Case Rerort

Indian Journal of Trauma and Emergency Pediatrics
Volume 10 Number 2, April - June 2018
DOI: http://dx.doi.org/10.21088/ijtep.2348.9987.10218.5

Posterior Reversible Encephalopathy Syndrome (PRES) with Pign - Case Report of a Real Emergency

Bijoy Patra[1], Riteish Yadav[2], Chidambaram Laxman[3], Sandip Roy[4], Satinder Aneja[5]

Abstract

Posterior reversible encephalopathy syndrome (PRES) is a reversible neurological entity characterized by seizure, headaches, visual symptoms, impaired consciousness and other focal neurological findings. It is caused by a wide variety of causes ultimately leading to a vasogenic cerebral edema of occipital and parietal lobes of the brain. A 10 Year Old Girl presented in Pediatric emergency with altered sensorium and intermittent tonic posturing. During examination her BP was noted to be 140/90mmHg (> 99th percentile for the patient).Urinalysis revealed hematuria. CT Cranium showed prominent ventricles and diffuse hypodensity in cerebellar region. Reversibility of the symptoms after institution of antihypertensivemeasures and MRI brain confirmed the diagnosis of PRES. Post streptococcal Glomerulonephritis (PSGN) was found to be the underlying etiology of PRES.

Keywords: Posterior Reversible Encephalopathy Syndrome; Post Infectious Glomerulonephritis; Calcineurin Inhibitors.

Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinico-radiological picture which presents with vomiting, severe headache, clouding of consciousness, behavioral changes, seizures and visual disorders and typical magnetic resonance imaging (MRI) findings. The radiological findings are frequently characterized with transient bilateral grey and white-matter changes compatible with vasogenic edema in the posterior cerebral hemispheres and cerebellum [1]. Although the entity is generally described in adult patients, it has been reported in children with a gradually increasing frequency [2]. In children, cases of PRES occuring with acute post-sterptococcal glomerulonephritis (APSGN), Henoch-Schönlein purpura (HSP), nephrotic syndrome (NS), lupus nephritis and use of calcineurin inhibitor (CNI) have been reported [3–5]. Hematuria during urinalysis gives the clue to the cause of hypertension. Although the pathogenesis is generally sudden increases in blood pressure, it may also be observed in relation with renal failure, fluid accumulation and cytotoxic effects of immunosupressive drugs on vascular endothelium [6].

Here,we report a pediatric patient who presented in the pediatric emergency with analtered sensorium and intermittent tonic posturing. Awareness and acess to neuroimaging led to the diagnosis of PRES. Appropriate and timely institution of anitihypertensive treatment led to a favourable outcome.

Case

A10 Year Old Girl presented in the pediatric emergency in the early morning with altered sensorium and intermittent tonic posturing. She had a history of fever for 5 days, and headache, vomiting and loose motion for 2 days. Since morning she was not recognizing parents and not responding to verbal commands. This was followed by episodes of intermittent posturing. On examination her BP was 140/90 mmHg (>99th percentile for the patient). She was in coma (GCS

Author Affiliation: [1]Consultant [2]Senior Resident [3]Post Graduate Resident, [4]Senior Medical Officer, [5]Director & Professor, Department of Paediatrics, Lady Hardinge Medical College and Associated kalawati Saran Children Hospital, New Delhi 110001, India.

Corresponding Author: Bijoy Patra, Consultant, Department of Paediatrics, Lady Hardinge Medical College and Associated Kalawati Saran Children Hospital, New Delhi 110001, India.

E-mail: bijoy_patra@yahoo.com

Received on 12.01.2018, **Accepted on** 23.04.2018

was E2 V1M3 -6/15), both pupils were equal and reacting, muscle tone was increased in all 4 limbs, DTRs were brisk, plantars were up-going. Meningeal signswere absent. Examination of all other systems were normal. Child was intubated and put on ventilator. Intravenous fluids, antibiotics, antiviral and antimalarial, mannitol and antihypertensives were started. ATT (Antitubercular treatment) was also commenced as she had history of Koch›s contact in her family and urgent CT showed dilated ventricles with cerebellar involvement (Fig. 1). She was catheterized. Urine output was adequate and Urine dipstic showed 2+blood. Blood glucose was 96 mg%. Her blood count were HB-10.8 gm%, hematoctrit: 34%, white blood cells: 17000/mm^3, platelets: 240 000/mm^3. Her KFTs and Electrolytes were urea: 68mg/dL, serum creatinine: 0.6 mg/dL, sodium: 136 mmol/L, potassium: 4.0 mmol/L, calcium: 9.2 mg/dL, phosphorous: 5.2 mg/dL.Fundus was normal. on lumbar puncture Cytology showed 30 cells (50% Neutrophil), Sugar/Protein - 97/30. Urine routine & microscopy shows protein nil, glucose nil, RBC 8-10/hpf, pus cells 1-3/hpf, no dysmorphic RBC. 24 hour urine protein was 280 mg/m2/h, and serum total protein: 6.3g/dL, albumin: 2.6 g/dL, triglyceride: 248 mg/dL, total cholesterol: 192 mg/dL. Peripheral smear for MP and RMAT was negative. Serum widal was negative. C3 was 24.1 mg/dL (N: 83–177) and ASO (Anti Streptolysin O) titre was raised (586 IU/L) C4 23.5 (n: 15–45). CRP was negative and ESR was 35mm/hr. On 5th day of admission patient was extubated.Post extubation patient developed RUL collapse, which resolved with position change and physiotherapy. ATT was stoped as tubercular workups were negative. (Mantoux test was 0X0 mm, Gastric aspirate showed no acid fast bacilli and Genexpert was negative). In view of persistent hypertension, further workup was done. Echo was normal with EF 58%. USG abdomen and renal Doppler study were normal. ANA, P-ANCA, C-ANCA, dsDNA were negative. Urinary VMA was normal. The electroencephalogram (EEG) performed was found to be normal. CT report shows prominent ventricles and diffuse hypodensity in cerebellar region. MRI showed T2 and Flair image hyperintensity in parieto occipital distribution. Diffusion weighted image was enhanced. ADC mapping was normal and there was no contrast enhancement with gladolinium. The findings were compatible with Posterior reversible encephalopathy syndrome (PRES). During followup, antihypertensive drugs were tapered and discontinued. Currently, the blood pressure is within the normal limits, neurological examination is normal and urinary findings are normal. Her cognitive functions are intact. The girl has gone back to her school and resumed her studies.

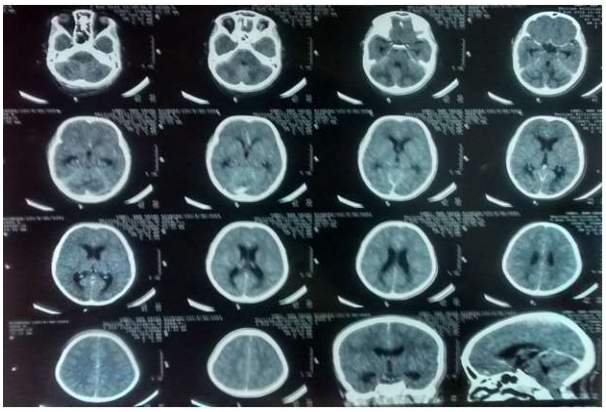

Fig. 1: CT Head showing cerebellar edema and dilatation of ventricles

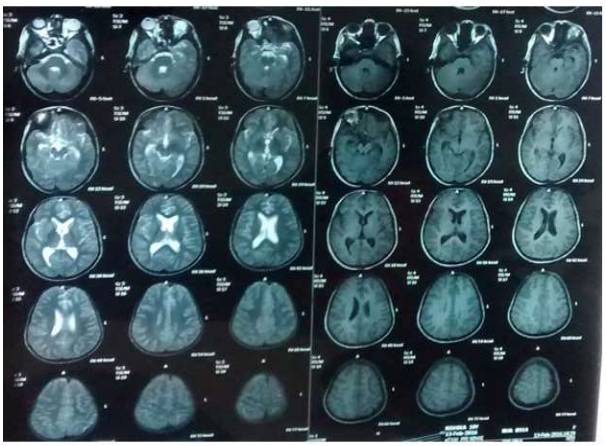

Fig. 2: MRI brain showing bilateral posterior occipito parietal oedema

Discussion

Since the first elucidation of Posterior reversible leucoencephalopathy syndrome in 1996 by Hinchey et al, many cases had been reported in children [1,2]. The awareness of this entity in pediatric age group, the presentation, noting of hypertension and the findings of prominent ventricles and diffuse cerebellar hypodensity inCT cranium were the reasons to consider the diagnosis and intervention of anitihypertensives and anticerebral edema measures mannitol. However as the child had a history of fever, vomiting and loose motion prior to hospitalization and a strong family history of tuberculosis, Lumbar puncture was considered to rule out CNS infection. As CSF cytology showed some 30 cells (50% lymphocytes) and CT cranium showed prominent ventricles, she was put on 4 drugs antitubercular treatment. After the child was extubated, MRI Brain was done on day 6 of her hospitalization which confirmed the diagnosis of PRES. ATT was discontinued. In view of persistent hypertensison and presence of

hematuria child underwent extenisve workup to findout an etiology. Raised ASO titre and low C3 pointed towards PSGN as the cause of hypertension in this child.

Posterior reversible encephalopahty syndrome is a kind of brain capillary leak syndrome. Sudden increases in the systemic blood pressure lead to exceeding of the autoregulatory capacity in the brain vessels. Consequently, vasoconstriction, subsequently vasodilatation, increase in vascular permeability, disruption in the brain-blood barrier, fluid transduction and petechial hemorrhages occur. The reason that the findings are mostly observed in the posterior hemispheres is the fact that sympathetic innervation which provides autoregulation in cases of increased blood pressure is found less in the posterior cerebral vessels [3]. Posterior reversible encephalopathy syndrome occurs as a result of increased blood pressure or endothelial damage which is observed in relation with acute glomerulonephritis, HSP, lupus, nephrotic syndrome or immunosupressive drug usage which lead to disruption of the blood-brain barrier [4,5].

In Posterior reversible encephalopathy syndrome, Neuroimaging detects edema. CT shows hypodensity and T2 images in MRI shows hyperdensity.Diffusion weighted (DW1) images detects movement of watermolecule while ADC mapping helps in differenting vasogenic edema from cytotoxic edema. There may or may not be contrast enhancement [5,6]. In the present case, CT cranium was showing diffuse hypodensity in Cerebelllum. Prominent ventricles might be because of brainstem and cerebellar edema leading to obstruction in CSF circulation [7].

Among the myriad causes of PRES, poststreptococcal glomerulonephritis is one of the common causes reported from developing countries [8]. PSGN was observed to be the cause in our present case. However sepsis or viral infections are also postulated to contribute or triger PRES [8,9]. This also explains vague illness of fever, loosemotion and vomiting in this child preceeding PRES.

In most cases, PRES resolves spontaneously and patients show both clinical and radiological improvements. The range of symptoms that can comprise the syndrome might be broader than usually thought. In its mild form, this disorder might cause only one clinical symptom (headache or seizure) and radiographically might show few areas of vasogenic oedema or even normal brain imaging in some rare cases. In severe forms, PRES might cause substantial morbidity and even mortality, most often as a result of acute haemorrhage or massive posterior fossa oedema causing obstructive hydrocephalus or brainstem compression [10]

Magnetic resonance imaging findings belonging to posterior reversible encephalopathy syndrome may be confused with glyomatosis cerebri, progressive multi-focal leukoencephalopathy, demyelinating conditions and infacts. This may lead to unnecessary tests and treatment which may arrive at biopsy [1,6]. Since early recognition of posterior reversible encephalopathy syndrome will provide administration of the most appropriate treatment and prevention of severe neurological sequelae, keeping PRES in mind in presence of clinical findings is very important [4].

Conclusion

Awareness of Posterior reversible encephalopathy syndrome is important in Pediatric emergency. Recording of High Blood Pressure in Children presenting with neurological symptoms can lead to the diagnosis which can be confirmed by neuroimaging. Simple urinalysis can give a clue towards its etiology. Timely institution of antihypertensives leads to reversibility of the symptoms and favourable outcome.

Conflict of interest: None

References

1. Hinchey J, Chaves C, Appignani B, et al. A reversible posterior leukoencephalopathy syndrome. N Engl J Med. 1996;334:494–5.
2. Jones BV, Egelhoff JC, Patterson RJ. Hypertensive encephalopathy in children. Am J Neuroradiol. 1997;18:101–6.
3. Wirrell EC, Hamiwka LD, Hamiwka LA, Grisaru S, Wei X. Acute glomerulonephritis presenting with PRES: a report of 4 cases. Can J Neurol Sci. 2007;34:316–21.
4. Pedraza Rodrigo,Marik Paul E., Varon Joseph. Posterior Reversible Encephalopathy Syndrome: A review. Critical Care & Shock 2009;12(4):135-43.
5. Legriel S, Pico F, Azoulay E.Understanding Posterior Reversible Encephalopathy Syndrome.In J-L Vincent (ed), Annual update in Intensive Care and Emergency Medicine. 2011DOI10.1007/978-3-642-18081-1. Springer Science+ Business Media LLC2011.
6. Endo A, Fuchigami T, Hasegawa M, et al. Posterior reversible encephalopathy syndrome in childhood:

report of four cases and review of the literature. Pediatr Emerg Care. 2012;28:153–7.

7. Prasad N, Gulati S, Gupta RK, et al. Spectrum of radiological changes in hypertensive children with reversible posterior leucoencephalopathy. Br J Radiol. 2007;80:422–9.
8. Gupta S, Goyal VK, Talukdar B. Reversible posterior leucoencephalopathy syndrome in post streptococcal glomerulonephritis. Indian Pediatr. 2010;47:274–6.
9. Bartynski WS, Boardman JF, Zeigler ZR, Shadduck RK, Lister J. Posterior reversible encephalopathy syndrome in infection,sepsis, and shock. Am J Neuroradiol. 2006;279(10):2179-90.
10. Yi Shiau Ng, Grainne S Gorman, Doug M Turnbull, Mika H Martikainen. The diagnosis of posterior reversible encephalopathy syndrome. The Lancet neurology. 2015;14(9):914–25.