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BRIEF REPORT

The spectrum of posterior reversible encephalopathy in systemic lupus erythematosus

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Abstract Our aim was to compare our South African cohort of systemic lupus erythematosus (SLE) and posterior reversible encephalopathy syndrome (PRES) with other published series. We reviewed the records of 10 patients with SLE and PRES seen over a 10-year period and their demographic data, clinical manifestations, laboratory tests, imaging findings, and outcome were recorded. We identified 10 females who included six Indians, three mixed ethnicity, and one African Black. Three patients had PRES at the onset of SLE. The most common manifestations at presentation were seizures (100 %), hypertension (80 %), and altered mental state (50 %). On neuroimaging, nine patients had bilateral involvement, and the occipital (90 %), parietal (90 %), and frontal lobes (50 %) were most commonly involved. The risk factors for PRES were disease activity (90 %), renal disease (80 %) and hypertension (80 %). Ninety percent of the patients were on immunosuppressive therapy. Immunosuppressive therapy was increased in six patients (60 %), continued in two and reduced in two patients after the diagnosis of PRES. Seven patients recovered completely and three patients died from co-morbidities. A review of the larger case series of SLE and PRES showed that the presentation and neuroimaging findings were similar; most patients had active disease at the time of PRES and the majority of patients required intensification of immunosuppressive therapy. We have shown that the majority of patients with SLE have active disease at the time of PRES, and they require an increase in their immunosuppressive therapy.

Keywords Systemic lupus erythematosus · Posterior reversible encephalopathy syndrome · Ethnicity · Asians · Immunosuppressive drugs · Disease activity

Introduction

The posterior reversible encephalopathy syndrome (PRES), also called the reversible posterior leucoencephalopathy syndrome, is a clinical syndrome of acute onset of headaches, seizures, hypertension, alteration in the mental state, and cortical blindness accompanied by characteristic findings of vasogenic oedema on neuroimaging studies.

In 2008, Mak et al. reviewed the literature on PRES, and noted that there were only 170 reported cases, 17 of whom were associated with systemic lupus erythematosus (SLE) [1]. Since then, 585 episodes of PRES have been reported in 569 patients in six large series, five from the USA, and one from Germany [2–7]. In 2013, Shaharir et al. reported an analysis of 87 cases of PRES in SLE [8]; they identified a further 27 patients in whom individual data was not reported [9, 10]. In a recent multicenter case-controlled study in Mexico, 48 episodes of PRES were reported in 43 patients with SLE [11].

In 1996, Hinchey et al. reported a series of 15 patients with PRES [12]. Seven of their patients were on immunosuppressive therapy (cyclosporine four and tacrolimus three), and four

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had hypertensive encephalopathy, two of whom had SLE [12]. All their patients recovered within 2 weeks with control of hypertension and seizures, and reduction in the dose of immunosuppressive therapy. Since then, immunosuppressive therapy has been recognized as a potential cause or risk factor for PRES.

The most common causes of PRES have included hypertension, pre-eclampsia/eclampsia, immunosuppressive therapy or post-chemotherapy, autoimmune disease, and infections [2–6]. A large number of other diseases and toxic agents have been also been reported as causes of PRES [13, 14]. When patients develop PRES after receiving calcineurin inhibitors for solid organ transplants, the general recommendation is to reduce the dose, stop treatment, or use alternate agents [5, 15]. However, when PRES occurs in SLE, a need to increase immunosuppressive therapy for active multisystem disease has been reported [8, 9].

Our aim was (a) to report our 10 patients with SLE and PRES seen in our multiethnic cohort of South African patients and compare our findings with other published series, (b) to compare PRES in unselected series of patients and series of patients with SLE, and (c) to analyze the use of immunosuppressive agents in SLE patients with PRES.

Patients and methods

We reviewed the records of patients with SLE who were seen in the Department of Rheumatology at Inkosi Albert Luthuli Central Hospital in Durban, South Africa from June 2003 to June 2013. Patients who were diagnosed as having SLE (according to the revised 1982 ACR criteria [16, 17]) and PRES were selected for inclusion in the study. A diagnosis of PRES was made on the basis of an acute presentation with seizures, hypertension, altered mental state, headaches, visual disturbance or the presence of focal neurological signs, and typical neuroimaging findings. The demographic data, clinical manifestations, co-morbid conditions, laboratory tests, imaging findings, treatment in the month preceding PRES and after the diagnosis of PRES, and outcome were recorded. As seizures are a feature of PRES, we calculated a modified systemic lupus erythematosus disease activity index (SLEDAI) score (SLEDAI-N) which excluded the neurological component of the SLEDAI. We defined disease activity based on the SLEDAI score with 0 being no disease activity, 1–5 as mild activity, 6–10 as moderate activity, 11–19 as high disease activity, and ≥20 indicating very high disease activity [18].

Ethical approval for the study was obtained from the Biomedical Ethics Review committee of the University of KwaZulu-Natal.

Results

Case series of patients with SLE and PRES

We identified 10 patients with SLE and PRES, and their findings are shown in Table 1. The diagnosis of PRES was made on MRI in nine patients and on CT scan in one patient (patient 3). Patient 2 had concomitant SLE and HIV infection and was reported previously in our series of patients with SLE and HIV [19]. Her CD4 count was 408 cells/mm^3, and she was not on antiretroviral treatment. Her neurological manifestations were attributed to SLE as she had moderate disease activity, positive ds-DNA antibodies, and hypocomplementaemia at presentation.

All the patients were female, including six Indians, three mixed ethnicity, and one African Black. The median age was 24 years (13–40 years), the median duration of SLE was 13 months (0–230 months), and the median number of criteria was 6 (4–9). Three patients developed PRES at the onset of SLE. The commonest manifestations were seizures (100 %) and hypertension (80 %), followed by altered mental state (50 %), focal weakness (40 %), headaches (30 %), and visual symptoms (30 %). The most common co-morbidities at presentation were hypertension (40 %) and hypothyroidism (30 %). The anti-nuclear antibodies were positive in all patients, and 70 % had positive ds-DNA antibodies. A positive lupus anticoagulant and or anticardiolipin antibodies were noted in five patients, but none had the anti-phospholipid antibody syndrome. The most common sites of involvement on neuroimaging were the occipital (90 %), parietal (90 %), and frontal lobes (50 %), followed by the cerebellum (30 %), temporal lobe (20 %), and brain stem (20 %). Nine patients (90 %) had bilateral involvement, except patient 2. The MRI findings at the onset of PRES and at follow-up 2 months later in patient 1 are shown in Fig. 1. The median SLEDAI-N score was 11.5 (0–21), with disease activity being moderate in three, high in two, and very high in four patients. One patient had inactive disease, apart from the neurological manifestations.

In our series, the most common risk factors were disease activity (90 %), lupus nephritis (80 %), hypertension (80 %), and raised creatinine >132 μmol/l (50 %). The majority (90 %) of the patients were on immunosuppressive therapy. Eight patients were on oral corticosteroids with a median dose of 40 mg (5–60 mg), three of whom also received intravenous methylprednisolone. Five patients were on mycophenolate mofetil, one of whom also received intravenous cyclophosphamide. One other patient was on azathioprine.

After the diagnosis of PRES, the management involved control of seizures, blood pressure, and disease activity. The immunosuppressive therapy was increased in six patients, who had active lupus at the time of developing PRES, and continued in two patients (patients 3 and 7) who were already on high doses of immunosuppressive therapy. The dose was

Table 1 Demographic data, manifestations, treatment, and outcome of PRES

Patient number	1	2	3	4	5	6	7	8	9	10
Age (years)	13	18	25	31	17	19	23	31	32	40
Race / gender	Indian / F	Indian /F	Indian /F	Colored /F	Colored / F	African /F	Colored / F	Indian /F	Indian /F	Indian /F
SLE duration (mo.)	1	Onset	86	Onset	24	Onset	2	139	65	230
Number of criteria	7	5	4	5	9	7	7	5	8	4
SLEDAI-N	21	7	10	20	12	9	21	11	20	0
Clinical features	S, HT, V, H, FW	S, V	S, HT, AMS	S, HT, AMS, FW	S, HT	S, AMS	S, HT, AMS	S, HT, V, H	S, HT, H, FW	S, HT, AMS, FW
Vasogenic oedema sites	O, P, F	P	O, CSO	O, P, F	O, P, T, BG, BS, CB	O, P, F	O, P, F	O, P	O, P, T, CB	O, P, F, CB, BS, CSO
Co-morbidities		HIV	HT, atrial myxoma				HT, hypothy.	HT, IHD	Hypothy	HT, hypothy, AVN hip, RF on HD
Positive auto-antibodies	ANA, Sm, RNP, ds-DNA, SSA, SSB, RNP, ACA	ANA, ACA, ds-DNA, LA	ANA, SSA, LA	ANA	ANA, SSA, ds-DNA, RNP, ACA	ANA, Sm, ds-DNA, RNP	ANA, Sm, ds-DNA, SSA	ANA, Sm, ds-DNA, SSA, RNP, LA	ANA, ds-DNA	ANA
Risk factors and treatment before PRES										
Hypertension	+	−	+	+	+	−	+	+	+	+
Creatinine (μmol/l)	259	53	385	586	98	43	412	103	117	513
Lupus nephritis	+	−	+	+	+	−	+	+	+	+
Immunosuppressive therapy	IVMP (×3), OCS 40 mg, MMF	Nil	IVMP (×3), OCS 60 mg	OCS 20 mg	IVMP (×1), OCS 40 mg, MMF	OCS 40 mg	OCS 60 mg, IVCYP, MMF	OCS 5 mg, MMF	OCS 15 mg, AZA	MMF
Other	*S. aureus* empyema						Gram negative septicemia			Septicemia erythropoietin
Therapy after PRES diagnosed										
Immunosuppressive therapy Drugs	IVMP X 1, OCS 50 mg, IVCYP, MMF, P/pheresi	Oral CS 45 mg	OCS 60 mg	IVMP ×3, OCS 50 mg, IV CYP	IVMP ×3, OCS 50 mg, IV CYP	OCS 30 mg	OCS 50 mg, IV CYP	OCS 40 mg, MMF dose ↑	OCS 40 mg, MMF	Nil
Therapy change	Increased	Increased	Continued	Increased	Increased	Decreased	Continued	Increased	Increased	Decreased
Seizures	+	+	−	−	+	+	+	−	+	−
Hypertension	+	−	+ (NGI)	+ (NGI)	+ (NGI)	−	+	+ (NGI)	+	+ (NGI)
Recovery	Complete	Complete	Complete	Demised	Demised	Complete	Complete	Complete	Complete	Demised

F female, *mo.* months, *S* seizures, *HT* hypertension, *V* visual disturbance, *H* headaches, *FW* focal weakness, *AMS* altered mental state, *O* occipital, *P* parietal, *F* frontal, *CSO* central semi-ovale, *T* temporal, *BG* basal ganglia, *BS* brain stem, *CB* cerebellum, *hypothy* hypothyroidism, *IHD* ischaemic heart disease, *AVN* avascular necrosis, *RF* renal failure, *HD* hemodialysis, *ANA* anti-nuclear antibody, *ACA* anticardiolipin antibody, *LA* lupus anticoagulant, *IVMP* intravenous methylprednisolone, *OCS* oral corticosteroids, *MMF* mycophenolate mofetil, *IVCYP* intravenous cyclophosphamide, *AZA* azathioprine, *NGI* nitroglycerin infusion

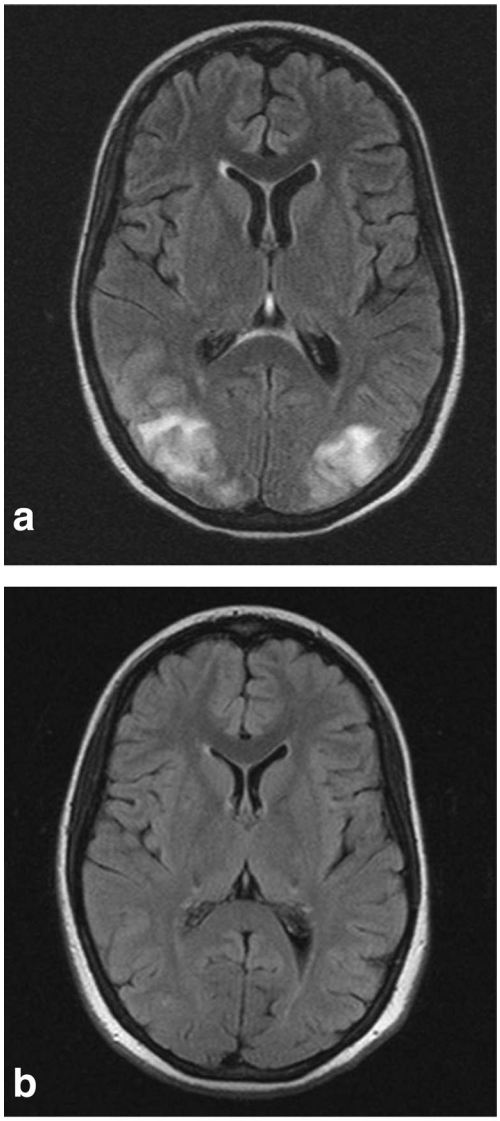

Fig. 1 The MRI findings in patient 1 show **a** hyper-intense lesions in the parieto-occipital regions with **b** resolution on repeat MRI 2 months later

reduced in two patients, one of whom (patient 6) was on a tapering steroid regimen, and the other (patient 10) had inactive SLE and subsequently developed septicemia. Seven patients showed a complete recovery, and three patients died during their hospitalization. Patient 4 showed initial improvement but eventually demised from pulmonary alveolar hemorrhage, intracranial hemorrhage, and infection. Patient 5 required ventilation to control seizures and died from multiple nosocomial infections. Patient 10 showed partial improvement but died following pulmonary embolism.

Repeat MRI was performed in five patients (patients 1, 6–9); they showed complete resolution in four and significant improvement in one. Two patients (patients 2 and 3) recovered completely and did not have repeat scans. The three patients who died did not have repeat MRI, but patient 4 had a CT scan which showed intracranial hemorrhage.

Comparison of the manifestations, risk factors, and outcomes in PRES associated with SLE

We compared the 87 patients reviewed by Shaharir et al. and a further 122 patients from six larger case series, including our 10 patients [8, 9, 11, 20–22]. The mean and median age was 27.9 years or less, and 89.6 to 100 % of the patients were females. PRES occurred at first presentation in 20 to 30 % [20, 21]. The prevalence of PRES ranged from 0.69 % of 3746 patients in Taiwan to 2.0 % of 740 patients in Korea [21, 22]. In the study by Baizabal-Cavallo, PRES accounted for 1.5 % of 1425 emergency room consultations for SLE [9]. Shaharir et al. noted that the ethnic composition of the patients they reviewed was Asians 74.2 %, Hispanics 16.4 %, and Caucasians 6.6 %. We noted that a further 48 (39.3 %) of 122 patients in Table 2 were also Asians [8, 20–22]. The most common risk factors for PRES were active disease with high SLEDAI and SLEDAI-N scores, hypertension (68–93.3 %), presence of renal disease (80.0–93.3 %), and elevated serum creatinine (20–80 %). There were 61.5–95.5 % of patients who were on immunosuppressive therapy. We found that in 36 (62.1 %) of the 58 episodes in three recent studies, blood transfusion was a risk factor [9, 20, 22]. A complete recovery was noted in 61.5 to 100 % of patients. The mortality in SLE ranged from 4.5 to 30.0 % and was usually due to involvement of other organ systems or co-morbidities such as infection.

The spectrum of manifestations in unselected patients with PRES

We reviewed the findings from six large studies of 585 episodes of PRES in 569 unselected patients [2–7]. The mean age of the patients ranged from 33.5 to 50 years and females accounted for 52.6 to 68.4 %. The most common primary causes of PRES were hypertension (22.4–64.4 %), calcineurin inhibitors (22.7–50 %), infections (4–24.3 %), eclampsia/pre-eclampsia (6–22.7 %), and the use of chemotherapy, immunosuppressive drugs, or cytotoxic agents (2.6–22 %). Hypertension, malignancy, infection, renal disease, and autoimmune disease were considered to be the primary cause in some studies and co-morbidities in other studies. Patients in the unselected series had serious co-morbidities such as organ transplantation and malignancies, and a complete recovery was noted in only 65–67.3 % [5, 6].

Immunosuppressive therapy and PRES in SLE

The majority (61.5–95.5 %) of patients with PRES and SLE received immunosuppressive therapy prior to the onset of PRES. Most of the patients were on high doses of oral corticosteroids or intravenous methylprednisolone, while others received cyclophosphamide, azathioprine, or mycophenolate

Table 2 Comparison of findings of series of PRES and SLE

	Shaharir et al. [8]	Baizabal-Carvalho et al. [9], Mexico	Liu et al. [20], China	Jung et al. [21], Korea	Lai et al. [22], Taiwan	Merayo-Chalico et al. [11], Mexico	Current study – 2015, S. Africa
Patients (episodes)	87	21 (22)	10	15	23 (26)	43 (48)	10
Prevalence		1.5 %[a]	(10/732) 1.4 %	(15/740) 2.0 %	(23/3746) 0.69 %		
Study period (months)	NR	64	30	156	85	185	121
Females (%)	95.4	95.5	100	100	95.7	89.6 (Episodes)	100
Age in years	26.3±8.8[b]	24.9±8.6[b]	22.9±2.5[b]	27 (17–51)[c]	26.4±7.6[b]	27.9±1.05[b]	24 (13–40)[c]
SLE duration in months	4.4±1.3 years[b]	61.8±53.6[b]	20.8±12.8[b]	6.1 (0–18.6 years)[c]	5.9±5.8 years[b]	5.42±0.82[b]	13 (0–230)[c]
Clinical features at presentation							
Hypertension	91.7 % (*n*=85)	81.8 %	80 %	93.3 %	88.5 %	68 %	80 %
Headaches	60.9 %	86.3 %	30 %	60 %	76.9 %	NR	30 %
Seizures	78.2 %	95.5 %	80 %	93.3 %	57.7 %	NR	100 %
Altered mental state	54.5 %	27.3 %	70 %		61.5 %	NR	50 %
Visual disturbance	47.1 %	45.4 %	20 %	33.3 %	57.7 %	NR	50 %
Focal weakness		9.1 %				NR	40 %
Risk factors							
Active disease	97.5 % (*n*=79)	SLEDAI 25.7±6.2[b]	SLEDAI 25.8±5.7[b]	SLEDAI-N 17(5–22)[c] SLEDAI 25(13–45)[c]	SLEDAI-N 16±5[b]	SLEDAI 17.6±0.8[b]	SLEDAI-N 11.5 (0–21)[c]
Renal disease	85.1 % (*n*=84)		80 %	93.3 %	84.6 %	NR	80 %
Raised creatinine [d]	235.6±173.2 μmol/l	72.7 %	20 %	80 %	73.1 %	304.1±43.3 μmol/l[b]	50 %
APLS / APL antibody	10.5 %	36.3 % / 57.1 %	0 % / 40 %	0 % / 33.3 %	7.7 %	10 %	0 % / 40 %
Other		BTF 9 (40.9 %)	BTF 10(66.7 %)		BTF 17(65.4 %)		EPO 1(10 %)
Sites of vasogenic oedema							
Occipital		81.8 %	100 %	93.3 %	100 %	84 %	90 %
Parietal		59.0 %	80 %	80 %	76.9 %	NR	90 %
Frontal		36.3 %	40 %	66.7 %	53.8 %	NR	50 %
Temporal		40.9 %	50 %	60 %	65.4 %	NR	20 %
Brainstem		18.1 %	10 %	6.7 %		NR	20 %
Cerebellum		54.5 %	20 %	26.7 %	34.6 %	NR	30 %
Basal ganglia		9.0 %	10 %	13.3 %		NR	10 %
Immunosuppressive therapy							
Before PRES	64.9 %	95.5 %	80 %	73.3 %	61.5 %	NR	80 %
After PRES	NR	100 %	100 %	100 %	100 %	NR	90 %
Drug therapy before PRES							
OCS	50 %	95.3 %	IVMP 60 %[e]	100 %		Mean 31.5±3mg[b]	80 %
CYP	22 %	13.6 %	CYP 80 %[e]	46.7 %		1385±475mg[b]	10 %

Table 2 (continued)

	Shaharir et al. [8]	Baizabal-Carvalho et al. [9], Mexico	Liu et al. [20], China	Jung et al. [21], Korea	Lai et al. [22], Taiwan	Merayo-Chalico et al. [11], Mexico	Current study − 2015, S. Africa
Other	CYCS 12 %, MMF 8 %, AZA 8 %, rituximab 2 %	AZA 27.3 %		MMF 13.3 % mizoribine 13.3 % IVIG 6.67 %	Prior IS use in 61.5 %	MMF 180.8±78.1mg[b] AZA 17.5±5.3 mg[b]	IVMP 30 %, MMF 50 % AZA 10 %
Drug therapy after PRES							
OCS	NR	100 %	80 %	100 % dose ↑80 %	100 %	NR	90 %
IVMP	NR	68.2 %	80 %		15.4 %	NR	30 %
CYP	NR	36.6 %	80 %			NR	40 %
Other	NR	AZA 60 %	IVIG 80 %	NR	Other IS in 46.2 %	NR	MMF 30 %, PPh 10 %
Outcome							
Complete recovery	90.8 %	86.3 %	80 %	86.7%[f]	61.5 %	100 %	70 %
Partial recovery		9.1 %	10 %		11.5 %		
Death		4.5 %	10 %	6.7 %	26.9 %[g]	3 (7 %) at 12 months	30 %

NR not reported, *APLS* anti-phospholipid antibody syndrome, *APL* anti-phospholipid, *BTF* blood transfusion, *EPO* erythropoietin, *OCS* oral corticosteroids, *IVMP* intravenous methylprednisolone, *CYP* cyclophosphamide, *CYCS* cyclosporine, *MMF* mycophenolate mofetil, *AZA* azathioprine, *IVIG* intravenous immunoglobulin, *IS* immunosuppressive, *PPh* plasmapheresis

[a] PRES accounted for 1.5 % of all emergency room consultations for SLE

[b] Mean±SD

[c] Median (range)

[d] Creatinine >132 μmol/l or >1.5 mg/dL

[e] At the onset of the disease

[f] Outcome not established for one patient who had previous bipolar disorder and psychosis persisted

[g] Five patients died from septic shock and two from pulmonary hemorrhage

mofetil. Immunosuppressive therapy was used in 90–100 % of SLE patients after the diagnosis of PRES.

Discussion

We report our experience with a series of 10 patients with SLE and PRES in a multiethnic cohort of patients in Durban, South Africa. Our comparison of the findings in our patients with other series of patients in Table 2 shows that there was a marked similarity between the studies. In SLE patients, PRES was most common among young women; hypertension and seizures were the most common findings at presentation; the occipital, parietal, and frontal lobes were the most common sites affected, and the most common risk factors were disease activity, hypertension, and renal disease. Although the majority of the SLE patients were on immunosuppressive therapy prior to the onset of PRES, many of them required an increase in therapy to control disease activity. The reason for the relatively higher prevalence of PRES among Asians is not known but may be related to them having more severe disease [23].

We compared the findings of PRES in SLE patients with PRES patients from unselected series. In both groups, the most common manifestations at presentation were hypertension and seizures, and the most common sites of vasogenic oedema were the occipital, parietal, and frontal lobes. PRES patients with SLE were young women, while the patients in the unselected series were older and included more males. One of the main differences between PRES in SLE patients and PRES from other causes is that disease activity is the trigger in SLE. Although hypertension and renal disease were a common cause or co-morbidity in both series, the use of calcineurin inhibitors was a major cause of PRES in the unselected series. Among the drugs which are incriminated as precipitants for PRES are immunosuppressive agents for organ transplantation, chemotherapy, high doses of corticosteroids, and other agents such as cyclophosphamide [13]. The calcineurin inhibitors, tacrolimus and cyclosporine, are most widely reported as risk factors or causes of PRES. Since the original study by Hinchey, most authors have recommended a reduction, withdrawal, or change of immunosuppressive therapy after the onset of PRES [12]. In patients with SLE, there are several case reports of PRES occurring after the administration of high-dose corticosteroids or cyclophosphamide [24, 25]. Despite these reports, SLE patients usually require continuation or an increase in their immunosuppressive therapy to control lupus disease activity.

In conclusion, although SLE and PRES have been reported from all over the world, there have not been any case series from Sub-Saharan Africa. The presentation, neuroimaging findings, risk factors, and outcome in our patients are similar to other reported series. The trigger for PRES in SLE is disease activity. We found that although the majority of patients had active disease and were on immunosuppressive therapy prior to developing PRES, most patients still required an increase or continuation of their immunosuppressive therapy to control disease activity after the onset of PRES. An awareness and early diagnosis of PRES enables the clinician to institute appropriate therapy which includes control of blood pressure, management of seizures, and use of immunosuppressive therapy to control disease activity.

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