ATYPICAL POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME IN A PATIENT WITH SEPSIS

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PRESENTATION

64 year old lady transferred from outside facility in septic shock secondary to bacterial pneumonia and possible H1N1 influenza.

PATIENT BACKGROUND

Asthma
Essential Hypertension
Polymyalgia Rheumatica
Temporal arteritis
Urinary stress incontinence
Rheumatoid Arthritis

HOSPITAL COURSE

At presentation, the patient was in septic shock and multiorgan dysfunction including, liver, renal, pulmonary, coagulation and platelet dysfunction. The patient was intubated due to acute hypoxemic respiratory failure, transferred to ICU, and pressor, antibiotic and steroid therapy was started. After 7 days she was extubated and transferred to the medical floor.

24 hours after transferring to the floor, she was noticed to have sudden decline in her mental status and developed a seizure. The event lasted for a few minutes and her mental status was back to the baseline by the following morning. She did not experience any other neurological event and was discharged to home 2 days later in a stable condition. At 2 month follow up, she was doing well without any more seizures. A follow up MRI at 5 weeks demonstrated a complete resolution of the abnormal MRI findings.

LABORATORY FINDINGS

Creatinine: Primarily ranged from 2.3-2.9 at the time of admission and during ICU stay; later improved during the hospital course; it was 1.16 at the time of seizure
Calcium: 6.8-7.1
Hyperbilirubinemia and Transaminitis, resolved on day 6
INR:1.6 at the time of presentation
Pneumococcal urine antigen: Positive
Influenza A antigen: Positive

RADIOLOGIC FINDINGS ON BRAIN MRI

Focal regions of symmetric hemispheric edema manifested as abnormal T2 and FLAIR hyperintensity involving subcortical and deep white matter.

The lesions are predominantly seen on parietal/occipital area followed by inferior temporal-occipital region, frontal lobes and cerebellar hemispheres.

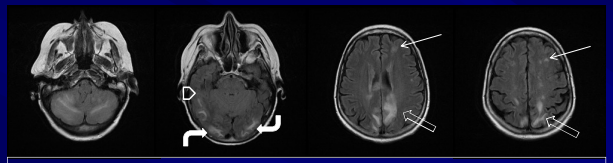

Fig.1 - T2-FLAIR sequences demonstrating, abnormal hyperintensity in occipital (curved arrows), superior frontal (arrows), paramedian parietal (open arrows), right posterior inferior temporal lobe (arrowhead) and posterior cerebellar hemispheres bilaterally

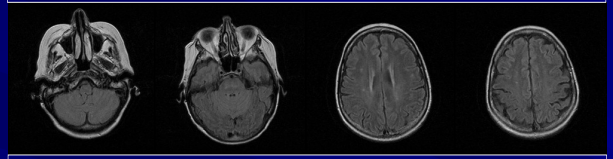

Fig.2 - Follow up MRI after 5 weeks shows resolution of findings in subcortical and deep white matter

IMAGING PATTERN AT MR PERFUSION

Reduced relative cerebral blood volume (rCBV)) in cortex and white matter.

Comparing anterior to posterior hemispheric flow demonstrates significant posterior hypoperfusion, increased mean transit time, reduced rCBV and reduced cerebral blood flow.

IMAGING PATTERN AT MR SPECTROSCOPY

Reduced ***N*-acetylaspartate:choline** and ***N*-acetyl aspartate:Creatine** ratios have been shown in area of PRES vasogenic edema as well as unaffected regions.

IMAGING PATTERN AT ANGIOGRAPHY AND MRI USING TIME OF FLIGHT TECHNIQUE(TOF)

Vasculopathy pattern, including focal vasoconstriction/vasodilation, string of bead pattern and diffuse vasoconstriction

PATHOPHYSIOLOGY

Elements of both vasoconstriction and vasodilatation

- **Severe hypertension** results in failed autoregulation ,hyperperfusion and vasogenic edema
- **Endothelial injury/dysfunction** mediated by TNF-α, IL-1β, Endothelin-1 and activated T-cells , results in vasoconstriction, hypoperfusion, ischemia and ultimately edema

References:
1. Posterior reversible encephalopathy syndrome in infection, sepsis, and shock. Bartynski WS, Boardman JF, Zeigler ZR, Shadduck RK, Lister J. AJNR Am J Neuroradiol. 2006 Nov-Dec;27(10):2179-90
2. Posterior reversible encephalopathy syndrome, part 2: controversies surrounding pathophysiology of vasogenic edema. Bartynski WS. AJNR Am J Neuroradiol. 2008 Jun;29(6):1043-9. Epub 2008 Apr 10
3. A reversible posterior leukoencephalopathy syndrome.Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A, Pessin MS, Lamy C, Mas JL, Caplan LR. N Engl J Med. 1996 Feb 22;334(8):494-500

CONDITIONS AT RISK

Severe Hypertension
Preeclampsia/Eclampsia
Cyclosporine/Tacrolimus therapy in patients with Allogenic BMT – Solid Organ Transplant
Sepsis/Infection/Shock (G+ organisms)
High dose corticosteroids
Autoimmune diseases (SLE-Scleroderma-Wegener's-PAN)

NEUROLOGICAL FINDINGS

Seizure
Encephalopathy
Altered Mental Status
Visual symptoms
Headache
Hemianopsia

PROGNOSIS and COMPLICATIONS

Overall PRES is associated with a good prognosis and self limiting if the primary etiology is no longer present. Recurrence has been reported in a few cases associated with severe hypertension, after allo-BMT, sickle cell disease with infection, allo-BMT with infection, atypical autoimmune diseases with infection.

DISCUSSION

Reversible Posterior Encephalopathy syndrome first described in 1996 as Posterior Reversible Leukoencephalopathy in 15 patients(3 with eclampsia, 7 with bone marrow transplant or solid organ transplant under treatment with Cyclosporin or Tacrolimus, 2 with SLE, 1 with hepatorenal syndrome, 1 with acute nephritis and 1 with melanoma on INF-α therapy.

In 2006, for the first time its occurrence in the setting of Infection/Sepsis/ shock was described in 25 patients by Bartynski et al; 84% of cases showed G+ organisms in culture

Reversible Posterior Encephalopathy syndrome is a neurotoxic state with unique radiologic appearance of bilateral white matter vasogenic edema, predominantly in parietal/occipital and temporal/occipital lobes. It may occur in superior frontal lobe and cerebellar hemispheres.

In 40% of patient with PRES due to sepsis, and overall in 25% of all patients with PRES, blood pressure is within normal range. Interestingly vasogenic edema is greater in normotensive patients. Recent reports also note PRES in the setting of post-streprococcal glomerulonephritis and Henoch-Schonlein purpura in young patients.

This patient demonstrates a case of atypical PRES (lesions in frontal lobe and cerebellar hemispheres) in the setting of sepsis due to bacterial pneumonia and H1N1 influenza, multiorgan dysfunction and recent corticosteroid use. Systolic ,diastolic and mean arterial blood pressure fluctuated between 115-170 mm/Hg,60-86 mm/Hg and 91-119 mm/Hg respectively. These BP findings are compatible with the normotensive group records in Bartynski et al study which interestingly showed greater edema on MRI compare to hypertensive group