

Case Report

A diagnostic challenge of a patient with postpartum seizures and thrombocytopenia: A case report of posterior reversible encephalopathy syndrome with immune thrombocytopenia

Dilushan Lasith Porawagamage, Harshani Fernando

National Hospital of Sri Lanka

Key words: diagnostic challenge, postpartum seizures, posterior reversible leukoencephalopathy syndrome, immune thrombocytopenia

Corresponding Author: Dilushan Lasith Porawagamage, E-mail:< dilulasith@gmail.com >  <https://orcid.org/0000-0003-4594-9773>

Received: 10 Oct 2021, Accepted: 12 Nov 2021, Published: 22 Mar 2022

Competing Interests: Authors have declared that no competing interests exist

© *Authors*. This is an open-access article distributed under a Creative Commons Attribution-Share Alike 4.0 International License (CC BY-SA 4.0), which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are attributed and materials are shared under the same license.



Introduction

New onset seizures in the postpartum period add a unique challenging facet to the diagnosis, as there are a number of causes with considerable overlap in their presentation. Correct identification of the underlying cause and making an accurate diagnosis is of paramount importance since the treatment will differ completely depending on the underlying aetiology.

Posterior reversible leukoencephalopathy syndrome (PRES) is a well-known cause of seizures in the postpartum period [1]. PRES is a neurological disorder characterized by a range of signs and symptoms, with distinctive neuroimaging findings reflecting vasogenic oedema [2]. We report a case of a woman in the postpartum period who presented with first episode of seizures. She had thrombocytopenia, as well, complicating the picture. Since there was a wide range of differential diagnoses, we describe here how we evaluated the patient carefully, excluding each differential, to come to the correct diagnosis.

Case Presentation

A 20-year-old female was admitted on day 21 postpartum following her first pregnancy with a history of four episodes of generalized tonic clonic (GTC) seizures within 2 hours. Each episode lasted 1-2 min and was followed by post-ictal drowsiness. She had a history of generalized headache which had started the day before the seizures and had lasted throughout the day. This headache was of gradual onset, moderate in severity and did not respond to simple analgesics. There was no fever, vomiting, photophobia or visual disturbances. There was no abdominal pain, haematuria, frothy urine or reduced urine

output. There was no history of head trauma. There was no history of photosensitive skin rash, oral ulcers, alopecia or joint pain. There was no past history of blood transfusion, intravenous drug abuse or sexual promiscuity.

She had a past history of bleeding tendency with an episode of prolonged bleeding following dental extraction at the age of 12 years. However, there was no history of muscle haematoma or haemarthrosis. However, she had had menorrhagia for previous four years for which she had not sought medical advice. There was no history of miscarriages or any history suggestive of deep vein thrombosis or pulmonary embolism. Her pregnancy was unplanned with a late booking visit at a period of amenorrhea (POA) of 24 weeks. Her antenatal records revealed normal blood pressure throughout the antenatal period. There was no gestational diabetes mellitus or proteinuria. Her full blood count at the first booking visit revealed a haemoglobin of 11.2 g/dl and a platelet count of 18000/cmm. This was managed as for immune thrombocytopenia. However prednisolone was not given. She had defaulted on follow up. She underwent an emergency caesarean section with platelet transfusion at a POA of 36 weeks due to fetal distress and delivered a healthy baby.

On examination, she was afebrile, drowsy with a Glasgow coma scale (GCS) of 14/15, conscious and oriented. There were two ecchymotic patches on her left arm. There was no alopecia, oral ulcers, joint swelling or ankle oedema. Neurological examination revealed bilateral equal and reactive pupils. Her fundi were normal. Cranial nerves and cerebellar examination was normal. Upper and lower limb examination was normal including normal reflexes and a flexor plantar response. Her pulse rate was 86 bpm with a blood pressure of 140/90 mmHg. There were no cardiac murmurs. The respiratory system and abdominal examinations were normal.

Her full blood count on admission showed a total white blood cell count of 13×10^3 /cmm, haemoglobin 11g/dl and platelet 43×10^3 /cmm. Blood picture revealed isolated thrombocytopenia with large platelets, suggestive of thrombocytopenia due to peripheral sequestration or destruction. No evidence of microangiopathic haemolytic anemia (MAHA) was seen. Her serum creatinine was 0.9 mg/dl. The urine full report (UFR) showed albuminuria without pus cells or red cells. The 24-hour urinary protein was 50mg/mmol (>30 mg/mmol). Her C-reactive protein level (CRP) was 10 and erythrocyte sedimentation rate (ESR) was 15mm per hour. Serum electrolytes, including ionized calcium and magnesium, were normal. Her liver function tests and coagulation profile were normal.

The cerebrospinal fluid (CSF) full report showed a protein level of 0.45g/l, without cells. CSF sugar was 80mg/dl (RBS 108mg/dl). CSF Gram stain was negative for organisms and CSF culture failed to grow any organism. The electroencephalogram (EEG) showed bilateral delta slow complexes suggestive of generalized brain dysfunction with increased seizure predisposition. Her magnetic resonance imaging scan (MRI) of the brain revealed a T2 FLAIR high signal areas in the bilateral occipital lobes involving the lingual gyrus, cuneus, calcarine sulcus. T2 FLAIR high signal areas were also noted in the left superior

parietal and angular gyrus. The appearance was suggestive of posterior reversible encephalopathy syndrome (PRES).

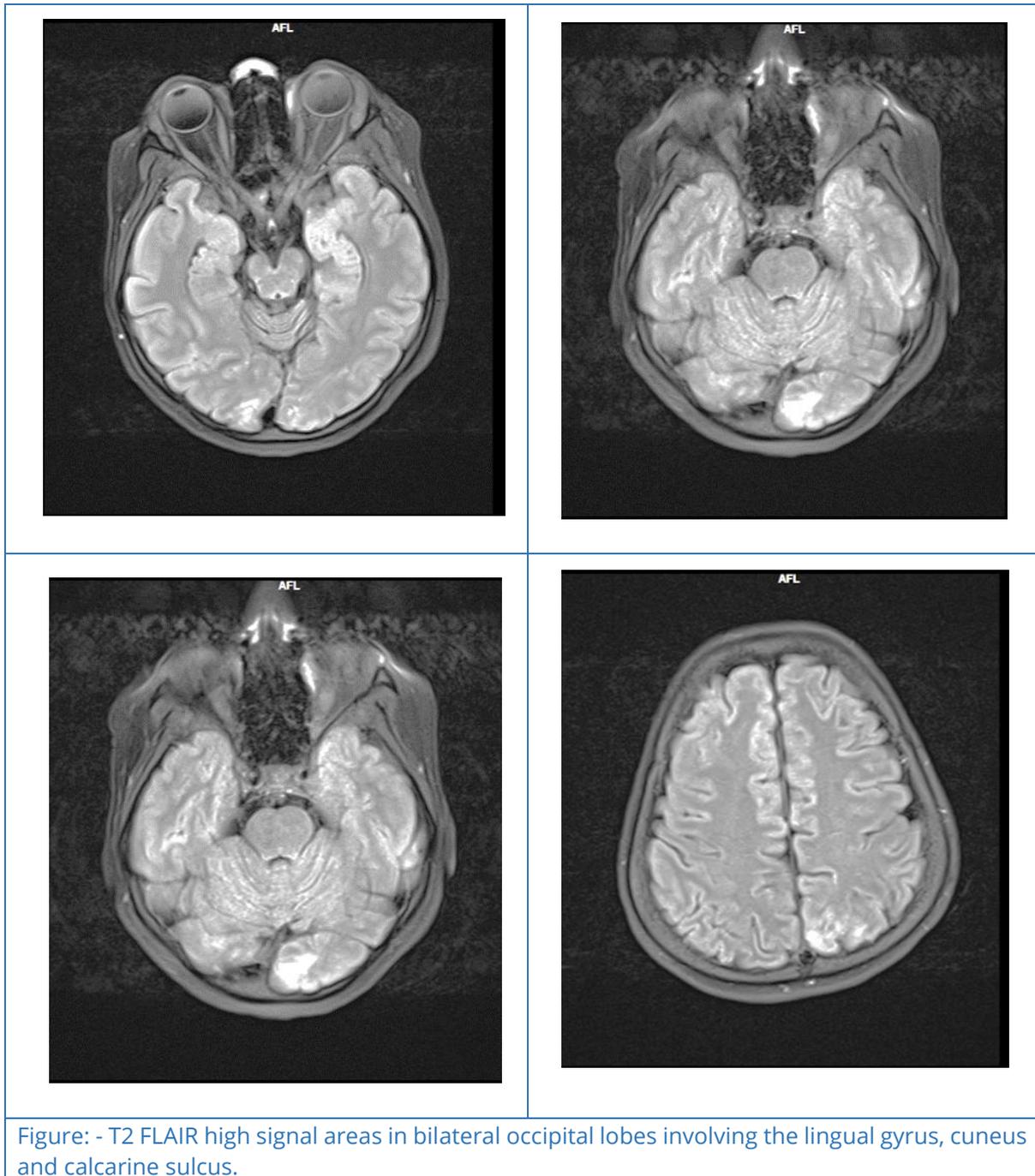


Figure: - T2 FLAIR high signal areas in bilateral occipital lobes involving the lingual gyrus, cuneus and calcarine sulcus.

She was evaluated for any underlying autoimmune disorder and the antinuclear antibody (ANA) was positive at a titre of 1/160. However anti-Ds DNA antibody and extractable nuclear panel involving Anti Ro and Anti La were negative. C3 and C4 levels were normal. Antiphospholipid antibody was negative. Hepatitis and HIV screening were negative.

After extensive evaluation, a final diagnosis of postpartum eclampsia with posterior reversible encephalopathy syndrome and underlying Immune thrombocytopenic purpura was made.

Discussion

When our patient presented with postpartum GTC seizures with a past history of thrombocytopenia, there was a wide range of differential diagnoses including immune thrombocytopenic purpura (ITP) with intracranial haemorrhage (ICH), postpartum eclampsia, thrombotic thrombocytopenic purpura, previously undiagnosed systemic lupus erythematosus (SLE) with cerebral lupus, antiphospholipid antibody syndrome (APLS) with cerebral venous sinus thrombosis, reversible cerebral vasoconstriction syndrome or posterior reversible encephalopathy syndrome.

Since the blood pressure was marginally elevated with proteinuria, we considered eclampsia as our first differential diagnosis. To exclude ICH on a background of ITP, we proceeded with non-contrast imaging of the brain which turned out to be normal. As there was a low platelet count, APLS which carries a thrombosis risk needed to be considered. In addition, cerebral venous sinus thrombosis is known to occur during the postpartum period [3]. Reversible cerebral vasoconstriction syndrome (RCVS) is considered as an important differential diagnosis as RCVS is commonly associated with pregnancy. There is a significant overlap of clinical and radiological features in RCVS and PRES [4]. However, RCVS usually manifests with sudden attacks of severe headache which are described as thunder cap headaches defined as “any severe headache peaking within 1 min” [5]. But our patient demonstrated only a typical headache gradual in onset and diffuse in nature which is described in PRES. This is generally characterized by gradual in onset and diffuse headache [6]. An angiogram may help to diagnose RCVS as there will be segmental narrowing and dilatation of large and medium cerebral arteries [5].

Since PRES is associated with eclampsia, we proceed with an MRI of the brain. The typical findings on MRI are symmetrical white matter oedema in the posterior cerebral hemispheres, particularly in the parieto-occipital region. This was observed in our patient.

Our patient had concurrent ITP. Thrombocytopenia is of concern as certain conditions which causes postpartum seizures can also have thrombocytopenia. They are thrombotic thrombocytopenic purpura, APLS and meningococcal sepsis. The blood picture is important to differentiate them. A diagnosis of primary immune thrombocytopenia is made in the absence of any underlying cause of the thrombocytopenia. Autoimmune mediated thrombocytopenia is commonly associated with connective tissue diseases. In those patients ANA may be positive.

However, there is a subset of patients with primary immune thrombocytopenia patients who have positive ANA but do not meet the classification criteria for any connective tissue diseases or undifferentiated connective tissue diseases. A retrospective study carried out by Yuan Liu et al. studying 226 patients with primary immune thrombocytopenia between 2012 and 2015 revealed that 51.7% of them had a positive ANA. However, most of these

patients had no autoantibodies against extractable nuclear antigen [7]. Therefore, the ANA positivity in immune thrombocytopenia as seen in our patient is a recognized entity.

We could not find any literature regarding an association between ITP and PRES. Therefore, we concluded that this case was a coexistence of two conditions ITP and PRES and plan to follow her up to detect any future development of connective tissue disease.

References

1. Fugate JE. et al. Posterior reversible encephalopathy syndrome: associated clinical and radiologic findings. *Mayo Clinic proceedings* [Internet]. 2010;85(5):427-32. Available from: <https://www.ncbi.nlm.nih.gov/pubmed/20435835>
<https://doi.org/10.4065/mcp.2009.0590>
2. Fischer M, Schmutzhard E. Posterior reversible encephalopathy syndrome. *Journal of Neurology*. 2017 Jan 4;264(8):1608-16. <https://doi.org/10.1007/s00415-016-8377-8>
3. Cantú C, Barinagarrementeria F. Cerebral venous thrombosis associated with pregnancy and puerperium. Review of 67 cases. *Stroke*. 1993 Dec;24(12):1880-4. <https://doi.org/10.1161/01.STR.24.12.1880>
4. Aracki-Trenkić A, Stojanov D, Trenkić M, Radovanović Z, Ignjatović J, Ristić S, et al. Atypical presentation of posterior reversible encephalopathy syndrome: Clinical and radiological characteristics in eclamptic patients. *Bosnian Journal of Basic Medical Sciences* [Internet]. 2016 Aug 1 [cited 2021 Oct 2];16(3):180-6 <https://doi.org/10.17305/bjbms.2016.1201>
5. Pilato F, Distefano M, Calandrelli R. Posterior Reversible Encephalopathy Syndrome and Reversible Cerebral Vasoconstriction Syndrome: Clinical and Radiological Considerations. *Frontiers in Neurology*. 2020 Feb 14;11. <https://doi.org/10.3389/fneur.2020.00034>
6. Maramattom BV. Cough headache as a presenting feature of posterior reversible encephalopathy syndrome (PRES). *Clinical Medicine* [Internet]. 2021 Mar 1 [cited 2021 Oct 3];21(2):e237-8. <https://doi.org/10.7861/clinmed.2020-1080>
7. Liu Y. et al. Clinical characteristics of immune thrombocytopenia associated with autoimmune disease: A retrospective study. *Medicine* [Internet]. 2016 Dec 1 [cited 2021 Sep 25];95(50):e5565. <https://doi.org/10.1097/MD.0000000000005565>