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 was Na-140 Meq, K-3.2 Meq, 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 hypodensity 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. Posssible diagnosis of (RPLS) Reversible posterior leukoencephalopathy syndrome was made and MRI was further suggested to evaluate suspicious brain stem hypodensities.
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 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).

Diagnosis of reversible posterior leucoencephalopathy 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.Nifidipine 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).

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 Preecampsia / 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 leucoencephalopathy syndrome is a clinicoradiologic 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 (6). 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
2) Distinct imaging patterns and lesion distribution in posterior
3) W.S Bartynski. Posterior Reversible encephalopathy
syndrome. Part 1: Fundamental imaging and clinical features. AJNR
4) Alexander M. McKinney 1, 2, James Short 1, 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
– 43.