International Journal of Research in Medical Sciences
Das M et al. Int J Res Med Sci. 2018 Jul;6(7):2551-2553
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20182819

Case Report

Posterior reversible encephalopathy syndrome in a patient with underlying mixed connective tissue disease

Malyaban Das, Prajit Mazumdar*, Divakar Kumar, Vaibhav Mathur

Department of Medicine, Rajendra Institute of Medical Sciences, Ranchi, Jharkhand, India

Received: 22 April 2018
Accepted: 22 May 2018

***Correspondence:**
Dr. Prajit Mazumdar,
E-mail: Prajitmazumdar@gmail.com

ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a condition which is characterized by symmetric involvement of posterior white matter on brain imaging and neurological impairments such as seizures, altered mental status, headache, and visual disturbances. This entity has been classically described with hypertension, renal failure and eclampsia but it can also been seen in cases with normal blood pressure especially in patients receiving immunosuppressive therapy, chemotherapy and in patients with underlying autoimmune disease. Although PRES has been reported with several autoimmune disorders, association of Posterior reversible encephalopathy syndrome (PRES) with mixed connective tissue disease (MCTD) is very rare, hence we report a case of Posterior reversible encephalopathy syndrome in a patient with underlying mixed connective tissue disease (MCTD).

Keywords: Mixed connective tissue disease (MCTD), Posterior reversible encephalopathy syndrome (PRES), Vasogenic oedema

INTRODUCTION

The Posterior reversible encephalopathy syndrome (PRES) was first described by Hinchey in 1996.[1] It is characterized by neurological features like headache, visual disturbances, seizures and vomiting along with bilateral symmetric reversible vasogenic oedema in white matter of the brain in parietal and occipital regions.[2,3] This entity has been classically described with hypertension, renal failure and eclampsia but it has also been seen in cases with normal blood pressure especially in patients receiving immunosuppressive therapy, chemotherapy and in patients with underlying autoimmune disease.

The most important part of management of posterior reversible encephalopathy syndrome (PRES) is early identification of the precipitating factors to prevent further damage to the central nervous system. Hypertension is one of the most important cause of Posterior reversible encephalopathy syndrome (PRES), it has been suggested that a compromised cerebrovascular autoregulation due to acute hypertension may play a important role. Hence, impaired cerebrovascular regulation ultimately leading to arteriole leakage and development of vasogenic edema. Although association between hypertension and posterior reversible encephalopathy syndrome (PRES) is remarkable, there are other possible mechanisms of posterior reversible encephalopathy syndrome (PRES) such as breakdown of blood-brain barrier following cytotoxic agents-induced endothelial toxicity, autoimmunity, and sepsis. It has been seen that endothelial cell damage resulted from different auto antibodies in mixed connective tissue disease (MCTD), leading to development of PRES.

Although PRES has been reported with several autoimmune disorders, its association to mixed

connective tissue disease mixed connective tissue disease (MCTD) is rare, hence we report this case.

CASE REPORT

A 20-year-old female was admitted to our hospital with history of headache and intermittent vomiting over last 3-4 days along with two episodes of generalized tonic clonic seizures 1 day back. On examination, the patient had stable vitals (pulse rate of 80/min and blood pressure of 116/80mmHg) with neck soft and Kerning's test negative. The remainder of systemic examination was also within normal limits.

A CT Scan brain was done which showed bilateral hypo densities in parietal and occipital region of the brain and a underlying posterior reversible encephalopathy syndrome was suspected.

A MRI Brain was done which showed biateral hyper intense signal in occipitial region on T2 weighted MRI (Figure 1) and FLAIR (Fluid attenuated inversion recovery scan) and bilateral hypo intense signal in T1 weighted MRI (Figure 2), thus further confirming the presence of Posterior reversible encephalopathy syndrome.

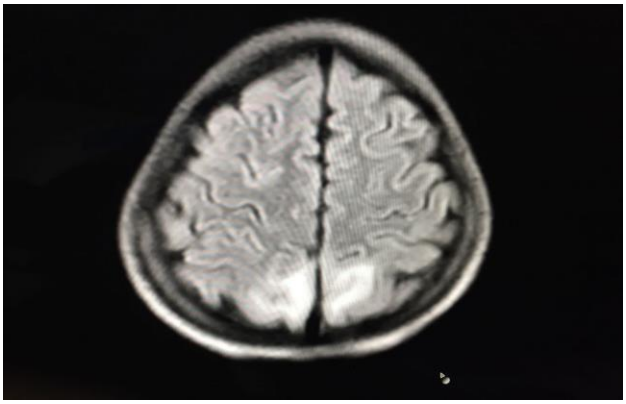

Figure 1: Biateral hyper intense signal in occipitial region on T2 weighted MRI.

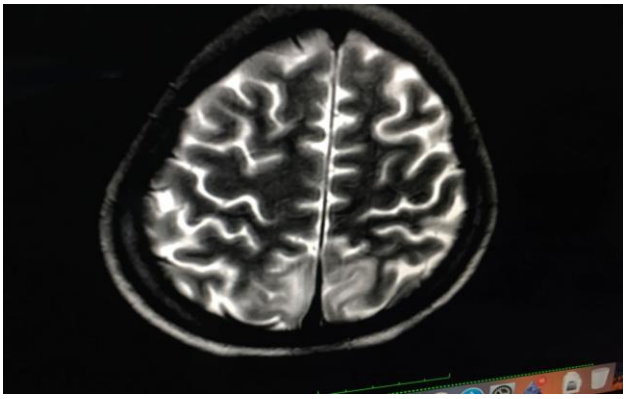

Figure 2: Biateral hypo intense signal in occipitial region on T1 weighted MRI.

Cerebro spinal fluid examination was normal, and remainder of systemic examination was normal except for low hemoglobin on complete blood count and proteinuria (protein+++) on routine urine examination.

A careful detailed history was taken after the patient recovered from post-ictial confusion. The patient gave history of joint pain mainly involving small joints of the hand and elbow and knee joint along with discoloration of her fingers on cold exposure for last 1 year. On detailed examination, patient had pallor and there was acrosclerosis and tender swollen PIP and MCP, along with tender elbow and knee joints were present.

Since the above clinical features are suggestive of a underlying connective tissue disease, a anti-nuclear antibody (ANA) was done. However, blood test for RA Factor and Anti CCP were negative. ANA profile of the patient is shown in Table 1.

Table 1: ANA profile of patient.

ANA profile of patient	
Antibody	Status
Antibody to SS-B	Positive
Antibody to Ro-52	Positive (+)
Antibody to PM-Sc1	Positive (+)
Antibody to Ku	Positive (++)
Antibody to U1-Sn RNP	Positive (+++)
Antibody to Histone	Positive
Antibody to Centromere	Negative
Antibody to Smith	Negative
Antibody to ds-DNA	Negative
Antibody to Scl-70	Negative

Hence a diagnosis of mixed connective tissue disease (MCTD) was made as our patient fulfilled the ALARCON-SEGOVIA diagnostic criteria-positive serology (high titre of anti-RNP) and three of the five clinical criteria-Raynaud phenomenon, Acro sclerosis, synovitis were positive.[4] The KAHN criteria of mixed connective tissue disease was also fulfilled as high titre of U1-RNP positivity and any two of 3 clinical features-Raynauds phenomena, synovitis were present. We treated the patient conservatively following which she recovered and on 12 th day a follow up MRI brain was done which was normal. Hence, a final diagnosis of posterior reversible encephalopathy syndrome in a patient with underlying mixed connective tissue disease was made.

DISCUSSION

Posterior reversible encephalopathy syndrome (PRES) is a condition which is characterized by symmetric involvement of posterior white matter on brain imaging and neurological impairments such as seizures, altered mental status, headache, and visual disturbances.[5] PRES has been described in association to:

- Hypertension,
- Eclampsia,
- Organ transplantation,
- Connective tissue disorders,
- Autoimmune disorders,
- Immunosuppressive therapy, chemotherapy, and HIV patients on antiretroviral treatment.

Two main theories have been proposed:[6]

- High blood pressure leading to loss of cerebral auto regulation which leads to cerebral hyper perfusion with endothelial damage and vasogenic oedema.
- Endothelial dysfunction leading to vasoconstriction and cerebral hypoperfusion, ultimately resulting in subsequent vasogenic oedema.

The neurological manifestations of MCTD are very less as compared to involvement of the other systems and mainly includes aseptic meningitis, trigeminal neuropathies and headache.[7] PRES in a patient with mixed connective tissue disease is rare. It has been seen that endothelial cell damage resulted from different auto antibodies in MCTD, leading to development of PRES.[8] In addition, the dysfunction of the autonomic system has been shown in MCTD.[9] After reviewing the literature, it has been found that PRES has been described in several autoimmune disorders (mainly lupus), but till date only one case has been associated to MCTD in the literature.

CONCLUSION

Posterior reversible encephalopathy syndrome has been described with many autoimmune disorders but its association with mixed connective tissue disease is rare. The pathogenesis appears to be endothelial cell damage resulted from different auto antibodies in MCTD, leading to development of PRES.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. 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.
2. Bartynski WS. Posterior reversible encephalopathy syndrome, part 2: controversies surrounding pathophysiology of vasogenic edema. AJNR Am J Neuroradiol. 2008;29(6):1043-9.
3. Bartynski WS. Posterior reversible encephalopathy syndrome, part 1: fundamental imaging and clinical features. AJNR Am J Neuroradiol. 2008;29(6):1036-42.
4. Amigues JM, Cantagrel A, Abbal M, Mazieres B. Comparative study of 4 diagnosis criteria sets for mixed connective tissue disease in patients with anti-RNP antibodies. autoimmunity group of the hospitals of toulouse. J Rheumatol. 1996;23(12):2055-62.
5. Liman TG, Bohner G, Heuschmann PU, Endres M, Siebert E. The clinical and radiological spectrum of posterior reversible encephalopathy syndrome: the retrospective Berlin PRES study. J Neurol. 2012;259(1):155-64.
6. Bartynski WS. Posterior reversible encephalopathy syndrome, part 2: controversies surrounding pathophysiology of vasogenic edema. AJNR Am J Neuroradiol. 2008;29(6):1043-9.
7. Ortega-Hernandez OD, Shoenfeld Y. Mixed connective tissue disease: an overview of clinical manifestations, diagnosis and treatment. Best Pract Res Clin Rheumatol. 2012;26(1):61-72.
8. Soltesz P, Bereczki D, Szodoray P, Magyar MT, Der H, Csipo I, et al. Endothelial cell markers reflecting endothelial cell dysfunction in patients with mixed connective tissue disease. Arthritis Res Ther. 2010;12(3):R78.
9. Stacher G, Merio R, Budka C, Schneider C, Smolen J, Tappeiner G. Cardiovascular autonomic function, autoantibodies, and esophageal motor activity in patients with systemic sclerosis and mixed connective tissue disease. J Rheumatol. 2000;27(3):692-7.

Cite this article as: Das M, Mazumdar P, Kumar D, Mathur V. Posterior reversible encephalopathy syndrome in a patient with underlying mixed connective tissue disease. Int J Res Med Sci 2018;6:2551-3.