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Case Report

Hypertensive Retinopathy as a Presenting Sign of Pheochromocytoma with Malignant Hypertension: A Child Case

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Keywords
Childhood tumors · Hypertensive retinopathy · Neuro-ophthalmic disease · Genetic disease · Congenital abnormalities

Abstract
A 13-year-old was admitted to our clinic complaining about a vision loss of over 2 weeks. Bilateral optic disc edema, peripapillary flame-shaped hemorrhages, macular star pattern exudates, and cotton wool spots were found in fundoscopic examination. The OCT exam showed bilaterally serous retinal detachments in sub-foveal region with intraretinal exudates. A pediatric examination found a high systemic hypertension of 230/140 mm Hg, and laboratory tests revealed increased levels of plasmatic and urinary catecholamines. An abdominal MRI showed a left suprarenal pheochromocytoma and the child underwent an adrenalectomy urgently. Informed patient consent was obtained from legal guardians to publish clinical images. Malignant hypertension (MHT) as a result of a pheochromocytoma is an extremely rare condition in childhood. MHT crisis represents a potential life-threatening event and an immediate treatment can prevent a multi-organ failure, including the eyes. An early diagnosis of MHT by fundus examination can lead to a completely favorable ophthalmological evolution and entirely functional recovery.

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Introduction

Hypertension in pediatric population is estimated to have a world-wide prevalence of 2–5% [1], while hypertension crisis, such as malignant hypertension (MHT), is an extremely rare condition [1, 2]. MHT in children older than 10 years is characterized by two major features: severe increase in systolic blood pressure (BP), diastolic BP or both (≥170 mm Hg systolic and ≥110 mm Hg diastolic) [3], and hypertensive retinopathy grades 3 or 4 according to the classification of Keith et al. [4]. Renal vascular diseases and primary hypertension are recognized as the main causes of pediatric hypertension in children older than 10 years [2, 5–7], while pheochromocytomas are extremely rare conditions [1, 2, 5].

Case Report

A 13-year-old boy, without previous medical history, was presented to our emergency department for evaluation because of progressive and painless vision loss in both eyes of over 2 weeks without any other systemic complain. BCVA was 20/50 in the right eye and 20/80 in the left eye (LE). The anterior segment of both eyes was normal in the biomicroscopic examination, and the bilateral intraocular pressure was 15 mm Hg. Fundus examination revealed signs of hypertensive retinopathy (grade 4) with bilateral optic disc swelling, peripapillary flame-shaped hemorrhages, loss of the normal A/V ratio due to arteriolar narrowing, vascular tortuosity and, in the LE, disseminated cotton wool spots on the posterior pole as well as perifoveal exudates in a macular star's pattern (Fig. 1). The OCT showed bilaterally serous retinal detachments in sub-foveal region with intraretinal exudates (Fig. 2).

Pupils were both isochoric, isocyclic, and reactive with no relative afferent pupillary defect. The child was orthophoric with no restriction of extraocular motility.

Thus, a multidisciplinary approach was adopted for the diagnosis and treatment. A pediatric examination found a high systemic hypertension of 230/140 mm Hg, and a cardiac ultrasound was performed showing a severe concentric hypertrophy of the left ventricle, while the chest X-ray and the renal echography were completely normal. Laboratory tests revealed elevated levels of noradrenaline (118,266 pmol/L), adrenaline (799 pmol/L), and dopamine (1,972 pmol/L), while urinary spots showed a high level of normetanephrine (17,179 nmol/L). The patient underwent systemic antihypertensive treatment with hydrochloride labetalol i.v. 0.25 mg/kg/h which led only to a partial reduction of the systemic BP to 190/129 mm Hg. Furthermore, an abdominal MRI showed a left suprarenal mass of 39 × 31 × 48 mm confirmed by the scintigraphy to be a pheochromocytoma. Based on these findings, a left suprarenal adrenalectomy was made on urgency the days after. One month after surgery, the systemic BP was stabilized at 130/90 mm Hg with a DBP <110/60 mm Hg and his vision improved to 20/20 in both eyes. The eye examination after 1, 3, 6, 9, and 12 months revealed a progressive and almost complete absorption of the macular exudates in both eyes (Fig. 3). The OCT confirmed a normal retinal profile, while only minor intraretinal exudates in the foveal region of the LE remained. A genetic analysis (*next-generation sequencing*) did not reveal any further endocrinal associated tumors, such as VHL or other hereditary syndromes. Ethical approval is not required for this study in accordance with local or national guidelines. Written informed consent was obtained from the parents of the patient for publication of the details of their medical case and any accompanying images.

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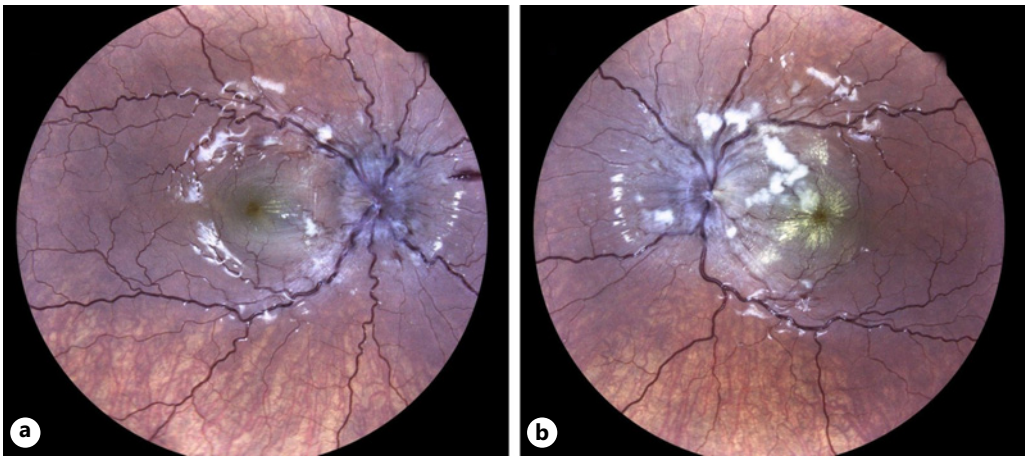

Fig. 1. Color fundus photographs of the RE and LE (**a**, **b**) show a bilateral optic disc swelling, peripapillary flame-shaped hemorrhages in the RE (**a**), arteriolar narrowing, vascular tortuosity, cotton wool spots, and perifoveal exudates in a macular star's pattern in the left eye (**b**).

Discussion

We describe a case of hypertensive retinopathy grade 5, according to the classification of Keith et al. [4] as a presenting sign in a 13-year-old child with malignant hypertension secondary to a left suprarenal pheochromocytoma [8–10]. It is known that hypertensive episodes associated with pheochromocytoma are produced by abnormally increased of sympathetic neuronal impulse frequency with excessive release of norepinephrine into the synaptic cleft with each impulse [1, 6] and it seems that it can determinate an optic disc ischemical damage [4, 5, 10]. Because of the vascular injury, a rapid reduction of high BP should avoid and prevent optic atrophy and permanent vision loss [4, 10] so they must be considered systemic emergencies.

Presentation with visual loss is rare and, in previous reports, returned to baseline after treatment of the pheochromocytoma [11, 12]. However, persistent visual loss from other causes of hypertensive retinopathy in children has been reported [13]. In a series of 4 patients younger than 16 years of age with malignant hypertension attributable to renal disease, 1 patient had persistent bilateral vision loss from ischemic optic neuropathy and another from a choroidal neovascular membrane with macular hemorrhage [14]. Another case report had persistent vision loss secondary to dense macular exudates [15]. Our clinical case is unique because despite presenting a very advanced clinical presentation with a stage 4 hypertensive retinopathy associated with papilledema and subretinal fluid on OCT, total visual recovery is possible with early diagnosis.

The major limitations of this study are the lack of OCTA or FA that were not performed. Therefore, information on retinal and choroidal ischemia is lacking. Further studies are needed to further evaluate the changes in choroidal and retinal perfusion in these findings.

In conclusion, MHT crisis represent a potential life-threatening condition, demanding prompt identification and immediate treatment to prevent a multi-organ failure, including the eyes [2]. This case shows how an early diagnosis of MHT with a pheochromocytoma in a child, made by ocular examinations, can lead to a completely favorable ophthalmological evolution and entirely functional recovery. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000531663).

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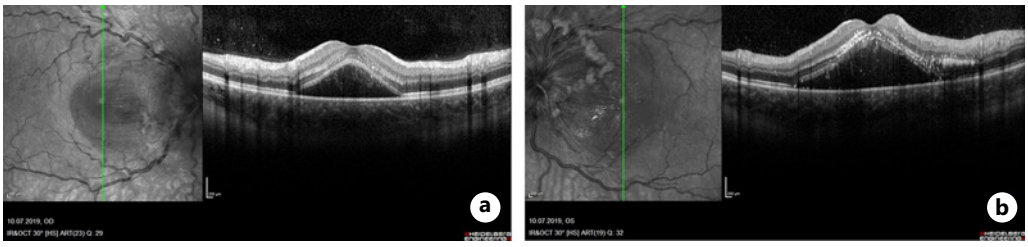

Fig. 2. Optical coherence tomography of the RE and LE (**a**, **b**) at presentation showing a bilateral serous retinal detachment in sub-foveal region associated with intraretinal exudates.

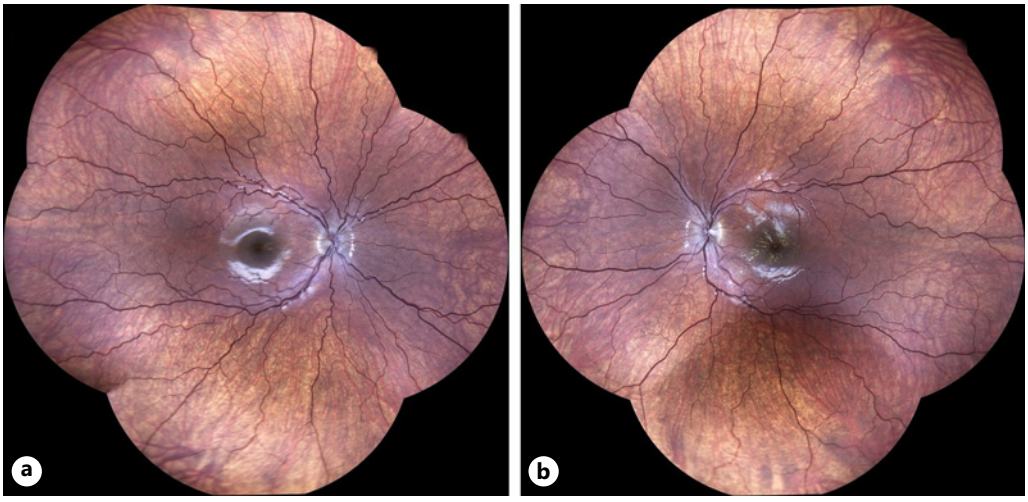

Fig. 3. Color fundus wide-field photographs of the RE and LE (**a**, **b**) 3 months later showing a progressive and almost complete absorption of the macular exudates in both eyes. RE, right eye.

Statement of Ethics

Ethical approval is not required for this study in accordance with local or national guidelines. Written informed consent was obtained from the parents of the patient for publication of the details of their medical case and any accompanying images.

Conflict of Interest Statement

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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Author Contributions

Feliciana Menna and Filippo Billia: substantial contributions to the conception or design of the work or the acquisition, analysis, or interpretation of data for the work. Nadia Palmieri: final approval of the version to be published. Andrea Maccari: agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. Andrea Consigli: drafting the work or revising it critically for important intellectual content. Patient perspective: received a good treatment.

Data Availability Statement

All data generated or analyzed during this study are included in this article and its online supplementary material files. Further inquiries can be directed to the corresponding author.

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