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Posterior Reversible Encephalopathy Syndrome: Typical and Atypical Findings on MR Imaging

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ABSTRACT

We report three cases of posterior reversible encephalopathy syndrome (PRES), caused by eclampsia or chemotherapeutic drugs. In all three cases, not only the occipital and/or parietal lobes but also the brain stem, cerebellum, basal ganglia and other lobes were widely involved on magnetic resonance (MR) imaging. In two cases, abnormal signals disappeared completely on follow-up MR imaging. However, partial irreversible change persisted in the third case, which showed atypical hyperintense signals on the initial diffusion-weighted (DW) images. We emphasize that PRES is not always completely reversible and DW imaging may be helpful in distinguishing irreversible lesions from typical, reversible lesions. Ryukyu Med. J., 26,3,4 167~171, 2007

Key words: posterior reversible encephalopathy syndrome, magnetic resonance imaging, diffusion-weighted imaging, eclampsia, hypersensitive encephalopathy

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES), also known as reversible posterior leukoencephalopathy syndrome, is defined as a neurologic disorder demonstrating characteristic radiological abnormalities found mainly in the occipital and parietal lobes and other posterior circulation territories. Several conditions and medications have been reported to cause PRES: hypersensitive encephalopathy, chemotherapeutic or immunosuppressive drugs, eclampsia-preeclampsia, and renal failure are frequently described. Magnetic resonance (MR) imaging has been reported as the best modality to demonstrate involved areas, usually resulting in hyperintense signals on T2-weighted or fluid-attenuated inversion recovery (FLAIR) images, and isointense signals on diffusion-weighted (DW) images, reflecting reversible vasogenic edema. Although the term PRES implies that the involved lesions are reversible, unusually advanced cases may develop irreversible changes that show up as hyperintense signals on DW images. Additionally, several studies have reported that PRES involves anterior circulation territories as well as posterior territories.

Here, we report three cases of PRES. Two of the cases showed multiple lesions in the anterior and posterior territories and achieved complete recovery. In the third case, on the other hand, only the occipital and parietal lobes were involved on initial MR imaging. However, follow-up MR imaging revealed partial, irreversible changes in accordance with the hyperintense signals on the initial DW images. This report aims to supplement previous articles of PRES, mainly by its distribution on MR imaging and the effectiveness of DW imaging to predict irreversible changes.

CASE REPORT

Case 1

A 39-year-old woman was undergoing a scheduled cesarean section for twin. When around the time of the operation, her blood pressure elevated to 160/100 mmHg and she complained of a mild
headache. Nine hours after delivery, she suddenly experienced a general convulsion that lasted one minute and resolved spontaneously. One hour later, a second convulsion occurred and persisted for three minutes. The patient received anticonvulsant drugs and was admitted into the intensive care unit. No other abnormal neurologic findings were detected. Blood examination revealed a declined platelet count of $6.3 \times 10^4$ /ul, an elevated aspartate aminotransferase level of 233 mg/dl, and an elevated alanine aminotransferase level of 153 mg/dl, which were consistent with HELLP syndrome. Brain MR imaging was performed the following day and demonstrated several hyperintense areas on FLAIR images found mainly in the basal ganglia, brain stem, right cerebellum, and parieto-occipital lobes (Fig. 1A). In addition, small cortical and subcortical lesions were also revealed in the frontal lobes (Fig. 1B). No obvious hyperintense signal was observed on DW images. After receiving medication for brain edema, she maintained free of seizure and her abnormal laboratory values improved. She was discharged 8 days post-surgery, and follow-up MR imaging one month later demonstrated complete recovery of the abnormal signals (Fig. 1C).

Case 2
A 47-year-old woman, who was previously diagnosed with multiple lung metastases of left adrenal cancer, was receiving chemotherapy using cisplatine, etoposide and doxorubicin. Her blood pressure gradually elevated from 140/80 mmHg to 170/100 mmHg during the chemotherapy. On the 25th day of the series, she suddenly demonstrated generalized tonic-clonic seizure, which resolved spontaneously in approximately one minute. Her blood pressure rose to 220/120 mmHg at the same
time. She had a second seizure on the same day, and received an intravenous anticonvulsant as well as a calcium-blocker to control her blood pressure. Physical examination did not reveal abnormal neurologic findings. Brain MR imaging was performed the following day. On T2-weighted images (Fig. 2A, 2B), small hyperintense lesions were primarily observed in the occipital lobes and cerebellum. Tiny lesions were also seen in the basal ganglia and thalami. No abnormal signaling was detected on DW images. After receiving the medication, the patient did not experience any further seizures and follow-up MR imaging two weeks later revealed complete disappearance of the lesions without degeneration (Fig. 2C).

Case 3

A 27-year-old woman with a right femoral Ewing sarcoma was receiving preoperative chemotherapy using cyclophosphamide, doxorubicin and vincristine. On the 10\textsuperscript{th} day of the series, she complained of sudden narrowing of her visual field. Physical examination revealed right hemianopia but no other neurologic abnormalities. Her blood pressure was slightly elevated at 150/100 mmHg. Brain MR imaging on the same day showed obvious hyperintense areas spreading bilaterally in the occipital and parietal lobes on T2-weighted images and FLAIR images, which contained both cortical and subcortical involvements (Fig. 3A). On DW images, partial hyperintense signals were seen in the parietal lobes (Fig. 3B), which were atypical as PRES. The patient received medication for brain edema and hypertension, and her hemianopia resolved completely within one week. Follow-up MR imaging was performed approximately one month later. Although almost all abnormal signals had disappeared, small irreversible degenerations were observed in the parietal lobes, which were consistent with the hyperintense areas seen on the initial DW images (Fig. 3C). However, the patients did not complain of any persistent visual disorders.

DISCUSSION

PRES was originally termed “reversible posterior leukoencephalopathy syndrome” by Hinchey \textit{et al.} in 1996\textsuperscript{1}. They emphasized the similarity of abnormal findings on CT and MR imaging for several conditions, including hypertensive encephalopathy, eclampsia, renal failure, and complications of immunosuppressive drug therapy (cyclosporine, tacrolimus). At first, it was thought that only white matter was involved; thus, the term “leukoencephalopathy” was frequently used. However, several subsequent reports, particularly those using FLAIR sequences, clarified that cortical gray matter, as well as subcortical white matter, was also involved\textsuperscript{5,10}. Therefore, “PRES” has recently become the more favored term. In addition to such classical conditions, many diseases and drugs have been reported to cause PRES, including autoimmune diseases such as systemic lupus erythematosus or rheumatoid arthritis, thrombotic thrombocytopenic purpura, interferon-alpha, and chemotherapeutic drugs such as cisplatin, cyclophosphamide, vincristine\textsuperscript{12,13,14,15}. PRES typically occurs following an elevation of blood pressure, as our report showed. However, several cases have been reported that do not show
hypsntension around the occurrence. In some exceptional patients, recurrent episodes of PRES have been reported

PRES presents clinically with headache, seizures, visual changes (cortical blindness), altered mental status, and occasionally focal neurologic signs. Although its true etiology is still unclear, it has been suggested that PRES is actually temporary vasogenic edema caused by loss of cerebral vascular autoregulation in the involved brain, and not cytotoxic edema or infarction. This theory has been supported by pathologic reports demonstrating only interstitial edema, microhemorrhages, and fibrinoid necrosis within the arteriole walls in involved lesions.

PRES is characterized by the distribution seen on MR imaging; bilateral and relatively symmetric hyperintense lesions mainly in the parieto-occipital regions on T2-weighted or FLAIR images. This characteristic distribution has been explained as vasogenic edema that occurs easily in the posterior cerebral artery territories. The same condition often occurs in other posterior circulation territories, including the brain stem and cerebellum. However, as two of our former cases showed, PRES can occur in the area supplied by the anterior or middle cerebral arteries, and is usually accompanied by posterior involvement. Furthermore, some exceptional cases have demonstrated only isolated anterior lesions without posterior involvement. Thus, although the occipital and parietal lobes and the infratentorial brain are the areas most frequently involved in PRES, it should be taken into consideration that even anterior circulation territories, including the basal ganglia, thalamus, and frontal or temporal lobes, can be involved.

Another imaging feature of PRES is reversibility. Classical PRES demonstrates hyperintense signals on T2-weighted and FLAIR images, and normal or slightly decreased/increased signals on DW images, which suggest the potential for reversible vasogenic edema. These abnormal signals on T2-weighted and FLAIR images usually disappear quickly within a few weeks if blood pressure is properly controlled or other offending factors are removed. On the other hand, acute brain infarction or cytotoxic edema appear as apparent hyperintense signals on DW images and result in irreversible degeneration. Thus, normal or intermediate DW signals have been thought to be an essential finding for the diagnosis of PRES. However, several recent reports have clarified that atypical hyperintense signals on DW images are relatively common and may account for up to 27% of all PRES patients. In our present report, one patient demonstrated hyperintense areas on initial DW images, which later turned into partial, irreversible changes on follow-up MR imaging. Given the fact that patients with abnormal hyperintense DW signals persisted irreversible degeneration (infarction) on follow-up MR imaging, PRES should be considered potentially irreversible or cytotoxic and require careful management.

Although DW imaging is thought to be a useful sequence for diagnosing irreversible changes in PRES, several exceptional cases have been reported that showed reversible lesions with hyperintense DW signals. This phenomenon is known as T2 shine through, which is caused by increased T2 signals affecting DW signals. In these cases, apparent diffusion coefficient (ADC) maps give further information on whether the lesions are truly reversible or not. Unfortunately, ADC mapping was not available at our institution during the time of study. We can currently evaluate the patients with a combination of DW imaging and ADC mapping, thus, further research will be performed with more accuracy.

In conclusion, PRES can involve not only the parietal and/or occipital lobes, but also the brain stem, cerebellum, basal ganglia, thalamus, and frontal or temporal lobes, as shown in our cases. Although DW signals are usually normal for lesions of reversible vasogenic edema, hyperintense DW signals may be seen in advanced cases that persist to irreversible changes. Physicians should be aware that involved areas are not always reversible in PRES and DW imaging can be helpful for distinguishing irreversible changes from true reversible lesions.

REFERENCES


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