



A RARE OCCURRENCE OF REVERSIBLE CYTOTOXIC EDEMA IN REVERSIBLE POSTERIOR LEUCOENCEPHALOPATHY SYNDROME.

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Abstract : Common imaging feature of reversible posterior leukoencephalopathy syndrome (RPLS) is with symmetric vasogenic edema involving parietooccipital region. The predilection for posterior circulation is thought to be due to relative lack of sympathetic innervation at the level of the arterioles supplied by the vertebrobasilar system compared with the anterior circulation. However involvement of frontal , temporal lobes, deep white matter, basal ganglia, cerebellum, brain stem all have been reported to varying extent. This is a rare case report of reversible cytotoxic edema along the pyramidal tracts in a patient with RPLS.

Keyword : Vasogenic edema, Facilitated diffusion, posterior circulation, Pyramidal tract

CASE REPORT:

A 22 year-old right-handed woman was transferred to our hospital at 36 weeks of gestation with imminent eclamptic symptoms of holocranial head ache ,vomiting,blurring of vision. She also had two episodes of generalised tonic clonic seizures each lasting for 10 min on admission. Her pulse rate and blood pressure at admission was 112 beats per min and 150/ 100 mmHg. Fundal examination revealed bilateral papilledema. Lab parameters like Complete blood count including platelet count was unremarkable. Liver enzymes are within normal limits. Blood Biochemistry values were Na-140 Meq , K-3.2 Meq , Sugar-130 mg/dl ,Urea-28 mg/dl, Creatinine -0.9 mg/dl . Urine dipstick test was positive for proteinuria.

Labetolol infusion was started along with Magsulf regimen to bring down the blood pressure. Emergency LSCS was done and healthy boy baby of 2.8 kg was delivered. Patient was Drowsy post surgery and from 2nd Post operative day patient had weakness of Right upper and lower limb with facial palsy. On examination patient had power of 3/5 in both right upper and lower limbs and bilateral pyramidal tract signs positive with extensor babinsky reflex. Clinical suspicion of cortical venous thrombosis or post ictal state was made and was referred for CT Brain. Non contrast CT of brain was obtained from vertex to skull base on the 2nd post operative day. Summary of CT findings are Bilateral diffuse asymmetric hypo density of periventricular and subcortical white matter predominantly in parieto occipital regions with accentuation of grey white matter interphase (**fig:1**). Bilateral external capsule , posterior limb of internal capsule , cerebellar white matter hypodensities, Bilateral asymmetric hypodense regions in caudate heads and lentiform nucleus (**fig:2**). There was

also suspicious hypo densities in brain stem (**fig:3**) ,whether it was true lesions or due to beam hardening artefact in posterior fossa could not be made out. No evidence of parenchymal haemorrhage or dural sinus thrombosis was observed in CT. Possible diagnosis of (RPLS) Reversible posterior leukoencephalopathy syndrome was made and MRI was further suggested to evaluate suspicious brain stem hypodensities.

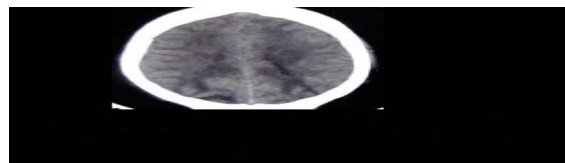


FIG:1 Non contrast CT Brain obtained on 2nd post partum day showing Bilateral Diffuse symmetrical confluent white matter hypodensities Predominantly in parieto-occipital regions with Accentuation of gray-white matter interphase.

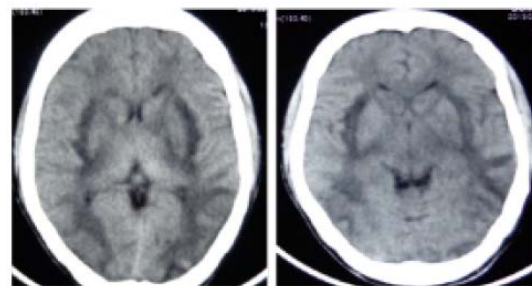


FIG:2 (Right) Non contrast CT Brain on 2nd post partum day showing Bilateral External capsule and posterior limb of internal capsule hypodensities. (Left) Asymmetric low attenuation regions in Bilateral basal ganglia.

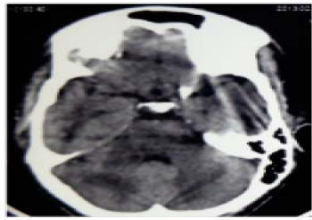


FIG:3 CT Brain obtained on 2nd Post partum day at the level of posterior fossa showing Suspicious hypodense regions in brain stem.Bilateral cerebellar hemisphere involvement.

MRI brain which included T1 Sagittal (TR-400, TE-15), T2 axial (TR-5550, TE-104), FLAIR coronal (TR-9000, TE-115, TI-150), Diffusion weighted images, Gradient, Time of flight MR angiogram and venogram images was obtained on the third post operative day. MR showed Bilateral confluent diffuse periventricular and subcortical white matter T2 and FLAIR hyperintensities with posterior predominance with non restricted diffusion and high ADC values consistent with vasogenic edema (fig:7). Bilateral basal ganglia, external capsule, Bilateral cerebellar white matter (fig:6) involvement was also seen. MR confirmed the suspicious CT abnormality of Brain stem with FLAIR hyper intense abnormalities in brain stem. Pyramidal tract involvement was represented by Bilateral posterior limb of Internal capsule and restricted diffusion along the corticospinal tract in brain stem (fig:8).

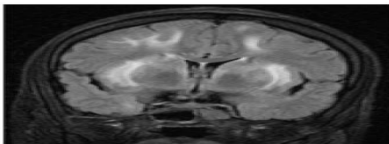


FIG:4 MRI FLAIR coronal image at the level of Basal ganglia shows Hyperintensities involving subcortical white matter regions, external capsule and caudate head

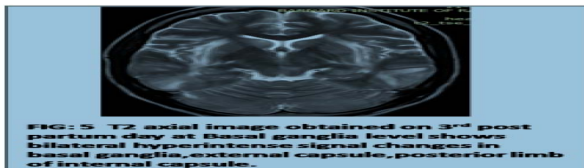


FIG: 5 T2 axial image obtained on 3rd post partum day at Basal ganglia level shows bilateral hyperintense signal changes in basal ganglia, external capsule, posterior limb of internal capsule.

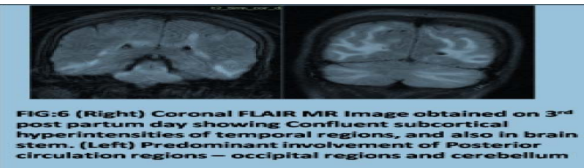


FIG:6 (Right) Coronal FLAIR MR Image obtained on 3rd post partum day showing Confluent subcortical hyperintensities of temporal regions, and also in brain stem. (Left) Predominant involvement of Posterior circulation regions – occipital regions and cerebellum

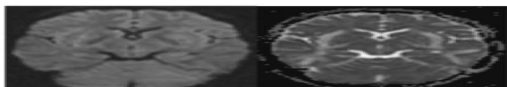


FIG:7 Diffusion Weighted imaging with corresponding ADC map at the level of basal ganglia shows No restricted diffusion with high ADC values suggestive of vasogenic edema

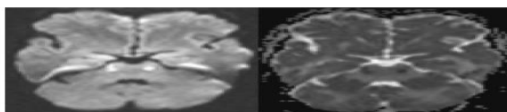


FIG:8 Diffusion Weighted imaging with corresponding ADC map obtained on 3rd post partum day showing Restricted diffusion along bilateral pyramidal tracts in brain stem suggestive of cytotoxic edema

Diagnosis of reversible posterior leukoencephalopathy syndrome with cytotoxic edema along the bilateral pyramidal tracts was made and follow up imaging was suggested after 2 weeks as most of the cases of PRES will resolve in this duration. Patient was put on Tab.Nifedipine to control blood pressure. There was marked clinical recovery from 6th Post operative day with power improving to nearly 5/5 on 2nd post operative week with no residual neurological deficit. Follow up MRI performed after 2 weeks revealed complete reversal of the abnormality (fig:9,10), No regions of restricted diffusion seen in brain stem (fig:11)

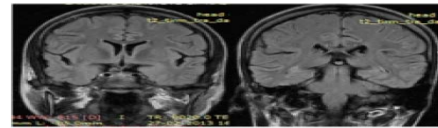


FIG:9 Follow up FLAIR MR images obtained after 2 weeks of initial MRI showing complete reversal of abnormality in white matter, Basal ganglia and brain stem

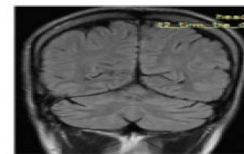


FIG:10 Follow up FLAIR MRI image obtained after 2 weeks of initial MRI showing complete reversal of abnormality in parieto-occipital region and cerebellum.

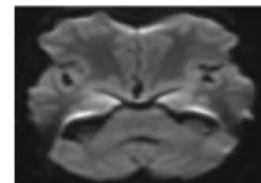


FIG:11 Follow up Diffusion Weighted image obtained after 2 weeks of initial MRI showing reversal of restricted diffusion along the bilateral pyramidal tracts.

Large retrospective study published in AJNR August 2007 28: 1320 - 1327 titled "(2)" Distinct imaging patterns and lesion distribution in posterior reversible encephalopathy syndrome" has reported lesions in occipital / parietal, Frontal, inferior temporal lobes, cerebellum, brain stem, Basal ganglia, Deep white matter, Splenium of corpus callosum. But PRES involving Bilateral pyramidal tracts with reversible cytotoxic edema is rarely reported which is the unique feature of our case.

DISCUSSION:

PRES is recognised in Preeclampsia / eclampsia, Allogeneic bone marrow transplantation, Organ transplantation, Autoimmune disease and High dose chemotherapy (3). Most cases of PRES occur with hypertension or immunosuppression. Reversible posterior leukoencephalopathy syndrome is a clinico radiologic entity characterized by headaches, altered mental status, seizures, and visual loss and is associated with white matter vasogenic edema predominantly affecting the posterior occipital and parietal lobes of the brain (5). The pathophysiology of PRES is under debate, but it is related to disordered cerebral autoregulation (1). Two pathophysiologic mechanisms have been proposed regarding cerebral Autoregulation—cerebral vasospasm, which results in cytotoxic edema, and vasodilatation, which results in vasogenic edema. The latter is more favored by most experimental and clinical data. The pathophysiology of PRES also implicates endothelial dysfunction, especially in

cases without severe hypertension, such as pre-eclampsia or cytotoxic therapies. The most characteristic imaging pattern in PRES (5) is the presence of edema involving the white matter of the posterior portions of both cerebral hemispheres, especially the parieto-occipital regions. PRES favors the posterior circulation, may be because of relative lack of sympathetic innervation at the level of the arterioles supplied by the vertebrobasilar system compared with the anterior circulation; this innervation presumably protects the brain from marked increases in intravascular pressure, such as with severe hypertension. However, other structures (such as the brain stem, cerebellum, and frontal and temporal lobes) may also be involved, and although the abnormality primarily affects the subcortical white matter, the cortex and the basal ganglia may also be involved. Although they are rare, gyriform signal enhancement or parenchymal hemorrhage can occur in complicated cases. Recently, studies with diffusion-weighted sequences and diffusion-tensor sequences have shown increased apparent diffusion coefficients (ADCs) in the involved regions accompanied by anisotropy loss, which suggests reversible vasogenic edema as an underlying pathophysiology. Therefore, early diagnosis and treatment is essential for the patients prognosis.

CONCLUSION:

Classical location of RPLS is bilateral parieto occipital regions, however radiologists should be aware of atypical presentations including involvement of pyramidal tracts . Diffusion weighted imaging can distinguish between cytotoxic and vasogenic edema both of which can occur in RPLS, however in appropriate clinical background diagnosis of RPLS should be made to prevent deleterious work-ups or therapies.

REFERENCES:

- 1)Sebastian Koch, Alejandro Rabinstein, Steven Falcone, and Alejandro I. Case Report:Diffusion-weighted Imaging Shows Cytotoxic and Vasogenic Edema in Eclampsia.AJNR Am J Neuroradiol,June/July 2001;22:1068–1070.
- 2)Distinct imaging patterns and lesion distribution in posterior reversible encephalopathy syndrome.AJNR August 2007 28: 1320 - 1327.
- 3)W.S Bartynski.Posterior Reversible encephalopathy syndrome,Part 1: Fundamental imaging and clinical features. AJNR June 2008 29: 1036 – 1042.
- 4)Alexander M. McKinney^{1, 2}, James Short¹, Charles L , et al .Original Research,Posterior Reversible Encephalopathy Syndrome: Incidence of Atypical Regions of Involvement and Imaging Findings.Neuroradiology,October 2007, Volume 189, Number 4.
- 5)Hyo-Jeong Lee et al . Posterior Reversible Encephalopathy syndrome: Appl Radiol. 2007;36(5): 42 – 43.

