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Reversible Posterior Leukoencephalopathy Syndrome in Children: MR Imaging Findings[1]

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Purpose: To find out the characteristic MR findings of reversible posterior leukoencephalopathy syndrome (RPLS) due to various causes in children.

Materials and Methods: Eight children with RPLS underwent MR imaging, and the findings were retrospectively analyzed. All eight were acutely hypertensive at the time of a neurotoxic episode. Three had intra-abdominal tumors (one adrenal pheochromocytoma, one para-aortic paraganglioma and one para-aortic ganglioneuroma encasing the left renal artery); three were being treated with cyclosporine; one was being treated with steroid; and one had hemolytic uremic syndrome. Initial cranial MR images were analyzed with particular emphasis on the distribution of the lesions. To assess possible sequelae, follow-up MR images were obtained in seven patients at least one week after the treatment of hypertension. Four underwent proton MR spectroscopy.

Results: Characteristic distribution of lesions in the occipital and posterior parietal lobes was identified in all cases regardless of the causes of RPLS. The cerebellum, basal ganglia, anterior parietal, and frontal lobe were involved in four, two, one, and one case, respectively. Cortical gray matter involvement was predominant in six and subcortical white matter involvement predominated in two patients. The distribution of lesions was bilateral and asymmetric. Gyriform enhancement was identified in six cases, and small hemorrhage was noted in one. In seven patients, the clinical and MR findings improved without sequelae on follow-up study. In one, proton MR spectroscopy demonstrated a high lactate peak at the time of the neurologic event. Near-normal spectra were noted in three children who underwent proton MR spectroscopy after recovery.

Conclusion: The MR findings of RPLS are characteristic in that lesions are distributed in the posterior region of the brain and they are reversible on follow-up study. In children with RPLS due to unknown causes, the possibility of intra-abdominal tumors should also be considered.

Index words : Brain, disease
Brain, MR
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Reversible posterior leukoencephalopathy syndrome (RPLS), a complex of cerebral disorders which includes headache, seizure, visual disturbances and other neurologic manifestations, is associated with a variety of conditions in which blood pressure rises acutely (1). Although the syndrome is usually reversible if hypertension is treated early, it may be fatal if unrecognized and if treatment is delayed (2). Occasionally, the clinical findings are nonspecific to the extent that the diagnosis is difficult to establish. For accurate diagnosis, imaging findings may thus be very important.

In this study, we tried to find out the characteristic MR imaging findings of RPLS due to various causes in eight children and the proton MR spectroscopic findings in four children.

Materials and Methods

Patients

During the two-year period from January 1998 to December 1999, eight children [six boys and two girls aged from 3 to 16 (mean, 10.4) years] underwent cranial MR imaging for the evaluation of various neurologic symptoms and signs after acute hypertension. Three of the eight had intra-abdominal tumors (one adrenal pheochromocytoma, one para-aortic paraganglioma, and one para-aortic ganglioneuroma encasing the left renal artery); three were being treated with cyclosporine for nephrotic syndrome, renal transplantation and Langerhan 's cell histiocytosis, respectively; one was being treated with steroid as a chemotherapeutic regimen for leukemia; and one had hemolytic uremic syndrome.

Presenting symptoms and signs were seizure (four children), headache (three), and visual disturbance (cortical blindness in one, blurred vision in one). Focal weakness was noted in those children with lesions in the basal ganglia.

MR Imaging

Initial cranial MR imaging was performed within 24 hours of the onset of neurologic symptoms and signs. To evaluate possible sequelae, follow-up MR imaging after the treatment of hypertension and/or removal of underlying causes was performed in seven children between 7 and 47 (mean, 21.5) days after the neurologic event.

For all sequences, a 1.5-T unit (either Signa Advantage; GE Medical Systems Milwaukee, U.S.A., or Vision Plus; Siemens, Erlangen, Germany) was used, set at the following parameters: repetition time msec/echo time msec of 466 - 665/11 - 15 for T1-weighted spin-echo imaging; 2,900 - 3,787/95 - 102 (effective) with an echo train length (ETL) of between 8 and 11 for T2-weighted fast spin-echo imaging; and 9,000 - 13,000/110 - 133 (effective) with an inversion time (TI) of 2,500 - 2,600 msec and an ETL of 11 to 32 for fluid-attenuated inversion recovery (FLAIR) imaging. Contrast enhanced T1-weighted images were obtained after intravenous injection of gadopentetate dimeglumine (Magnevist; Berlex

Table 1. Diagnostic Data in Eight Children with Reversible Posterior Leukoencephalopathy Syndrome

Patient No. /Age (y)/Sex	Diagnosis	Maximal BP* (MAP)	Baseline BP**(MAP)	Difference of MAP (Percentage of difference, %***)
1/ 14/M	Paraganglioma	250/160 (190)	120/60 (80)	110 (138)
2/11/M	Pheochromocytoma	240/150 (180)	120/70 (87)	93 (107)
3/3/M	Ganglioneuroma	180/100 (127)	135/85 (102)	25 (25)
4/8/M	Cyclosporine neurotoxicity	200/130 (153)	135/75 (95)	58 (87)
5/16/F	Cyclosporine neurotoxicity	180/110 (133)	115/75 (88)	45 (51)
6/13/M	Cyclosporine neurotoxicity	165/90 (115)	120/70 (87)	28 (32)
7/7/F	Steroid	150/95 (113)	110/65 (80)	33 (38)
8/11/M	Hemolytic uremic syndrome	185/115 (138)	130/95 (107)	31 (29)

Note.-BP = blood pressure. MAP = mean arterial pressure, calculated as follows: [systolic blood pressure + 2 (diastolic blood pressure)]/3.

*Maximal blood pressure measured during neurologic event

**Baseline blood pressure measured at least one week prior to the event or one month after improvement of neurologic symptoms and signs.

***Percentage of difference in mean arterial pressure between baseline and maximum levels, calculated as follows: [(maximal mean arterial pressure -baseline mean arterial pressure)/baseline mean arterial pressure] ×100%

Laboratories, Wayne, N.J., U.S.A.) at a dose of 0.1 mmol/kg of body weight. For each pulse sequence, the slice thickness was 6.0 - 9.0 mm with a 0.5-1.0-mm intersection gap, one to two signals were acquired, and a 256 ×192 or 264×512 matrix was used. The field of view was 180 - 240 mm, depending on head size and section planes.

Cranial MR images were analyzed with particular emphasis on the distribution of the lesions.

MR Spectroscopic Study

Hybrid two-dimensional proton MR spectroscopy was performed at the time of the neurologic event in one child (patient 4), and after improvement of neurologic symptoms and signs in three (patients 6, 7, and 8). For MR spectroscopy, a 1.5-T scanner with a standard head quadrature coil was used. 16×16 phase encoding steps were set on a field of view (FOV) of 20×20 cm^2, and hybrid volume of interest (VOI) inside FOV was set at 75× 75×15 mm^3. This combination yields good localization and excellent rejection of unwanted subcutaneous fat signal. Point-resolved spectroscopy (PRESS) (TR/TE=1,500 msec/135 or 270 msec) was employed with standard chemical shift selective saturation (CHESS) pulses for water suppression. Hybrid two-dimensional proton chemical shift imaging (CSI) raw data were then post-processed using CSIFT software (SUN SPARC 20, Numaris 3), with or without k-space zero filling.

Proton MR spectroscopic findings were evaluated with emphasis on the presence or absence of lactate peak. The peak heights of three major metabolites (N-acetyl aspartate, creatine and choline) were visually measured.

Results

For each child, the maximal arterial pressure obtained during the hypertensive crisis and the baseline arterial pressure obtained at least one week prior to the neurologic event or one month after the improvement of neurologic symptoms and signs are shown in Table 1. All children were acutely hypertensive at the time of the symptomatic episode. Average baseline blood pressure was 123/74 mm Hg (mean arterial pressure, 91±14 mm Hg), and at the time of the neurologic event, average blood pressure was 194/119 mm Hg (mean arterial pres-

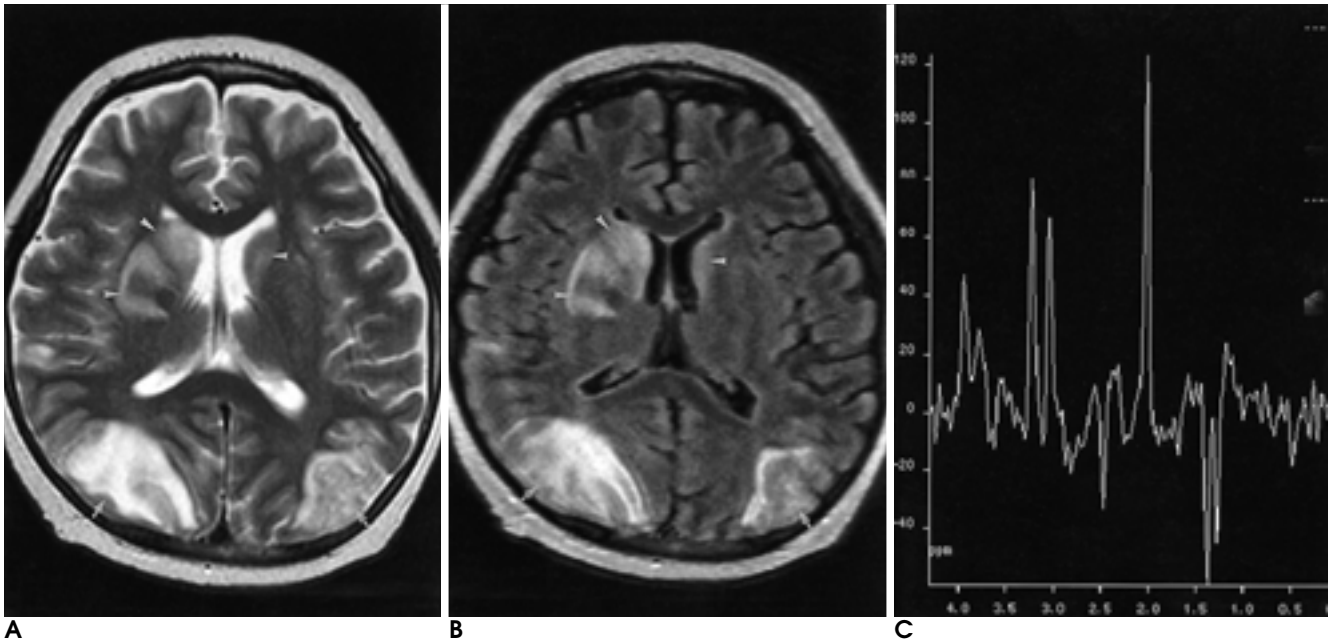


Fig. 1. An 8-year-old boy in whom reversible posterior leukoencephalopathy syndrome developed after treatment with cyclosporine for nephrotic syndrome.

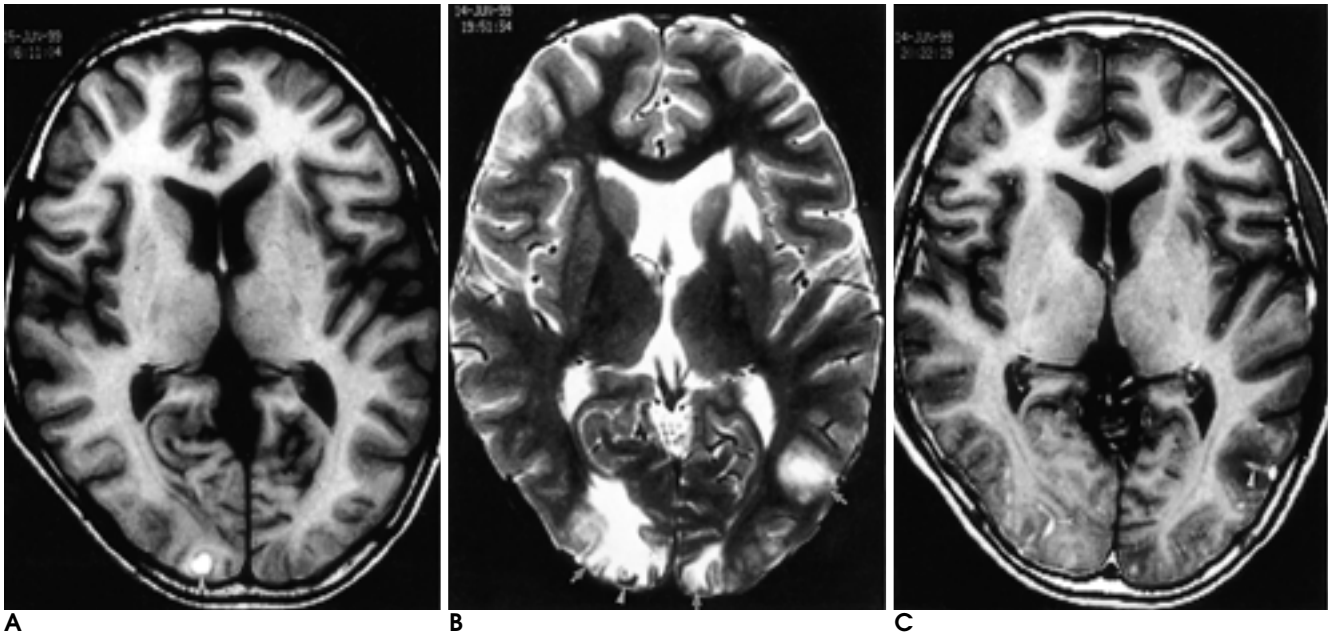
A. Axial T2-weighted (2,900/102, echo train length of eight) demonstrates bilateral hyperintense areas (arrows) in the occipital and parietal subcortical white matter and cortical gray matter. Also noted is asymmetric involvement of bilateral basal ganglia (arrowheads).

B. Fluid-attenuated inversion recovery (FLAIR) image (12,900/90; inversion time, 2,600) shows bilateral hyperintense lesions (arrows and arrowheads) in the same regions as T2-weighted image.

C. Proton MR spectroscopy (1,500/135) obtained from the right parietal white matter lesion reveals inverted lactate peak at 1.33 ppm with nearly normal N-acetyl aspartate, creatine and choline peaks.

sure, 152±38 mm Hg). Average fluctuation of mean arterial pressure was 71/45 mm Hg, representing a 57% increase.

Regardless of the causes of the RPLS, the characteristic distribution of lesions in the occipital and posterior parietal lobes was identified in all cases (Figs. 1 - 3). Additional involved areas included the cerebellum in four patients, the basal ganglia in two, the anterior parietal lobe in one, and the frontal lobe in one. In six cases, both the cortex and subcortical white matter were in-



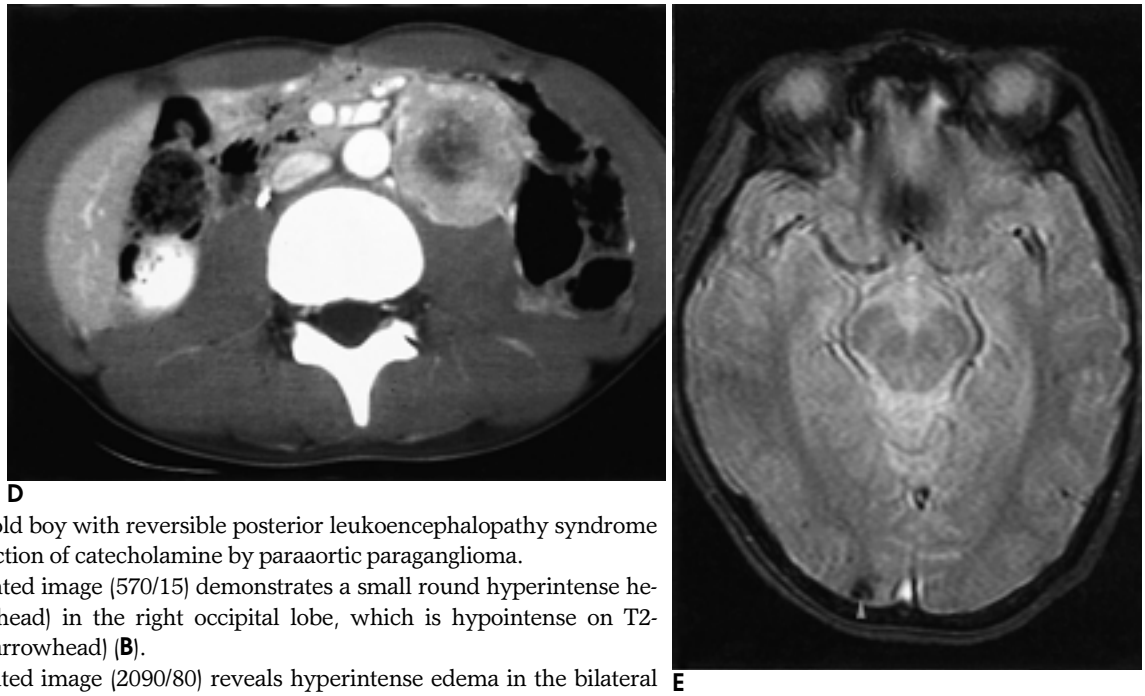


Fig. 2. A 14-year-old boy with reversible posterior leukoencephalopathy syndrome due to overproduction of catecholamine by paraaortic paraganglioma.
A. Axial T1-weighted image (570/15) demonstrates a small round hyperintense hemorrhage (arrowhead) in the right occipital lobe, which is hypointense on T2-weighted image (arrowhead) (**B**).
B. Axial T2-weighted image (2090/80) reveals hyperintense edema in the bilateral occipital and parietal lobes (arrows). Subcortical white matter is predominantly involved. Also noted is hyperintense lesion in the left lentiform nucleus.
C. Axial gadolinium-enhanced T1-weighted image (570/15) demonstrates gyral enhancement in the lesions of bilateral parieto-occipital regions (arrowheads).
D. Axial enhanced CT scan with 10-mm thickness reveals a round well enhancing soft tissue mass with central low density in the left paraaortic area.
E. Follow-up axial gradient-echo image (600/15; flip angle, 15°) obtained 47 days after initial exam demonstrates a focal hypointense hemosiderin deposit in the right occipital lobe (arrowhead).

volved (Figs. 1, 2); in four of these, lesions were predominantly cortical, and in two, subcortical white matter involvement predominated. In two further cases, only the cortex was involved (Fig. 3). The distribution of lesions was bilateral and asymmetric (Figs. 1, 2). These lesions showed high-signal-intensity on T2-weighted and FLAIR images, and low-signal-intensity on T1-weighted image (Figs. 1 - 3). Gyriform enhancement was identified in six cases (Fig. 2), and a small hemorrhagic focus was noted in one (Fig. 2).

In seven children who underwent follow-up MR imaging after the control of hypertension and/or the removal of underlying causes, these abnormalities were resolved. At that time, neurologic symptoms and signs improved, without sequelae. In one child with adrenal pheochromocytoma, a hemorrhagic focus that showed high signal intensity on T1-weighted image and low signal intensity on T2-weighted images was demonstrated on initial MR study (Figs. 2A, 2B). In this child, follow-up study after the normalization of blood pressure and the improvement of neurologic symptoms and signs revealed a small residual hemosiderin deposit (Fig. 2E).

In one child, in whom proton MR spectroscopic studies were performed at the time of the neurologic event, a high lactate peak and nearly normal N-acetyl aspartate, creatine and choline peaks were noted (Fig. 1E). In three children who underwent proton MR spectroscopic study after the recovery of neurologic symptoms and signs, MR spectra were nearly normal and showed no lactate peak.

Discussion

RPLS is a recently proposed cliniconeuroradiologic category which is characterized by seizure, visual abnormalities including blurring or cortical blindness, headache, nausea, vomiting, lethargy or confusion (3), and occurs in a setting of acute hypertension. The syndrome has been recognized in a wide variety of clinical conditions, including essential or chronic hypertension, renal disease, collagen vascular disease, mixed connective tissue disorders, endocrine abnormalities, eclampsia, and immunosuppressive treatment (most notably, involving the use of cyclosporine) for neoplasms or to prevent rejection after organ transplantation (3 - 7). Similar cases involving patients receiving the anticancer drugs cytarabine (8) and cisplatin (9) have been reported.

Hypertensive encephalopathy, first described in 1928 by Oppenheimer and Fishberg, is a specific clinical syndrome characterized by acute neurologic change in the setting of sudden and/or prolonged hypertension that overcomes the autoregulatory capacity of the cerebral vasculature (10). It is a subset of the more generalized reversible posterior leukoencephalopathy syndrome (3, 9). Although a wide variety of causes exist, hypertensive encephalopathy and RPLS have common clinical and imaging findings and probably a common pathophysiology in that acute hypertension is the most important trigger, regardless of the causes.

RPLS is rare in the pediatric population. When present,

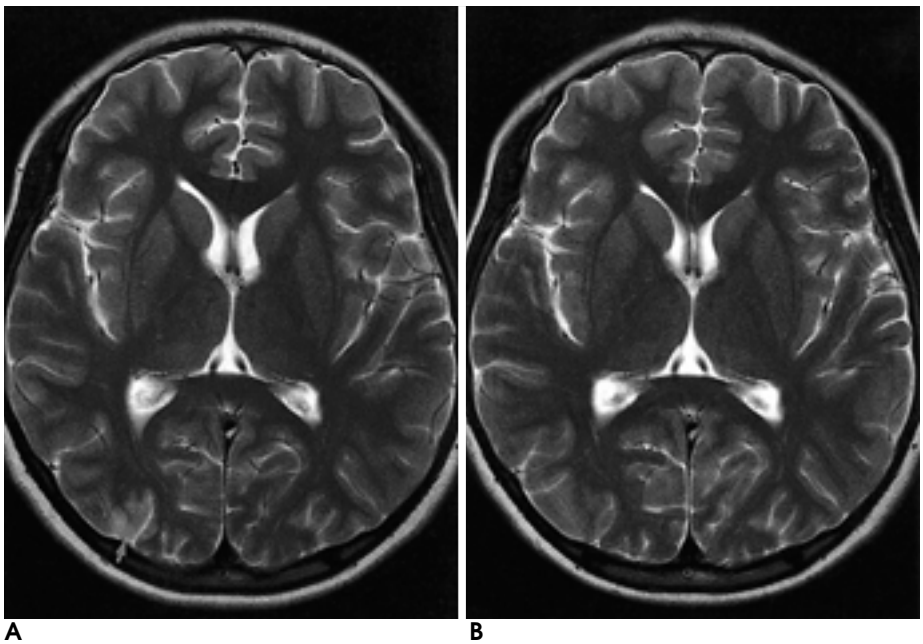


Fig. 3. An 11-year-old boy with reversible posterior leukoencephalopathy syndrome caused by hemolytic uremic syndrome.
A. Axial T2-weighted image (3787/99; echo train length, 11) demonstrates a focal area of gyral swelling in the right parietal lobe (arrow).
B. Follow-up T2-weighted image (1787/99; echo train length, 11) obtained 20 days after initial exam shows complete resolution of gyral swelling.

it is usually related to renal diseases such as acute glomerulonephritis, renovascular hypertension, or chronic renal failure from any cause (11, 12). Cyclosporine is a well-known cause of RPLS in children and adults (8, 9), and may induce sympathetic hyperactivity and, as a result, hypertension (13). Furthermore, it has a cytotoxic effect on vascular endothelium, leading to brain-capillary leakage and acute blood-brain barrier disruption, which may trigger vasogenic edema (4, 14, 15).

RPLS in patients with an abdominal tumor has rarely been reported in the literature (16), but we identified the syndrome in three children with an intra-abdominal tumor. The only initial clinical manifestations in these patients were due to RPLS. Laboratory findings failed to document abnormal renal function, and so abdominal US and CT were performed, demonstrating three intra-abdominal tumors: one adrenal pheochromocytoma, one para-aortic paraganglioma, and one para-aortic ganglioneuroma encasing the left renal artery. Excessive production of catecholamine was responsible for hypertension in two children, and renal artery obstruction caused this condition in one. These tumors were not palpated during physical examination, and in the absence of neurologic symptoms and signs of RPLS, might have remained undiagnosed.

Because hypertension is uncommon in the pediatric population and the findings of RPLS are reversible, it has been suggested that the syndrome is often unrecognized, and its true prevalence may thus be underestimated (12). It should be noted that in all our cases, clinical diagnosis depended upon recognition of the imaging findings that supported the diagnosis. The syndrome might otherwise have gone undiagnosed or diagnosis might have been delayed. In our first patient (case 7), who was undergoing a chemotherapeutic regimen involving the use of a steroid for the treatment of leukemia, the presence of hypertension was unnoticed at the time of initial presentation. The MR imaging findings, however, were consistent with hypertensive encephalopathy. Retrospective review of the patient's medical records showed that while baseline blood pressure was 110/65, at the time of neurologic event, a transient fluctuation of blood pressure (150/95) had occurred. No treatment was instituted and the hypertension resolved spontaneously.

Although systolic pressure greater than 250 mm Hg and diastolic pressure greater than 150 mm Hg are commonly encountered in patients with RPLS (12), it is fluctuation of blood pressure rather than its absolute value that is important for the development of RPLS.

The precise mechanism of cerebral edema in RPLS is still unknown (4). Two theories have been proposed to account for the clinical and radiologic abnormalities associated with hypertensive encephalopathy. The first postulates that this results from spasm of the cerebral vasculature in response to acute hypertension (i.e. overregulation), resulting in ischemia and cytotoxic edema involving mainly border-zone arterial regions (17, 18). A more recent hypothesis suggests that the syndrome results from the breakthrough of autoregulation, with passive overdistension of cerebral arterioles (19, 20, 21). Arterioles situated a short distance from the cortical surface are most affected, though sympathetic nervous activity affords protection from these effects. The posterior circulation has significantly less sympathetic innervation than the carotid circulation (22, 23), and this may explain why the majority of lesions seen in hypertensive encephalopathy are found in the vascular territory of the posterior circulation. Recent evidences with diffusion-weighted imaging have shown that these abnormal MR findings do not demonstrate restricted diffusion, and therefore most likely represent vasogenic edema (24, 25).

Unfortunately, the term "leukoencephalopathy" may be confusing to many neuroradiologists because there usually is no accompanying destructive process of the white matter. Furthermore, in our series, gray matter structures as well as white matter were involved. Since the clinical and radiologic findings generally resolve after the normalization of blood pressure, a better term might be "reversible posterior cerebral edema syndrome"(26).

The clinical picture is associated with reversible white matter edema on CT and MR examinations (4). In uncomplicated cases, images reveal the presence of cerebral edema, mostly in the cortex and subcortical white matter of the posterior regions of the brain (occipital lobes, posterior parietal and temporal lobes, and posterior fossa structures), though involvement of the frontal lobe has also been reported (9). In our study, cortical gray matter rather than subcortical white matter was more frequently and/or predominantly involved. Intracerebral hemorrhage may occur, especially in patients with thrombocytopenia or in those in whom hypertension cannot be controlled. In one patient in our study (case 1), intracerebral hemorrhage occurred, but follow-up study showed that it had resolved without significant neurologic sequelae (case 1).

Sengar et al. (27) reported the MR spectroscopic findings of brain in ten adults with eclampsia, noting that in nine with reversible imaging changes, NAA levels had decreased slightly and the decrease persisted even after two weeks. In one patient, the presence of lactate along with markedly decreased NAA and creatine suggested infarction. The marked decline in NAA in this patient correlated well with the development of cerebral atrophy and poor clinical outcome, which probably resulted from gross neuronal damage. In one of our patients, a high lactate peak, thought to be due to the transient derangement of energy metabolism, was noted at the time of the neurological event, but N-acetyl aspartate, creatine and choline peaks were nearly normal. Near-normal N-acetyl aspartate level probably represents an absence of neuronal damage, and in RPLS is associated with a favorable prognosis. The reversibility of this syndrome can also be explained by the nearly normal spectroscopic findings seen on follow-up after the improvement of neurologic symptoms and signs in three children. Thus, proton MR spectroscopy may play a role in determining the prognosis of RPLS, though further prospective evaluation is needed.

The treatment of RPLS involves the control of hypertension and the removal of underlying causes. With adequate treatment, all abnormalities resolve within a few weeks. Whatever the cause of RPLS, early recognition of the syndrome, revealed by the presence of posterior parietal parasagittal areas of high signal intensity on T2-weighted images, allows appropriate clinical management with targeted treatment of hypertension. In most cases of RPLS, the causes of hypertension can be determined clinically. In three children with RPLS caused by intra-abdominal tumors, the causes of hypertension were not initially established. Thus, in cases of RPLS due to unknown causes, the possibility of intra-abdominal tumors also should be considered.

In conclusion, the MR imaging findings of RPLS are characteristic in that lesions are distributed in the posterior region of the brain and they are reversible on follow-up study.

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소아에서의 가역성 후 백질뇌병증: 자기공명영상 소견[1]

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목적: 소아에서 다양한 원인에 의한 가역성 후 백질뇌병증의 자기공명영상 소견에 대해 알아보고자 하였다.
대상과 방법: 가역성 후 백질뇌병증으로 진단된 8명의 환아를 대상으로 자기공명영상 소견을 후향적으로 분석하였다. 모든 환아에서 신경학적 증상 발현시 급성 고혈압이 동반되었다. 이들 중 3예에서는 복강 내 종양 (부신 크롬친화세포종, 대동맥주위 부신경절종, 대동맥주위 신경절신경종, 각각 1예)이 있었고 3예에서는 cyclosporine, 1예에서는 steroid 치료를 받은 병력이 있었으며, 1예는 용혈성 요독 증후군 환아 였다. 초기 뇌 자기공명영상 소견은 병변의 분포를 중심으로 분석하였으며. 7예에서는 후유증의 유무를 평가하기위해 고혈압을 치료한 후 1주일 이상 경과한 후에 추적 자기공명영상을 시행하였다. 4예에서 양성자 자기공명분광법을 시행하였다.
결과: 가역성 후 백질뇌병증의 병변은 모든 예에서 원인에 관계없이 특징적으로 후두엽과 후 두정엽에 분포하였다. 소뇌에 병변이 보였던 경우가 4예였으며 대뇌기저핵이 2예, 전 두정엽과 전두엽이 각각 1예이었다. 대뇌 피질의 침범이 현저한 경우가 6예였으며 피질 하 백질의 침범이 현저한 경우가 2예이었다. 병변의 분포는 양측성이었으며 비대칭적이었다. 대뇌 이랑 형태 (gyriform)의 조영증강이 6예에서 보였으며 1예에서는 소량의 출혈소견이 있었다. 추적 검사를 시행한 7예에서 후유증 없이 임상 소견 및 자기공명영상 소견이 호전되었다. 신경학적 증상 발현시기에 양성자 자기공명분광법을 시행한 1예에서는 높은 lactate 최고점을 보였으나 신경학적 증상이 소실된 후 시행한 3예에서는 정상에 가까운 스펙트럼을 보였다.
결론: 병변이 뇌의 후방에 분포하며 추적 검사에서 병변이 호전되는 것은 가역성 후 백질뇌병증의 특징적 자기공명영상 소견이라 할 수 있으며, 원인 불명의 가역성 후 백질뇌병증 환아의 경우 복강내 종양에 대한 가능성도 고려하여야 할 것으로 생각된다.