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Abstract

We report the case of a 4-year-old female who presented with high-grade fever and acute left-sided weakness. Initial non-contrast CT suggested acute encephalitis, leading to a provisional diagnosis of partially treated meningoencephalitis and illustrating the potential for diagnostic misinterpretation. However, contrast-enhanced MRI revealed leptomeningeal enhancement, cortical atrophy, and dilated deep medullary veins, consistent with atypical Sturge–Weber syndrome Type III. The patient improved with supportive management. This case highlights the diagnostic challenge of atypical SWS and underscores the crucial role of contrast-enhanced MRI when CT findings are nonspecific or misleading.

Keywords: Sturge–Weber syndrome, Atypical Sturge–Weber syndrome, Leptomeningeal angiomas, Magnetic resonance imaging, Pediatric neurocutaneous disorders

Key Clinical Message

Atypical Sturge–Weber syndrome Type III can present without facial port-wine stain or ocular involvement, leading to diagnostic delay. Contrast-enhanced MRI is crucial for establishing the diagnosis in pediatric patients presenting with acute focal neurological deficits and inconclusive CT findings. Increased clinical awareness of this rare variant can prevent misdiagnosis and improve neurological outcomes.

1.Introduction

Sturge–Weber syndrome (SWS) is a rare, sporadic neurocutaneous disorder caused by a developmental vascular anomaly involving the leptomeninges, skin, and eyes. It is classically characterized by facial port-wine stain, leptomeningeal angiomas, and ocular abnormalities such as glaucoma, with neurological manifestations including seizures and hemiparesis presenting early in life [1]. However, SWS exhibits a broad clinical spectrum, and atypical presentations without cutaneous manifestations have been increasingly recognized. In such cases, diagnosis becomes challenging, as patients may present with nonspecific focal neurological deficits or stroke-like episodes, often leading to misdiagnosis as infectious or vascular conditions [2]. Conventional non-contrast CT may be unrevealing in early or atypical disease. In contrast, contrast-enhanced MRI is the key diagnostic modality for detecting leptomeningeal angiomas, cortical atrophy, and abnormal venous drainage, including dilated deep medullary veins, facilitating early and accurate diagnosis [3].

Although atypical Sturge–Weber syndrome Type III has been reported previously, presentation with acute focal neurological deficits in the absence of both facial port-wine stain and ocular involvement remains uncommon and poses a significant diagnostic challenge. The present case illustrates how an atypical clinical presentation and nonspecific initial CT findings resulted in a provisional diagnosis of acute encephalitis before contrast-enhanced MRI established the correct diagnosis. By reporting this case, we aim to highlight the importance of maintaining a high index of suspicion for atypical Sturge–Weber syndrome and the pivotal role of contrast-enhanced MRI in children presenting with unexplained acute focal neurological deficits.

2. Case Presentation

A 4-year-old female child, one of twins, with previously normal developmental milestones, presented with a four-day history of high-grade fever followed by sudden-onset left-sided weakness. The weakness was acute in onset, non-progressive, and not associated with vomiting, seizures, slurring of speech, visual disturbances, or bowel or bladder incontinence. The child was otherwise active, independent, and attending school regularly. Antenatal, perinatal, and postnatal histories were unremarkable. There was no prior history of seizures, neurological illness, or hospital admissions. Family history was negative for neurological or genetic disorders. The patient was initially evaluated at a local healthcare facility where she received an undocumented injection, resulting in partial symptomatic improvement. Due to persistent symptoms, she was referred to a secondary care hospital for further evaluation, where a non-contrast CT scan of the brain was performed. Only the written CT report was available at the time of presentation, as the original CT images were not provided. On admission to a tertiary care hospital, the patient was febrile but conscious, alert, and oriented, with a Glasgow Coma Scale score of 15/15. Speech and hearing were intact. Neurological examination revealed normal motor power (5/5) on the right side and mild weakness (4/5) on the left side involving both upper and lower limbs. Cranial nerve examination was normal. No seizures were observed or documented during the hospital stay. Based on the history of high-grade fever, acute focal neurological deficit, and the initial non-contrast CT findings suggestive of acute encephalitis, the provisional diagnosis at admission was partially treated meningoencephalitis.

Laboratory investigations revealed hemoglobin of 10.9 g/dL, white blood cell count of 7.5/mm³, hematocrit of 31.5%, and reticulocyte count of 0.2%. Lymphocytes comprised 58.5% of the total

leukocyte count. Liver function tests showed mildly elevated AST (66.7 U/L), while ALT was within normal limits. PT/INR values were normal. Chest radiograph and echocardiography did not reveal any abnormalities. Neuroimaging included a non-contrast CT scan of the brain followed by contrast-enhanced MRI of the brain. Electroencephalography and ophthalmological evaluation were also performed. Cerebrospinal fluid (CSF) analysis was not performed.

The non-contrast CT brain demonstrated multiple variable-sized patchy to confluent hypodense areas involving the deep and subcortical white matter of both cerebral hemispheres (Fig.1), distributed asymmetrically in the bilateral parietal, occipital, and right frontal lobes (Fig.3). Posterior fossa structures and brainstem were normal, with no evidence of hydrocephalus, intracranial hemorrhage, or midline shift. These findings were interpreted as suggestive of acute encephalitis. Contrast-enhanced MRI of the brain revealed leptomeningeal enhancement, mild cortical atrophy in the right frontoparietal cortex, and dilated deep medullary veins(Fig. 4), findings consistent with atypical Sturge–Weber syndrome Type III (Fig. 5). FLAIR imaging also demonstrated bilateral periventricular white matter hyperintensities (Fig. 2). EEG was normal. Ophthalmological examination showed no evidence of glaucoma or visual abnormalities. No cutaneous manifestations, including port-wine stains, were present. Given the initial working diagnosis of partially treated meningoencephalitis, empirical treatment with intravenous ceftriaxone, vancomycin, acyclovir, dexamethasone, and supportive care was initiated pending further diagnostic evaluation. During a hospital stay of 7–8 days, gradual improvement in left-sided weakness was observed. At follow-up, the patient remained seizure-free, continued to demonstrate improvement in left-sided weakness, and was able to attend school and perform activities of daily living independently.

5. Discussion

The differential diagnosis in this patient included acute encephalitis, ischemic stroke, cerebral vasculitis, and other neurocutaneous disorders presenting with focal neurological deficits in childhood. The history of high-grade fever together with the initial non-contrast CT findings favored an infectious etiology, resulting in a provisional diagnosis of partially treated meningoencephalitis. However, the absence of seizures, progressive neurological deterioration, altered consciousness, and the characteristic ophthalmological or cutaneous manifestations of other neurocutaneous syndromes made these alternative diagnoses less likely. Contrast-enhanced MRI subsequently demonstrated leptomeningeal enhancement, mild cortical atrophy involving the right frontoparietal cortex, and dilated deep medullary veins, findings highly characteristic of Sturge–Weber syndrome Type III. FLAIR imaging also demonstrated bilateral periventricular white matter hyperintensities. Although these white matter changes were noted, they were nonspecific and were not considered the primary basis for the diagnosis. In contrast to acute encephalitis, ischemic stroke, or cerebral vasculitis, these imaging findings reflect leptomeningeal angiomas and abnormal venous drainage, which are characteristic neuroimaging features of SWS. This case highlights the diagnostic challenge posed by atypical Type III SWS, where the absence of classical clinical features and misleading initial CT findings may delay recognition. It also emphasizes that contrast-enhanced MRI, interpreted in conjunction with the clinical presentation, remains the cornerstone for establishing an accurate diagnosis in atypical pediatric cases.

Published reports of Sturge–Weber syndrome Type III remain limited because of the rarity of this subtype. The published cases summarized in Table 1 demonstrate considerable clinical heterogeneity despite the common absence of facial port-wine stain and ocular involvement. Patients presented with diverse neurological manifestations, including focal and generalized

seizures, migraine-like episodes, visual disturbances, hemiparesis, and cognitive impairment [4–7]. Despite these differences in clinical presentation, contrast-enhanced MRI consistently demonstrated characteristic leptomeningeal angiomatosis and associated vascular abnormalities, establishing the diagnosis in each case [4–7]. Compared with these reports, our patient presented with acute febrile hemiparesis without seizures, initially mimicking an infectious neurological disorder, further emphasizing the diagnostic complexity of Type III SWS and the importance of early contrast-enhanced MRI for accurate diagnosis.

Table 1. Comparison of the clinical characteristics of published cases of Sturge–Weber syndrome Type III without facial port-wine stain.

Author (Year)	Age / Sex	Neurologic Presentation	Neuroimaging Findings	Facial Nevus / Ocular Findings	Management / Outcome
Siri et al., 2013 [4]	5 patients (6–20 years; 3F/2M)	Predominantly focal epilepsy, generalized seizures, febrile seizures, migraine-like attacks, learning difficulties, attention deficit	Unilateral cortical calcifications, leptomeningeal/pial enhancement, cortical atrophy (mostly parieto-occipital involvement)	No facial port-wine stain; ocular examination normal	Two patients underwent epilepsy surgery with good seizure control; others showed variable seizure outcomes
Jordan et al., 2016 [5]	9-year-old male	Status migrainosus, focal seizures,	Left occipital–parietal pial angiomatosis with	No facial nevus; no glaucoma	Improved with antiepileptic medication,

		transient visual field defect	cortical calcification and atrophy		aspirin and migraine management
Manohari et al., 2023 [6]	Middle childhood / Female	Focal seizures with hemiparesis, initially suspected ischemic stroke	Gyriform calcifications, leptomeningeal enhancement, cerebral hemiatrophy, enlarged choroid plexus	No facial nevus	Diagnosis established by MRI; improved with medical treatment
Laaraje et al., 2025 [7]	11-year-old female	Drug-resistant focal epilepsy, mild hemiparesis, learning difficulties (ADHD)	Cortical calcifications, hemispheric atrophy, enlarged choroid plexus, dilated deep medullary veins	No facial nevus; no ocular involvement	Persistent epilepsy despite multiple antiseizure medications

ADHD, attention-deficit/hyperactivity disorder; F, female; M, male; MRI, magnetic resonance imaging

In contrast to the published cases summarized in Table 1, our patient presented with acute febrile hemiparesis without seizures, initially leading to a provisional diagnosis of partially treated meningoencephalitis based on non-contrast CT findings. The combination of fever, acute focal neurological deficit, misleading initial CT findings, absence of cutaneous and ocular manifestations, and subsequent diagnosis by contrast-enhanced MRI further expands the limited spectrum of reported pediatric Type III Sturge–Weber syndrome presentations. This case highlights the importance of maintaining a high index of suspicion for Type III SWS in children presenting with unexplained focal neurological deficits despite the absence of classical phenotypic features and reinforces the pivotal role of contrast-enhanced MRI in establishing the diagnosis [4–7].

Ocular involvement is a well-recognized component of classical Sturge–Weber syndrome and frequently contributes to early diagnosis. However, the published Type III cases summarized in Table 1 consistently lacked the characteristic facial port-wine stain, while ocular involvement

was absent or minimal in most patients [4–7]. In contrast, the complete absence of ocular manifestations in our patient strongly supports a diagnosis of Type III SWS and underscores the diagnostic challenge posed by this variant. The lack of ophthalmological findings removes another major clinical indicator, further delaying recognition and reinforcing reliance on contrast-enhanced MRI for diagnosis.

Management of Type III SWS is individualized and depends primarily on neurological manifestations rather than the presence of cutaneous disease. The majority of reported Type III cases were managed with antiseizure medications, low-dose aspirin when clinically indicated, and supportive neurological care [4–7], whereas surgical intervention was generally reserved for patients with medically refractory epilepsy [8,9]. In contrast, our patient demonstrated gradual neurological recovery with conservative management and remained seizure-free during follow-up, highlighting that favorable outcomes may be achieved when atypical Type III SWS is recognized early and managed appropriately.

Severe neurological involvement in SWS may necessitate surgical procedures such as posterior quadrant disconnection, particularly in patients with intractable epilepsy or progressive hemispheric dysfunction [8]. Such interventions underscore the potential morbidity associated with delayed diagnosis or uncontrolled disease. The favorable clinical course observed in our patient, characterized by motor recovery and absence of seizures, contrasts sharply with these severe presentations and reinforces the prognostic benefit of early recognition and appropriate monitoring.

At the extreme end of the disease spectrum, hemispherectomy has been reported in pediatric patients with devastating neurological impairment and refractory seizures due to extensive leptomeningeal involvement [9]. These cases illustrate the potential long-term consequences of

SWS when diagnosis is delayed or disease progression is unchecked. In comparison, our case represents a milder phenotype, further supporting the concept that SWS encompasses a continuum of disease severity.

Collectively, the published cases summarized in Table 1 and our patient illustrate the considerable clinical heterogeneity of Type III Sturge–Weber syndrome [4–7]. Despite differences in clinical presentation, contrast-enhanced MRI consistently proved to be the key diagnostic modality for establishing the diagnosis, particularly in patients lacking the classical facial and ocular manifestations [4–7]. While non-contrast CT imaging may be useful for excluding acute intracranial hemorrhage, it lacks sensitivity for detecting leptomeningeal angiomas and may lead to misdiagnosis, whereas early utilization of contrast-enhanced MRI is essential for accurate diagnosis. By documenting this case, we add to the limited body of literature on pediatric Type III SWS and reinforce the need for imaging-driven diagnosis in atypical presentations.

6. Conclusion

Atypical Sturge–Weber syndrome Type III is an uncommon entity that may present without classical cutaneous or ocular manifestations, leading to diagnostic delay. This case highlights the importance of considering Sturge–Weber syndrome Type III in pediatric patients presenting with acute focal neurological deficits in the absence of classical cutaneous or ocular manifestations. Contrast-enhanced MRI is crucial for accurate diagnosis, as non-contrast CT may be non-specific or misleading in atypical presentations. Increased awareness of this rare variant may facilitate timely recognition, appropriate management, and improved neurological outcomes.

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Consent

Written informed consent was obtained from the patient's parents for publication of this case report and accompanying images.

Conflict of Interest

No conflict of interest.

Ethics approval and consent to participate

Not applicable. This manuscript reports a single case and no separate research procedures were performed beyond routine clinical care.

Availability of data and materials

The data supporting the findings of this case report are available from the corresponding author on reasonable request, subject to applicable privacy and confidentiality requirements.

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Authors' contributions

A.F.M, S.A.S, M.S.K, U.S contributed to the conception and design of the case report, F.N, U.I,

A.S collected and interpreted the clinical data, and U.I, F.N , P.S drafted the manuscript. All authors reviewed and approved the final manuscript.

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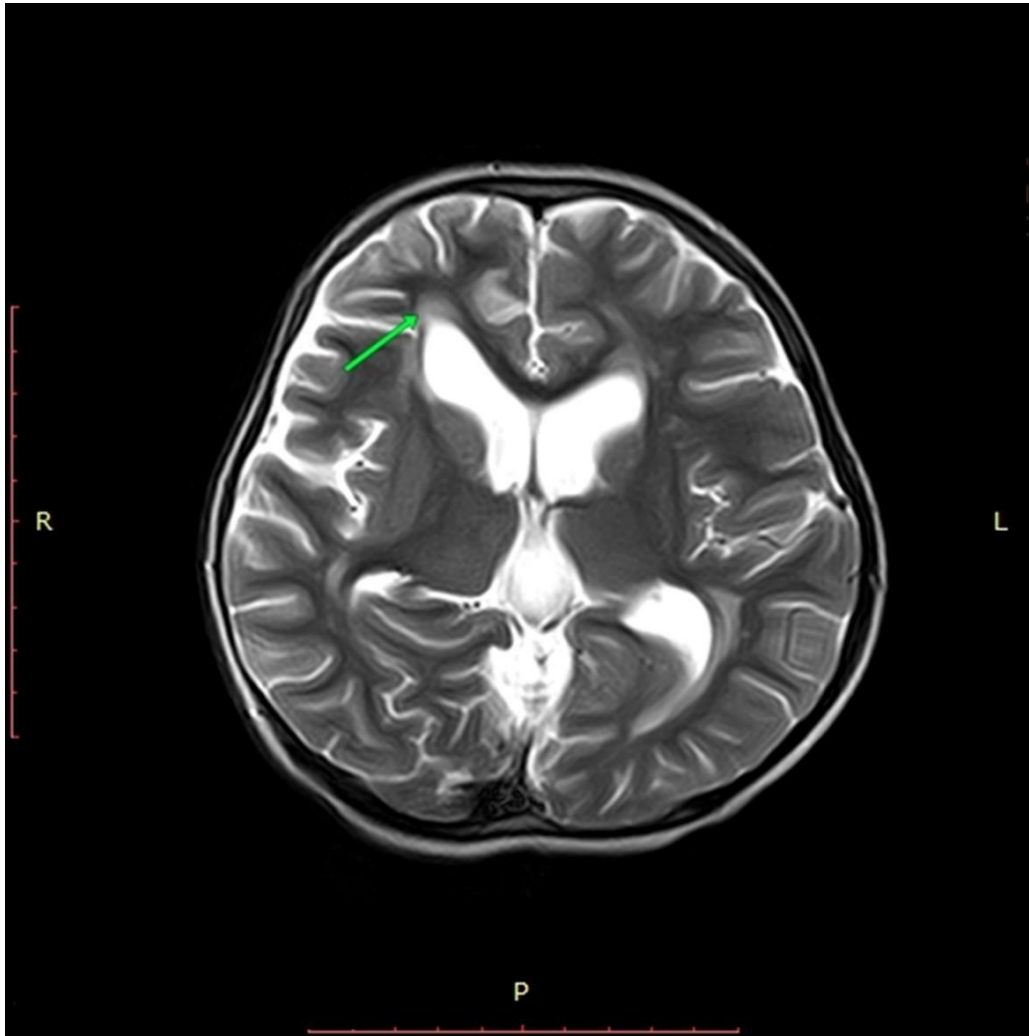


Figure 1: Axial T2-weighted MRI of the brain demonstrating mild cortical atrophy involving the right frontoparietal cortex with subtle hyperintense signal changes in the adjacent subcortical white matter (arrow).

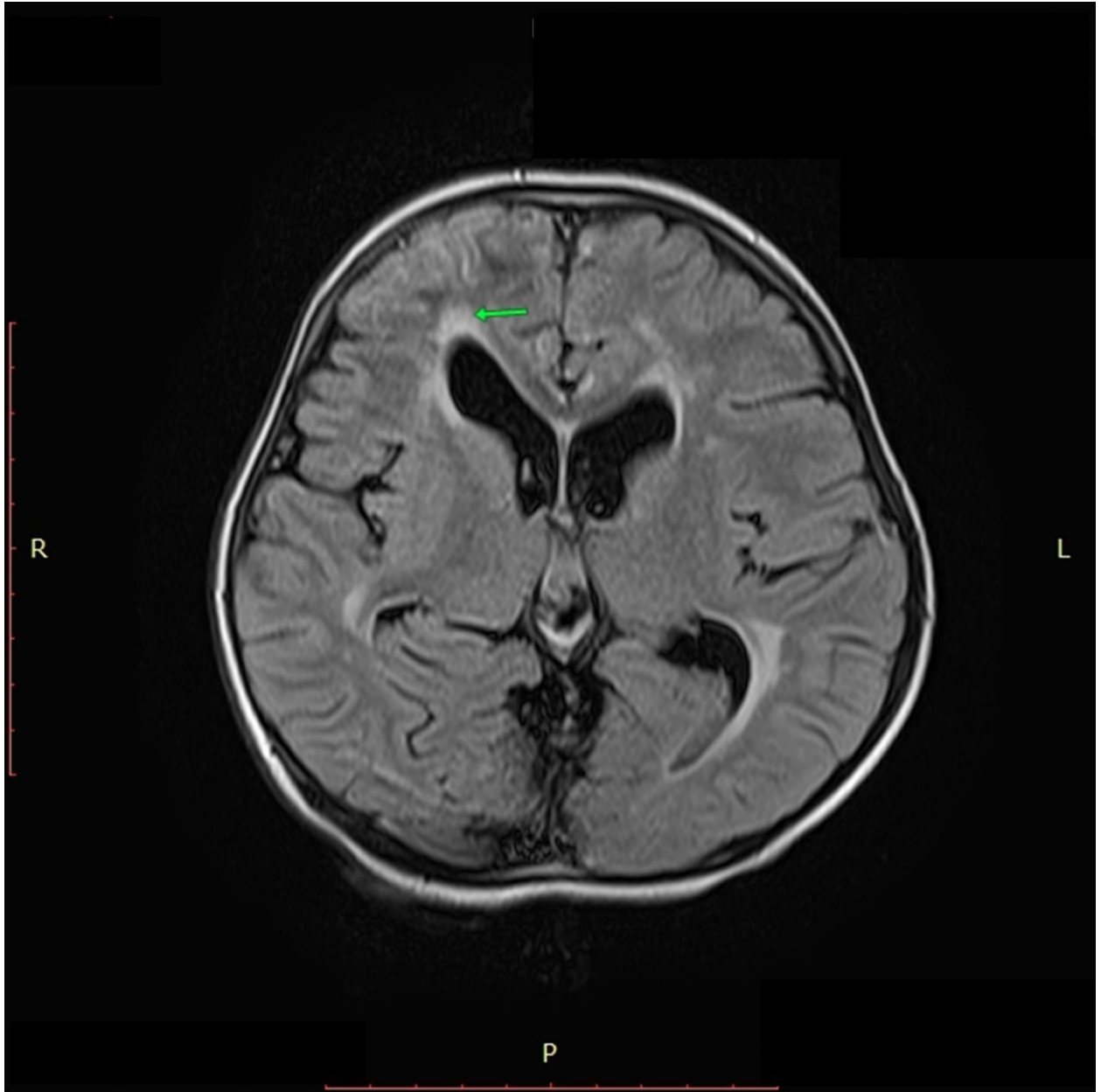


Figure 2: Axial FLAIR MRI sequence showing mild cortical atrophy involving the right frontoparietal cortex with more conspicuous hyperintense signal intensity in the subcortical white matter (arrow), better delineated compared to the T2-weighted image.



Figure 3: Axial contrast-enhanced T1-weighted MRI revealing gyriform (pial) enhancement involving the right frontoparietal cortex (arrows), consistent with leptomeningeal angiomatosis characteristic of Sturge–Weber syndrome.



Figure 4: Coronal contrast-enhanced T1-weighted MRI demonstrating the extent of gyriform pial enhancement within the right frontoparietal cortex (arrow), further supporting the diagnosis of leptomeningeal angiomatosis.

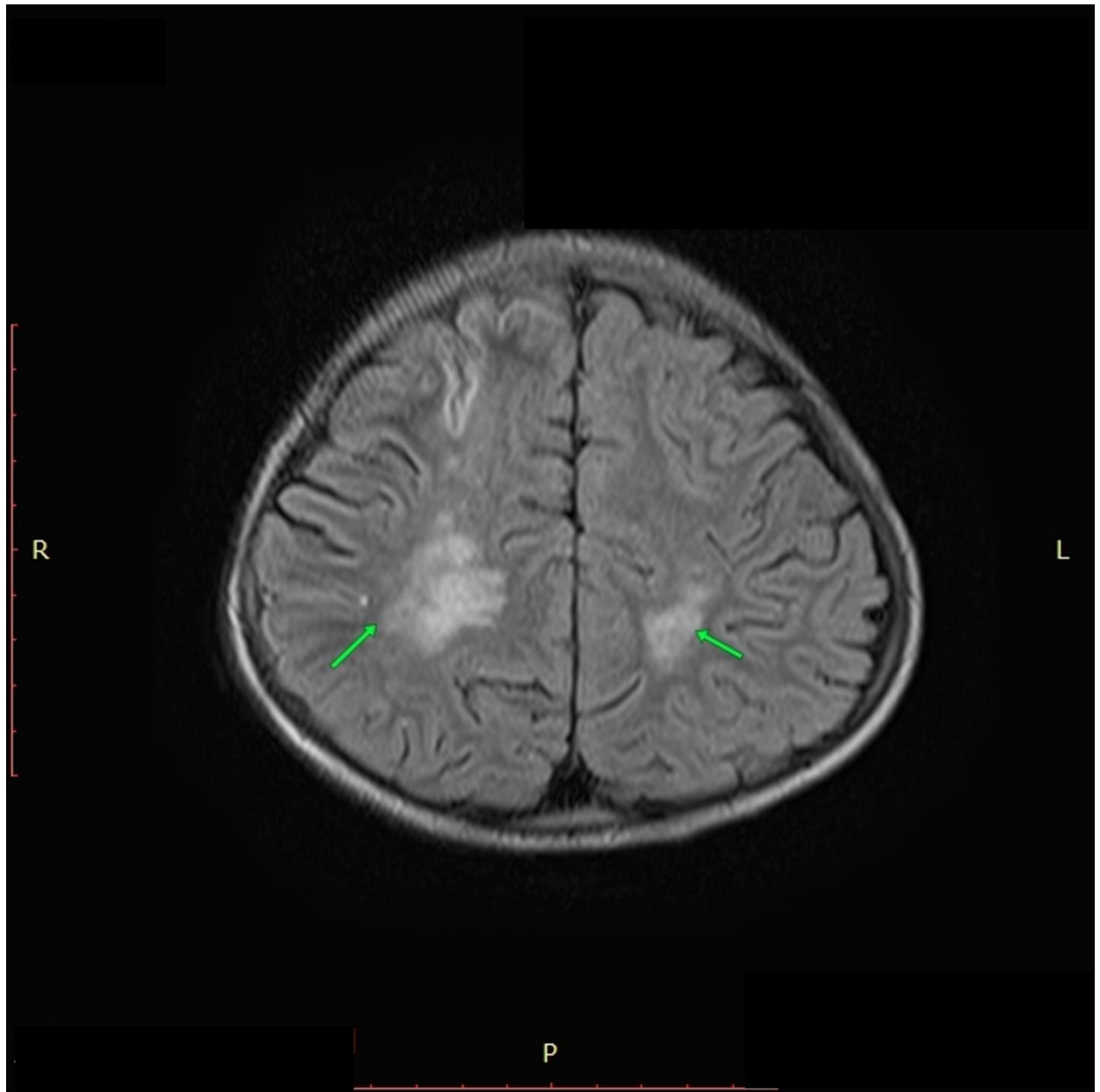


Figure 5: Axial FLAIR MRI (higher slice) demonstrating bilateral periventricular white matter hyperintensities (arrows).