54 mg/dl
Cr	7.8 mg/dl
Na	139 meq/l
K	3.8 meq/l
Hb	11.2 g/dl
WBC	5100 mm^3
PLT	175000 mm^3
PT	12 sec.
PTT	34 sec.
INR	1

Upon the admission, the patient was hospitalized in ICU and initial managements were done for her. The patient's seizure was controlled. Heparin 5000 units subcutaneous injected twice a day and antihypertensive drugs started. Within 2 weeks the patient gradually regained her consciousness and she was separated from the ventilator. Visual impairment and hemiparesis gradually recovered so that after this period, the patient was able to walk on her own. In the MRI performed 2 months after, the radiologic findings had been resolved.

Discussion

PRES is a disorder of cerebrovascular autoregulation with multiple underlying etiologies and it is commonly associated with increase in BP.[5]PRES has been associated with chronic renal failure,hypertensive encephalopathy, eclampsia , use of some immunosuppressive and cytotoxic drugs.[3]It so thought that the sudden elevation in BP lead to disruption of the autoregulatory mechanisms

in the central nervous system vasodilatation and vasoconstriction resulting in a breakdown of the blood brain barrier and vasogenic edema.[2, 3, 6]However it is documented in some cases the BP may be only minimally elevated even normal blood pressures.[2, 7, 8]

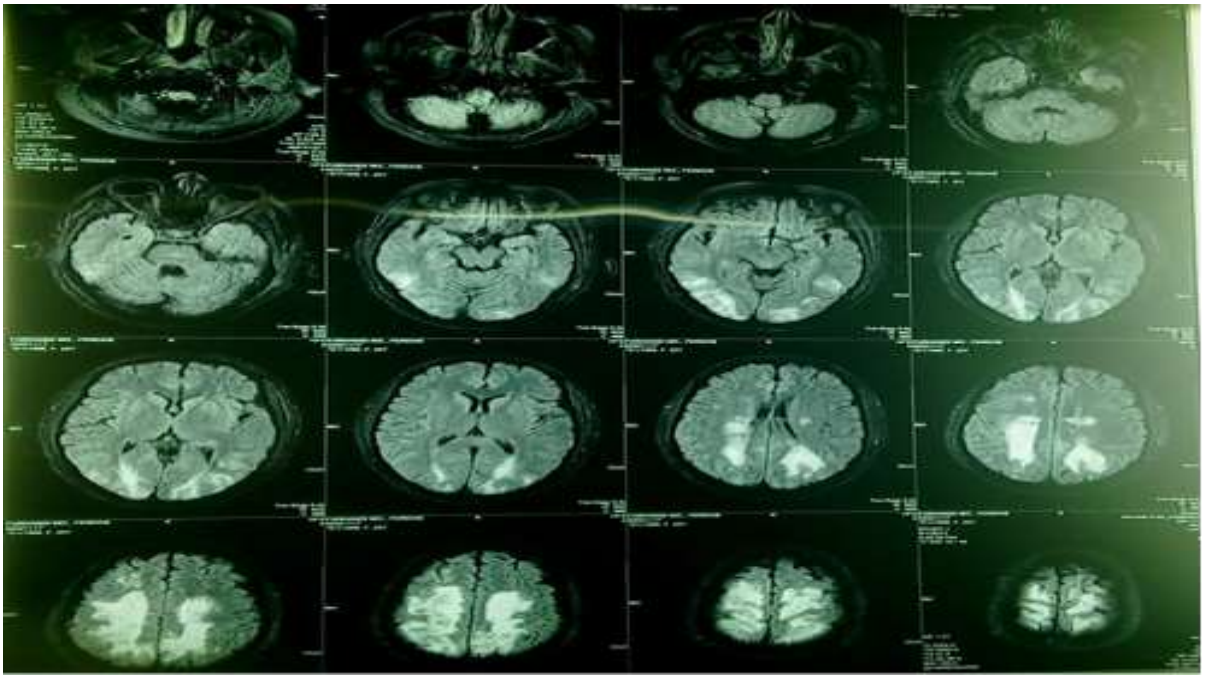

Figure 1:MRI : bilateral symmetrical high signal intensity change in white matter of parieto-occipital lobes

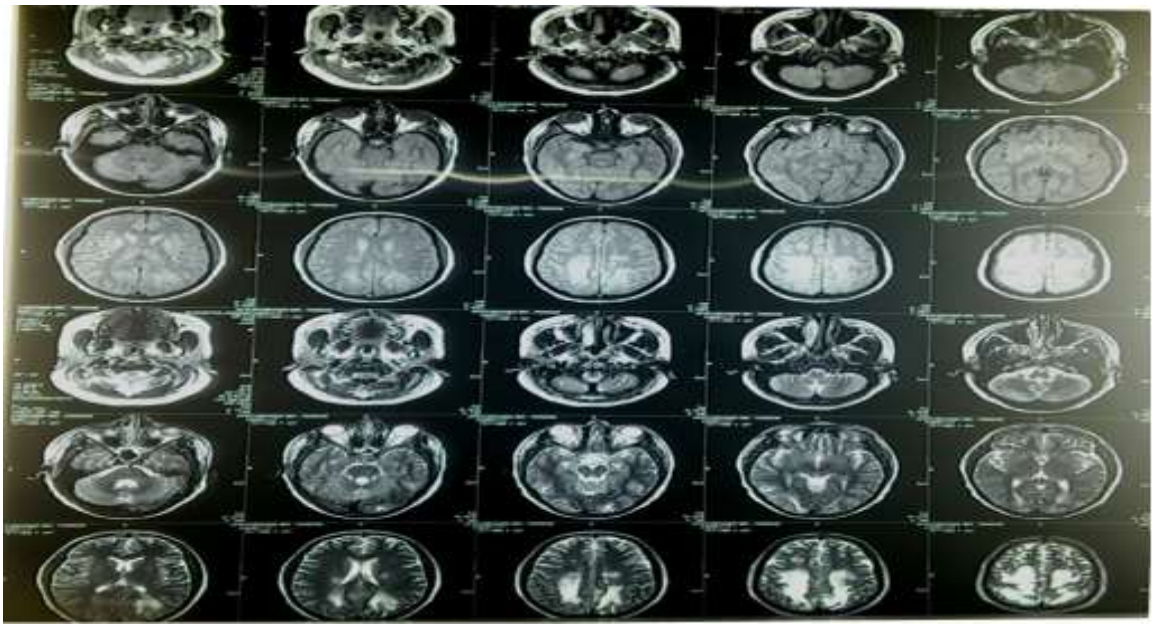

Figure 2:MRI: bilateral symmetrical high signal intensity change in white matter of parieto-occipital lobes

The diagnosis of PRES can be made via CT scan but MRI is a more sensitive imaging .The most common abnormalities on CT or MRI scans are focal regions of vasogenic edema involving the white matter in the posterior cerebral hemispheres, often asymmetrically and most commonly involving the parieto-occipital lobes bilaterelly.the medial occipital lobe structures are spared, which distinguishes PRES from bilateral posterior cerebral artery infarcts.[7]On MRI ,lesions are hyperintense on FLAIR and T2 wighted images .The lesions ,best seen on T2 pulse sequences, are characterized by high signal abnormalities in the occipital white matter with some encroachments on gray matter .[3] Clinical and radiologic aspects of this syndrome is typically reversible after removal of the inciting factor and control of the blood pressure.[2]

Early recognition of PRES is important for timely institution of therapy, which typically consists of gradual blood pressure control and treatment of underlying conditions . Although reversible by definition, secondary complications, such as status epilepticus (SE), intracranial hemorrhage, and massive ischemic infarction, can cause substantial morbidity and mortality.[2, 9]

In this patient considerable point was development of PRES by intermediate elevated BP and she never had high BP during the hospitalization.

Finally we recommand attention to PRES in every patient who has history of renal failure and presented Loss of consciousness with intermediate elevated BP .

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