

# POSTERIOR REVERSIBLE LEUKOENCEPHALOPATHY (PRES) IN ACUTE INTERMITTENT PORPHYRIA (AIP): A RARE NEUROLOGICAL COMPLICATION

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## ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is now being increasingly documented in clinical practice because of increased use of DWI in MRI. PRES is seen in various systemic illnesses, eclampsia and has been well documented. It has specific imaging features on MRI which lead to diagnose this condition and to differentiate it from acute stroke as both entities are managed differently.<sup>1</sup>

Acute intermittent porphyria (AIP) is one of the porphyrias which results in excessive secretion of porphyrins and porphyrin precursors in urine involving defects in heme metabolism. The presenting complaints range from abdominal pain, constipation and neuropathies. Here we present a case of young male with AIP who was found to have typical PRES findings on MRI.

**Keywords:** PRES, AIP, DWI

## Case Report

A 20 years old male presented to our ER with acute abdominal pain. On examination he was found afebrile with BP 140/100 mmHg. He had generalized tenderness all over the abdomen. His complete blood count, serum urea electrolytes, liver function tests and ultrasound abdomen were all within normal limits. After 06 hours of admission he developed tonic clonic convulsions and became semiconscious. There was a progressive deterioration of the consciousness. An urgent CT scan brain showed bilateral symmetrical hypodensities in occipital and parasagittal locations. He was transferred to intensive care unit and was put on ventilator because of deteriorating consciousness. Next day his MRI brain was performed which showed hyperintensities on T2/FLAIR sequences showing predominantly facilitated diffusion with few areas showing restricted diffusion on DWI/ADC mapping in bilateral occipital and parasagittal

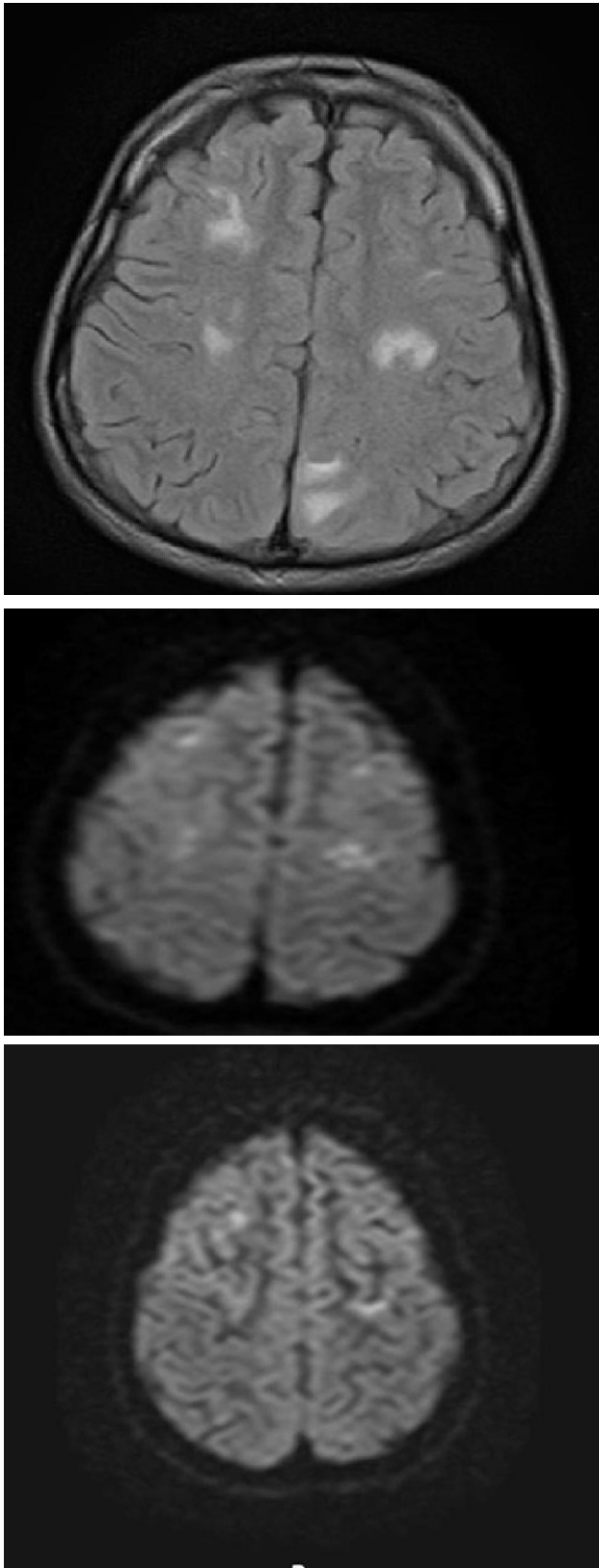
locations. There was no contrast enhancement (Fig.1). MRA of the brain was normal but, as it could not detect and exclude spasm in small size vessels. The findings were in favor of PRES. His urine sample was analyzed for PBG (urine porphobilinogen) which was raised 100 micromol/L (normal 8.8 micromol/L) which led us to diagnose AIP. His condition improved and after a week repeat MRI showed reversal of lesions in occipital lobes but those in parasagittal location were reduced but still some of them showed restricted diffusion which suggested ischemia.

## Discussion

Acute intermittent porphyria (AIP), is an autosomal dominant disease, constituting a varied group of inborn errors of metabolism that are characterized by specific inherited enzyme defects in haem bio-

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**Figure 1:** MRI images on admission: Bi-occipital and parasagittal hyperintensities showing facilitated diffusion on DWI

synthesis. The patients usually present with hypertension, tachycardia and abdominal pain. The central nervous system is involved in 10% of AIP cases with symptoms of autonomic or peripheral neuropathy or central nervous system dysfunction.<sup>2</sup> There are few reports on the neuroimaging findings of AIP in the literature. There is usually reversible, symmetrical cortical and subcortical involvements of the occipital and parietal lobes.<sup>1,3</sup> These lesions predominately show facilitated diffusion on DWI/ADC mapping secondary to vasogenic edema. The exact etiology is not known but it is postulated that it may be due to relative sparseness of sympathetic innervation in the posterior part of the brain which leads to failure in autoregulation in hypertension.<sup>4</sup> These lesions are reversible and show good prognosis. Sometimes few lesions are associated with cytotoxic edema which may be due to vasospasm<sup>5</sup> leading to restriction on DWI/ADC mapping and has been associated with infarction and irreversible brain destruction.<sup>5</sup>

In our patient, whom we followed with serial brain scans, lesion distribution and AIP with hypertension strongly suggest PRES. However the areas which showed restricted diffusion may be a result of PRES being complicated by ischemic infarction.

PRES is still poorly understood; upon literature review there are two hypotheses on its mechanism. The older hypothesis states that during hypertension there is autoregulatory vasoconstriction and hypoperfusion which later caused ischemia and cerebral edema.<sup>5</sup> However; the newer hypothesis suggests that the autoregulatory limits of the cerebral vasculature are exceeded by severe hypertension which then causes breakthrough of the blood-brain barrier, fluid leakage, and reversible vasogenic edema.<sup>5</sup>

## Conclusion

AIP is a multisystem disease of varied clinical spectrum; neurological manifestations are however very rare. Posterior reversible encephalopathy syndrome (PRES) is a neurotoxic state which develops in patients with complex systemic

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conditions such as eclampsia, after transplantation, in infection/sepsis/shock and autoimmune disease, and after cancer chemotherapy. The lesions commonly involve parietal-occipital lobe or are predominantly "posteriorly" located. All these lesions show facilitated diffusion on DWI/ADC mapping secondary to vasogenic edema; however few areas of restricted diffusion are also encountered. During last few years many cases are diagnosed as PRES secondary to increased awareness among radiologists about this unique entity which leads to good clinical workup and better management of patients.

## References

1. Hinchey J, Chaves C, Appignani B, et al. A reversible posterior leukoencephalopathy syndrome. *N Engl J Med* 1996; **334**: 494-500.
2. Desnick RJ. The Porphyrrias. In: Kasper DL, Braunwald E, Fauci AS, et al, editors. *Harrison's principles of internal medicine*. New York: McGraw-Hill, 2005: 2303-8.
3. Celik M, Forta H, Dalkılıç T, Babacan G. MRI reveals reversible lesions resembling posterior reversible encephalopathy in porphyria. *Neuroradiology* 2002; **44**: 839-41.
4. Schaefer PW, Buonanno FS, Gonzalez RG, Schwamm LH. Diffusion-weighted imaging discriminates between cytotoxic and vasogenic edema in a patient with eclampsia. *Stroke* 1997; **28**: 1082-5.
5. Bartynski WS. Posterior reversible encephalopathy syndrome, part 2: controversies surrounding pathophysiology of vasogenic edema. *Am J Neuroradiol*. 2008; **29(6)**: 1043-9.