IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 19, Issue 11 Ser.6 (November. 2020), PP 47-50
www.iosrjournals.org

Malignant Hypertensive Retinopathy in a Child as Initial Presentation of Pheochromocytoma

Wafae JOUIDI, Kaoutar NAYA, Hélène NyinkoAboughé, Elhassan ABDELLAH, Amina BERRAHO
Ophthalmology B department CHU Avicenne of Rabat, Morocco. Mohammed V university of Rabat

Abstract
We report a case of a 14-year-old boy who presented with blurry vision in both eyes and headaches of 2 weeks' duration. He was not on medication and denied any health problems. Fundus examination showed bilateral but markedly asymmetric macular exudates and optic disk edema. The blood pressure measurements was markedly elevated.Computed tomography revealed unilateralleftsuprarenal tumorand led with laboratory studies to a diagnosis of pheochromocytoma, a rare catecholamine-secreting tumor. Resection of the tumor along with left total adrenalectomy were performed. Blood pressure and visual acuity returned to normal after surgery.
Key words: *hypertensive retinopathy,Pheochromocytoma, , child, bilateral.*

Date of Submission: 06-11-2020 Date of Acceptance: 19-11-2020

I. Introduction:

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 but also 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]. We reporta case of bilateral pheochromocytoma complicated by malignant hypertensive retinopathy in a 14-year-old boy. We noted spectacular improvement in his visual acuity after tumor resection.

II. Case Report:

A 14-year-old boy visited our ophthalmic emergency department because of a 3-day history of blurred vision and headache. General malaise, sweating and frequency in urination were noted. His medical history was unremarkable. His family history was negative for hypertension. Hypertensive retinopathy was suspected and he was admitted to our hospital for further investigation. On examination, the patient had mild respiratory distress. On ophthalmological evaluation, visual acuity was counting fingers at 5 feet in the right eye and 0.4 in the left eye. he had no afferent pupillary defect, and anterior segment examination was normal. Dilated fundus examination revealed 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 but more marked on the right eye (Fig. 1).

Tests for Bartonella henselae, Bartonella Quintana, Lyme disease, toxoplasmosis, toxocariasis, syphilis, and tuberculosis were negative. The findings of a chest radiograph were normal. Given a concern for hypertensive retinopathy, a pediatrician examined him: the heart rate was 137 beats/minute, and the respiration rate was 23/minute. Wide fluctuations in blood pressure were noted, varying from maximal systolic and diastolic pressures of 220 mmHg and 140 mmHg to minimal systolic and diastolic pressures of 125 mmHg and 70 mmHg. Auscultation were normal and there was no palpable mass in the abdomen. Laboratory testing confirmed the diagnosis, with markedly elevated levels of catecholamines (dopamine 77 pg/mL; normal range, 0–20 pg/mL) ; The urinary vanillymandelic acid (VMA) level was 29.7mg/24 hr (normal range, 2–7 mg/24 hr).

Computed tomography of the abdomen demonstrated unilateral left noncalcified, well-defined lobulated adrenal tumor, 6 x4,5 cm (Fig. 2). Bilateral adrenal pheochromocytoma complicated by hypertensive retinopathy was diagnosed based on these findings.

The patient underwent an exploratory laparotomy with resection of a capsulated 6 *4,5 * 3.5 cm left adrenal mass with focal necrosis. Tumor pathology confirmed the diagnosis of pheochromocytoma.

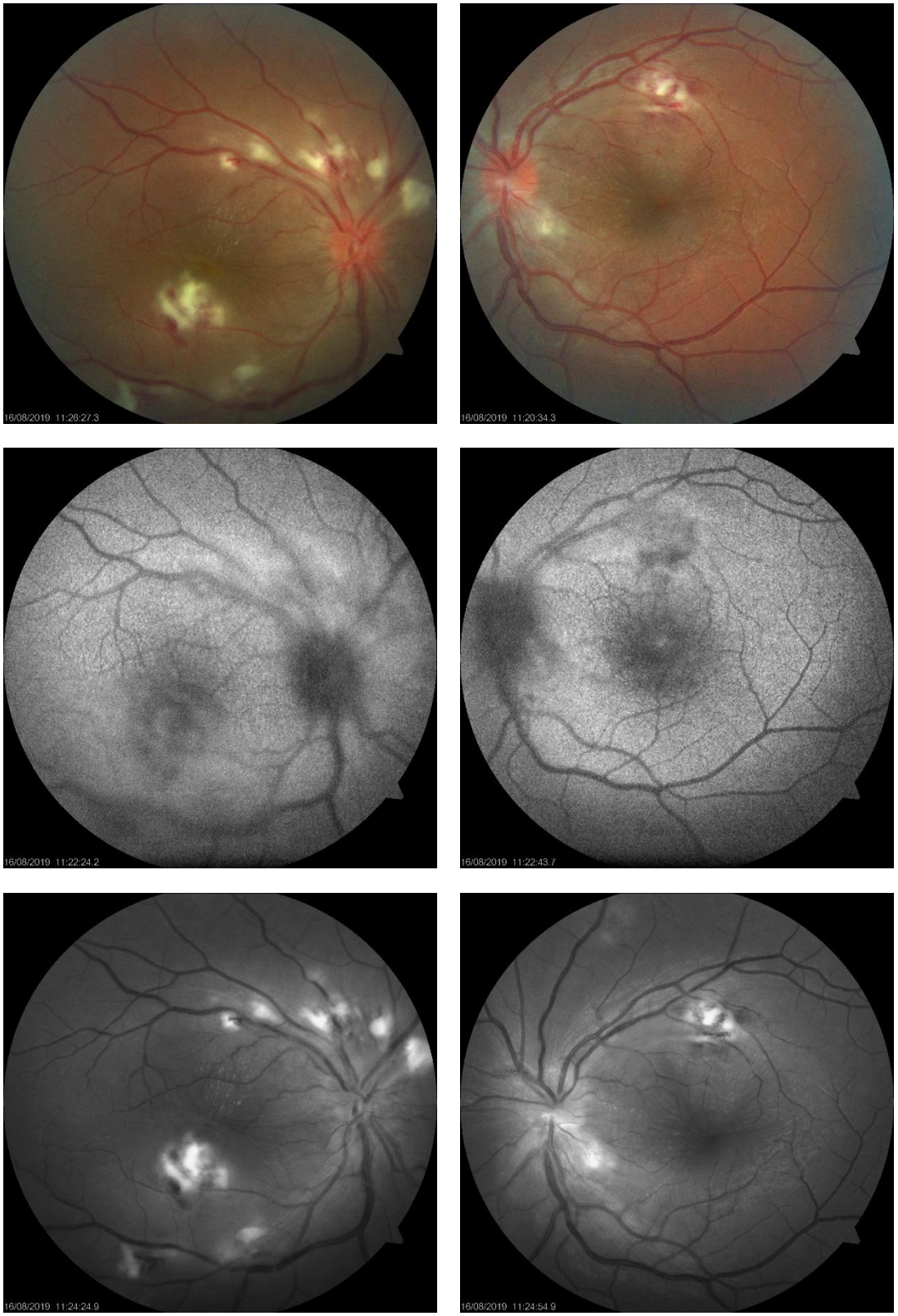

Figure1: Fundus, Autofluorescence and Red-free photographs of the right (A; C ; E) and left (B; D ; F) eye. Macular edema in both eyes, more marked in the right eye in the form of macular star. Vascular tortuosity, narrowing of the arteries and retinal edema, especially the right eye retina. Flame-shaped hemorrhages, cotton wool spots and optic disc neuropathy. There is blurring of the optic disk margin, along with cotton wool spots, retinal hemorrhages, and retinal hard exudates

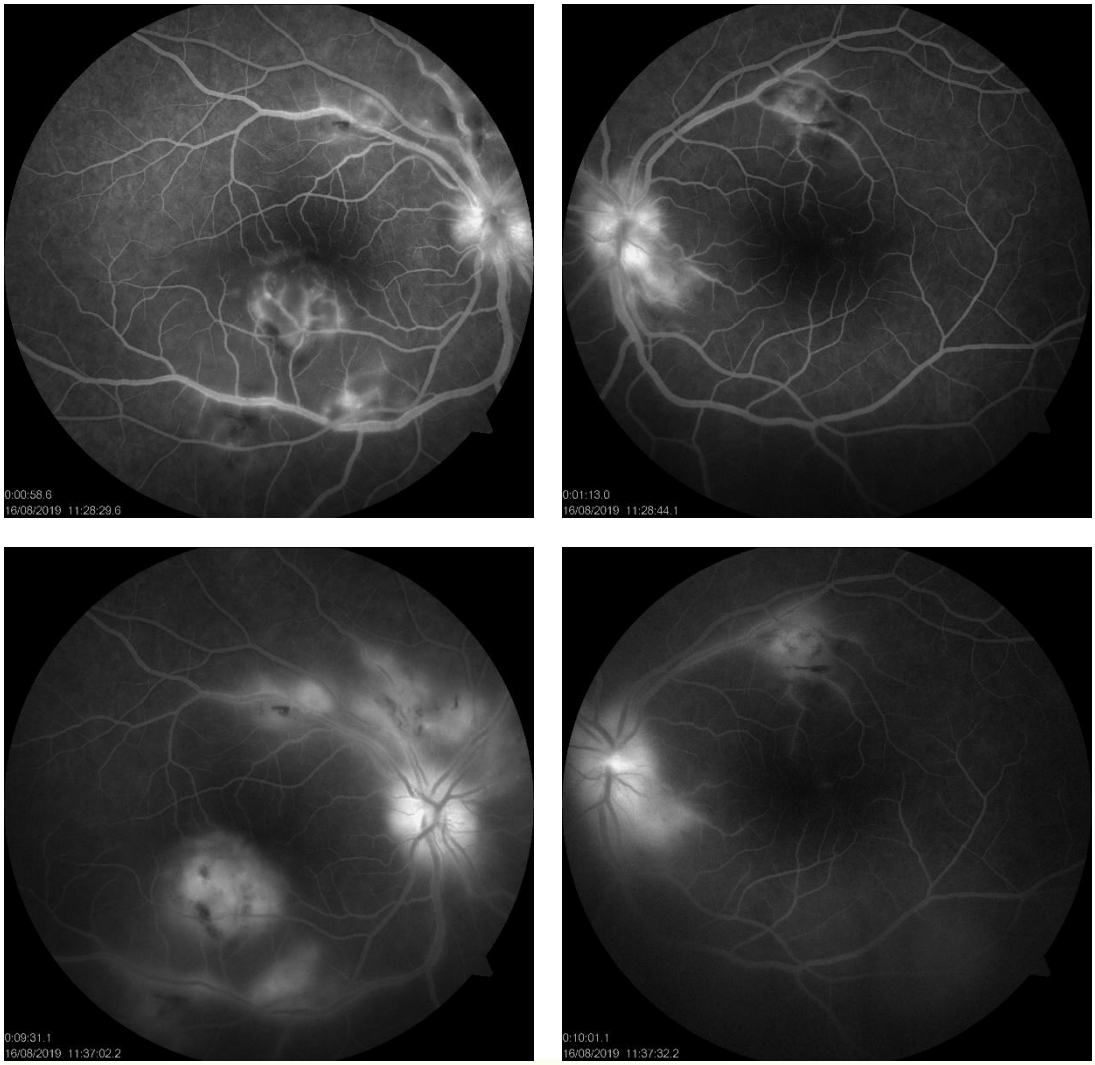

Figure 2: Fluorescein angiography right eye (**A**;**C**) and left eye (**B**; **D**). Delayed filling of the superior and inferior temporal artery corresponding to the edema in the superior retina. Presence of areas of hypofluorescence caused by retinal hemorrhages.Progressively intense hyperfluorescencearound the optic disc and adjacent areas.

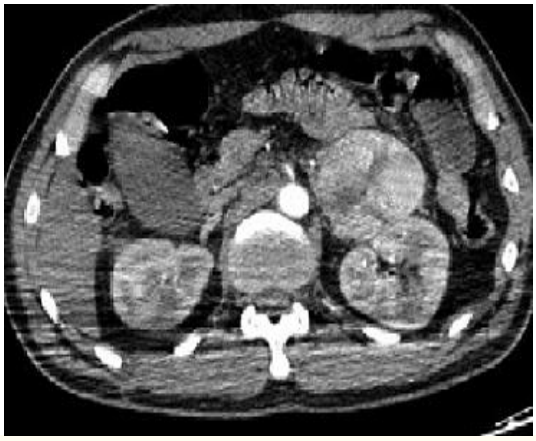

Figure 3: Computed tomography scan of the abdomen of the patient demonstrates a left well-defined adrenal tumeur

III. Discussion:

In cases with hypertensive retinopathy, besides the retina, both the choroid and the optic nerve may be involved [5, 6, 7]. In fact, malignant hypertension is a rare cause for presentation to a pediatric ophthalmologist. Systemic hypertension can cause optic disk edema and macular exudates, but these fundus findings have a broad

differential diagnosis that also includes papilledema, compressive or ischemic optic nerv disease, as well as neuroretinitis from infectious causes such as Lyme disease, cat scratch disease, syphilis, tuberculosis, and toxocariasis[8]. However, because the symptoms and signs of pheochromocytoma are not specific in children, early diagnosis is difficult. Hypertension is rare in children and should always prompt a thorough evaluation for secondary causes [9]. In one series, 98% of patients less than 14 years old had an identifiable cause for hypertension [10]. Visual prognosis is generally good. Vision loss may result from prolonged papilledema or retinal pigment changes due to serous retinal detachment [11]. Pheochromocytoma, athough a rare tumor is one of many causes of secondary hypertension [12, 13]. Pheochromocytomas are rare tumors arising in chromaffin cells of the adrenal medulla or paraganglia. 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 [14]. Additionally, it may coexist with von Hippel–Lindau (VHL) disease and with cutaneous neurofibromatosis [15]. From this brief discussion we can clearly conclude the role of the ophthalmologist in the diagnosis of pheochromocytoma and associated conditions such as MEN and VHL disease. The existence of a very large variety of symptoms and signs encountered in patients with pheochromocytoma gave the title of "Great Masquerader" to this tumor. Whenever the diagnosis is in doubt, biochemical testing can always establish the presence or absence of a pheochromocytoma [13, 16, 17], and localization with computed tomography or magnetic resonance imaging, is possible [16, 15]. In fact, In more than 95% of patients, the diagnosis can be established by an increased urinary concentration of VMA [19].

Once the diagnosis is made, surgical removal is the treatment of choice. Adrenalectomy carries a mortality of about 2% to 4%, but the morbidity may be as high as 40% [18]. No morbidity was encountered in the present case. The long-term prognosis for most patients with pheochromocytoma after unilateral adrenalectomy is excellent [19].

In conclusion, severe hypertensive retinopathy with optic neuropathy may be a consequence of malignant hypertension due to a pheochromocytoma. It is reversible after ablation of the tumor. Early diagnosis is of vital importance and relies on hormonal investigation and immunohistochemistry.

References:

[1]. Sisson JC, Shapiro B, Beierwaltes WH, et al: Locating pheochromocytomas by scintigraphy using 131Imetaiodiobenzylguanidine. CA Cancer J Clin 1984;34:86–92.
[2]. Fernandez-Calvet L, Garcia-Mayor RV: Incidence of pheochromocytoma in South Galicia, Spain. J Intern Med 1994; 236:675–7.
[3]. DeGraeff J, Horak BJ: The incidence of pheochromocytoma in the Netherlands. Acta Med Scand 1964; 176:583–93.
[4]. Abramson SJ: Adrenal neoplasms in children. Radiol Clin North Am 1997;35:1415–53
[5]. Hayreh SS, Servais GE. Retinal hemorrhages in malignant arterial hypertension. Int Ophthalmol 1988; 12: 137–145
[6]. Sathyan P, Myint K, Singh G et al. Acute painful visual loss from accelerated hypertensive choroidopathy. Acta Ophthalmol Scand 2007; 85: 337–338
[7]. Wall M. Optic disk edema with cotton-wool spots. Surv Ophthalmol 1995; 39: 502–508
[8]. Ghauri RR, Lee AG. Optic disk edema with a macular star. Surv Ophthalmol 1998;43:270-74
[9]. Sinaiko AR. Hypertension in children. N Engl J Med 1996;335: 1968-73.
[10]. Wyszynska T, Cichocka E, Wieteska-Klimczak A, Jobs K, Januszewicz P. A single pediatric center experience with 1025 children with hypertension. Acta Paediatr 1992;81:244-6
[11]. Spencer CG, Lip GY, Beevers DG. Recurrent malignant hypertension: a report of two cases and review of the literature. J Intern Med 1999; 246: 513–516
[12]. Abul-Ezz SR, Bunke CM, Singh H et al. Arterial hypertension. In: Andreoli TE, Bennett JC, Carpenter CCJ, Plum F (ed). Cecil's essentials of medicine. Philadelphia: W. B. Saunders, 1997; 4th edn: 227–231
[13]. Bravo EL, Tagle R. Pheochromocytoma: state-of-the-art and future prospects. Endocr Rev 2003; 24: 539–553
[14]. Mannelli M: Diagnostic problems in pheochromocytoma. J Endocrinol Invest 1989;12:739–57.
[15]. Manger WM. An overview of pheochromocytoma: history, current concepts, vagaries, and diagnostic challenges. Ann N Y Acad Sci 2006; 1073: 1–20
[16]. Eisenhofer G, Goldstein DS, Sullivan P et al. Biochemical and clinical manifestations of dopamine-producing paragangliomas: utility of plasma methoxytyramine. J Clin Endocrinol Metab 2005; 90: 2068– 2075
[17]. Lenders JW, Pacak K, Walther MM et al. Biochemical diagnosis of pheochromocytoma: which test is best? JAMA 2002; 287: 1427–1434
[18]. Werbel SS, Ober KP: Pheochromocytoma: update on diagnosis, localization, and management. Med Clin North Am 1995;79:131–53.
[19]. Barvo EL: Evolving concepts in pathophysiology, diagnosis, and treatment of pheochromocytoma. Endocr Rev 1994;15:356–68

Wafae JOUIDI, et. al. "Malignant Hypertensive Retinopathy in a Child as Initial Presentation of Pheochromocytoma." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, 19(11), 2020, pp. 47-50.