CASE REPORT

Reversible Posterior Leucoencephalopathy Syndrome in Post Streptococcal Glomerulonephritis

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Reversible posterior leukoencephalopathy syndrome is characterized by an acute, usually reversible encephalopathy, with radiological findings that mainly involve the white or grey matter of the parieto-occipital lobes. We report a case of post streptococcal glomerulonephritis presenting as reversible leukoencephalopathy syndrome. Immediate control of hypertension resulted in rapid and complete neurological recovery.

Key Words: *Hypertension, Post streptococcal glomerulonephritis, Reversible posterior leukoencephalopathy*

Reversible posterior leucoencephalopathy (RPLS) is a clinico-radiological syndrome, manifesting with headache, confusion, seizures, visual disturbances and radiological findings of bilateral grey and white matter abnormalities suggestive of edema in the posterior regions of cerebral hemispheres(1). Described mainly in adults, it has also been reported in children(2,3,4). We report a rare case of post streptococcal glomerulonephritis manifesting as reversible posterior leucoencephalopathy syndrome.

CASE REPORT

A 12 year female presented to us with complaints of headache, vomiting for two days and loss of vision for 1 day. Headache was mild, bi-frontal, continuous, not interrupting sleep pattern, non-radiating and relieved with vomiting, which was non projectile and non bilious. There was no past or family history of headache. There was no history of fever, altered urine color, decreased urine output, any preceding skin or upper respiratory infection. On examination, patient was conscious, afebrile, with a pulse rate of 120/min regular, respiratory rate 24/min regular, BP 160/130 mmHg and no edema. On CNS examination, fundus was normal and pupils were bilateral central, circular, reacting to light, but perception to light was absent, bilateral deep tendon reflexes were brisk and plantars were extensor. Rest of the systemic examination was normal.

The blood counts, serum electrolytes, and liver function tests were normal. Blood urea was 54 mg/dL, serum creatinine 1.31 mg/dL and serum uric acid 7.04 mg/dL. Urine examination was normal on the first two days of hospitalization: however, on the third day of hospitalization, urine examination showed 6-8 pus cells/ hpf and plenty of RBCs. Urine culture was sterile. Ultrasound examination showed bilateral loss of cortico-medullary differentiation with normal sized kidneys. ASO titer was raised i.e. 800 U/L. Antinuclear antibody and antineutrophil antibody titers were negative. No pathogenic bacteria were grown in throat culture. Complement C 3 level was 9 mg/dL (normal: 90-180 mg/dL).

CECT brain showed bilateral extensive ill defined symmetrical hypodensities involving the white matter of high parietal and occipital lobes.

(***Fig.* 1**) Patient was started on furosemide and sodium nitroprusside, the blood pressure was gradually reduced. She regained vision on third day of hospitalization and by the seventh day of illness, her abnormal neurological findings and vision was normal. She was discharged on nifedipine. On follow-up after 2 weeks, serum complement (C3) level, electroencephalogram, ultrasound and CT head were normal.

DISCUSSION

Reversible posterior leucoencephalopathy syndrome in children has been shown to be associated with ganglioneuroma, Henoch-Schönlein purpura, acute lymphoblastic leukemia, steroids, hemolytic uremic syndrome, Addison's disease, hypertension, intrabdominal neurogenic tumors, porphyria and bone marrow transplant(2-6). In our patient, headache was the main presenting complaint, followed by nausea, vomiting, and visual disturbance in the form of loss of vision. An almost similar clinical picture was reported earlier(1,3). Other reported clinical symptoms and signs include altered alertness and behavior, seizures, altered speech and visual perception like blurred vision, hemianopsia, and visual neglect(5). In patients of RPLS, seizures (frequently new onset, secondary generalized occipital lobe seizure) almost always occurs during the course of illness(7). Our case did not present with disturbance in consciousness or seizures. Prolonged seizures, hypertension or both may result in permanent neurological deficits and cerebral infarctions, if not treated properly(5). A few of them may not recover completely and develop neurodevelopmental sequelae(2).

The diagnosis of RPLS was established due to the presence of hypertension, typical bilateral CT findings, and reversibility of the lesions. The most common abnormality on neuroimaging is edema involving the white matter in the posterior portions of cerebral hemispheres, especially in the parieto-occipital regions(3). Our case also had the same findings. Kwon, *et al*.(2), in their study of 12 children with RPLS, found that cortical grey matter was predominantly involved in four patients and in two patients, only grey matter was involved. Since it involves both grey and white matter, some authors have suggested that it should be renamed as occipital parietal encephalopathy syndrome(8). Involvement of brain stem, cerebellum, basal ganglion, and frontal lobes has also been reported(3,9). The calcarine and paramedian occipital lobe structures are usually spared and that distinguishes RPLS from bilateral infarction of the posterior cerebral artery territory(5). An important characteristics of RPLS is reversibility of the imaging abnormalities, as seen in our case.

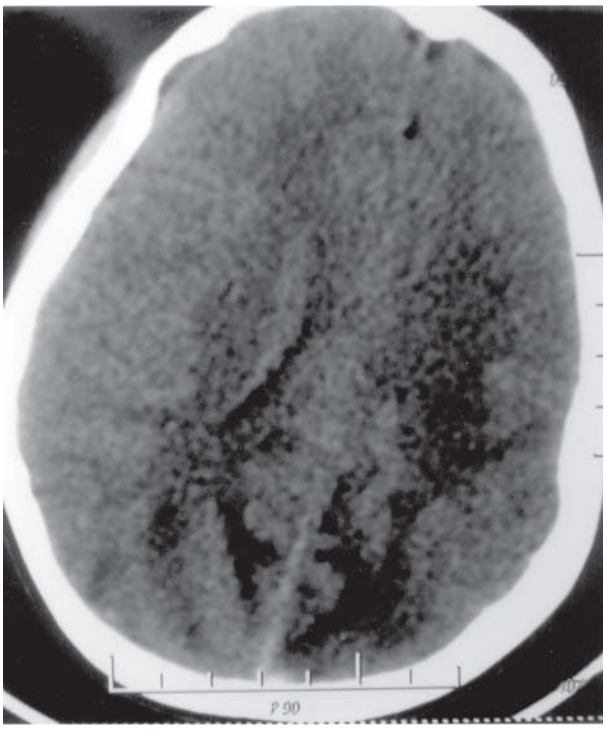

FIG. 1 *Symmetrical hypodensities involving the posterior regions of cerebral hemispheres.*

The pathophysiology of RPLS is complex. It is probably a brain capillary leak syndrome related to hypertension, fluid retention and possibly the cytotoxic effects of immunosuppressive agents on the vascular endothelium(5). The relative paucity of sympathetic innervation in the posterior brain leads to susceptibility to hyperperfusion and vasogenic edema during acute blood pressure elevations which may explain the presence of majority of the lesions in the vascular territory of posterior circulation(6). Children develop RPLS at a lower absolute pressures than adults owing to the relative "left shift" of their range of cerebral blood flow autoregulation(6)

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REFERENCES

1. Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A, *et al*. A reversible posterior leucoencephalopathy syndrome. N Engl J Med 1996; 334: 494-500.
2. Kwon S, Koo J, Lee S. Clinical spectrum of reversible posterior leucoencephalopathy syndrome. Pediatr Neurol 2001; 24: 361-364.
3. Prasad N, Gulati S, Gupta RK, Sharma K, Gulati K, Sharma RK, *et al*. Spectrum of radiological changes in hypertensive children with reversible posterior leukoencephalopathy. Br J Radiol 2007; 80: 422-429.
4. Singhi P, Subramanian C, Jain V, Singhi S, Ray M. Reversible brain lesions in childhood hypertension. Acta Paediatr 2002; 91: 1005-1007.
5. Özcakar ZB, Ekim M, Fitoz S, Teber S, Hizel S, Acar B, *et al*. Hypertension induced reversible posterior leukoencephalopathy syndrome: a report of two cases. Eur J Pediatr 2004; 163: 728-730.
6. Jones BV, Egelhoff JC, Patterson RJ. Hypertensive encephalopathy in children. Am J Neuroradiol 1997;18: 101-106.
7. Garg RK. Posterior leucoencephalopathy syndrome. Postgrad Med J 2001; 77: 24-28.
8. Pavlakis SG, Frank Y, Kalina P, Chandra M, Lu D. Occipital- parietal encephalopathy: A new name for an old syndrome. Pediatr Neurol 1997; 16: 145-148.
9. Schwartz RB, Jones KM, Kalina P, Bajakian RL, Mantello MT, Garada B, *et al*. Hypertensive encephalopathy: findings on CT, MR imaging, SPECT imaging in 14 cases. Am J Roentgenol 1992; 159: 379-383.