Posterior Reversible Encephalopathy Syndrome

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KEYWORDS

- Posterior reversible encephalopathy syndrome (PRES)
- Reversible posterior leukoencephalopathy syndrome (RPLE)
- Hypertensive encephalopathy • Preeclampsia • Eclampsia • Cyclosporine
- Transplant • Chemotherapy

HOSPITAL MEDICINE CLINICS CHECKLIST

1. Posterior reversible encephalopathy syndrome (PRES) is characterized by headache, altered consciousness, visual disturbances, and seizures, along with distinctive findings on MRI.
2. Conditions particularly associated with the disorder include hypertensive emergency, preeclampsia/eclampsia, posttransplantation status, and chemotherapy for malignancy.
3. MRI is the imaging modality of choice for assisting in the diagnosis of PRES.
4. Treatment includes aggressive blood pressure reduction in most cases, along with reversal of the underlying predisposing state.
5. Prompt recognition and treatment of PRES are vital to prevent long-term neurologic sequelae and death.

What is posterior reversible encephalopathy syndrome (PRES)?

PRES, sometimes referred to as reversible posterior leukoencephalopathy syndrome (RPLE), is a clinical neurologic syndrome associated with characteristic imaging findings. Although scattered case reports before 1996 clearly delineate the entity in retrospect,[1–4] the disorder remained poorly characterized until a seminal case series of 15 patients published in the *New England Journal of Medicine*.[5] The original name RPLE was coined because of the observation of posterior circulation white matter changes

Conflicts of interest: The authors declare that they have no commercial or financial conflicts of interest concerning this article.
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Hosp Med Clin 4 (2015) 358–367
http://dx.doi.org/10.1016/j.ehmc.2015.03.007
2211-5943/15/$ – see front matter

on neuroimaging and evidence of clinical and radiographic reversibility within several weeks.[5] However, subsequent imaging studies have revealed that brain lesions are not confined solely to white matter,[6,7] so the term PRES may be more accurate.[8]

Which patients are at risk for the condition?

Case series consistently show a 2:1 female/male predilection[4,8,9]; all ages are potentially affected.[10] Risk factors include hypertension, preeclampsia or eclampsia, renal disease, immunosuppressive therapy, and conditions associated with endothelial dysfunction (**Box 1**).[11]

HYPERTENSION

Hypertension is present in about three-quarters of cases of PRES.[6,8,12] In hypertensive encephalopathy, acute sustained increase in blood pressure can exceed the upper limits of cerebral blood flow autoregulation.[13,14] The accompanying constellation of symptoms and neuroimaging findings can be indistinguishable from PRES.[1,4] The degree of increase in blood pressure required to exceed cerebral blood flow autoregulation depends on baseline blood pressure. Normotensive individuals can show encephalopathic changes at systolic and diastolic blood pressures of 160 mm Hg and 100 mm Hg, respectively. In contrast, abrupt increases in blood pressure exceeding 220/110 mm Hg may be required to engender hypertensive encephalopathy in chronic hypertension.[14]

PREECLAMPSIA/ECLAMPSIA

Preeclampsia and eclampsia are well-established risk factors for PRES.[5,6,12] Typically occurring in the third trimester of pregnancy, preeclampsia manifests with the triad of hypertension, proteinuria, and edema and is associated with both endothelial dysfunction and hypomagnesemia; two additional conditions that may foster the development of PRES.[6,13] Eclampsia, further defined as onset of convulsions with gestational hypertension or preeclampsia,[15] shows considerable clinical and radiographic overlap with PRES.[16–18] PRES has also been reported in the context of delayed eclampsia (ie, onset after the puerperium)[19] and following resection and chemotherapy for hydatidiform mole.[18]

ORGAN/TISSUE TRANSPLANTATION

Patients having transplants are likewise at increased risk for developing PRES; the risk extends both to patients having solid organ transplants and allogeneic bone marrow or stem cell transplants.[6,20,21] Reported incidences range from between 0.4% and 6% after solid organ transplantation to 16% with high-dose myeloablative regimens for allogeneic bone marrow transplantation.[6] Peak incidence is within 1 month of allogeneic bone marrow transplantation, within 2 months of liver transplantation, and significantly later for renal transplantation.[6] Donor-recipient human leukocyte antigen mismatch, graft-versus-host disease, and transplant rejection all seem to be correlated with development of PRES,[6] suggesting underlying immune-mediated mechanisms. Patient population risk may also be related to commonly used immunosuppressive drugs known to be independently associated with PRES, such as cyclosporine and tacrolimus (discussed later).