Posterior reversible encephalopathy syndrome

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- Evaluating Risk Factors for Lethality in Posterior Reversible Encephalopathy Syndrome following Hematopoietic Stem Cell Transplantation in Pediatric Patients: A Systematic Review and Individual Participant Data Meta-analysis

Posterior Reversible Encephalopathy Syndrome (PRES) is a rare neurological condition characterized by a variety of symptoms including headache, altered mental status, seizures, visual disturbances, and focal neurological deficits. The syndrome was first described in 1996 by Hinchey et al., and its name reflects the typical pattern of brain regions affected on neuroimaging.

Despite its name, PRES can affect both the posterior and anterior regions of the brain. The characteristic feature on imaging studies, such as MRI, is reversible vasogenic edema predominantly involving the posterior cerebral hemispheres, though it can also involve other brain regions.

The exact cause of PRES is not fully understood, but it is often associated with conditions that lead to rapid increases in blood pressure, such as hypertensive crises, eclampsia (a complication of pregnancy), immunosuppressive therapy, kidney disease, autoimmune disorders, and certain medications. The mechanism underlying PRES is believed to involve endothelial dysfunction and disruption of the blood-brain barrier, leading to cerebral edema.

Treatment typically involves managing the underlying cause, such as controlling blood pressure or discontinuing offending medications. In most cases, with prompt recognition and appropriate management, symptoms of PRES are reversible, and the prognosis is generally favorable. However, if left untreated, PRES can lead to more severe complications, including permanent neurological deficits or death.

AKA reversible posterior leukoencephalopathy syndrome (RPLS). A group of encephalopathies with a characteristic pattern of widespread vasogenic brain edema seen on CT or MRI with some predominance in the parietal and occipital regions [1].

The most common PRES pattern involves watershed zones with the involvement of the cortex, subcortical, and deep white matter to a variable extent [2].

A small number of patients with PRES will go on to infarction.

Patients may present with headaches, seizures, mental status changes, and focal neurologic deficits.

Intracerebral hemorrhage (ICH) and SAH may occur in up to 15% [3].

Posterior reversible encephalopathy syndrome (PRES) is a constellation of neurologic symptoms-seizures, headaches, altered mental status, and visual changes-associated with characteristic brain magnetic resonance imaging findings seen on T2 and fluid-attenuated inversion recovery sequences.

Etiology

The etiology of this entity includes a sudden increase in blood pressure, renal failure, immunosuppressive drugs, infections, and intravenous immunoglobulin (IVIG).

In a systematic review, antibiotic-associated PRES was more frequent in female patients (83.3%). Metronidazole and fluoroquinolones were the most reported antibiotics (33.3% each). Clinical and radiological features were comparable to those of PRES due to other causes [4]

The development of PRES after surgical resection of posterior fossa tumors has mostly been linked to the pediatric neurosurgical practice.

Associated findings and conditions

Includes:

1. hypertensive encephalopathy: commonly seen in the setting of subacute blood pressure elevations (as may occur with malignant hypertension). Imaging studies show symmetric confluent lesions with mild mass effect and patchy enhancement primarily in the subcortical white matter of the occipital lobes which may produce cortical blindness

a) moderate to severe hypertension is seen in ≈ 75% of patients with PRES although the upper limits of autoregulation are often not reached

b) in addition to hemispheric patterns of edema, isolated brainstem and cerebellar edema have been described. Posterior fossa edema has been reported to cause obstructive hydrocephalus in a severe case

2. preeclampsia/eclampsia associated with cerebral edema. The condition is often temporary, but (permanent) infarctions also occur. Restricted diffusion on MR imaging is seen in 11–26% of cases. Abnormal DWI areas on MRI may be associated with a worse prognosis

a) may present (e.g., with blindness) during pregnancy complicated by preeclampsia or eclampsia

b) may develop 4–9 days post-partum and may be associated with vasospasm even in patients not meeting clinical criteria for the diagnosis of eclampsia

c) toxemia is attributed to the placenta. Delivery and removal of the placenta are felt to be curative 3. infection, sepsis, and shock: blood pressure was normal in 40% (edema was greater in the normotensive patients). Gram positive organisms predominate

4. autoimmune disease: PRES has been described in patients with lupus, scleroderma, Wegener's granulomatosis, and polyarteritis nodosa. These patients often receive regimens of immunosuppressive medications (tacrolimus, cyclosporine), which have also been linked to cases of PRES

5. cancer chemotherapy: PRES occurs in patients receiving multi-drug high-dose chemotherapy most commonly for hematopoietic malignancies

6. transplantation: PRES has been reported both with bone marrow and solid organ transplantation

a) incidence: 3–16% with bone marrow transplantation depending on the preconditioning regimen and whether or not it is myeloablative

b) highest incidence in the first month following allogeneic bone marrow transplant

c) lower incidence following solid organ transplants. Occurs earlier following liver transplantation, usually within 2 months. Occurs later in renal transplants

7. cyclosporine post-transplant neurotoxicity

Clinical

Patients may present with headache, seizures, mental status changes and focal neurologic deficit. Intracerebral hemorrhage (ICH) and SAH may occur in up to 15% [5].

Diagnosis

Classically, magnetic resonance imaging (MRI) findings show a symmetric reversible vasogenic edema in the parietooccipital lobes. PRES can involve the brainstem and cerebellum and sometimes can leave irreversible lesions but it can also recur, which is a very rare presentation.

Treatment

Disordered autoregulation mandates tight control of blood pressure to reduce the risk of ICH. The underlying cause needs to be addressed (i.e., control HTN, hold immunosuppressives or chemotherapeutics, delivery of the placenta, etc.).

Prognosis

About one third of the cases were admitted to the intensive care unit, but almost all subjects (90.0%) had a complete or almost complete clinical and radiological recovery after prompt cessation of the causative drug. Antibiotic-associated PRES appears to share most of the characteristics of classic PRES. Given the overall good prognosis of the disease, it is important to promptly diagnose antibiotic-associated PRES and discontinue the causative drug [6)].

Case reports

A 55-year-old female patient with PRES occurring one day after administration of metronidazole and showing elevated serum neurofilament light chain protein levels and favorable outcome. In our systematic review, antibiotic-associated PRES was more frequent in female patients (83.3%). Metronidazole and fluoroquinolones were the most reported antibiotics (33.3% each). Clinical and radiological features were comparable to those of PRES due to other causes. Regarding the prognosis, about one third of the cases were admitted to the intensive care unit, but almost all subjects (90.0%) had a complete or almost complete clinical and radiological recovery after prompt cessation of the causative drug. Antibiotic-associated PRES appears to share most of the characteristics of classic PRES. Given the overall good prognosis of the disease, it is important to promptly diagnose antibiotic-associated PRES and discontinue the causative drug [7)]

Two cases of patients with SCI who developed PRES from AD. Each patient was correctly diagnosed, leading to appropriate treatment of the factors leading to their AD and subsequent resolution of their PRES symptoms. Conclusions/Clinical Relevance: In SCI patients who present with new seizures, visual deficits, or other neurologic signs, PRES should be considered as a part of the differential diagnosis as a good outcome relies on rapid recognition and treatment of AD [8)].

Hage et al., from the Saint George Hospital University Medical Center, Beirut, Lebanon report a case of recurrent PRES with cerebellar involvement associated with noncommunicating hydrocephalus in a 2-year-old child with renal failure on peritoneal dialysis after receiving Etoposide for macrophage activation syndrome [9)].

2016

Quarante et al report 2 new pediatric cases of posterior reversible encephalopathy syndrome (PRES) that developed after surgical resection of a posterior fossa tumor. Appropriate management includes supportive measures, antihypertensive agents, and antiepileptic drugs, if needed. Full recovery is the most likely outcome in line with previous articles [10)].

2015

Sorour et al. report the first case of PRES after resection of a giant vestibular schwannoma in an adult patient. This 57-year-old female patient underwent a retrosigmoid approach for total resection of her left-sided giant tumor. On the second postoperative day, she developed the classic clinical and radiologic characteristics of PRES. She was treated aggressively with antihypertensive and anticonvulsant medications and showed complete recovery without sequelae. Conclusion PRES is a potential yet rare complication of surgeries to posterior fossa tumors that are compressing the brainstem. Rapid diagnosis and aggressive management are essential for achieving the best outcome [11].

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