DOI: 10.7860/JCDR/2024/67676.19822

Case Report

Internal Medicine Section

Posterior Reversible Encephalopathy Syndrome with Addison's Disease in a Known Case of Arnold Chiari Malformation

SATBIR KAUR MALIK[1], SANGRAM S MANGUDKAR[2], MAHABIR MISHRA[3], SANKET GENUJI SHINDE[4]

ABSTRACT

Posterior Reversible Encephalopathy Syndrome (PRES) is a life-threatening neurological disease. The aetiology of Addison's disease has been modified from infectious to autoimmune pathology. Underlying pathogenesis is insufficient production of glucocorticoids and mineralocorticoids. Case series with PRES syndrome and Cushing's syndrome have been reported but PRESS with adrenal insufficiency is rarely published. We hereby report an operated case of Arnold Chiari malformation presenting with first episode of generalised tonic clonic seizure and diarrhoea for four days. Postictal confusion was present for two hours. She was previously diagnosed with Addison's disease. Patient had hypotension and dyselectrolytaemia. Systemic examinations were within normal limits. MRI brain revealed features of PRES. The authors encountered certain challenges in her management which included persistent hypokalaemia, hypocalcaemia and hypotension. Patient is on regular follow-up and she is currently asymptomatic. Prompt diagnosis of PRES and intensive case treatment is a must for the betterment of prognosis.

Keywords: Autoimmune, Dyselectrolytaemia, Hypotension

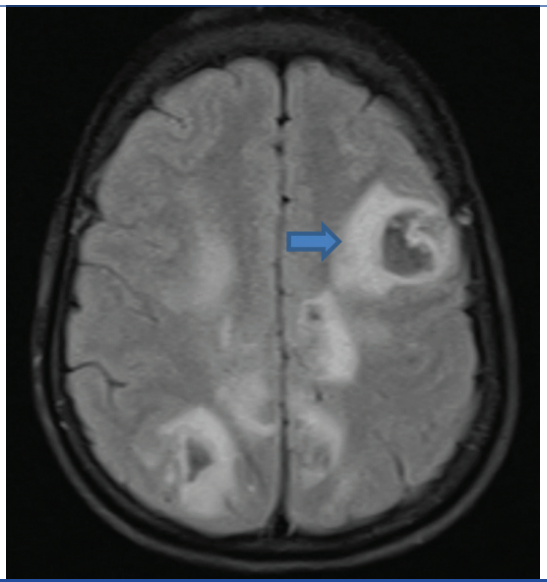
CASE REPORT

A 41-year-old female came with the chief complaints of diarrhoea since four days, which was three episodes per day. It was associated with abdominal distension and first episode of generalised tonic clonic seizure, along with up rolling of eyes. Postictal confusion was present for two hours. Patient was known case of Addison's disease which was diagnosed two months prior to the current admission. She was taking tablet Prednisolone 5 mg once a day. No other co-morbidities were present. In 2017, she underwent decompression of foramen magnum for Arnold Chiari Malformation.

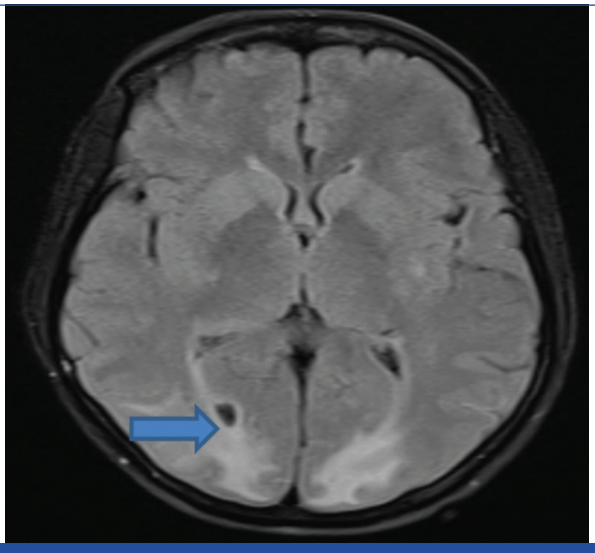
On examination, pulse rate was 80 beats/min, blood pressure was 90/70 mmHg, no postural drop noticed. Neurological assessment revealed Glasgow Coma Scale of 10. Biceps, triceps, supinator, knee and ankle reflexes were normal and Babinski sign was positive bilaterally. Bilateral nystagmus in the horizontal direction was noted. Routine blood investigations revealed haemoglobin of 8.40 gm/dL, serum potassium was 2.31 mmol/L magnesium was 1.10 mg/dL, serum calcium was 6.30 mg/dL, serum Vitamin D was 4.70, urinary potassium was 18.20 mmol/L and 8 am cortisol was 9.80 mcg/dL. Patient had persistent hypokalaemia despite of multiple corrections. MRI brain was suggestive of PRES.

The above findings were suggestive of ill-defined hyperintense areas in pons, midbrain, cerebellar hemispheres, occipital lobes, frontal lobes, parietal lobes [Table/Fig-1-3]. Multiple areas of haemorrhage (acute to early subacute stage) were noted in cerebellar hemisphere, bilateral frontal and parietal appearing isointense to slightly hyperintense on T1W, hypointense on T2W showing blooming on Gradient Restricted Echo-Gradient (GRE) with diffusion restriction on DWI.

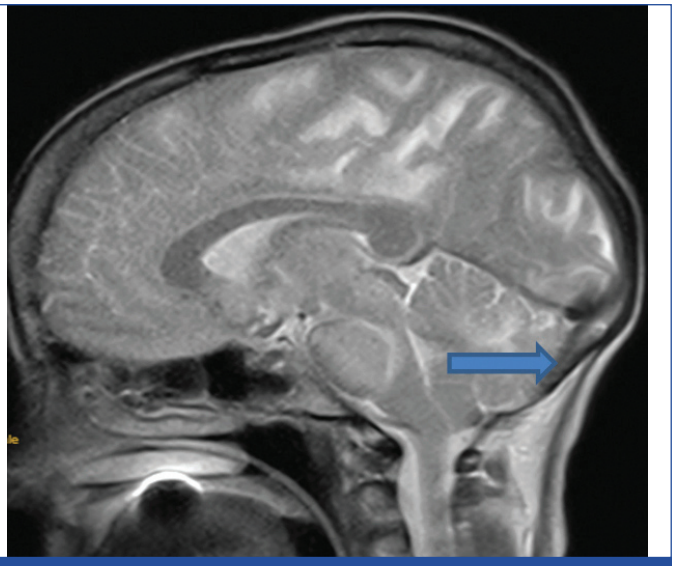
There was no evidence of papilloedema. She was started on Injection piperacillin tazobactam at the dose of 4.5 mg intravenous (i.v) three times a day, Injection metronidazole at the dose of 100 mL i.v. three times a day, Injection dexamethasone 4 mg i.v. three times a day followed by a tapering dose after two weeks, Injection levetiracetam 500 mg i.v. twice a day and Tablet fludrocortisone 100 µg once a day. Calcium and potassium corrections were given. Patient clinically improved in the form that generalised Tonic-clonic seizure has settled and metabolic



[Table/Fig-1]: Hyperintense areas are seen involving frontal and parietal areas.



[Table/Fig-2]: Hyperintense areas are seen involving occipital areas.



[Table/Fig-3]: Ill-defined hyperintense areas involving cerebellum.

parameters were normal postdischarge. Patient is on regular follow-up and she is currently asymptomatic.

DISCUSSION

The PRES is a neurotoxic state presenting with symmetrical subcortical vasogenic oedema, preferentially parieto-occipital region [1]. Pathophysiology behind PRES includes conditions causing either hyper perfusion leading to increased intracranial pressure or endothelial injury secondary to toxic or autoimmune aetiology [2]. Commonly, encountered causes include hypertensive encephalopathy, pre-eclampsia, drugs such as calcineurin inhibitor toxicity, methotrexate, azathioprine. Hypertension is the most common trigger. The patient had hypotension making the diagnosis complicated. Symptoms include blurring of vision, altered sensorium, headache and tonic clonic seizures [3]. Out of these, in the current case patient had seizures. It is a clinico-radiological diagnosis. Characteristic MRI findings of Posterior Reversible Encephalopathy Syndrome are characterised by the presence of symmetrical vasogenic oedema that often affects the subcortical white matter and can extend to the cortex.

These features are prominently evident on MRI brain with Fluid-Attenuation Inversion Recovery sequence (FLAIR) [4]. Contradictory to the present case, Hansberry DR et al., had presented a case wherein the patient who presented with left upper and lower limb paresthesia, underwent MRI of the brain and cervical spine which showed a Chiari I malformation with tonsillar descent beyond the level of the C1 lamina. She was operated for Chiari malformation 1 and further workup showed evidence of PRES [4]. The patient was diagnosed previously as a case of Addison's disease and currently had persistent hypokalaemia as a manifestation. At times, patients may experience severe consequences, such as status epilepticus or coma, which require urgent care treatment to prevent life-threatening outcomes [5,6].

Various studies have demonstrated that there are insufficient evidences that exist about the risk of seizures following the resolution of PRES. Hence, we still do not know the best length of time to treat with antiepileptic drugs [7-9]. Our patient who presented with features of PRES, was also a known case of Addison's disease and operated case of Arnold Chiari malformation. Hence, it was a difficult case to be managed. It was challenging to treat her but patient showed dramatic improvement with treatment.

CONCLUSION(S)

Patient presenting with such varied neurological pathologies involving PRES and Arnold Chiari malformation are difficult to be approached. Such patients don't present with specific symptoms making it difficult to diagnose. Management of such neurological diseases rather than Diagnosis is arduousness. Clinically, it is difficult to identify the precise diagnosis. Radiological assessment aids in such patient's evaluation. Intensive care is of utmost importance in the management.

REFERENCES

[1] Hou X, Xu J, Chen Z, Li G, Jiang H. Posterior reversible encephalopathy syndrome with involvement of the cervical cord and medulla: A case report. J Clin Diagn Res. 2015;9(1):CD01.
[2] Triplett JD, Kutlubaev MA, Kermode AG, Hardy T. Posterior reversible encephalopathy syndrome (PRES): diagnosis and management. Practical Neurology. 2022;22(3):183-89.
[3] Casey SO, Sampaio RC, Michel E, Truwit CL. Posterior reversible encephalopathy syndrome: Utility of fluid-attenuated inversion recovery MR imaging in the detection of cortical and subcortical lesions. American Journal of Neuroradiology. 2000;21(7):1199-206.
[4] Hansberry DR, Agarwal N, Tomei KL, Goldstein IM. Posterior reversible encephalopathy syndrome in a patient with a Chiari I malformation. Surgical Neurology International. 2013;04:130.
[5] Gokhale A, Kimona A, Kantor S, Prakash S, Manhas Y. Posterior reversible leukoencephalopathy syndrome (PRES) in intensive care unit–Case series. Indian Journal of Critical Care Medicine: Peer-reviewed, Official Publication of Indian Society of Critical Care Medicine. 2017;21(11):772.
[6] Strother R, Wong H, Miller NE. Posterior reversible encephalopathy syndrome secondary to hypertensive encephalopathy brought on by a MAO inhibitor: A case report. J Prim Care Community Health. 2019;10:2150132719869539.
[7] Datar S, Singh T, Rabinstein AA, Fugate JE, Hocker S. Long-term risk of seizures and epilepsy in patients with posterior reversible encephalopathy syndrome. Epilepsia. 2015;56(4):564-68.
[8] Fischer M, Schmutzhard E. Posterior reversible encephalopathy syndrome. Journal of Neurology. 2017;264(8):1608-16.
[9] Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A, et al. A reversible posterior leukoencephalopathy syndrome. New England Journal of Medicine. 1996;334(8):494-50.

PARTICULARS OF CONTRIBUTORS:
1. Assistant Professor, Department of Internal Medicine, Dr. D. Y. Patil Medical College, Dr. D. Y. Patil Vidyapeeth, Pimpri, Pune, Maharashtra, India.
2. Professor, Department of Internal Medicine, Dr. D. Y. Patil Medical College, Dr. D. Y. Patil Vidyapeeth, Pimpri, Pune, Maharashtra, India.
3. Resident, Department of Internal Medicine, Dr. D. Y. Patil Medical College, Dr. D. Y. Patil Vidyapeeth, Pimpri, Pune, Maharashtra, India.
4. Resident, Department of Internal Medicine, Dr. D. Y. Patil Medical College, Dr. D. Y. Patil Vidyapeeth, Pimpri, Pune, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Sanket Genuji Shinde,
Dr. D. Y. Patil Boys Hostel, Pimpri, Pune-411018, Maharashtra, India.
E-mail: sanket20834@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Sep 23, 2023
- Manual Googling: Jan 03, 2024
- iThenticate Software: Jul 15, 2024 (24%)

ETYMOLOGY: Author Origin

EMENDATIONS: 8

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Sep 23, 2023**
Date of Peer Review: **Nov 30, 2023**
Date of Acceptance: **Jul 17, 2024**
Date of Publishing: **Sep 01, 2024**