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SHORT COMMUNICATION

Case report

Polypoidal choroidal vasculopathy associated with Doyne's familial choroiditis: treatment with thermal laser

I.D. LADAS, D.A. KARAGIANNIS, I. GEORGALAS, A.A. ROUVAS, M.M. MOSCHOS, M. APOSTOLOPOULOS

Department of Ophthalmology, Medical School of Athens University, Athens - Greece

PURPOSE. *To report the unusual occurrence of polypoidal choroidal vasculopathy (PCV) in a patient with Doyne's familial honeycomb choroiditis (DFHC) and its course after laser treatment.*

DESIGN. *Interventional case report.*

METHODS. *Indocyanine green (ICG) angiography guided laser was performed on active polypoidal lesions.*

RESULTS. *A 45-year-old man with a 15-year history of bilateral DFCH and a scarred macular choroidal neovascularization in the right eye (RE) was referred to us with exudative maculopathy in the left eye (LE). His best-corrected visual acuity (BCVA) was 20/800 in the RE and 20/40 in the LE. ICG angiography revealed a picture that was characteristic for PCV in both eyes. ICG guided argon green laser was performed on the active parapapillary and perifoveal polypoidal lesions of the LE. Eight months after the laser photocoagulation treatment, the macular exudative lesions had subsided and the BCVA improved to 20/20. The favorable anatomic and functional results have remained stable over 3 years.*

CONCLUSIONS. *This is, to our knowledge, the first case of a PCV that occurred secondary to DFHC. (Eur J Ophthalmol 2004; 14: 264-8)*

KEY WORDS. *Polypoidal choroidal vasculopathy, Doyne's familial honeycomb choroiditis, Indocyanine green angiography, Choroidal neovascularization*

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INTRODUCTION

For about 15 years, polypoidal choroidal vasculopathy (PCV) has been recognized as a new distinct exudative maculopathy characterized by a peculiar network of fine branching vessels external to choriocapillaris and multiple terminal aneurysmal dilations at the border of the vascular network. It is generally accepted that indocyanine green (ICG) angiography is the examination of choice for identification of the vascular polypoidal lesions of PCV with great sensitivity and specificity (1-6).

Doyne's familial honeycomb choroiditis (DFHC) is one of the dominantly inherited drusen syndromes characterized by multiple large yellow-white soft drusen all over the posterior pole that appear at a relatively early age in more than one member of the same family and occasionally may lead to choroidal neovas-

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cularization (CNV) (7-9). We report, to our knowledge, the first case of PCV in a patient with DFHC and its course after thermal laser treatment.

Case report

A 45-year-old man with a 15-year history of DFCH was referred to us with the diagnosis of exudative maculopathy secondary to macular CNV in his left eye (LE). His family history was positive for DFCH (his 36-year-old brother presented confluent soft drusen in the macula of both eyes). He was in otherwise good health and taking no medication. His right eye (RE) was diagnosed with macular CNV on the ground of DFHC 6 years ago. Best-corrected visual acuity (BCVA) was 20/800 in the RE and 20/40 in the LE.

Fundus examination of the RE revealed an extensive geographic choroidal atrophy in the macula that had occurred on the ground of an old scarred CNV (Fig. 1a). Results of fundus examination of the LE showed a serosanguineous macular neurosensory detachment with hard exudates parapapillary and two attendant perifoveal detachments of the retinal pigment epithelium. All these lesions had been developed on the ground of multiple large soft drusen confluents in the macula area (Fig. 2a). Fluorescein angiogram (FA) delineated the above lesions (Fig. 2b). However, ICG angiography of the LE revealed, just nasal to the optic disc, a peculiar network of vessels at the level of choriocapillaris with multiple terminal aneurysmal dilations (Fig. 2d). Some more vascular polypoidal lesions were detected in the LE just inferior to the optic disc, as well as in the perifoveal area (Fig. 2, c-e). Results of ICG angiography of the RE showed some similar polypoidal lesions close to the superior margin of the optic disk and at the temporal border of the geographic choroidal atrophy (Fig. 1, b and c).

ICG-guided argon green laser photocoagulation was performed directly on the active vascular polypoidal lesions of the LE according to a similar technique that we have described previously in treating patients with diabetic macular edema (10). Particularly, the sites of laser application were mapped on the ICG angiogram image and transferred to the red-free fundus photograph using the Topcon Imagenet 2000 Digital Imaging System with Imagenet 2000 (version 2.14) software and the TRC-50IA fundus camera (Fig. 3a). We performed the laser treatment looking back and forth between the color image of the patient's fundus through the biomicroscope of the laser and the processed red free fundus image displayed on the computer monitor adjacent to the laser. The desired goal of the laser

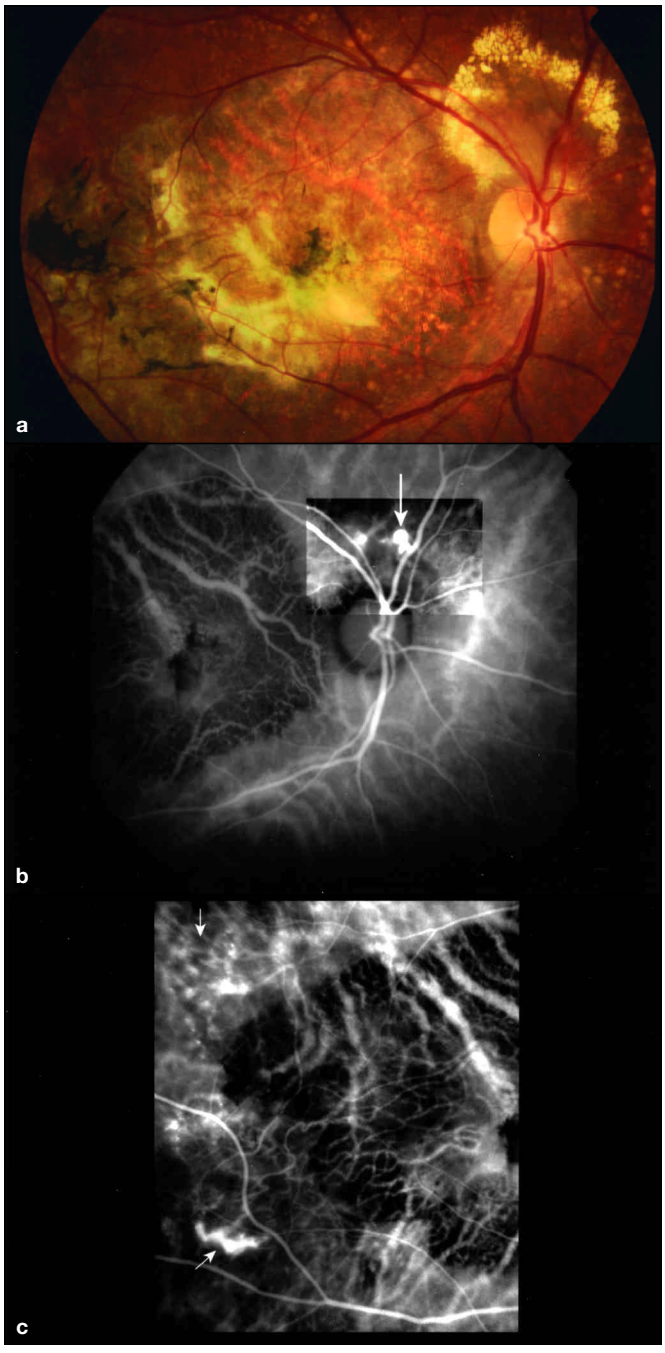

Fig. 1 - *Right eye.* **a)** *Color fundus photograph. Chorioretinal scar in the central retina.* **b, c)** *Indocyanine green angiograms. Vascular polypoidal lesions just superior to the optic disk and temporal to the macular scar (arrows).*

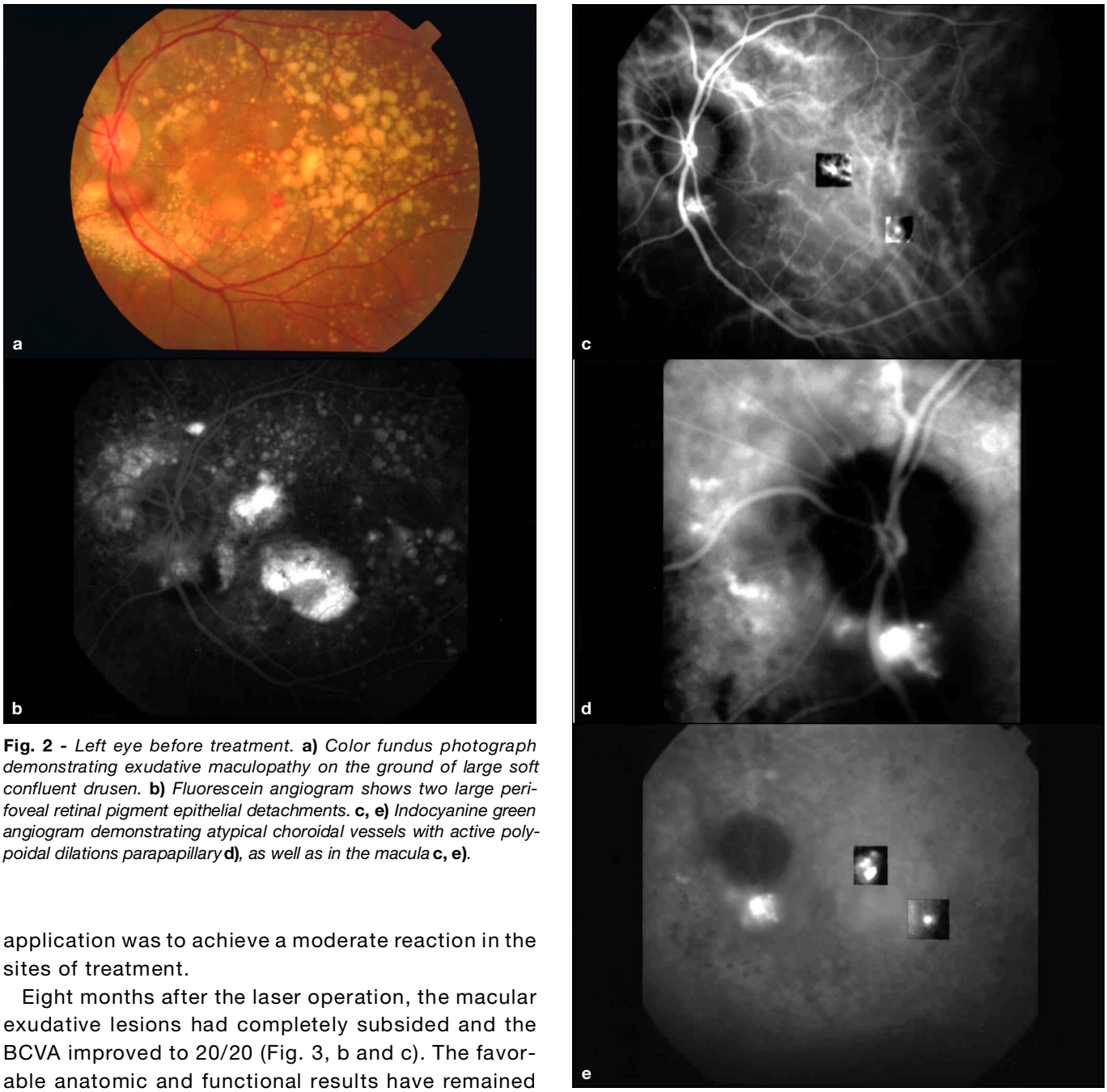

Fig. 2 - *Left eye before treatment.* **a)** *Color fundus photograph demonstrating exudative maculopathy on the ground of large soft confluent drusen.* **b)** *Fluorescein angiogram shows two large perifoveal retinal pigment epithelial detachments.* **c, e)** *Indocyanine green angiogram demonstrating atypical choroidal vessels with active polypoidal dilations parapapillary* **d)**, *as well as in the macula* **c, e)**.

application was to achieve a moderate reaction in the sites of treatment.

Eight months after the laser operation, the macular exudative lesions had completely subsided and the BCVA improved to 20/20 (Fig. 3, b and c). The favorable anatomic and functional results have remained stable over 3 years.

DISCUSSION

DFHC is an autosomal dominant disease characterized by multiple yellow-white large soft drusen that accumulate beneath the retinal pigment epithelium in the macula and they may lead to pigmentary changes and/or to macular CNV (7-9). The importance of this disease is due in large part to its close similarity to age-related macular degeneration, especially when the macular CNV develops in patients older than 50 years.

Our case concerns a 45-year-old man with a 15-year history of DFHC and an old macular fibrovascular scar in the RE, who presented an exudative maculopathy

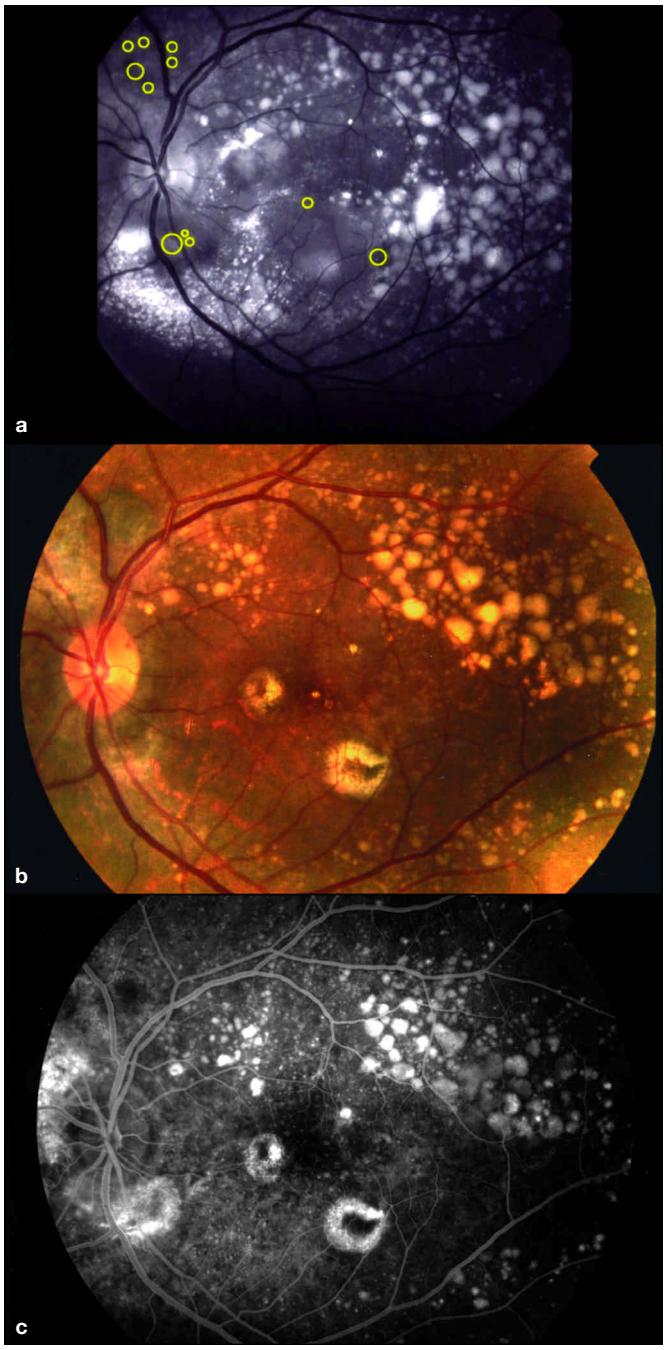

Fig. 3 - *Left eye.* **a)** *Processed red free fundus image used to guide laser treatment. Yellow traces: Active polypoidal lesions (laser spots must be applied on them).* **b, c)** *Color fundus photograph* **b)** *and fluorescein angiogram* **c)** *demonstrating complete resolution of the exudative lesions of the macula 8 months post laser treatment.*

suggesting the development of a macular CNV in the other eye. Although the patient was referred to us with the initial diagnosis of macular CNV, he was ultimately diagnosed with PCV. The role of ICG angiography to set the correct diagnosis was decisive. With ICG angiography we could identify the polypoidal vascular lesions and distinguish them from the CNV with great sensitivity and specificity (Fig. 1, b and c, and Fig. 2, c-e). Further, ICG angiography was very important as a guide to the laser photocoagulation treatment precisely on the active polypoidal lesions.

The results of the argon green laser photocoagulation in our patient were very satisfactory. Eight months after the laser treatment, the macular exudative lesions showed complete resolution (Fig. 3) and the BCVA had improved to 20/20. These favorable anatomic and functional results have remained unchanged during a long follow-up period.

Our patient is, to our knowledge, the first reported case of PCV on the ground of DFCH. ICG-guided argon laser treatment of the active polypoidal lesions had excellent results.

Reprint requests to:
Ioannis D. Ladas, MD
8 Meg. Alexandrou Str.
GR-152 36 P. Penteli
Athens, Greece
ladas@ath.forthnet.gr

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