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CASE REPORT

# Posterior reversible encephalopathy syndrome associated with Henoch–Schönlein purpura

**Henoch-Schönlein purpurası ilişkili posterior reversible ensefalopati sendromu**

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***SUMMARY***

***Introduction:*** *Henoch–Schönlein purpura (HSP) is an acute vasculitis that primarily affects children. Central nervous system (CNS) involvement and the appearance of neurological complications preceding the characteristic rash are rare. Posterior reversible encephalopathy syndrome (PRES) and cerebral vasculitis are neurological complications of HSP.*

***Case Presentation:*** *We describe a case of 5-year-old girl who presented with seizures, loss of consciousness, and total loss of vision. Her HSP was atypical because the presenting symptoms were neurological complications rather than the characteristic rash, and her radiological features were compatible with PRES. She did not have renal impairment or hypertension. Her abnormal findings resolved following a standard course of corticosteroids.*

***Conclusion:*** *Our case is atypical because of the presence of CNS involvement before the characteristic rash with the absence of hypertension. The patient's HSP presented as a complication of cerebral vasculitis.*

***Keywords:*** *Henoch–Schönlein purpura, cerebral vasculitis, posterior reversible encephalography syndrome*

***ÖZET***

***Giriş:*** *Henoch-Schönlein purpurası (HSP) primer olarak çocukları etkileyen akut bir vaskülittir. Merkezi sinir sistemi tutulumu ve karakteristik döküntülerden önce nörolojik komplikasyonların ortaya çıkması nadirdir. Posterior reversibl ensefalopati sendromu (PRES) ve serebral vaskülit HSP nın nörolojik komplikasyonlarıdır.*

***Olgu:*** *Konvülziyon, bilinç kaybı ve total görme kaybı ile başvuran 5 yaşındaki bir kız olgusu sunulmuştur. Nörolojik semptomlar ön planda olup, karekteristik döküntü yoktu. Radyolojik bulguları PRES ile uyumluydu. Klinik bulgular HSP için atipikti. Böbrek tutulumu ve hipertansiyon yoktu. Klinik bulgular kortikosteroid tedavisi ile düzeldi.*

***Sonuç:*** *Hastamız tipik HSP döküntülerinden önce santral sinir sistemi bulgularının ortaya çıkması ve hipertansiyonun olmaması nedeniyle ilginçtir. HSP nin santral sinir sistemi komplikasyonu ile başvurmuştur.*

***Anahtar Kelimeler:*** *Henoch-Schönlein purpurası, serebral vaskülit, posterior reversibl ensefalopati sendromu*

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## INTRODUCTION

Henoch–Schönlein purpura (HSP) is an acute vasculitis that primarily affects children. The clinical features of HSP are palpable purpura, arthralgia or arthritis, abdominal pain, and glomerulonephritis. Purpura is generally the first sign of HSP and is mandatory for its diagnosis (1). Arthralgias and gastrointestinal involvement are common. Renal involvement is responsible for the long-term morbidity and mortality associated with the condition. Neurological symptoms and signs have been described in cases of HSP. Although headache and behavioral changes are relatively common, severe neurological manifestations such as seizures, intracerebral hematoma, hemiplegia, and encephalopathy are rare and potentially serious complications. Central nervous system (CNS) symptoms are rare and CNS involvement complicates the diagnosis of HSP.

Seizures occur in 53% of the patients with neurological manifestations and may be the first sign of HSP (4). CNS involvement may stem directly from the extension of vasculitis into the CNS or indirectly via systemic involvement, particularly via renal impairment and hypertension (4, 5). Here, we present a case of HSP in which neurological complications without renal involvement or hypertension presented before the onset of the characteristic HSP rash.

## CASE

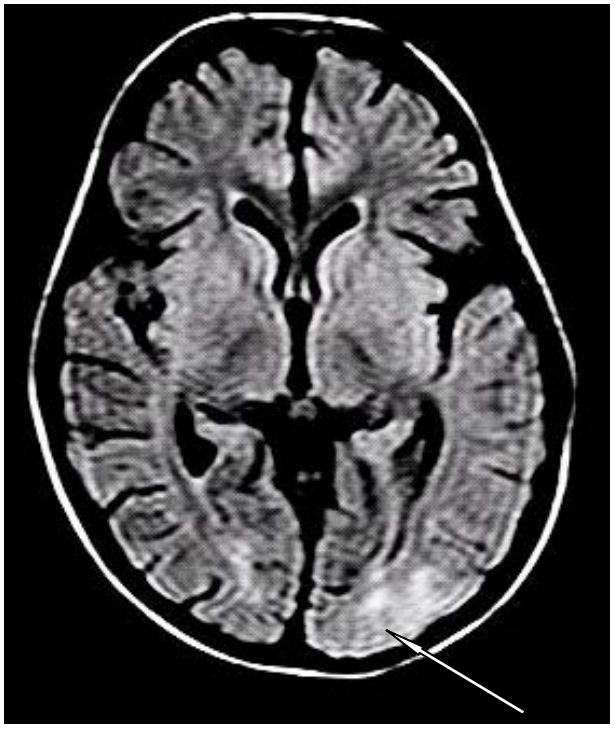
A 5-year-old girl presenting with severe headache, generalized tonic-clonic seizure, and loss of consciousness was admitted to our pediatric emergency clinic. On physical examination, her blood pressure was 108/70 mmHg (95th percentile, 110/72 mmHg) and her temperature was normal. In the emergency clinic, she complained of total loss of vision. After a short period of time, the patient experienced a generalized tonic-clonic seizure and became confused. Midazolam (0.1 mg/kg) was administered. Her vital signs, pupillary reflexes, and neurological

examination were normal. She had no history of toxic substance exposure. We learned that the patient had experienced abdominal pain and vomiting for 2 days before admission. Cranial computed tomography was normal. Examination of the cerebrospinal fluid (CSF) revealed pleocytosis (25 × 106 leukocytes/L) with normal glucose and protein. Since CNS infection could not be ruled out, we administered intravenous ceftriaxone and acyclovir. CSF cultures were negative. The blood count revealed hemoglobin 13.5 gr/dL, total leucocyte count 29.2 × 109/L, platelet count 578 × 109/L, erythrocyte sedimentation rate 13 mm/h, and C-reactive protein 15 mg/dL. Her urinalysis was normal. The serum electrolytes, urea, creatinine, serum C3 and C4 complement levels, serum lipid profile, liver function tests, coagulation parameters, and serum immunoglobulins were normal. Although she complained of severe abdominal pain and vomiting, her abdominal ultrasonography was normal.

On the second day, the patient regained consciousness and her vision recovered. Her blood pressure was normal. Cranial magnetic resonance imaging (MRI) revealed a signal abnormality in the left and right parieto-occipital regions predominantly affecting the cortical and subcortical white matter on the T2 fluid attenuated inversion recovery imaging (FLAIR) sequences (Fig. 1).

Her electroencephalogram (EEG) showed generalized slow-wave activity.

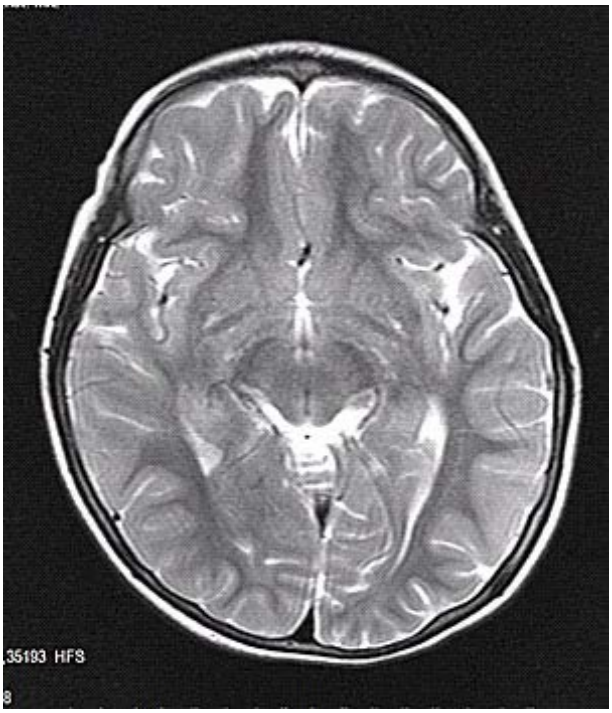
**Figure 1:** The initial brain magnetic resonance imaging (MRI) revealed a signal abnormality in the left and right parieto-occipital regions predominantly affecting the cortical and subcortical white-matter on T2 FLAIR sequences.



On the fourth day, she developed persistent palpable purpura and petechiae on the dorsal aspect of her feet and legs. The skin biopsy was consistent with leukocytoclastic vasculitis. A diagnosis of HSP was then made. The fecal occult blood test was positive. We administered steroids (2 mg/kg/day for 10 days) to treat her severe abdominal pain. The patient's neurological symptoms resolved on the seventh day, and her gastrointestinal symptoms regressed on the 10th day. She was followed after discharge from the hospital with no recurrence of seizures, visual loss, or abdominal pain. The cranial MRI and EEG findings were normal at the 6 week follow-up examination (Fig. 2).

The patient continues to be seen by a pediatric neurologist.

**Figure 2:** Follow-up MRI repeated 6 weeks after discharge from the hospital showed complete resolution of the previous cerebral lesions.




## DISCUSSION

Diagnosing HSP is difficult when the condition presents with symptoms other than purpura, such as occurs in 25–50% of cases. In one series, seizure or confusion was identified retrospectively in 17 of 244 (6.9%) cases of HSP, and one review identified CNS manifestations preceding onset of the rash in 16% of cases (6). Posterior reversible encephalopathy syndrome (PRES), described by Hinchey in 1996 (7), is a clinical and radiological spectrum that presents with vomiting, severe headache, altered sensorium, seizures, and visual disturbances. The pathophysiology of PRES is complex; however, two potential mechanisms are of interest: the loss of cerebral autoregulation resulting from uncontrolled hypertension, volume overload, or

endothelial injury resulting from various systemic conditions (8); and CNS vasculitis, which is suspected in patients with HSP who develop encephalopathy in the absence of severe hypertension or renal insufficiency. Indirect evidence, such as pleocytosis in the CSF and skin biopsy findings of leukocytoclastic vasculitis, suggests that CNS vasculitis can cause PRES in patients with HSP (2, 3, 5). PRES is diagnosed by the characteristic MRI and clinical findings. The neuroimaging is characterized by transient bilateral grey- and white-matter changes compatible with vasogenic edema in the posterior cerebral hemispheres, parieto-occipital areas, and cerebellum (7). In our patient, the clinical and imaging findings were remarkably similar to PRES as described by Hinchey: headaches, generalized seizures, sudden visual loss, and parieto-occipital white-matter abnormalities on MRI. Moreover, our patient did not have hypertension or renal insufficiency. Three previous studies have described cases of PRES related to HSP with normal blood pressure and neurological findings (2, 3, 9), and in one study, abdominal pain and neurological complications preceded the characteristic HSP rash, as in our patient (3). Renal involvement and CNS findings associated with hypertension have been reported in patients with PRES related to HSP (5, 10). We believe that, in our patient, PRES was attributable to CNS vasculitis rather than hemodynamic changes caused by severe hypertension. Previous investigators have suggested that cerebral vasculitis and PRES are closely related (3). The clinical and imaging features of the conditions are similar, and they affect the same brain regions (3, 4).

In these patients, the postictal EEG findings generally showed focal or generalized slow-wave activity; however, an epileptic focus was observed in a few cases (4). Our patient's EEG showed generalized slow-wave activity.

Henoch–Schönlein purpura is a self-limiting disease in most patients, and it is not clear whether steroid treatment affects the neurological complications. Rapid neurological recovery has been reported following high-dose intravenous steroid treatment (3). Our patient's neurological findings regressed following the administration of low-dose steroids and methylprednisolone pulses were not necessary. We used steroids to treat the patient's gastrointestinal symptoms. The diagnosis of HSP was not straightforward in our patient and we initially considered CNS diseases. PRES MRI findings might be confused with gliomatosis cerebri, progressive multifocal leukoencephalopathy, demyelinating conditions, and infarcts leading to unnecessary tests and treatment (7, 11). Our patient's gastrointestinal signs were pronounced and her neurological manifestations regressed quickly; therefore, we did not investigate other conditions.

Our patient recovered rapidly and her MRI findings and EEG changes resolved within 6 weeks. Previous investigators have reported that the MRI findings returned to normal in 4 to 6 weeks (2, 3).

In conclusion, we propose that HSP can present as a reversible neurological manifestation preceding the appearance of the characteristic rash in the absence of renal involvement or hypertension. MRI is central to the diagnosis. A pulse dose of methylprednisolone may be used as treatment. Furthermore, conservative treatment using a course of steroids may be effective, as was the case in our patient.

*The English in this document has been checked by at least two professional editors, both native speakers of English. (Textcheck Refnum 15032209).*

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