# IMAGING OF PERSISTENT BRAIN DAMAGE FROM HYPERTENSIVE ENCEPHALOPATHY IN A CHILD WITH NEUROPSYCHIATRIC SYSTEMIC LUPUS ERYTHEMATOSUS (NPSLE) AND ALFA THALASSEMIA TRAIT, A CASE REPORT.

**Siriporn HIRUNPAT,[1] Prayong VACHVANICHSANONG,[2]**

**Chakree HIRUNPAT.[3]**

## ABSTRACTS

An unusual case of a neuropsychiatric systemic lupus erythematosus (NPSLE) child with hypertensive encephalopathy is presented. Despite the rare presentation of hypertensive encephalopathy in children, characteristic lesions in the brain should be recognized as possible manifestation of this condition. In contrast to other reports in which the brain damage was almost always reversible, we present a serial follow up images which demonstrate both reversible and irreversible brain damages. Brain lesions associated with hemorrhage, which are seldom described in most reports, have less likelihood reversibility.

**Keywords:** Encephalopathy, Hypertension, NPSLE, Child, Thalassemia

## INTRODUCTION

Hypertensive encephalopathy (reversible posterior leukoencephalopathy) is an important clinical entity to be recognized because of the clinical and imaging findings, as the name implies, are usually reversible.

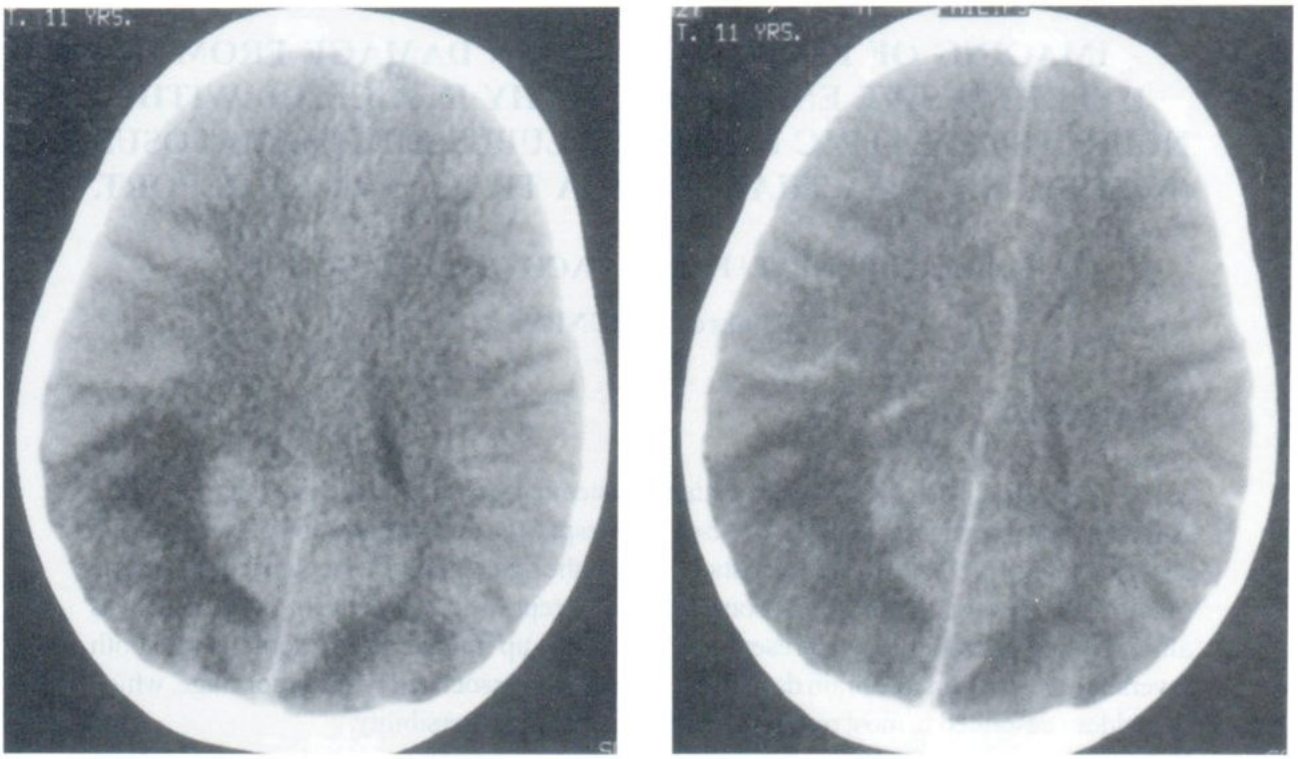
The clinical signs and MRI findings are characteristic. The condition is rarely reported in children and is usually seen in association with other systemic diseases.[1,2] Its true prevalence may be underestimated. We add a case report of a boy who had typical imaging of hypertensive encephalopathy in the serial imaging follow up from the first attack of the disease until recovery. Both reversible and irreversible changes in the brain were seen in this patient.

## CASE REPORT

This 11 year old boy with SLE and Alfa Thalassemia trait had acute cortical blindness and drowsiness during admission for cholecystectomy. His blood pressure was 160/110 mmHg. SLE is diagnosed on the basis of thrombocytopenia, positive ANA (1: 640), nephritis (biopsy = lupus nephritis) and CNS involvement.

His CT scan at 1 day after the onset of acute cortical blindness showed low density areas in the subcortical white matter of both the occipital and parieto-occipital regions, more prominent at the right side, without significant enhancement. Minimal hypodensity at the subcortical white matter of the left frontal lobe was also noted. ( Fig 1 A&B)

[1] Department of Radiology,
[2] Department of Pediatric,
3 Department of Ophthalmology Faculty of Medicine, Prince of Songkla University, Hat-Yai, Songkhla, Thailand
**Correspondence:** S. Hirunpat, Department of Radiology, Faculty of Medicine, Prince of Songkla University, Hat-Yai, Songkhla, Thailand 90110 Fax:66-74-429927. E-mail:Hirsirip@ratree.psu.ac.th




1a 1b

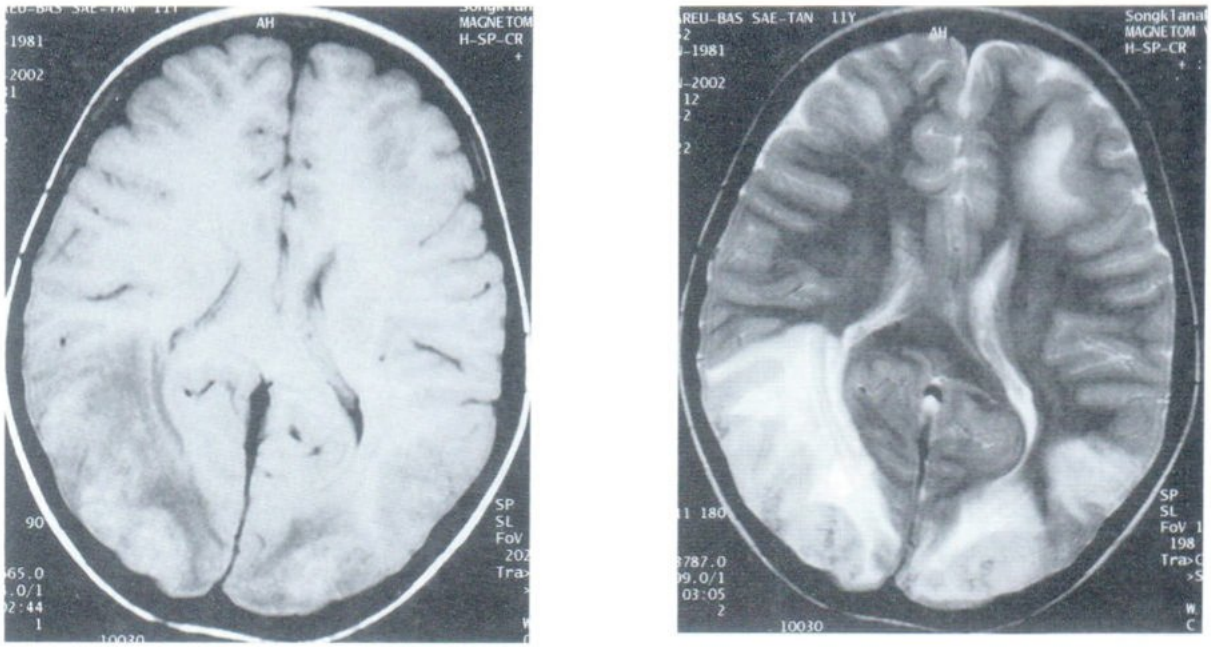
**Fig.1** **a.** Plain axial CT and, **b.** Post contrast axial CT scan reveal low density areas in the subcortical white matter of both the occipital and parieto-occipital regions, more prominent at the right side, without significant enhancement. Minimal hypodensity at the subcortical white matter of the left frontal lobe was also noted.

Two days later, an MRI revealed abnormally low signal intensity on T1W and high signal intensity on T2W in both occipital and parieto-occipital subcortical white matter and at the left frontal subcortical white matter. Edematous adjacent cortical gyri were also seen, with multiple small hemorrhagic foci seen as high signal intensity on T1W and low signal intensity on T2W along both parieto-occipital and occipital gyri indicating more severe damaging brainauese from hypertensive encephalopathy. (Fig 2A &B)

The patient was treated with high doses of corticosteroids and anti-hypertensive drugs. His clinical signs rapidly improved. Visual acuity returned to normal (VA 20/20) within a few days and follow-up MR imaging 2 weeks after the first scan also showed marked resolution of the abnormal findings. However, small hemorrhagic foci along both occipital and parieto-occipital cortical gyri persisted. (Fig 3A&B)

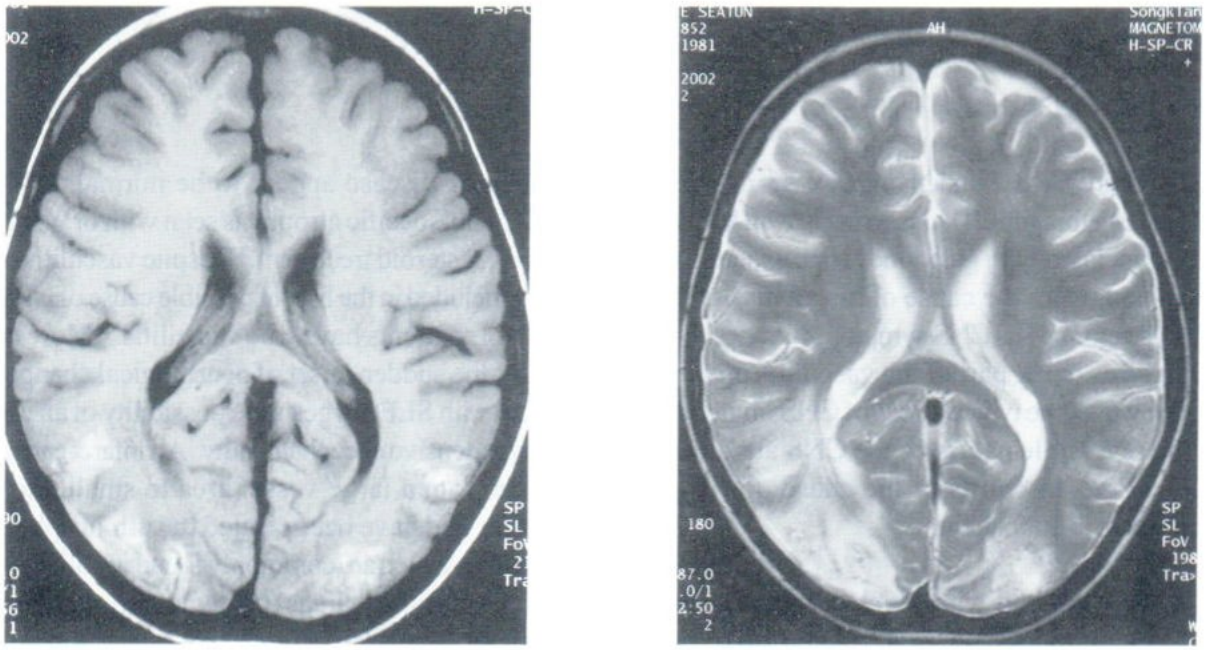
Finally, the patient's neurologic symptoms were all resolved. A follow up MRI at 6 months after the first scan revealed an almost completely normal brain except for small gliotic areas at right parieto-occipital and right occipital cortex which indicated the more severe and irreversible brain damaged areas.

The MRI study showed no residual brain edema, but a small area of gliotic change at the previous hemorrhage sites at the right parieto-occipital cortex. (Fig. 4A& B) The smaller areas of previous hemorrhage at the left occipital and parieto-occipital cortex showed no residual abnormality.



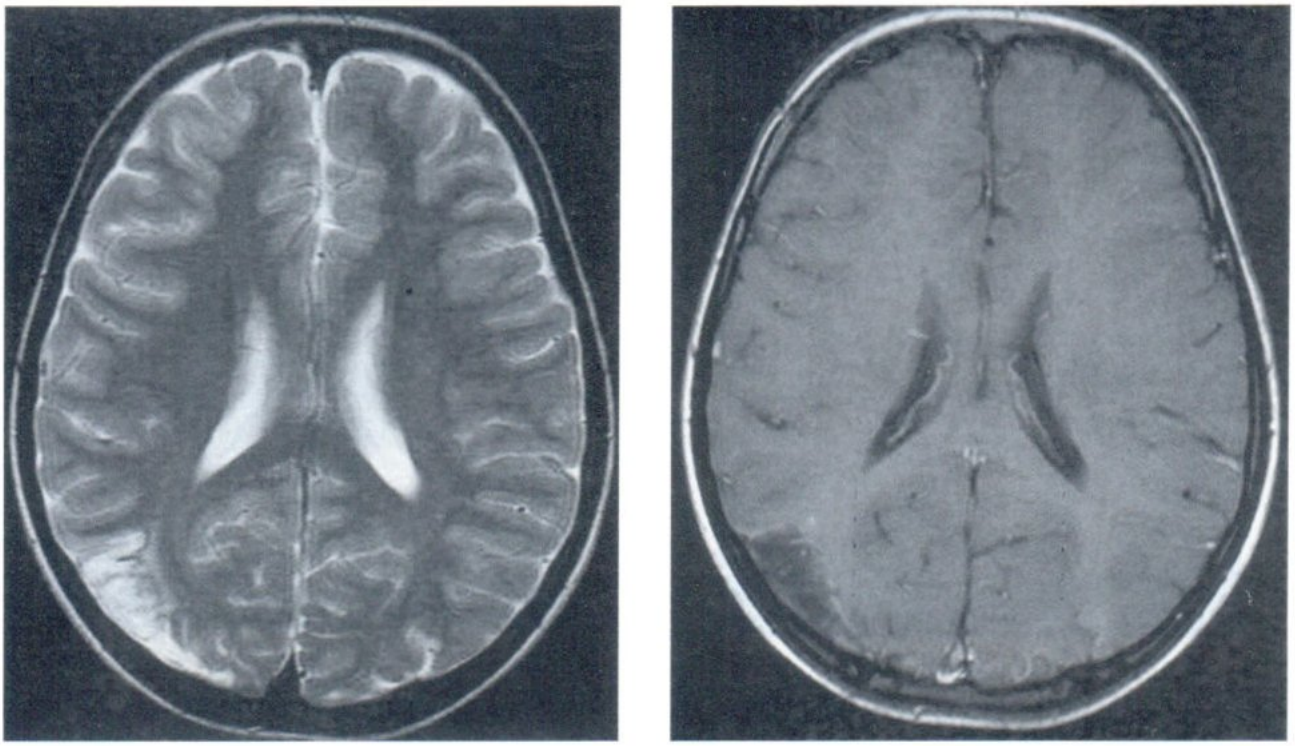
2a 2b

**Fig.2** **a.** Axial SE T1W and, **b.** axial SE T2W images reveal low signal intensity on T1W and high signal intensity on T2W in both occipital and parieto-occipital subcortical white matter and at the left frontal subcortical white matter. Edematous adjacent cortical gyri were also seen, with multiple small hemorrhagic foci seen as high signal intensity on T1W and low signal intensity on T2W along both parieto-occipital and occipital gyri indicating more severe damaging brain areas



3a 3b

**Fig.3** **a.** Axial SE T1W and, **b.** axial SE T2W images reveal marked resolution of the abnormal findings. However, small hemorrhagic foci along both occipital and parieto-occipital cortical gyri persisted.



4a 4b

**Fig.4** **a.** Axial SE T1w and, **b.** axial SE T2W images reveal no residual brain edema, but a few small areas of gliotic changes at the previous hemorrhage sites at the right parieto-occipital cortex.

## DISCUSSION

This child had underlying diseases of SLE and alfa Thalassemia trait. Thalassemia is well known as being associated with gallstones such as seen in this patient and, were the cause of his admission for a cholecystectomy. His CNS symptoms were originally explained as being part of the vasculitis process in SLE. However the typical areas of CNS involvement and the reversible nature of the CNS abnormality excluded the vasculitis as the cause of the CNS involvement in this patient.

SLE patients who develop symptoms in the CNS system have been called Neuropsychiatric Systemic Lupus Erythematosus (NPSLE). The underlying pathological basis of brain damage is not yet clear. MRI scans in many patients with clinically confirmed disease appear to be normal. In other cases, non-specific atrophy is seen with or without a history of steroid treatment.[3] Despite vasculitis often being included in the lists of possible cause of NPSLE, few SLE patients have true vasculitis.[4] More commonly, the sudden onset of neurological change in a patient with SLE suggests the possibility of an infarct or hypertensive encephalopathy. An infarct can range in size from a large vessel area to small, lacunar infarcts, and have been related to antiphospholipid antibodies which are commonly found in patients with SLE.[4] Multifocal areas of edema, especially in the posterior cerebral hemispheres, are the result of hypertensive encephalopathy which is common in SLE due to associated renal disease. Imaging findings are similar to other conditions such as eclampsia.

CT and MRI images of this patients were typical for hypertensive encephalopathy and also correlated well with his high blood pressure (160/110 mmHg). The cause of hypertension was thought to be his renal disease (lupus nephritis). Areas of low density on the CT, and abnormal signal intensity on the MRI, were thought to be due to elevated blood pressure exceeding the autoregulatory capacity of the brain vasculature, typically seen in bilateral subcortical white matter of regions supplied by posterior circulation (occipital, parietal, posterior temporal).

However, it may also have involved the frontal lobes and corpus callosum. Although hemorrhagic foci are commonly encountered in autopsy studies,[5] they are infrequently seen in imaging except in patient with chronic hypertension[6-7] or thrombocytopenia[2] as in this patient. Contrast enhancement may also be seen in the region of signal abnormality in a small number of cases. Typically the clinical and imaging findings are reversible after control of blood pressure, and these imaging findings can establish a diagnosis without the need for a biopsy. Most of the brain lesions in this patient reversed to normal, except for a few small areas at the right parieto-occipital and occipital lobe, which showed a more severe degree of cortical hemorrhage than the other parts of the brain. Hemorrhagic foci seen in the image, to our knowledge, may have been the indicators of more severely brain damaged regions which were less likely to fully recover.

Diffusion weighted image may be normal supporting the concept of increased interstitial fluid in the white matter and not ischemia.[8-9] However, in cases of prolonged seizures or hypertension, frank ischemia or infarction may result.[4] Preliminary reports of perfusion MR images have shown preserved or increased perfusion in affected regions of the brain, where acute ischemia is associated with decreased perfusion.[10] Because a history of hypertension or seizure may not always present, the characteristic imaging findings should allow the radiologist to suggest the diagnosis and subsequent clinical management should be focused on the treatment of the hypertension and its underlying causes.[1] Follow up imaging in 1-2 weeks will usually show improvement.[2]

## ACKNOWLEDGMENTS

We thank Dr. Wiwatana Tanomkiat for his kind suggestions during the preparation of this manuscript.

## REFERENCES

1. Jones BV, Egelhoff JC, Patterson RJ(1997) Hypertensive encephalopathy in children. AJNR,18:101-106.
2. Nusbaum AO, Fung KM, Atlas SW (2002) White matter disease and inherited metabolic disorders. In: Atlas SW (eds) Magnetic Resonace Imaging of the brain and the spine. 3rd edn. Vol. 1 Lippincott Williams&Wilkins, Philadelphia, pp510-511
3. York DH Jr (2002) Magnetic resonance imaging of CNS disease, A teaching file. 2nd edn. Mosby, Missouri
4. Hart B, Sibbitt W, Kornfeld M, Schmidt P, Brooks W (2002) Neuropsychiatric Manifestations of lupus: Radiologic findings and pathological correlates; presented as a scientific exhibit at the 40 th Annual Meeting of the ASNR, Vancouver, Canada, May 13-17.
5. Richards A, Graham D, Bullock R (1998) Clinicopathological study of neurological complications due to hypertensive disorders of pregnancy. J Neurol Neurosurg Psychiatry, 51:416-421.
6. Sanders TG, Clayman DA, Samchez-Ramos L, Vines FS, Russo L(1991) Brain in eclampsia: MR imaging with clinical correlation. Radiology, 80:475-478.
7. Schwartz RB, Jones KM, Kalina P, et al (1992) Hypertensive encephalopathy: findings on CT, MR imaging, and SPECT imaging in 14 cases. AJR,159:379-383.

8. Schwartz RB, Mulkern RV, Gudbjartsson H, et al (1998) Diffusion weighted MR imaging in hypertensive encephalopathy: clues to pathogenesis. AJNR,19:859-862.
9. Coley SC, Poter DA, Calamante F, et al (1999) Quantitative MR diffusion mapping and cyclosporine-induced neurotoxicity. AJNR, 20:1507-1510.
10. Engelter SR, Petrella JR, Albert MJ, et al (1999) Assessment of cerebral microcirculation in patient with hypertensive encephalopathy using MR perfusion imaging. AJR, 173: 1491-1493.