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Bilateral Hypertensive Retinopathy: A Rare Presentation of Pheochromocytoma

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Abstract

Pheochromocytomas are rare tumors of childhood usually presenting as the clinical triad of headaches, tachycardia, and sweating. It can also present with unusual findings like hypertensive retinopathy. We present a case of pheochromocytoma in an adolescent boy, who presented with diminution of vision due to hypertensive retinopathy as shown by fundus examination.

Keywords: Hypertensive Retinopathy; Pheochromocytoma.

Sir,

A 14 year old boy came to us with complain of diminution of vision since 5 days. He had visual acuity of 6/60 in both eyes. Slit lamp examination and intraocular pressure were normal. Fundus examination showed disc edema with cotton-wool hemorrhages at the retina along with macular edema in each eye [Figure 1]. On examination, heart rate was 120 /minute and a functional systolic murmur over left middle sternal border was appreciated. BP was elevated with wide fluctuations: maximal of 210/150 mmHg and minimal of 140/100 mmHg. There was a palpable abdominal mass in right upper abdomen. Routine urine analysis was normal. Complete cell count, blood urea nitrogen, creatinine, and electrolyte levels of blood were within normal limits. Ultrasound revealed a right suprarenal mass with compression of right kidney. Computed tomography of the abdomen demonstrated a right retroperitoneal, suprarenal tumor of 7.1 ×5.4 cm in size with tortuous veins and displacement of right renal artery and inferior vena cava. Urinary vanillyl mandelic acid level was highly raised [46.5 mg/24 hr (normal; 2-7 mg/24 hr)] and plasma renin activity was 9.02 ng/mL/hr during supine rest (normal; 0.2 to 3.3 ng/ml/hr). Two-dimensional echocardiography revealed left ventricle hypertrophy suggestive of long standing hypertension. Based on the bio-chemical and CT findings a diagnosis of phaeochromocytoma was made. The patient was put on a combination of phenoxybenzamine and a calcium channel blocker; and was advised surgery. Unfortunately the child's parents did not come for follow up.

Pheochromocytomas are rare tumors in children with a prevalence of 0.3-0.95% characterized by episodic headaches, tachycardia, and sweating; but retinopathy is rarely the presenting feature. Earlier case reports showed that patients presented after 7-30 days of experiencing blindness [1-4]. One child was diagnosed after a failed vision screening at school, the care givers did not even go to the clinician [1]. Such reports are rare and have increased in last two years which may be due to better work up. Other diseases which presented with hypertensive retinopathy are advanced renal disease, bilateral renal artery stenosis and systemic lupus nephritis [4, 5]. In

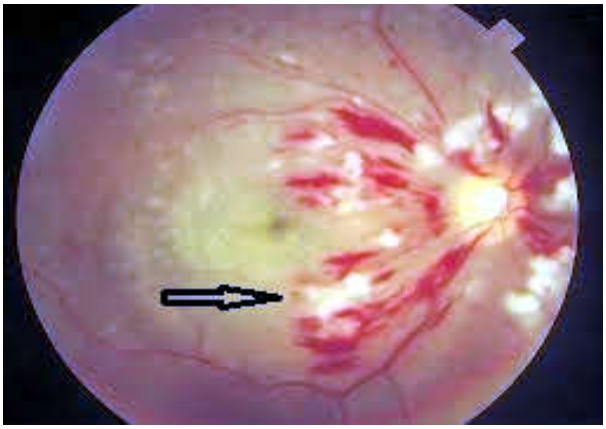

Fig. 1: Arrow showing cotton wool hemorrhages

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present case, histology is not available but biochemical diagnosis is enough for diagnosis [6]. Early identification can significantly reduce morbidity and mortality so a rapid work-up should be done when hypertensive retinopathy is encountered. Further, public awareness programs should be undertaken so that the caregivers should know the importance of early diagnosis and seek the clinical advice without delay.

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