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A Rare Case of Hypertensive Chorioretinopathy with Posterior Reversible Encephalopathy Syndrome (PRES) in a Teenage Girl

***Abstract*—**Hypertensive chorioretinopathy is a rare eye manifestation due to acute elevation in blood pressure. In this case, we reported a young hypertensive patient who presented with hypertensive chorioretinopathy and posterior reversible encephalopathy syndrome (PRES). The purpose of this case report is to raise awareness among clinicians to identify, aggressively treat and monitor paediatric patients with hypertension to prevent life and vision-threatening complications. A 14-year-old Malay girl with a history of uncontrolled hypertension and end-stage renal failure presented with the sudden onset of a generalised tonic-clonic seizure preceded by a headache for two days. There was a progressive blurring of vision in both eyes for four months before the presentation. Visual acuity over the right eye was counting fingers (CF) and hand movement (HM) in the left eye. Both fundi revealed features of hypertensive chorioretinopathy with Elschnig spots and Siegrist streaks. Systemic blood pressure during presentation was 235/152 mmHg. A computed tomography (CT) scan of the brain suggestive features of PRES. Six weeks later, with controlled hypertension, her visual acuity improved to 6/60 in both eyes. Hypertensive chorioretinopathy is usually the sign of an acute, dramatic increase in systemic blood pressure in a young person. Achievement of target blood pressure is important to prevent vision-threatening and neurological complication

Keywords*—Hypertension, chorioretinopathy, encephalopathy*

1 INTRODUCTION

Hypertension in young children is rarely seen and usually is attributed to a secondary cause like renal disease or endocrine disorder [1]. Accelerated hypertension may affect the eye in several ways, which include developing hypertensive retinopathy, optic neuropathy and choroidopathy. Foster et al. reported that in mild hypertensive retinopathy, only 3 out of 35 (8.6%) children were diagnosed with hypertension [2]. However, hypertensive choroidopathy is uncommon in paediatric patients and is rarely reported. It usually occurs in association with severe acute hypertension. Hypertensive chorioretinopathy is a term for hypertensive retinopathy seen with a combination of choroidopathy [3].

We are reporting a case of a teenage girl who presented with a hypertensive emergency with hypertensive chorioretinopathy and posterior reversible encephalopathy syndrome (PRES). The purpose of this case report is to raise awareness among clinicians to identify, aggressively treat and monitor paediatric patients with hypertension to prevent life and vision-threatening complications.

2 CASE REPORT

A 14-year-old Malay girl with underlying uncontrolled hypertension and end-stage renal disease secondary to glomerulonephritis on continuous ambulatory peritoneal dialysis (CAPD) presented with a sudden onset of a generalised tonic-clonic seizure while playing. The seizure lasted for about 10 minutes and aborted spontaneously. Upon further history, she complained of headaches two days before the seizure attack with a history of progressive blurring of vision in both eyes for four months. There was a worsening of vision for two days before the presentation. The burring of vision was not associated with eye redness, eye pain, diplopia, floaters, or flashes of light. There was no pre-existing ocular trauma or eye surgery.

Premorbid, the patient had clear vision in both eyes. The patient was diagnosed with hypertension with underlying glomerulonephritis seven months before the presentation. However, she has poor compliance with antihypertensive medications.

On arrival at the emergency department, the patient had another fitting episode. Intravenous valium 5 mg was given immediately, followed by intravenous infusion of phenytoin 1 g to be completed in one hour. The seizure attack was aborted after 3 minutes, and soon after that, the patient regained consciousness. Her vital signs showed elevated blood pressure of 235/152 mmHg and tachycardic with a pulse rate of 113 beats per minute (bpm). She was treated with a stat dose of tab captopril 25 mg to lower the blood pressure. Otherwise, body temperature, respiratory rate and oxygen saturation remained stable. His general physical examination was normal.

Immediate basic blood investigations revealed a normal complete blood count and liver function test. However, the creatinine level was raised to 1215 mg/dl. Her electrocardiogram showed normal sinus rhythm. Neurological and systemic examinations were normal, with no sign of meningeal irritation. A brain computed tomography (CT) scan was done within an hour. It showed ill-defined white matter hypodensity at the left posterior parietal lobe (Fig. 1A) and bilateral occipital lobe (Fig. 1B), suggesting white matter oedema. There was a well-defined hypodensity lesion at the right lentiform nucleus. However, there was no brain mass or signs suggestive of intracranial bleeding. The patient was transferred to the ward after stabilisation of her blood pressure. Subsequently, she was referred to the ophthalmology team for an eye assessment in view of a history of poor vision.

Ophthalmic examination revealed visual acuity of counting fingers in the right eye and hand movement in the left eye. Her pupils were reactive without an afferent pupillary defect. Confrontation visual field testing revealed vague left temporal homonymous hemianopia. Both eyes were orthophoric with normal extraocular motility. A slit lamp examination revealed normal anterior segments with normal intraocular pressure (18 mmHg) in both eyes. Fundus examinations in the right eye showed a feature of chorioretinopathy with optic disc swelling, arteriolar attenuation, extensive hard exudate surrounding the optic disc and macular area with macular oedema, Elschnig spots and Siegrist streaks (Fig. 2A). Examination of the left eye revealed similar retinal findings with slight tortuous and dilated retina vessels, optic disc swelling, extensive hard exudate surrounding the optic disc and macular area with macular oedema, and Siegrist streaks (Fig. 2B).

Optical coherence tomography (OCT) of the macula was attempted to monitor the status of macular oedema (presence of intra-retinal and sub-retinal fluid). However, because of poor vision, it was challenging to obtain a good-quality macular image due to poor signal strength. Fluorescein angiography was planned in this patient to look for delayed choroidal filling, delayed retinal arterial filling with areas of retinal capillary nonperfusion and features of Elschnig spots which will appear as areas of early hyperfluorescence with late subretinal leakage. However, fluorescein angiography was not performed in this case as the parents refused to give consent to perform the procedure. Based on clinical findings and findings from CT scan of the brain, the patient was diagnosed as having a hypertensive emergency with hypertensive chorioretinopathy and posterior reversible encephalopathy syndrome (PRES).

During hospitalisation, the patient continued her previous antihypertensive medication (tablet felodipine 10 mg 12 hourly and tablet telmisartan 80 mg daily). Her blood pressure was reduced, ranging from 135-145 mmHg systolic and 80-90 mmHg diastolic. The patient was counselled on compliance with medications upon discharge. In view of end-stage renal disease secondary to glomerulonephritis, CAPD was continued with the help of family members.

CT scan of the brain was repeated five weeks later. There was patchy resolution of white matter oedema with patchy hypodensity on the left posterior parietal region (Fig. 1C) and patchy hypodensity at the bilateral occipital region (Fig. 1D). Six weeks after discharge, her blood pressure was controlled with the same antihypertensive drugs. There has been no seizure attack since the last admission. Her visual acuity has improved to 6/60 in both eyes. Both fundi showed partial resolution of optic disc swelling. However, there were persistent extensive hard exudates surrounding the optic disc and macular area in both eyes (Fig. 2C and 2D). The vision in both eyes remained the same at the one-year follow-up.

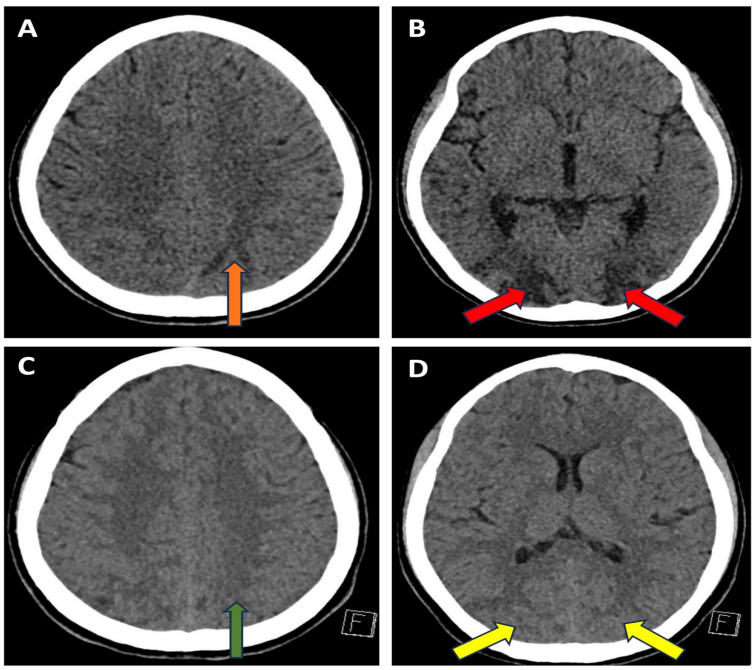

Figure 1. Non-contrast-enhanced computed tomography scan of the brain at presentation (above) and at five weeks follow-up (below): Non-contrasted computed tomography scan of the brain at presentation showed ill-defined white matter hypodensity at the left posterior parietal lobe (orange arrow) (A) and bilateral occipital lobe (red arrow) (B). A repeated computed tomography of the brain at five weeks follow-up showed patchy hypodensity on the left posterior parietal region (green arrow) (C) and hypodensity at the bilateral occipital region (yellow arrow) (D).

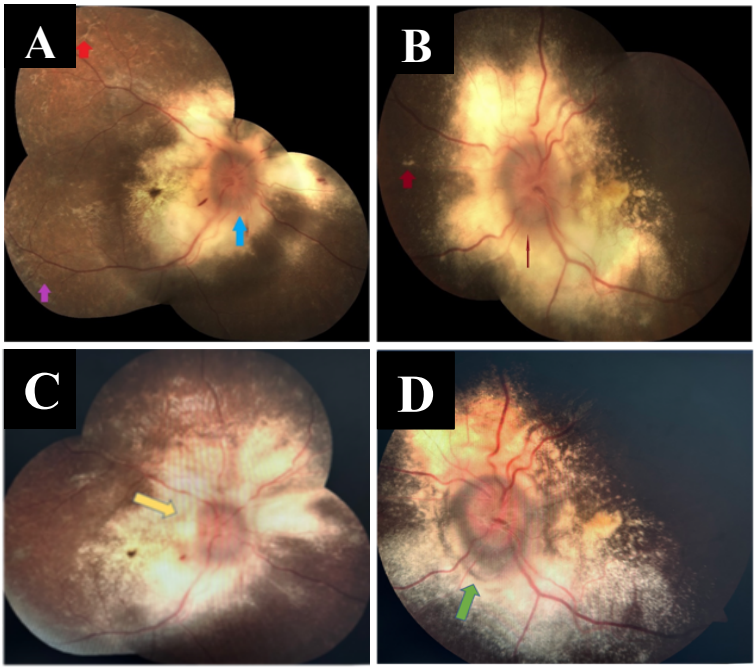

Figure 2. Fundus photography at presentation (above) and at six weeks follow-up (below): Fundus photography at presentation showed features of hypertensive chorioretinopathy with extensive hard exudate surrounding the optic disc and macular area in both eyes. Right eye (A) showed disc swelling (blue arrow), Elschnig's spot (purple arrow) and Siegrist's streak (red arrow). Left eye (B) showed disc swelling (green arrow) and Siegrist's streak (red arrow). Fundus at six weeks follow-up showed persistent extensive hard exudates surrounding the optic disc in the right eye (yellow arrow) (C) and left eye (green arrow) (D).

3 DISCUSSION

The incidence of hypertension in children is between 1-2 % of the population. In Malaysia, the prevalence of hypertension in primary school children was 13.4% [4]. However, the prevalence of malignant hypertension in children is extremely rare. Childhood hypertensive encephalopathy is an uncommon manifestation of hypertension and usually occurs in the presence of other serious medical problems [5].

Hypertension is associated with profound, often asymptomatic, multisystemic effects, and the eye is not spared from the elevated blood pressure. However, the eye is distinctive because it allows the direct sequelae of elevated blood pressure to be visualised early, mainly changes in the retinal microvasculature [6]. Untreated systemic hypertension is associated with retinopathy, choroidopathy and optic neuropathy [5]. Hypertensive choroidopathy has been reported in preeclampsia, renal disease, pheochromocytoma, and malignant hypertension and less observed than hypertensive retinopathy [7].

Choroidal ischemia may arise due to accelerated hypertension and ophthalmic or ciliary artery blockage. Without affecting the retinal arteries, a compromised ciliary artery can result in choroidal ischemia, producing hypertensive choroidopathy with or without optic neuropathy. However, complete ophthalmic artery occlusion compromises the retinal and choroidal circulations [7]. Choroidal involvement in malignant hypertension usually leads to a more guarded visual prognosis than only retinal involvement. The retinal pigment epithelium becomes necrotic; this may result in areas of serous retinal detachment. Two manifestations of hypertensive choroidopathy are Elschnig spots and Siegrist streaks [8]. Elschnig spots are the changes in the retinal pigment epithelium from nonperfused areas of the choriocapillaris [9]. Their appearance is pale or yellow, and they have defined margins. Once a scar develops, a central pigment is seen within the atrophic region. Siegrist streaks are linear hyperpigmented streaks over choroidal arteries, and fluorescein angiography confirms ophthalmoscopic suspicion of hypertensive choroidopathy [7]. With immediate antihypertensive therapy initiation, typical retinal abnormalities may disappear within two to three months with prompt initiation of antihypertensive treatment [10]. However, not all retinopathies are completely reversible and visual impairments in malignant hypertension can occur due to optic nerve infarction from rapid blood pressure reduction [11]. Although cotton-wool spots resolve within one month of blood pressure control, they are clinically significant as they represent permanent nerve fibre layer defects. Intraretinal hyperreflective dots have persisted for more than six months in some patients. Poor visual recovery on follow-up, despite the resolution of intra-retinal or subretinal fluid, has been attributed to photoreceptor defects which is better appreciated in OCT as focal loss of interdigitation zone and ellipsoid zone [10,12]. Our patient had only a slight improvement in visual acuity after six weeks. However, fundi findings remained almost the same despite well-controlled blood pressure.

PRES is a potentially reversible neurotoxic state. According to the widely accepted vasogenic theory, cerebral autoregulation failure caused by increased cerebral blood failure results in hyperperfusion, increased capillary hydrostatic pressure, vasodilation, and vasogenic oedema [13]. Most affected patients are in their fourth or fifth decade of life, and it rarely occurs in children [9]. PRES aetiologies include hypertensive emergency, cytotoxic medications, preeclampsia or eclampsia, and autoimmune and systemic conditions, including sepsis [14]. Gupta et al. found that renal disease could be the most common cause of PRES in paediatric patients [15].

PRES is a neuro-clinical condition that has distinct imaging manifestations. Headache, seizures, encephalopathy, visual abnormalities, and focal neurologic impairments are some of the clinical signs and symptoms in PRES [16]. Children with PRES exhibit a variety of non-specific symptoms [17]. In PRES, CT brain may be normal (22%) or indicate vague findings (33%). However, in up to 45% of cases, bilateral parenchymal hypodensities indicative of vasogenic oedema secondary to PRES may be seen [18]. Classic imaging patterns usually reveal vasogenic oedema involving the parieto-occipital regions, which are bilateral, subcortical, and symmetrical in appearance [19]. CT scan examination is usually the initial imaging test for acute neurological symptoms and may demonstrate white matter hypoattenuation in the affected region [17]. Paediatric patients had more abnormal magnetic resonance imaging (MRI) findings at diagnosis. In the acute periods of

PRES, vasogenic oedema usually resolves [20]. As a result of the hypertensive emergency, our patient also has PRES in addition to hypertensive chorioretinopathy.

According to Malaysia Clinical Practise Guidelines (CPG), the goal of therapy in children and adolescents with hypertension is to keep the blood pressure within their range. Once pharmacologic therapy is initiated in children, the aim is to achieve the goal of treatment, which is to reduce blood pressure to <90th percentile (systolic and diastolic) and <130/80 mmHg in adolescents ≥13 years old. For hypertensive children with proteinuric chronic kidney disease (CKD), the target blood pressure is <50th percentile. Medication consists of angiotensin-converting enzymes or angiotensin receptor blockers are preferred in children with proteinuric CKD. Clinicians should provide diet advice and recommend moderate to vigorous physical activity to help reduce blood pressure when diagnosing hypertension in children [21]. In our case, the patient was treated with felodipine and telmisartan before the episode of hypertensive emergency. However, compliance and logistic issues may play a role of visual and neurological complications related to hypertensive emergency.

4 CONCLUSION

Hypertensive choroidopathy in children is uncommon and rarely occurs in the setting of a hypertensive emergency. It is usually the sign of an acute, dramatic increase in systemic blood pressure in a young person. Hypertensive emergency requires multi-disciplines involvement. The participation of paediatricians, nephrologists, ophthalmologists, and dieticians play crucial roles in managing children with hypertension. Preventive measures must be taken to prevent not only visual-threatening complications but also neurological damage.

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