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CORRESPONDENCE

Is poor outcome predictable in posterior reversible encephalopathy syndrome? A case series

Posterior reversible encephalopathy syndrome (PRES) was first described by Hinchey *et al*. in 1996, as a distinct clinico-radiological disease with clinically presence of acute onset seizures, encephalopathy, headache and/or visual disturbances together with radiological findings of vasogenic brain edema typically in the parieto-occipital white matter.[1,2] The prognosis of PRES is generally benign. However, it can result in poor outcome and even death.[3-5] We aimed to analyze the clinical findings of PRES patients followed at our institution and identify the possible unfavorable prognostic factors. We retrospectively reviewed the hospital charts of patients diagnosed with PRES in the last two years.

There were 13 patients (10 female, 3 male; age: 19-70 years). The characteristics of our patients are summarized in Table 1.

All patients had vasogenic edema findings in their initial brain MRI. Six (46%) of them showed diffusion restriction; 3 of them showing bleeding foci in the PRES related edema area on SWI sequences. The patients with no diffusion restriction on MRI (n=7), recovered fully. However, the outcome was poor in 4 of 6 patients with diffusion restriction on MRI (3 patients died, 1 patient had a squelae of homonymous hemianopia). The common features of the 3 patients who died were that their MRIs showed diffusion restriction, and they had an underlying serious disease that required the use of chemotherapeutic agents. In surviving patients with diffusion restriction, PRES was associated with suddenly increased blood pressure, and these patients did not have a serious underlying disease.

Among those patients with diffusion restriction on MRI, the patient with a gross cerebral bleeding focus died and two patients with micro hemorrhage recovered without sequelae.

Despite the syndrome's name, clinical and structural abnormalities may not be reversible and PRES can cause morbidity and mortality.[3-5] In the Berlin PRES study, data of 151 patients with PRES were analyzed retrospectively.[4] The authors reported 17 (11.2%) of their patients died and elevated CRP levels, altered coagulation, altered mental status and subarachnoid hemorrhage were independently associated with in-hospital death. Cytotoxic edema was present in 34% of patients with nonfatal outcome and in 56% of patients with fatal outcome; however, the difference was not statistically significant.

A meta-analysis reported in 2018, included 6 studies with 448 cases.[3] They mentioned that hemorrhage was associated with high risk and toxemia of pregnancy was associated with reduced risk of poor outcome. The pooled OR for cytotoxic edema was 2.59 but did not show statistical significance. Nevertheless, the researchers interpreted the results as suggesting that bleeding or cytotoxic edema may be associated with poor prognosis. Schweitzer *et al*. investigated possible characteristics in PRES associated with clinical outcome. In this study 99 cases of PRES were analyzed retrospectively.[5] Extensive vasogenic edema, hemorrhage with mass effect, or diffusion restriction was associated with worse clinical outcome.

Looking at Table 1, all of the patients without cytotoxic edema on MRI (n=7) showed complete clinical recovery, while half of those with cytotoxic edema (n=6) died.

In terms of etiology, 5 of our patients were in the peri-partum state. All, including the patient with cytotoxic edema on MRI, had a complete clinical recovery. Six of our patients had a history of serious illness (malignancy or autoimmune disease) and use of immunosuppressive drugs or chemotherapeutic agents. Two of them did not have cytotoxic edema on MRI and showed full recovery, while 3 of 4 patients with cytotoxic edema on MRI, died.

This is a case series with 13 patients presented. Together with considering the results of previous studies, PRES in the context of toxemia of pregnancy may have a good prognosis even in the presence of cytotoxic edema, Secondly, cytotoxic edema on MRI seems to indicate a poor prognosis; however, this still needed to be confirmed. Lastly, presence of micro hemorrhage is not likely to contribute to poor prognosis.

Although the interest in PRES has improved since it was defined about 30 years ago, it is clear that there are still questions to be answered. Why do some patients develop cytotoxic edema while others do not? Could inflammation be associated with cytotoxic edema and/or poor prognosis? Can

Table 1: Clinical and radiological features of the patients. The patients with poor prognosis are shown in the first 4 lines and the patients with diffusion restriction on brain MRI are shown in the first 6 lines (marked with bold characters)

No	Age/ gender	Underlying Disease	Acute elevation in BP	Confusion	Seizure	Headache	Visual signs	Localization of vasogenic edema on MRI	Diffusion restriction on MRI	Hemor-rhage on MRI	Outcome
1	54/F	NHL	+	+	+	-	-	Bifrontoparieto-occcpital, bilateral thalamus	+	-	Exitus
2	54/M	NBD	-	+	-	-	-	Bilateral parieto-occipito-temporal	+	Gross	Exitus
3	35/F	Liver tx	-	+	-	-	-	Bilateral occipital and cerebellar	+	-	Exitus
4	55/F	HT	+	+	+	-	+	Bilateral occipital (right>>left)	+	-	Sequel *
5	70/F	GastricCA	+	+	+	-	+	Bilateral occipital, right frontal, left parietal	+	Micro	FR
6	32/F	C/S	+	+	-	-	+	Bilateral parieto-occipital,cerebellar and bilateral basal ganglia	+	Micro	FR
7	53/M	GastricCA	-	-	-	-	+	Bilateral parieto-occipital	-	-	FR
8	19/F	Leukemia	**-**	**-**	**+**	**-**	**+**	Bilateral fronto-parieto-occipital	-	-	FR
9	25/F	C/S	**+**	**+**	**+**	**-**	**-**	Bilateral occipital, bilateral basal ganglia	-	-	FR
10	36/F	C/S	**+**	**-**	**+**	**-**	**-**	Bilateral fronto-parieto-occipital	-	-	FR
11	32/F	C/S	**+**	**+**	**+**	**-**	**-**	Bilateral fronto-parieto-occipital, bilateral basal ganglia	-	-	FR
12	32/F	C/S	**+**	**-**	**+**	**+**	**-**	Bilateral fronto-parieto-occipital, bilateral cerebellar	-	-	FR
13	68/M	HT	**+**	**+**	**+**	**-**	**+**	Bilateral occipital	-	-	FR

Tx: transplantation, HT; Hypertension, C/S: Cesarean Section, NBD: Neuro Behçet's Disease, NHL: Non-Hodgkin Lymphoma, BP: Blood Pressure, MRI: Magnetic Resonance Imaging, CA: Cancer, FR: Full recovery
* Homonymous hemianopia

it enable us to develop treatment strategies, such as steroid therapy or plasmapheresis that may have a positive effect on clinical outcome?

*Seyda Erdoğan *MD*, *Zerin Özaydın Aksun *MD*
**S Erdogan and ZO Aksun contributed equally to this work and are joint first author*

Neurology Department, Faculty of Medicine, Ankara University, Ankara, Turkey

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Address correspondence to: Seyda Erdoğan, MD Ankara University School of Medıcıne, Department of Neurology, Ibn-i Sina Hospital, 06100, Ankara, Turkey. e-mail: dr_seyda@yahoo.com

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