ATYPICAL POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME IN A PATIENT WITH SEPSIS

Mohammad Bahador MD, Thomas A Kim MD, Humera Ahsan MD
Department of Radiology, University of Missouri Health Care
Department of Radiology, Carle Foundation Hospital

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

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)

POGNOSIS and COMPLICATIONS
Reversible Posterior Encephalopathy syndrome first described in 1996 as Posterior Reversible Encephalopathy 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 hepaticenral 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 and was 84% of cases showed Gv organisms in culture.

DISCUSSION

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, hyperperfusion, ischemia and ultimately edema

RADIOLOGIC FINDINGS ON BRAIN MRI
Focal regions of symmetric hemispheric edema manifested as abnormal T2 and FLAIR hyperintensity involving subcortical and deep white matter.

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 1.46 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

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/vasodilatation, string of bead pattern and diffuse vasoconstriction

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 1.46 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

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 hypoxic 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.

Fig. 1- T2-FLAIR sequence demonstrating, abnormal hyperintensity in occipital (curved arrow), superior frontal (arrow), paracentral-parallel (open arrow), LTP posterior temporal lobe (arrowhead) and posterior-unilateral hemispheres (elaborated).

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

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.

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.

IMAGING PATTERN AT ANGIOGRAPHY AND MRI USING TIME OF FLIGHT TECHNIQUE(TOF)
Vasculopathy pattern, including focal vasoconstriction/vasodilatation, string of bead pattern and diffuse vasoconstriction

REFERENCES