World Journal of Advanced Research and Reviews
eISSN: 2581-9615 CODEN (USA): WJARAI
Cross Ref DOI: 10.30574/wjarr
Journal homepage: https://wjarr.com/

(CASE REPORT)

Check for updates

Atypical posterior reversible encephalopathy syndrome presenting with cortical laminar necrosis: A case report

Zineb Essolaymany [1, 2, *], Zineb Ezzoulali [1, 2], Hajar Ouazzani [2], Ismail Chaouche [1], Amal Akammar [2], Nizar El Bouardi [1], Badreddine Alami [1], Siham Bouchal [3], Naima Chtaou [3], Faouzi Belahsen [3], Meryem Boubbou [2], Mustapha Maâroufi [1] and Moulay Youssef Alaoui Lamrani [1]

[1] *Department of Radiology, Faculty of Medicine and Pharmacy, Sidi Mohammed Ben Abdellah University, Hassan II University Hospital, Fez, Morocco.*
[2] *Department of Mother and Child Radiology, Faculty of Medicine and Pharmacy, Sidi Mohammed Ben Abdellah University, Hassan II University Hospital, Fez, Morocco.*
[3] *Department of Neurology, Faculty of Medicine and Pharmacy, Sidi Mohammed Ben Abdellah University, Hassan II University Hospital, Fez, Morocco.*

World Journal of Advanced Research and Reviews, 2025, 27(02), 224-230

Publication history: Received on 1 June 2025; revised on 30 July 2025; accepted on 02 August 2025

Article DOI: https://doi.org/10.30574/wjarr.2025.27.2.2780

Abstract

Posterior reversible encephalopathy syndrome (PRES) represents a neurological disorder with varied clinical presentation and typical imaging findings of predominant vasogenic edema in the parieto-occipital regions. The imaging abnormalities found in PRES are usually reversible. In some rare cases, it can lead to irreversible damage to the cerebral parenchyma, leading to cortical laminar necrosis, which is an atypical feature of PRES. We describe a unique case of a patient admitted for an atypical PRES presenting with generalized tonic-clonic seizures and imaging features of cortical laminar necrosis. To our knowledge, our case is only the third one reported of PRES carrying imaging features of cortical laminar necrosis.

Keywords: Posterior Reversible Encephalopathy Syndrome; Cortical Laminar Necrosis; CT; MRI; Case report

1. Introduction

Posterior reversible encephalopathy syndrome (PRES) is a distinctive neurotoxic condition with a non-specific clinical presentation and unique neuroradiological findings [1, 2]. The pathophysiologic mechanism behind it hasn't been fully clarified. Major theories focus on disruption of the blood-brain barrier due to elevation of intracranial pressures and endothelial injury, resulting in vasogenic edema with a parieto-occipital white matter predominant pattern [2]. PRES is seen in situations of hypertensive emergencies, renal diseases, autoimmune disorders and use of cytotoxic medications [3]. The term may be misleading as PRES can concern or expand beyond the parieto-occipital regions and can lead to permanent brain lesions. The target of the treatment is the underlying cause, with reversibility of symptoms and imaging abnormalities in most cases [2]. Infrequently, it can result in irreparable injury to the cerebral parenchyma, leading to cortical laminar necrosis (CLN). We describe a rare case of a patient admitted for an atypical PRES presenting with imaging features of cortical laminar necrosis. To our knowledge, our case is only the third one reported of PRES carrying imaging features of cortical laminar necrosis.

* Corresponding author: Z. Essolaymany

2. Case presentation

58-year-old female patient, treated for diabetes for 12 years, admitted to the emergency room (ER) for management of a generalized tonic-clonic seizure followed by a post-ictal state.

Anamnesis revealed that the patient had left-sided cataract blindness and had undergone an appendectomy 8 days prior to her admission to the ER. The post-operative course was marked by the onset of a severe headache followed by a generalized tonic-clonic seizure, concomitant with an episode of arterial hypertension. Clinical assessment revealed a conscious but mildly confused patient with complete blindness. Blood pressure was correct (120/60 mmHg). Biological tests showed hyperglycemia, renal failure with uremia at 1.47 g/l and creatininemia at 21 mg/l. It also showed hyponatremia at 127 mmol/l and a positive infectious workup, notably urine culture that was positive for multi-resistant E. Coli.

On the first line of investigations, a Head CT was performed. It demonstrated bilateral and symmetrical white and gray matter hypoattenuation involving the posterior occipitoparietal regions, as well as the posterior regions of the cerebellum, consistent with vasogenic edema. There was no CT evidence of hemorrhage or calcifications (**Figure 1**). There was no pathological contrast enhancement in the affected regions and the dural venous sinuses were permeable.

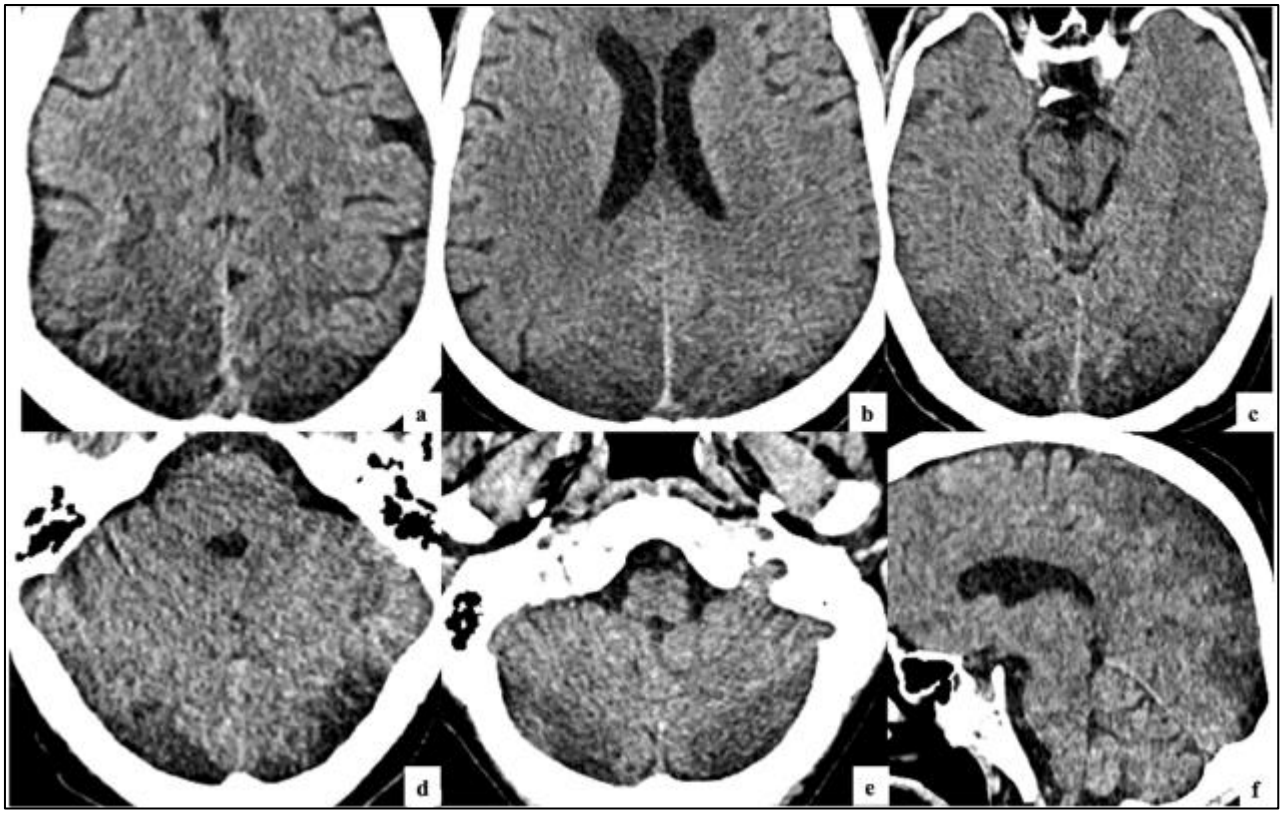

Figure 1 (a – f). Axial and sagittal non-contrast CT scan images : Symmetrical abundant vasogenic edema involving the posterior occipitoparietal regions and the posterior cerebellum, without obvious hemorrhage or calcifications

Complementary MRI showed abundant edema in the posterior frontal and occipitoparietal regions, as well as in the posterior cerebellum, with a hyperintense signal on diffusion-weighted imaging (DWI), consistent with cortical infarction (**Figure 2**). It also revealed gyriform cortical T1WI hyperintensity with GRE hypointense signals, compatible with cortical laminar necrosis with CT negative petechial hemorrhage. Additional 3D Time-of-flight MRA did not demonstrate any vascular occlusion (**Figure 3**).

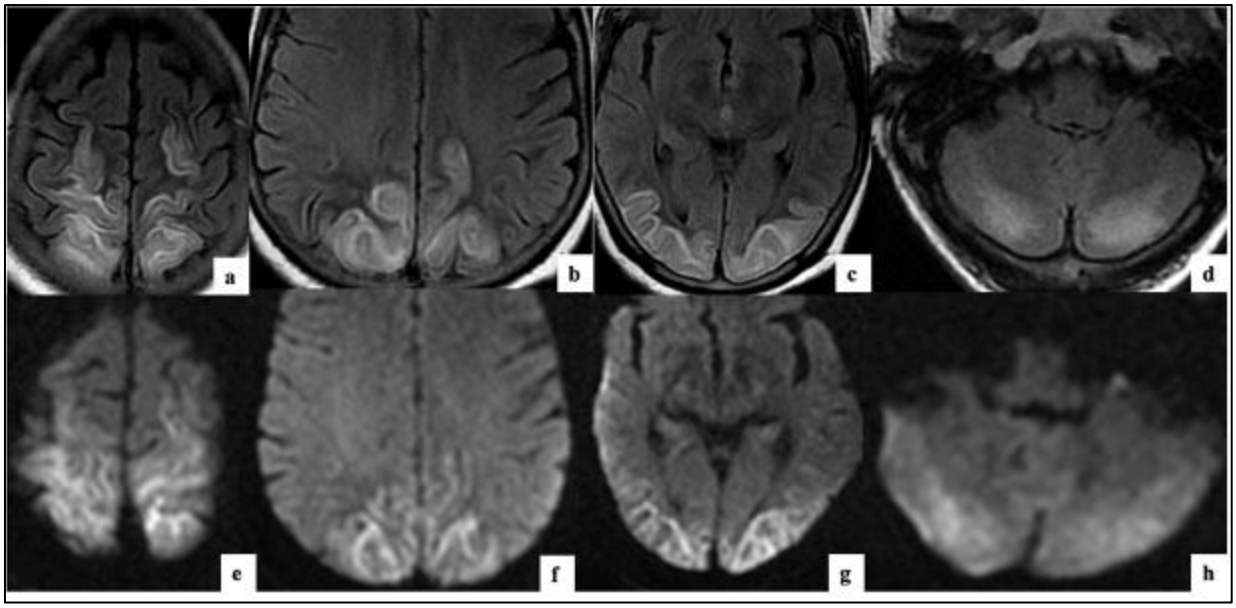

Figure 2 (a – h). Axial MRI, FLAIR and DWI images : Hyperintense FLAIR (a – d) signal in the posterior frontal and occipitoparietal regions, as well as in the posterior cerebellum, with a hyperintense signal on DWI (e – h), consistent with cortical infarction

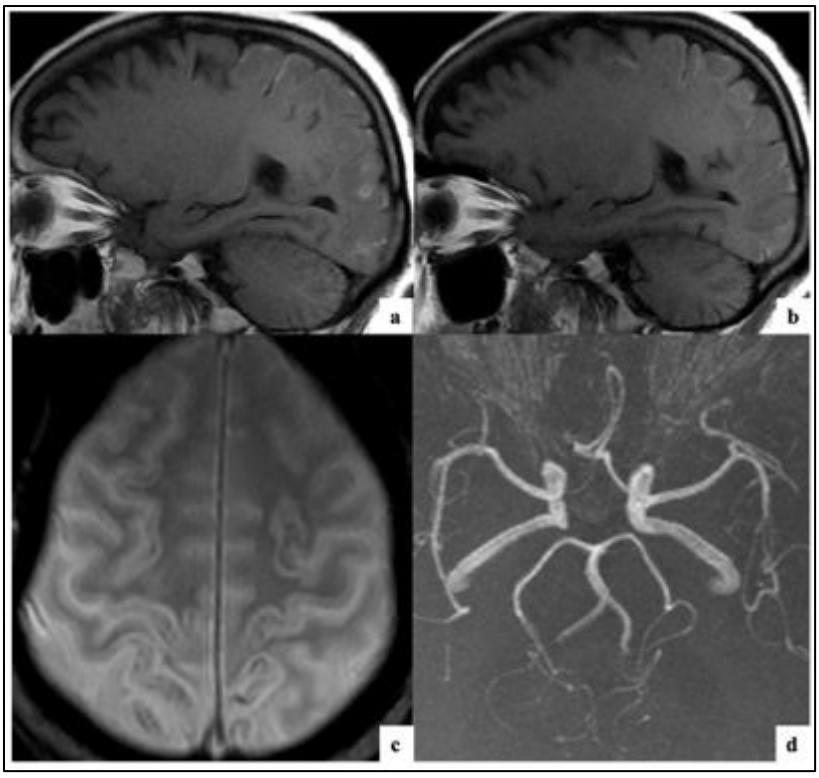

Figure 3 Sagittal T1WI (a, b), axial GRE (c) and 3D TOF MRA (d) images : Gyriform cortical T1WI hyperintensity with GRE hypointense susceptibility signals, compatible with cortical laminar necrosis, with no vascular occlusion

The patient was put on anticonvulsants and on intravenous antibiotics for her urinary tract infection. She has not had a convulsive seizure since, with improved visual acuity in the right eye. The continuous monitoring of vital parameters permitted the exclusion of hypertension or other dysautonomic features that, if present, could have justified PRES. Her follow-up day 14 head CT shows slightly increased cerebral edema. It was again negative for obvious hemorrhage (Figure 4).

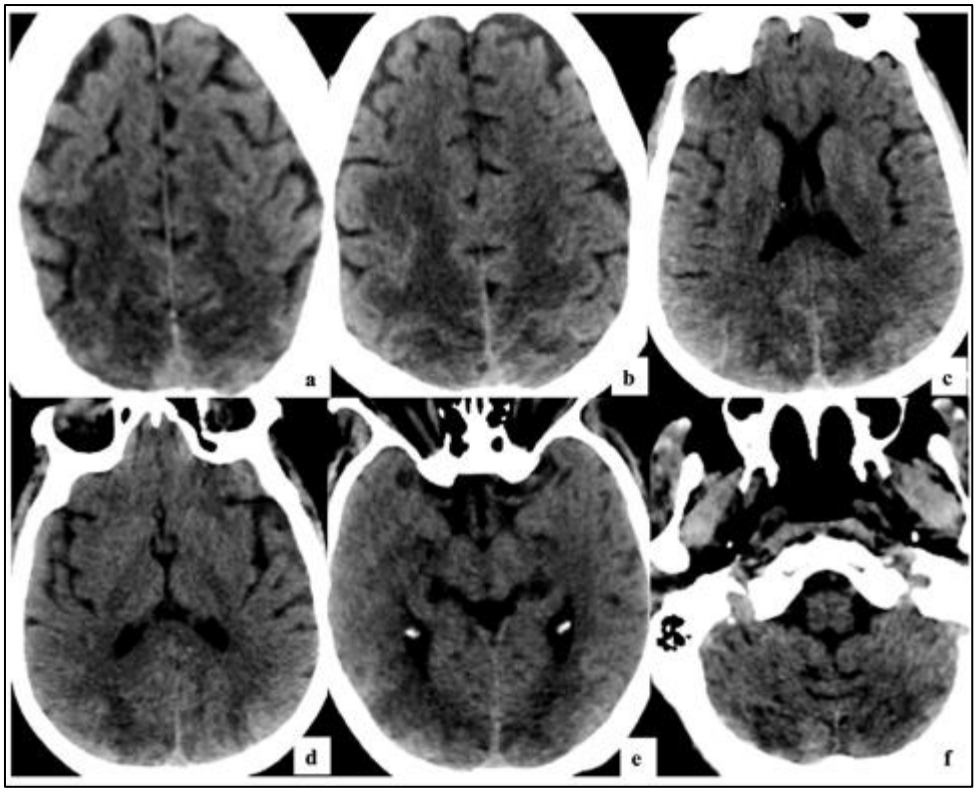

Figure 4 (a – f). Axial non-contrast CT scan images : Follow-up day 14 head CT shows slightly increased cerebral edema, with no obvious hemorrhage

The final diagnosis was atypical PRES associated with cortical laminar necrosis and CT negative hemorrhage. After management of her urinary tract infection and her acute renal failure secondary to dehydration, the patient was discharged in a stable condition, on anticonvulsant therapy. The patient will be in close follow-up in the Department of Neurology.

3. Discussion

PRES can be present in multiple clinical scenarios, but is most frequently observed with arterial hypertension, eclampsia, renal failure, sepsis, autoimmune diseases and use of certain cytotoxic medications. The pathophysiologic mechanisms behind this condition remain controversial and have not been fully elucidated yet [3], but are thought to build up to altered integrity of the blood-brain barrier. There are currently two main precipitant theories. The breakthrough theory proposes designate high arterial pressure as the key factor [2] ; elevated arterial blood pressure will eventually disrupt the cerebral autoregulation, leading to blood-brain barrier dysfunction and vascular leakage with resultant vasogenic edema [4]. The posterior cerebral regions are more exposed to the risk since the basilar arterial vasculature lacks sympathetic tone [2]. Dysfunction of the endothelium is the basis of the second theory, which may be secondary to circulating toxins, justifying the absence of elevated arterial pressures in some patients with PRES [2, 5].

PRES can have a wide range of clinical manifestations, ranging from headaches to altered mental status, depending on the area of the brain that is affected. Seizures usually reveal this syndrome and are described in 74 – 87% of patients ; generalized tonic-clonic seizures are the most common type [6]. Encephalopathy has been found in 28 – 94% of patients, ranging from confusion, cognitive impairment to unresponsiveness and coma [6]. Visual disturbances have been reported in 39% of patients, including altered visual acuity, diplopia, visual field deficits, cortical blindness and visual hallucinations [6]. Focal neurological deficits have been observed in 19% of patients [6, 7]. Myelopathic symptoms have rarely been demonstrated [8].

Typical PRES is seen on imaging studies as bilateral and symmetrical white matter vasogenic edema, predominating in the posterior cerebral regions [3]. The parietal and occipital lobes are frequently involved, but PRES can also be found in a non-posterior distribution, including the brainstem, the cerebellum, the corpus callosum and other cerebral areas, such as the frontal lobes (up to 68%) and the inferior temporal lobes (up to 40%) [2, 9]. Vasogenic edema can also involve, in addition of subcortical white matter, the cerebral gray matter.

A head CT scan is frequently the first-line imaging modality in the setting of an acute neurological presentation and demonstrates white matter hypoattenuation [3]. MRI plays a critical part in the diagnosis of this syndrome, by detecting vasogenic edema that is clearly identified on the T2-weighted and T2 FLAIR (fluid-attenuated inversion recovery) sequences [10]. PRES can be differentiated from acute cerebral infarction (notably involving posterior circulation) with a hyperintense signal on DWI and a decreased signal on ADC, orienting towards cytotoxic edema. In case of PRES, vasogenic edema can show hyperintense signal on DWI, with no corresponding decreased signal on ADC [2]. Less commonly, atypical PRES may progress to cortical infarction ; Diffusion restriction, consistent with an infarct, is present in 11 – 26% of the cases [1].

The radiological anomalies found in PRES are generally reversible, as the name suggests, once the triggering event has been treated. In some rare cases, areas of restricted diffusion can ultimately result in irreversible injury to the brain parenchyma, leading to cortical laminar necrosis [2]. CLN is an atypical feature of PRES. Cortical laminar necrosis (CLN) is a selective damage to the most metabolically active cerebral cortical cell layers. It is believed to reflect pan-necrosis of the different cells in the affected area, with reactive gliosis and deposition of lipid-rich macrophages. The preserved glial cells and microvessels produce certain macromolecules or minerals in the affected laminae [11]. Features of cortical laminar necrosis on CT can be subtle, manifesting as changes in attenuation in a gyriform pattern, that can be hypodense or hyperdense, depending on timing. Hemorrhage and calcifications are not seen in the early stages [12]. On MRI, the presence of a gyriform T1 hyperintense signal in the absence of gradient susceptibility is typical for CLN. Hemorrhage with CLN is extremely rare. This T1 high signal is believed to be caused by the accumulation of denatured proteins in dying cells and/or lipid-rich macrophages [13]. T1 curvilinear hyperintensities can be detected in the first three to five days, but become more evident after two weeks, with a peak of intensity after one month, and then slowly fade afterwards [12, 13]. To our knowledge, our case is one of very few reported cases of PRES carrying imaging features of cortical laminar necrosis and CT negative hemorrhage [14, 15].

List of abbreviations

- PRES : Posterior reversible encephalopathy syndrome
- CLN : Cortical laminar necrosis
- ER : Emergency room
- CT : Computerized Tomography
- MRI : Magnetic Resonance Imaging
- T1WI : T1 weighted image
- T2WI : T2 weighted image
- FLAIR : Fluid-attenuated inversion recovery
- GRE : Gradient-recalled Echo
- DWI : Diffusion-weighted imaging
- ADC : Apparent diffusion coefficient
- MRA : Magnetic Resonance Angiography

4. Conclusion

Atypical imaging findings may be encountered with PRES. MRI plays an important role in the correct diagnosis of atypical PRES, particularly with cortical laminar necrosis and CT negative hemorrhage.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare that they have no competing interests.

Statement of ethical approval

The study was conducted in accordance with ethical principles. Ethical approval was not required for this case report, as per institutional policy. Written informed consent was obtained from the patient.

Statement of informed consent

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

Author's contribution

- ZE is the corresponding author, she participated in the organization and writing of the article and studying the case with ZE.
- Professor NE, BA and MM supervised working and validated the figures.
- Professor of radiology YL read and allowed the article for publication.

References

[1] Bartynski WS. Posterior reversible encephalopathy syndrome, part 1: fundamental imaging and clinical features. AJNR Am J Neuroradiol. 2008 Jun;29(6):1036-42. doi: 10.3174/ajnr.A0928. Epub 2008 Mar 20. PMID: 18356474; PMCID: PMC8118828.

[2] Anderson RC, Patel V, Sheikh-Bahaei N, Liu CSJ, Rajamohan AG, Shiroishi MS, Kim PE, Go JL, Lerner A, Acharya J. Posterior Reversible Encephalopathy Syndrome (PRES): Pathophysiology and Neuro-Imaging. Front Neurol. 2020 Jun 16;11:463. doi: 10.3389/fneur.2020.00463. PMID: 32612567; PMCID: PMC7308488.

[3] Bartynski WS. Posterior reversible encephalopathy syndrome, part 2: controversies surrounding pathophysiology of vasogenic edema. AJNR Am J Neuroradiol. 2008 Jun;29(6):1043-9. doi: 10.3174/ajnr.A0929. Epub 2008 Apr 10. PMID: 18403560; PMCID: PMC8118813.

[4] Paulson OB, Waldemar G, Schmidt JF, Strandgaard S. Cerebral circulation under normal and pathologic conditions. Am J Cardiol. 1989 Feb 2;63(6):2C-5C. doi: 10.1016/0002-9149(89)90396-2. PMID: 2643850.

[5] Marra A, Vargas M, Striano P, Del Guercio L, Buonanno P, Servillo G. Posterior reversible encephalopathy syndrome: the endothelial hypotheses. Med Hypotheses. 2014 May;82(5):619-22. doi: 10.1016/j.mehy.2014.02.022. Epub 2014 Mar 1. PMID: 24613735.

[6] Hinduja A. Posterior Reversible Encephalopathy Syndrome: Clinical Features and Outcome. Front Neurol. 2020 Feb 14;11:71. doi: 10.3389/fneur.2020.00071. PMID: 32117030; PMCID: PMC7034490.

[7] Fugate JE, Claassen DO, Cloft HJ, Kallmes DF, Kozak OS, Rabinstein AA. Posterior reversible encephalopathy syndrome: associated clinical and radiologic findings. Mayo Clin Proc. 2010 May;85(5):427-32. doi: 10.4065/mcp.2009.0590. PMID: 20435835; PMCID: PMC2861971.

[8] De Havenon A, Joos Z, Longenecker L, Shah L, Ansari S, Digre K. Posterior reversible encephalopathy syndrome with spinal cord involvement. Neurology. 2014 Nov 25;83(22):2002-6. doi: 10.1212/WNL.0000000000001026. Epub 2014 Oct 29. PMID: 25355822.

[9] McKinney AM, Short J, Truwit CL, McKinney ZJ, Kozak OS, SantaCruz KS, Teksam M. Posterior reversible encephalopathy syndrome: incidence of atypical regions of involvement and imaging findings. AJR Am J Roentgenol. 2007 Oct;189(4):904-12. doi: 10.2214/AJR.07.2024. PMID: 17885064.

[10] Hugonnet E, Da Ines D, Boby H, Claise B, Petitcolin V, Lannareix V, Garcier JM. Posterior reversible encephalopathy syndrome (PRES): features on CT and MR imaging. Diagn Interv Imaging. 2013 Jan;94(1):45-52. doi: 10.1016/j.diii.2012.02.005. Epub 2012 Jul 24. PMID: 22835573.

[11] Takahashi S, Higano S, Ishii K, Matsumoto K, Sakamoto K, Iwasaki Y, Suzuki M. Hypoxic brain damage: cortical laminar necrosis and delayed changes in white matter at sequential MR imaging. Radiology. 1993 Nov;189(2):449-56. doi: 10.1148/radiology.189.2.8210374. PMID: 8210374.

[12] Komiyama M, Nishikawa M, Yasui T. Cortical laminar necrosis in brain infarcts: chronological changes on MRI. Neuroradiology. 1997 Jul;39(7):474-9. doi: 10.1007/s002340050448. PMID: 9258922.

[13] Elgendy A, Walizai T, Campos A, et al. Cortical laminar necrosis. Reference article, Radiopaedia.org (Accessed on 08 Apr 2025) https://doi.org/10.53347/rID-27802

[14] Ahmad I, Kralik S, Ho CY, Ammar T, Douglas AC. Cortical Laminar Necrosis and CT Negative Hemorrhage in Posterior Reversible Encephalopathy Syndrome. A Case Report. Neuroradiol J. 2012 Dec 20;25(6):671-5. doi: 10.1177/197140091202500605. Epub 2012 Dec 20. PMID: 24029179.

[15] MacLeod C, Bireley JD, Huang MM, Kiczek MP, Buletko AB. Atypical Posterior Reversible Encephalopathy Syndrome Presenting With Cortical Laminar Necrosis. Neurology. 2024 Nov 26;103(10):e210026. doi: 10.1212/WNL.0000000000210026. Epub 2024 Oct 28. PMID: 39467230.