Reversible Hypertensive Retinopathy in a Child with Bilateral Pheochromocytoma after Tumor Resection

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Abstract: Pheochromocytoma is very rare in children. We report a case of bilateral pheochromocytoma in a 12-year-old boy who had blurred vision due to hypertensive retinopathy. Abdominal ultrasound and computed tomography revealed bilateral suprarenal tumors. Resection of the bilateral tumors along with right total and left subtotal adrenalectomy were performed. Blood pressure and visual acuity returned to normal after surgery.

Key words: pheochromocytoma, hypertensive retinopathy, child

Pheochromocytoma, a rare tumor arising from the chromaffin tissue, can produce and secrete catecholamines: epinephrine, norepinephrine, and dopamine. Most pheochromocytomas arise from the adrenal medulla. However, pheochromocytoma can arise from a nest of chromaffin tissue outside the adrenal medulla, such as in the bladder, or along the abdominal and thoracic sympathetic chain [1]. The incidence of pheochromocytoma was reported at 0.4 to 2.06 per million per year [2, 3]. Most cases are unilateral and found in adults; less than 5% of cases occur in children [4].

Pheochromocytoma may present as a component of familial multiple endocrine neoplasia type II. This autosomal dominantly inherited syndrome also carries the risk of developing parathyroid hyperplasia and medullary thyroid carcinoma. Sporadic pheochromocytoma is rare in children [5]. Herein, we present an unusual case of bilateral pheochromocytoma complicated by hypertensive retinopathy in a 12-year-old boy. His visual acuity dramatically improved after resection of the tumors.

Case Report

A 12-year-old boy visited our outpatient clinic because of a 5-day history of blurred vision. Headache, sweating, weight loss, and general malaise were also noted. His medical history was unremarkable. His family history was negative for hypertension. Hypertensive retinopathy was suspected at our clinic and he was admitted to our hospital for further investigation.

On examination, the patient was thin (body weight 38 kg; 25th percentile) and had mild respiratory distress. Visual acuity was 0.05 in both eyes. Slit lamp examination results and intraocular pressure were normal. Ophthalmoscopy showed hyperemia of the optic disc, macular star around the fovea, intra-retinal hemorrhage, and cotton-wool spots at the posterior pole and the mid-peripheral retina in both eyes (Fig. 1A). The heart rate was 145 beats/minute, and the respiration rate was 24/minute. Wide fluctuations in blood pressure were noted, varying from maximal systolic and diastolic pressures of 249 mm Hg and 175 mm Hg to minimal systolic and diastolic pressures of 131 mm Hg and 70 mm Hg, respectively. Bounding pulses were found in all limbs. Auscultation revealed no gallop rhythm, abnormal heart sounds, or murmurs. There was no palpable mass in the abdomen. The rest of the examination was unremarkable. The urine analysis results, complete cell count, and blood urea nitrogen, creatinine, and electrolyte levels were within normal limits. Urine culture showed no growth. The urinary vanillymandelic acid (VMA) level was 37.9 mg/24 hr (normal, 2–7 mg/24 hr). The plasma renin activity was 181 mg/mL (normal 7–76 mg/mL).

Chest roentgenography and electrocardiography revealed no abnormalities. Two-dimensional echocardiography revealed hypertrophy of the interventricular septum and left ventricular posterior wall. Ultrasonography and computed

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Computed tomography of the abdomen demonstrated bilateral noncalcified, well-defined adrenal tumors, 7 x 5 cm on the right, 5 x 4 cm on the left (Fig. 2). Bilateral adrenal pheochromocytoma complicated by hypertensive retinopathy was diagnosed on the basis of these findings.

The patient was treated with phenoxybenzamine, an alpha-adrenergic blocking agent. The dosage of phenoxybenzamine was 20 mg/day initially and increased to 30 mg/day after 1 week of therapy, to control the systolic blood pressure within the range 130 mm Hg to 150 mm Hg. Surgery was arranged after hypertension was brought under control. Tumor resection, total right adrenalectomy, and subtotal adrenalectomy of the left adrenal gland with some cortical sparing were performed. The operative course was uneventful except for transient hypertension during manipulation of the tumors. The final pathologic diagnosis confirmed adrenal pheochromocytoma.

Five days after surgery, the patient’s urinary VMA concentration and blood pressure returned to normal, and no cortical deficiency was noted. One month after surgery, his vision improved to 1.0 in both eyes; the appearance of the optic discs was normal on ophthalmoscopy 5 months postoperatively (Fig. 1B). He remained normotensive, with normal visual acuity and no recurrence of symptoms at 6 months’ follow-up.

Discussion

The classic triad of headache, palpitation, and sweating is reported in 89% of cases of pheochromocytoma, and more than 90% of patients with pheochromocytoma have hypertension [6]. Hypertension is the most prominent feature of pheochromocytoma, and may be paroxysmal or persistent; it is caused by secretion of one or more of the catecholamine hormones. In addition, headache, tachycardia, diaphoresis, postural hypotension, nausea, vomiting, abdominal or chest pain, weight loss, shortness of breath, polydipsia, polyuria, acrocyanosis, convulsions, and visual disturbances are common [5]. However, because the symptoms and signs of pheochromocytoma are not specific in children, early diagnosis is difficult. Therefore, clinical alertness to hypertension is important in identifying pheochromocytoma.

Pheochromocytoma is very rare, but it is an important cause of severe hypertension in children. Evaluation of a child for suspected pheochromocytoma should include determination of 24-hour norepinephrine, epinephrine, total metanephrines, and VMA concentrations in the urine. In more than 95% of patients, the
diagnosis can be established by an increased urinary concentration of VMA [7]. Plasma catecholamine determination is generally less useful than urinary assessment, except in hypertension crisis.

Once the diagnosis is made, surgical removal is the treatment of choice. Surgical complications may occur owing to excessive and abrupt release of catecholamines [7]. Good control of hypertension is important prior to surgery and could effectively reduce morbidity and mortality. Phenoxybenzamine, a non-competitive alpha-adrenoreceptor blocker, is useful in controlling hypertension in patients with pheochromocytomas [5]. It can reduce the hypertensive crisis during induction, intubation, or surgical manipulation of the tumor. However, patients who have adequate phenoxybenzamine are prone to orthostatic hypotension [8]. Therefore, this preoperative treatment should be given in the hospital and patients should be instructed on the need to maintain an upright position and should be assisted to maintain this position to prevent complications.

Adrenalectomy carries a mortality of about 2% to 4%, but the morbidity may be as high as 40% [5]. Potential morbidity includes intraoperative injury to an adjacent structure, postoperative infection, thromboembolism, and adrenal insufficiency [5]. No morbidity was encountered in the present case. The cortical-sparing left adrenalectomy was effective in preserving mineralocorticoid activity.

The long-term prognosis for most patients with pheochromocytoma after unilateral adrenalectomy is excellent, but some patients may require corticosteroid therapy following bilateral adrenalectomy [9]. In our patient, cortical-sparing adrenalectomy was successfully performed; thus, chronic hormone replacement and the risk of Addisonian crisis were avoided.

Hypertensive retinopathy may occur in chronic essential hypertension, toxemia of pregnancy, or malignant hypertension. Hypertensive retinopathy is uncommon in children. The disease is not a benign condition in children, and delay in diagnosis may result in permanent visual damage.

Histologically, the ocular manifestations of systemic hypertension are divided into three distinct entities: retinopathy, choroidopathy, and neuropathy [10]. Retinopathy is more common than choroidopathy and neuropathy in patients with hypertension. The fundus picture shows generalized or focal retinal arteriolar constriction in the early stages. As the disease progresses, superficial flame-shaped hemorrhages and small white or gray superficial foci of retinal ischemia (cotton-wool spots) develop. In severe hypertension, the optic disc may become congested and edematous. If elevated blood pressure is controlled promptly with medication or surgery, the retinal blood vessels may return to a normal state with no permanent pathologic changes [11, 12]. In our patient, though ophthalmoscopy of the fundus revealed intraretinal hemorrhage and cotton-wool spots not in the early stage, his visual acuity dramatically returned to normal after resection of the tumors. This demonstrates that hypertensive retinopathy in children may be reversible if early correction is possible.

Although rare, hypertensive retinopathy may be induced by pheochromocytoma in children. In the present case, preoperative stabilization of blood pressure and cortical-sparing adrenalectomy resulted in uneventful recovery.

References