

## CORTICAL BLINDNESS IN A BOY WITH ACUTE GLOMERULONEPHRITIS

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**Abstract:** Post-infectious acute glomerulonephritis with hypertensive encephalopathy is characterized by episodic hypertension with headache, vomiting, and hematuria. The association between hypertensive encephalopathy and cortical blindness in children with acute glomerulonephritis is extremely rare. We report the case of a 10-year-old boy with acute glomerulonephritis who presented with gross hematuria, headache, vomiting, and oliguria, and developed transient cortical blindness as a complication of hypertensive encephalopathy. No occurrence of seizure was observed during the clinical course. T2-weighted cranial magnetic resonance imaging showed a high-intensity signal over cortical and subcortical areas of bilateral occipital regions. His vision recovered fully, 2 days after receiving antihypertensive therapy. The patient was well without complaint at 1-year follow-up. This case highlights the possibility that cortical blindness may develop as a complication of acute glomerulonephritis in children. Prevention of the occurrence of neurological deficits in children with acute glomerulonephritis and hypertensive encephalopathy requires careful evaluation and appropriate management of hypertension.

**Key words:** Glomerulonephritis; Infection; Blindness, Cortical; Hypertensive encephalopathy; Magnetic resonance imaging

*J Formos Med Assoc* 2003;102:52-4

Cortical blindness is defined as a loss of vision in the presence of normal papillary reflexes without significant ophthalmological disease. The development of acute cortical blindness in children may follow cardiac arrest, hypoxia, status epilepticus, head trauma, infection of the central nervous system, cerebral infarction, carbon monoxide poisoning, and chemotherapy.<sup>1,2</sup> Although post-infectious acute glomerulonephritis (AGN) is frequently seen in children and adolescents, and is usually associated with a favorable prognosis, cortical blindness as a complication of AGN in children is extremely rare. Our review of the literature found that only 4 cases of pediatric AGN with the complication of cortical blindness have been reported.<sup>1,3-5</sup> All of them presented with hypertensive encephalopathy and transient cortical blindness. Here we report a case of pediatric AGN with the complication of cortical blindness and recovery of sight after treatment with antihypertensive therapy.

### Case Report

A 10-year-old boy was referred to our hospital because of persistent puffy eyes and gross hematuria. He had been well and active until one week earlier, when an upper respiratory tract infection with symptoms of cough, rhinorrhea and sore throat developed, followed 7 days later by puffy eyes, gross hematuria and intermittent headache. He received supportive treatment at a local hospital. Because symptoms persisted, he was referred to our hospital for further management. He had no previous history of renal disease, hypertension, or diabetes mellitus.

On the day of admission, he had gross hematuria, headache, vomiting, and oliguria. On physical examination, he was acutely ill and lethargic. Body temperature was 37.2°C, pulse rate 120/min, respiratory rate 26/min, and blood pressure 142/92 mm Hg (compared with a normal level of < 115/75 mm Hg

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Received: 6 June 2002      Revised: 9 August 2002      Accepted: 5 November 2002

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for his age). Body weight was 39 kg (3 kg above the dry weight). The eyelids were puffy and the neck was supple without meningeal signs. There was no rash or lymphadenopathy. There was no pitting edema of the lower extremities. Coarse rales were heard scattered over both lung fields but there was no heart murmur. The abdomen was distended.

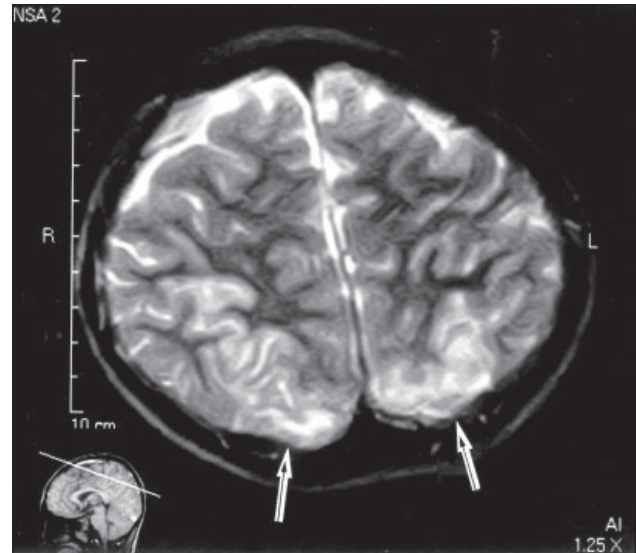
Hemoglobin was 98 g/L, hematocrit 29.8% and platelet count  $271 \times 10^9/L$ . White cell count was  $9.2 \times 10^9/L$  with 70% segmented neutrophils, 7% monocytes and 22% lymphocytes. The urine sample was yellow-colored and turbid in appearance. Microscopic examination revealed numerous red cells with 15–18 white cells per high-power field. No red blood cell casts were seen. Dipstick testing revealed 3+ protein and 3+ occult blood. Electrolytes were within normal limits. Serum creatinine was  $53.0 \mu\text{mol/L}$ , blood urea nitrogen (BUN)  $5.4 \text{ mmol/L}$ , triglycerides  $1.36 \text{ g/L}$ , cholesterol  $4.87 \text{ mmol/L}$ , and serum albumin  $31 \text{ g/L}$ .

Testing for antinuclear antibody was negative. Antistreptolysin O titer was 125 Todd IU. Measurement of serum immunoglobulin levels revealed an elevated immunoglobulin (IgG) level of  $20.6 \text{ g/L}$ , a normal IgA level of  $2,340 \text{ mg/L}$  and a normal IgM level of  $1,980 \text{ mg/L}$ . Serum complement revealed a depressed C3 of  $80 \text{ mg/L}$ , and a normal C4 of  $237 \text{ mg/L}$ . Twenty-four hour urine protein was  $1.4 \text{ g}$ . Renal ultrasound demonstrated swollen kidneys with increased echogenicity of the cortex. Chest x-ray showed bilateral hyperinfiltration of the perihilar area of the lung.

After admission, treatment was initiated with intravenous furosemide ( $0.5 \text{ mg/kg}$  8-hourly) and strict fluid restriction. Systolic pressure ranged from 120 to 147 mm Hg and diastolic pressure ranged from 76 to 98 mm Hg during the first 3 days after admission. But sudden onset of blindness developed on awakening during the morning of the 3rd day of admission. His consciousness remained clear and he was alert and oriented. He was able to walk with support, and he had normal spontaneity of speech. Anton's syndrome did not develop and no seizure was noted.

Subsequent neurological examination revealed no other cranial nerve dysfunction except bilateral blindness. The doll-eye's phenomenon was normal. Tendon reflexes in the limbs were slightly increased. Ophthalmological examination revealed no light perception. The pupils were not dilated, and were equal in size with normal direct and indirect light reflex.

Fundoscopic examination demonstrated clear disc margins without swelling or hemorrhage. Bilateral macula were normal, and there was no retinal exudation or neovascularization. Both neurological and ophthalmological consultations concurred with



**Fig.** T2-weighted MRI (spin echo, 4000/120) showing a high-intensity signal in cortical and subcortical areas of bilateral occipital regions (arrows).

the diagnosis of cortical blindness. T2-weighted magnetic resonance imaging (MRI) of the brain showed a high-intensity signal over the cortical and subcortical areas of bilateral occipital regions (Fig.). These changes on MRI were consistent with the clinical presentation of cortical blindness. Electroencephalography showed diffuse suppression of background rhythm and activity without focal lesion or epileptiform discharge. His blood pressure progressively decreased to normal levels on the 4th day after admission. His sight recovered fully, 2 days after the blood pressure had returned to normal. Blood pressure stabilized at 110/70 mm Hg. Percutaneous renal biopsy was performed on the 10th day of admission and histological study showed diffuse proliferative glomerular disease consistent with post-infectious AGN.

He was discharged on the 12th hospital day without any visual impairment or hypertension. At follow-up 3 weeks later, blood pressure was 110/70 mm Hg, and serum C3 level was  $1250 \text{ mg/L}$ . Urine was yellow-clear in appearance with 6 to 10 red cells and 3 to 5 white cells per high-power field. Dipstick protein was negative. The patient was well without complaint at 1-year follow-up.

## Discussion

Hypertensive encephalopathy is a neurological syndrome characterized by rapidly progressive symptoms and signs, including headache, nausea, vomiting, seizure, visual disturbance, altered mental status, and focal neurological signs in conjunction with elevated

blood pressure. The development of hypertensive encephalopathy is usually associated with abrupt elevation of blood pressure. The pathogenesis of hypertensive encephalopathy is not completely understood but has been hypothesized to result from a failure of cerebral autoregulation triggered by abrupt onset of hypertension. According to such a mechanism, interstitial extravasation of fluid and protein would ensue, producing local vasogenic edema in the surrounding brain parenchyma.<sup>6</sup>

Both adult and pediatric patients can develop an acute reversible posterior leukoencephalopathy syndrome presenting with headache, altered mental status, seizures, and visual disturbance.<sup>7,8</sup> The edema mostly involves the white matter in the posterior portions of the cerebral hemispheres, especially in bilateral parietal-occipital regions. The clinical and radiological features of hypertensive encephalopathy are indistinguishable between adults and children. It has been postulated that hypertensive encephalopathy may be the cause of reversible posterior leukoencephalopathy syndrome or be only one part of a greater syndrome.<sup>8</sup>

Cortical blindness as a complication of hypertensive encephalopathy is infrequently seen in children with AGN. Only 4 other pediatric cases have been reported, involving 3 males and 1 female.<sup>1,3-5</sup> The first reported case involved a 9-year-old boy with AGN and transient cortical blindness who initially presented with marked hypertension and severe azotemia.<sup>1</sup> He received peritoneal dialysis and developed grand mal seizures resulting from hypocalcemia. Blindness developed postictally. However, the present case and the other 3 reported cases were not complicated by azotemia and none of them required dialysis. Serum electrolyte levels in these cases were within normal limits. Abrupt onset of blindness within 3 days after admission was observed in all 4 of these cases. Our patient's vision recovered fully 2 days after the onset of cortical blindness. The time to recovery of sight in the other reported cases ranged from 2 to 12 days. No visual sequelae were reported in any of these cases.

Deal et al<sup>9</sup> observed that rapid blood pressure reduction in children with hypertensive emergencies was associated with a higher incidence of permanent neurological sequelae. To prevent the occurrence of permanent neurological deficits in these children with severe hypertension, they recommended that the blood pressure should be reduced in a controlled manner over the first 96 hours of admission. The blood pressure of our patient was monitored closely during his treatment, and sudden drops in blood pressure did not occur. His blood pressure had returned to normal levels by the 4th day after admission. The possibility of a rapid blood pressure

reduction contributing to the development of cortical blindness in our patient could thus be excluded.

Schwartz et al<sup>10</sup> reported the radiological findings of hypertensive encephalopathy on computed tomography (CT), MRI and single-photon emission computed tomography (SPECT) imaging. The characteristic findings of their studies included areas of reversible hypodensity on CT and increased T2-weighted signal on MRI, usually located in the occipital lobes; increased perfusion in these regions was also found by SPECT. Our patient showed similar T2-weighted signal-intense lesions involving bilateral occipital regions on brain MRI. These changes on MRI are likely to reflect the extravasation of fluid and protein into the surrounding brain parenchyma, and resolution of the lesions may reflect their reabsorption.

In summary, the MRI findings indicate that the complication of reversible cortical blindness in this boy with AGN and hypertensive encephalopathy was not caused by ocular lesions. The brain lesions on MRI findings of the patient were correlated with the presentation of cortical blindness. The prevention of neurological sequelae in children with AGN and hypertension requires early diagnosis, careful investigation and appropriate management.

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