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LETTER TO THE EDITOR

Management of capillary non-perfusion associated with retinal racemose hemangioma

Non-perfusion capillaire associée à un hémangiome racémeux rétinien

Introduction

Retinal racemose hemangioma is a vascular malformation in which a large retinal vessel directly communicates with a retinal vein without intervening capillary bed [1]. It can also occur as an isolated phenomena or be part of the Wyburn-Mason syndrome, characterized by similar systemic vascular malformations [1]. The clinical appearance is characterized by dilated and tortuous retinal vessels [2]. Fluorescein angiogram may show rapid filling of the arteriovenous communications (AVC) [3]. Most lesions are stationary and do not need treatment. However, the visual prognosis is poor [3]. Numerous complications may occur leading to severe visual impairment [1].

Case report

A 14-year-old boy was referred following a routine exam with the diagnosis of optic nerve tumor. He reported a decreased visual acuity in his left eye for nearly 2 months. His medical history was otherwise unremarkable with no ocular past or family history of malformations. The examination of the right eye was normal with a visual acuity of 10/10 [+0.25 (—0.5 on 160°)] (Fig. 1). On the left one, best-corrected visual acuity was 0.5/10 with an optic correction of +0.75 (—1.25 on 165°); P14. The intraocular pressure and ocular media were normal. Iris slit-lamp examination was unremarkable. Fundus bio-microscopy disclosed an AVM involving the whole retina with tortuous and enlarged retinal vessels distributed over all four quadrants of the left fundus (Fig. 2). The arteries were well individualized, located at the center of the posterior pole, bright red, very dilated and tortuous. A whitish, engagement surrounded them with yellowish deposit in some seats (Fig. 2). The optic disc was obscured by large vascular loops. An epimacular membrane extending from the papilla to the temporal periphery exerted a tractional effect on the retinal structures (Fig. 2). The venous component was easily separated,

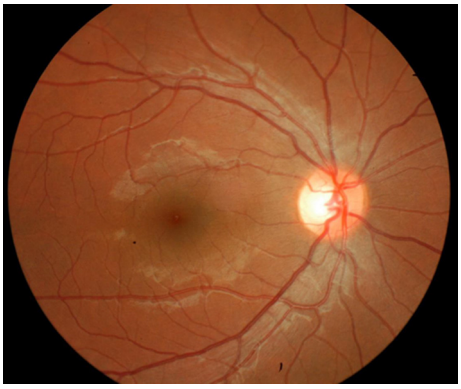

Figure 1. Fundus photograph of the right posterior pole. Normal aspect.

with less dilated and protruding wall in the vitreous cavity and more periphery rested seat compared to arteries (Fig. 2). A third vascular contingent with more disorganized and well-individualized architecture, as if placed upon the voluminous arteries, seems to plunge into the mooring fibrous tissue (Fig. 2). Furthermore, exophthalmos was not found (Fig. 3), and the ocular motility was normal.

Fluorescein angiography showed rapid filling of the arteries, with dilated and thickened wall without diffusion as the angiogram progressed (Fig. 4). The veins were more granular with sinuous path and delay in filling. At a later time, a third type of anastomosing vessels of superficial localization impregnated with dye leakage (Fig. 4). At the retinal periphery, the vessels were of hazy appearance, with sinuous path and hyper-fluorescent walls. Signs of marked non-perfusion were noted on the peripheral capillary bed (Fig. 4). A retinal diffusion at the posterior pole and peri-papillary area was observed at the late angiogram and well documented at the spectral domain optical coherence tomography (OCT-SD) (Fig. 5). The OCT imaging revealed a structural disorganization with thickening of the inner layers (Fig. 5). A thick epimacular membrane extended from the papilla to the retinal periphery, covering the whole vascular structures. The vascular section showed dilated loops, with thickened hyper-reflective walls, protruding into the vitreous cavity. A fibrous tissue, well individualized from the membrane described above, seems to connect the vessels to each other (Fig. 5). The internal boundary layer was irregular and thickened with structural changes in the nerve fiber

Abbreviations: AVC, arteriovenous communications; CRAVC, congenital retinal arteriovenous communications; OCT-SD, spectral domain optical coherence tomography; MRA, magnetic resonance angiography; MRI, magnetic resonance imaging.

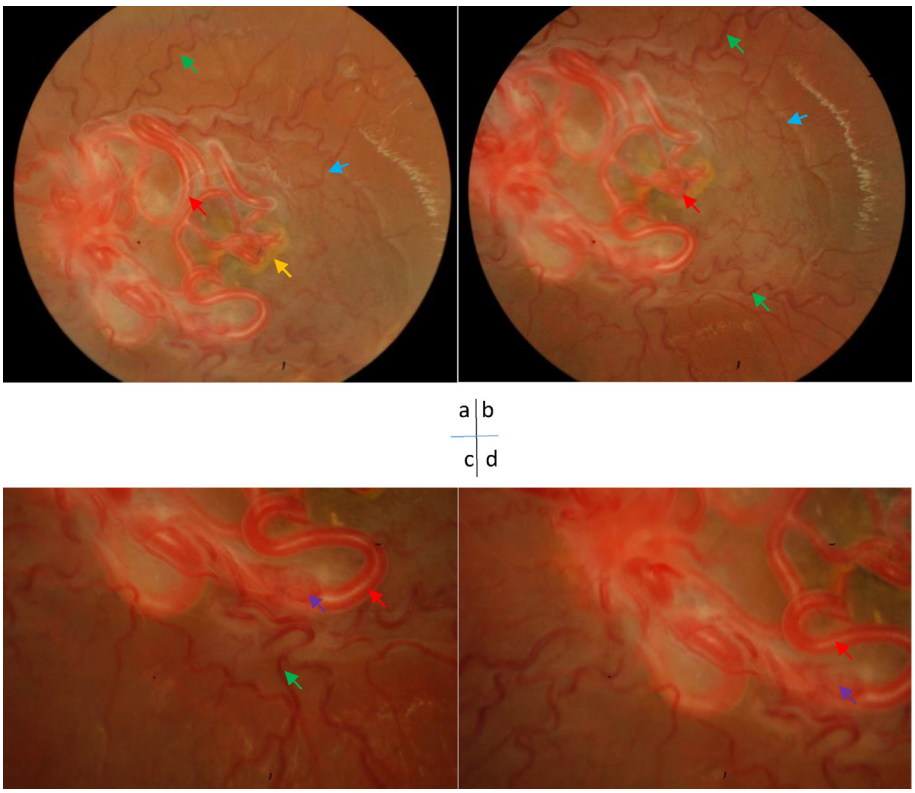

Figure 2. Fundus photograph of left posterior pole. Note dilation of both the arterial and venous systems distributed from the optic disc over four quadrants in the left eye. Most of the optic disk is covered by thickened vessels. Arteries are located in the central pole, more dilated and bright red (red arrows). A whitish, engagement surrounded them with yellowish deposit in some seats (yellow arrow). Venous component easily separated, with less dilated and protruding wall in the vitreous cavity and more periphery rested seat compared to arteries (green arrows). Retinal arteriovenous malformations are numerous, more disorganized and well-individualized architecture, as if placed upon the voluminous arteries seems to plunge into the mooring fibrous tissue (purple arrows). An epimacular membrane extending from the papilla to the temporal periphery, exerted a tractional effect on the retinal structures (blue arrows).

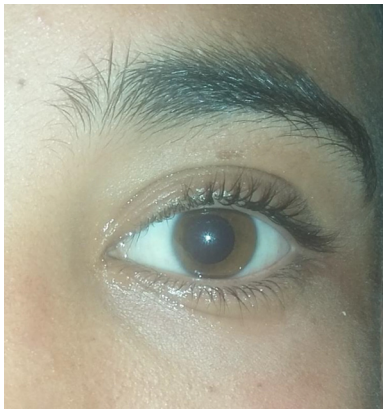

Figure 3. No exaphtolmos nor vascular conjunctival dilatation.

layer. A vitreous tyndall was well individualized in the posterior vitreous as hyper-reflective points (Fig. 5). The vascular structures of the choroid was thick compared to the right eye (Fig. 5).

Moreover, imaging studies included ultrasonography with color Doppler study, demonstrating a high-throughput arteriovenous flow (Fig. 6). Neurological examinations were unremarkable without any history of epilepsy, and no changes were found in the facial skin (Fig. 3). MRI studies were performed, comprising gadolinium enhanced T1, T2 sequences, as well as magnetic resonance angiography (MRA). This showed no intracranial midbrain extension or communication with cerebral vessels (Fig. 7). The patient was clinically followed up. Fluorescein angiography was remade every 6 months and OCT-SD every 3 months. No active treatment was indicated. The patient has been stable for 3 years and remains under long-term review.

Comments

Our patient presented with an unusual unilateral sporadic isolated retinal vascular anomaly. The clinical presentation and multimodal imaging appearance was concordant with the diagnosis of a CAVRC group 3. They are malformations in which blood passes directly from arteries to veins. Although being congenital, no hereditary predisposing factor has been described [4,5]. Since 1943, association with vascular abnormalities of the face and brain was recognized as a special entity by Bonnet-Dechaume-Blanc [6] and then characterized by Wyburn-Mason [7]. This entity is also considered to be one of the phakomatosis, or neuro-oculocutaneous syndromes [2,8]. CAVRC, associated or not with cutaneous or cerebral involvement are rare. In a review of the literature up to 2008, 122 cases were reported, including two bilateral malformations, with 57 isolated retinal disorder cases [9]. These congenital malformations may be due to