POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME IN OUR SETUP

Imran Ahmad, Usama Bin Zubair, Humza Mumtaz, Muhammad Ali Yousaf, Wasim Wali Muhammad*

Military Hospital Rawalpindi, *Combined military Hospital Lahore

ABSTRACT

Objective: To assess the clinical presentation and neuroimaging abnormalities in a series of patients diagnosed as posterior reversible encephalopathy syndrome at Military Hospital Rawalpindi.

Study Design: Case series study.

Place and Duration: Study was carried out at Military Hospital Rawalpindi from December 01st , 2011 to May 31st, 2012.

Patients and Methods: Study included all the cases of the Posterior reversible encephalopathy syndrome (PRES) admitted in the wards and intensive care unit (ITC). Neuroimaging was done and all the studies were reviewed by independent neuroradiologist. Different clinical and laboratory variables were also studied and correlated with neuroimaging. Follow up was done to look for the prognosis.

Results: Of the seven patients labelled as PRES two were male and five were female. Two patients were over 50 years of age, out of them one was male and one was female. One patient had end stage renal disease (ESRD) secondary to diabetes mellitus (DM) and hypertension (HTN), one had eclampsia, one had pregnancy-induced hypertension (PIH) and one had just uncontrolled HTN. Peak spontaneous bacterial peritonitis (SBP) in 5 cases was 210 mm of Hg, four of which had seizures. Rest two had spontaneous bacterial peritonitis (SBP) of 160 out of which one developed seizures. Total out of 7, 5 experienced seizures and altered conscious state, rest two only had confusion. One patient had papilloedema. Follow up was done after 06 weeks, 02 patients died, 05 remained alive and symptoms of PRES had vanished.

Conclusion: PRES is a neurological emergency, presents with a variety of symptoms and has a specific neuroimaging pattern. Early recognition and prompt treatment result in a good neurological outcome.

Keyword: Cerebral edema, Neuroimaging, PRES, Seizures.

INTRODUCTION

First described in 1996 by Hinchey and colleagues, later on in 2000 Casey and colleagues proposed its current name, agreeing that PRES is transient, reversible, posterior cerebral edema that can extend into the temporal or frontal lobes[1,2].

PRES is a clinically recognizable entity that presents with neurological signs and symptoms (headaches, altered consciousness, visual abnormalities and seizures) in conjugation with the unique neuroimaging findings of vasogenic edema involving the posterior circulation[1]. Risk factors include malignant hypertension, eclampsia, medications such as immunosuppressants (including tacrolimus and cyclosporine), chemotherapy, biotherapy, and renal failure[1]. Etiology is unclear, thought to be secondary to endothelial damage in the setting of hypertension and failure of cerebrovascular autoregulation with subsequent vasogenic edema. The posterior circulation is more sensitive to the effects of HTN. PRES is reversible with prompt and aggressive use of antihypertensive agents, hydration, and anticonvulsants. However, if not recognized and treated promptly and appropriately, this syndrome can progress to ischemia and hemorrhage, with permanent deficits[2].

A retrospective study carried out at in the Mayo clinic Rochester, USA, showed that PRES and seizures of eclampsia are pathophysiologically related as all patients of eclampsia who underwent neuroimaging

Correspondence: Lt Col Imran Ahmad, Classified Medical Specialist, MH Hospital, Rawalpindi.
Email: drimran72@yahoo.com
Received: 27 Sep 2012; Accepted: 10 Dec 2012

displayed clinical and radiological findings of PRES[3].

Another retrospective study was done on MRI's and CT Scans performed at the New England Medical Centre in Boston and Hospital Saint Anne in Paris revealed that PRES commonly occurs in patients with renal insufficiency, HTN or who are immune compromised and characteristic neuroimaging finding of sub cortical edema without infarction[2].

Atypical region involvement was described in a study carried out in the USA which described that large series of PRES cases show that a typical distribution and image manifestations of PRES have a higher incidence than commonly perceived and alcohol with drawl and contrast related anaphylaxis are potential new causes of PRES[3].

Our study aimed to look for the clinical presentation along with distribution and extent of neuroimaging abnormalities among patients diagnosed as PRES. In addition to this we followed up the patients after six months to look for the prognosis and mortality of the disease.

PATIENTS AND METHODS

With the approval of ethical committee Military Hospital Rawalpindi, this case series was carried between Dec 01, 2011 and May 30th, 2012. All the patients with age more than 13 years with characteristic neuroimaging of PRES admitted with or had during admission manifestation like mental status change, headache, visual disturbance and seizures supported with characteristic abnormalities in neuroimaging were included in the study. All the patients underwent CT Scan and MRI. Patients in which neuroimaging cannot be done were excluded from the study. A protocol performa was designed which included particular of the patient, peak blood pressure values, relevant laboratory findings and neurological findings. Follow up was done after 06 weeks to look for prognosis and repeat neuroimaging.

The incidence of PRES and a variety of presentations was noted. Relation of peak blood pressure, underlying disease and relevant laboratory investigation were made. The pattern of changes as well as evidence of permanent neurological abnormalities was noted.

RESULTS

A total of 7 patients were included in the study out of which 2 males (28.5%) and 5 females (71.4%). Age of patients ranged from 27 to 77 years with mean age of 43.71 years. Two patients were over 50 years of age, out of them one was male and one was female. Three patients were picked up from the gynaecology ward as a complication of pregnancy. One male patient had ESRD secondary to DM and HTN, and another one had just uncontrolled HTN. One female patient had post operative PRES and one had uncontrolled hypertension. Peak SBP at arrival in 5 (71.45%) cases was 210 mm of Hg , four of which had seizures. Rest 2 (28.5%) had SBP of 160 out of which one had developed seizures. Total out of 7, 5 experienced seizures and altered conscious state, rest two only had confusion. One patient had papilloedema.

Neuroimaging findings consist of T2 and flair hyperintense white matter signal (indicating white matter edema) localized to parieto occipital region in all seven cases, other location included one patient with deep white matter (14.2%), frontal lobes (14.2%). None of the lesion took intravenous contrast and there was no diffusion restriction on DWI.

Follow up was done after 06 weeks, 02 patients died, 05 remained alive and the symptoms of PRES had vanished. Follow up neuroimaging was possible in 3 patients only showing a complete resolution of symptoms.

DISCUSSION

According to Vaughan and colleagues, while the "exact mechanism" of PRES is not yet clear, "it does seem to be associated with a failure in cerebral blood flow autoregulation combined with endothelial dysfunction[3]. Cerebral blood vessels are able to constrict and dilate to control

cerebral blood flow, but the autoregulatory mechanism can be overwhelmed by a sudden spike in blood pressure. Dilated arterioles may allow hyperfusion, causing a breakdown in the blood–brain barrier[4].

Table-1: Description of laboratory and neuroimaging findings in patients of posterior reversible encephalopathy syndrome (n=7).

Patient I.D	Age (yrs)	Gender	Medical History	Peak SBP	Peak DBP	Peak Urea	Peak Creatinine
1	77	M	DM,ESRD,HTN,CVA	160	100	25.2	481
2	42	M	HTN, SOB	210	150	12.1	159
3	27	F	HTN	210	120	10.7	594
4	34	F	G4P2T1 Eclampsia	210	130	20	366
5	56	F	Malignant HTN, Pulmonary edema, bilateral vocal cord paralysis	210	140	21.2	219
6	35	F	Pregnant G1 P1	230	130	22.9	303
7	35	F	PIH	160	100	13	600

Systolic Blood Pressure (SBP), Diastolic Blood Pressure (DBP), Diabetes Mellitus(DM),End Stage Renal Disease(ESRD),Hypertension(HTN),Shortness of breath(SOB), Pregnancy-Induced Hypertension (PIH).

PRES in a rare neurological emergency, there is scarcity of published data from Pakistan on this issue. Most of the series are mostly case reports, there is only case series but it is only restricted to cases of eclampsia and hence restricted to only limited population.

This is for the first time a study was done to see the spectrum of PRES cases in our setup and to form a baseline data. Secondly our study had seven patients which is at present is the largest set of patients from Pakistan. This high incidence could be explained due to the reason as we being a tertiary care center for Pakistan army and cover a large drainage area including whole of AJk, NWFP, Gilgit Baltistan and upper Punjab, secondly being a government sponsored set up all relevant investigations and treatment is done free of cost and in time, this has led to enormous input of patients from even remote and far flung areas.

Similarly the high mortality of patients as compared with western studies is due to above mentioned reasons as patient presents very late in critical state because of the long distance involved in travelling lack of awareness in health care providers about this issue and thus late referrals from primary care centers.

One study focuses on the incidence and behavior of PRES in one set up. It is a rare neurological diagnosis characterized by an acute increase in blood pressure, and by headaches, altered mental status, seizures, and visual loss. It is usually seen on computed tomographic scans as white-matter vasogenic edema predominantly affecting the posterior occipital and parietal lobes of the brain.

As Hitchy and companions studied in 1996 the predisposing factor for PRES maybe immunodeficiency, renal insufficiency or HTN[1]. In our patients one patient had all these varieties except immunodeficiency, one had ESRD, One had Eclampsia, one had PIH and other had uncontrolled HTN episodes out of which one was not a previously diagnosed case of HTN. Another study by O'Hara McCoy H, in 2008 shows that PRES is less common in people who live with chronic hypertension, because they have adapted to the elevation in their blood pressure over time[4].

Pregnancy related complications are one of the major causes of PRES in our setup[6-7], however other causes include ESRD secondary to DM and hypertension, uncontrolled hypertension, both these patients were male.

Key symptoms of one patient included seizures and altered consciousness which were present in 5 of the 7 cases. Episodes of seizures have been usually frequent as in a study in intensive care setup shows that a single seizure is infrequent and multiple seizures are frequently reported[8].

Characteristic radiological findings of PRES demonstrate a distinctive pattern of white matter edema[9]. Patterns of changes in our set up were typical including white matter oedema mostly localized to the parieto occipital lobes in all the seven cases however in two patients they have extensive involvement of frontal lobes and basal ganglia also none of the lesion took up the gadolinium contrast when administered, on repeat follow up scan in 3 patients there was complete resolution of changes. Different studies in the world show that the typical lesions are located in the parieto-occipital and posterior, frontal, cortical, and sub cortical white matter, although less commonly the brain stem, basal ganglia, and cerebellum are involved[9-11].

Prompt recognition and treatment can make the difference between reversal of the condition and permanent neurologic deficits such as chronic epilepsy. White-matter edema may cause hemorrhage or infarction of brain tissue resulting in permanent neurological disablity[13] .

CONCLUSION

PRES is a real neurological emergency with varied etiologies. Presents with variable symptoms and specific neuroimaging. It is completely reversible both clinically and radiologically, provided it is diagnosed early and aggressive medical management done in time. However failure to recognize the syndrome and delay in medical management may result in potential morbidity and death in some case.

REFERENCES

1. Casey SO, Sampaioa RC, Michela E , Charles L. Truwita .Posterior reversible encephalopathy syndrome: utility of fluid-attenuated inversion recovery MR imaging in the detection of cortical and subcortical lesions AJNR Am J Neuroradiol. 2000; 21(7):1199–206.
2. Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A et al. A reversible posterior leukoencephalopathy syndrome N Engl J Med. 1996; 334(8):494–500.
3. Vaughn C, Zhang L, Schiff D. Reversible posterior leukoencephalopathy syndrome in cancer Curr Oncol Rep. 2008; 10(1):86–91.
4. O'Hara McCoy H. Posterior reversible encephalopathy syndrome: an emerging clinical entity in adult, pediatric, and obstetric critical care J Am Acad Nurse Pract. 2008; 20(2):100–6.
5. Servillo G, Bifulco F, De Robertis E, Piazza O, Striano P, Tortora F et al. Posterior reversible encephalopathy syndrome in intensive care medicine Intensive Care MED. 2007;33(2):230–6.
6. Fitzgerald-Hines J, King ML. Posterior reversible encephalopathy syndrome: a case study J Neurosci Nurs. 2006; 38(5):338–41 73.
7. Hagemann G, Ugur T, Witte O W Fitzek C. Recurrent posterior reversible encephalopathy syndrome (PRES) J Hum Hypertens. 2004;18(4):287–
8. Naqi R, Azeemuddin M. Posterior reversible encephalopathy syndrome. J Pak Med Assoc Jul 2012; 62(7):657-60
9. Javed MA, Sial MSH, Lingawi S, Alfi A, Lubbad E. Etiology of posterior Reversible Encephalopathy Syndrome (PRES) . Pak J Med Sci Apr- jun 2005; 21(2):149-54
10. Hashmi M, Siddiqi SA, Saleem F, Mustafa MS. Posterior reversible leucoencephalopathy. J Pak Med Assoc Sep 2007;57 (9):468-70
11. Naqi R, Ahsan H, Azeemuddin M. Posterior reversible encephalopathy syndrome : a case series in patients with eclampsia. J Pak MED Assoc May 2010; 64 (5): 397-7
12. Siddiqui TS, Haq IU, Rehman B,Kumar M,Iqbal N, Posterior reversible encephalopathy syndrome(PRES) J Coll Physicians Surg Pak Mar 2012; 22 (3):168-70
13. Ehtisham S, Hashmi HA. Posterior reversible encephalopathy syndrome. J Coll Physicians Surg Pak Jun 2012;22(6):398-400