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Indocyanine green angiography in the juvenile haemorrhagic choroidopathy

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Abstract

Juvenile haemorrhagic choroidopathy (JHC) is an idiopathic syndrome marked by macular choroidal neovascularization (CNV) in patients under the age of 50. We used fluorescein angiography (FA) and indocyanine green angiography (ICGA) to examine 17 patients with macular CNV and JHC. CNV was always unilateral. On ICGA examination the CNV were weakly fluorescent in 59% of cases, hyperfluorescent in the remaining 41%.

ICGA showed up the following alterations: a) areas with diffuse choroidal hyperfluorescence at the posterior pole or in the peripapillary region in 11 affected eyes (65%), in the fellow eye too in 5 patients; b) areas of choroidal hypofluorescence at the posterior pole but also outside the vascular arcades in 2 affected eyes (12%).

In conclusion, ICGA does not appear indispensable for detecting CNV in JHC but this method does show up diffuse choroidal alterations not detectable with FA. The pathogenetic implications of the ICGA findings are discussed.

Introduction

Juvenile hemorrhagic choroidopathy (JHC) is an uncommon syndrome characterized by a macular hemorrhagic lesion associated with choroidal neovascularization (CNV) in patients under the age of 50. This syndrome is usually unilateral and more frequently affects females. Its etiopathogenesis is still unknown but it has been suggested that JHC is secondary to choroidal inflammation, although no association has ever been found with current pathology, or with anything in the patient's history or detectable with serum tests [1, 2].

Indocyanine green angiography (ICGA) has proved useful in many conditions including various inflammatory or autoimmune diseases such as serpiginous choroiditis, multifocal choriocapillaritis [3], acute multifocal epitheliopathy [4], and HIV retinal syndrome [5]. The aim of our study was to investigate choroidal alterations in juvenile hemorrhagic choroidopathy utilizing indocyanine green angiography.

Patients and methods

34 eyes of 17 consecutive patients all under the age of 50 years (mean 30.9, range 14–48), were prospectively evaluated. Eleven were women and six men. All had unilateral CNV. Their mean refraction was -2.6 D (range +1 / -6 D). Eyes with degenerative myopia (more than -6 D and/or myopic alterations at the posterior pole) were excluded, as well as eyes with other lesions at the posterior pole, in the peripapillary region or retinal periphery.

Each patient underwent a complete clinical examination (including visual acuity), biomicroscopic fundus examination with a Goldmann contact lens or a Volk 90-diopter lens, fluorescein angiography (FA). ICGA was performed utilizing the Topcon IMAGE-net System after intravenous injection of 25 mg of indocyanine green, followed by a flush of 5 ml of balanced saline solution.

Table 1. Fluorangiografic characteristics of the patients.

	Patient	Age	CNV	O.N. Hyper	Fellow eye
1	TS	21	Yes	No	
2	TM	14	Yes	No	-
3	GG	27	Yes	No	-
4	DDL	42	Yes	No	-
5	BM	27	Yes	No	-
6	DMA	22	Yes	Yes	-
7	VS	23	Yes	Yes	RPE dystrophy
8	RR	33	Yes	No	RPE dystrophy
9	CG	37	Yes	No	-
10	BL	46	Yes	No	RPE dystrophy
11	PN	48	Yes	No	-
12	NV	39	Yes	No	-
13	LFC	14	Yes	No	-
14	PM	35	Yes	No	-
15	PP	32	Yes	No	-
16	AP	36	Yes	No	-
17	DDV	29	Yes	No	-

O.N. Hyper = Optic Nerve Hyperfluorescence
RPE = Retinal Pigment Epithelium

Table 2. Indocyanine green angiography characteristics.

	Patient	Affected eye				Fellow eye
		CNV early phases	CNV late phases	Hyper areas	Hype areas	Hyper areas
1	TS	Hypo	Hypo	No	No	No
2	TM	Hyper	Hyper	No	No	No
3	GG	Hypo	Hyper	No	No	No
4	DDL	Hypo	Hypo	Yes	No	Yes
5	BM	Hypo	Hypo	Yes	No	No
6	DMA	Hypo	Hyper	No	Yes	No
7	VS	Hypo	Hypo	Yes	Yes	Yes
8	RR	Hyper	Hypo	Yes	No	No
9	CG	Hypo	Hypo	Yes	No	Yes
10	BL	Hyper	Hyper	Yes	No	No
11	PN	Hyper	Hyper	Yes	No	No
12	NV	Hypo	Hypo	Yes	No	No
13	LFC	Hyper	Hyper	No	No	No
14	PM	Hypo	Hypo	Yes	No	No
15	PP	Hyper	Hyper	Yes	No	Yes
16	AP	Hypo	Hypo	Yes	No	Yes
17	DDV	Hyper	Hyper	No	No	No

Results

None of the eyes had any sign of anterior or posterior inflammation. All patients had a well defined CNV at FA. Two eyes presented hyperfluorescence at the optic disk. In three cases, the fellow eye presented limited areas of dystrophy of the retinal pigment epithelium. The characteristics of patients and eyes are shown in Table 1.

ICGA findings

CNV were detected by ICGA in all the cases. 10 CNV (59%) were hypofluorescent in the early phases; 8 of them remained unchanged throughout the examination, and 2 showed some staining during the late ICGA phases (30–40m). 7 CNV (41%) appeared fluorescent in all the phases (Fig. 1) and 4 of them showed marked staining during the late angiographic phases. Furthermore ICGA detected diffuse choroidal areas of hyperfluorescence in 11 affected eyes (65%) and in 5 fellow eyes (29%) during the medium-late phases (Figs 2–3). Hyperfluorescence was localized at the posterior pole or in one cases in the peripapillary region. 2 eyes had patches of choroidal hypofluorescence at the posterior pole and also beyond the arcades (Fig. 4), seen during the middle phases of the examination and becoming more evident in the later phases. These were the 2 patients who had shown hyperfluorescence of the optic disk at the FA examination. Table 2 summarizes the ICGA features.

Discussion

ICG is a water-soluble contrast medium 90% of which binds to plasma proteins, particularly albumin and alpha-lipoprotein. ICGA has been widely used in ophthalmological practice only in the last few years, but has proved useful in certain pathologies, particularly age-related macular degeneration. In this disorder-ICGA is extremely useful in diagnosing occult neovascularizations, choroidal newvessels in areas of pigment epithelium detachment, and recurrent neovascularization when the lesions are poorly visible on retinal FA [6]. ICGA is less useful in pathologies in which CNV are clearly visible on retinal FA, such as pathological myopia [7] and angioid streaks.

Currently ICGA is also used for the diagnosis of other diseases such as tumours [8], central serous chorioretinopathy [9, 10] and many forms of choroiditis

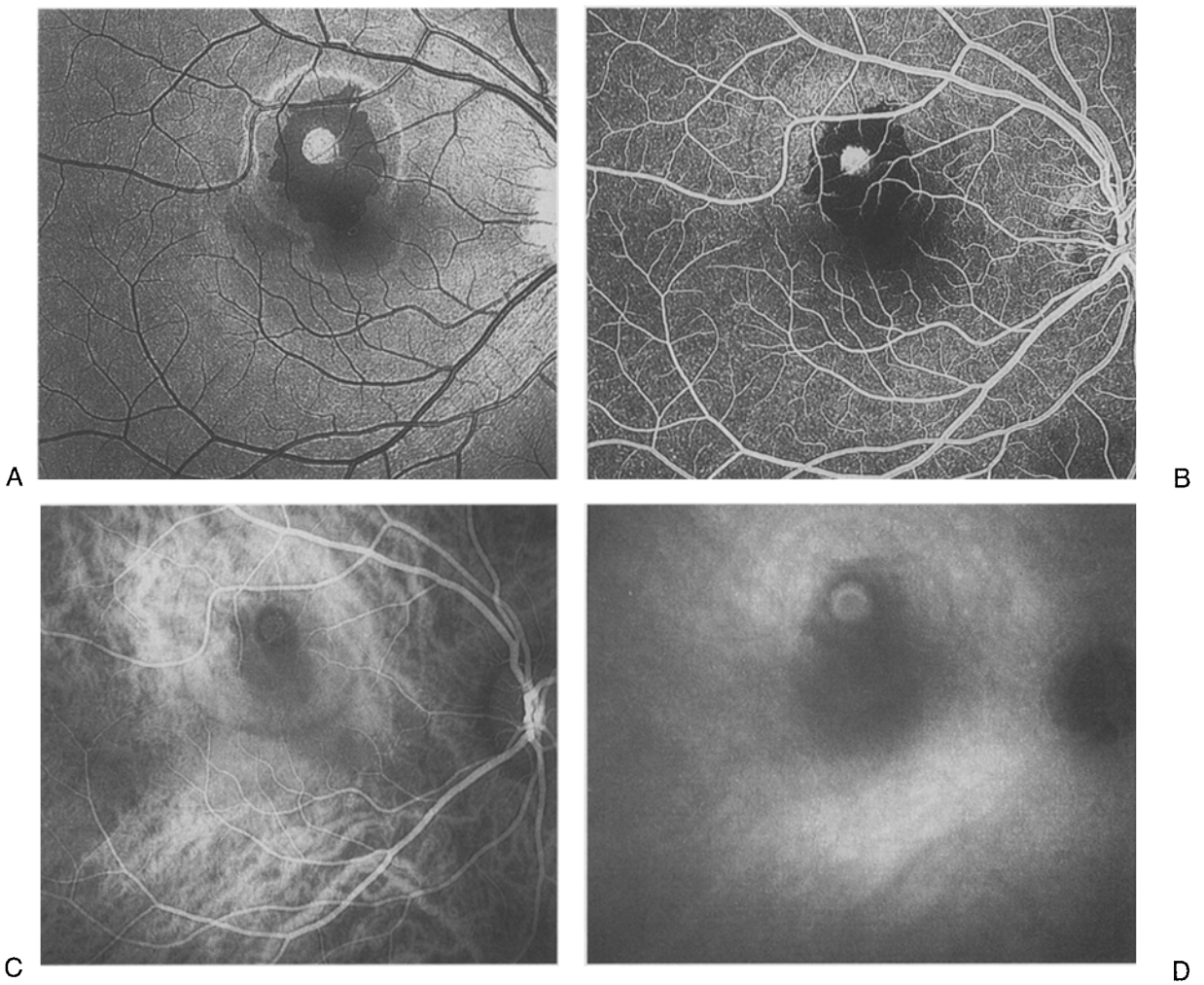

Fig. 1. A large serous haemorrhagic detachment of the neurosensorial retina is well delineated on red-free retinography (*A*) and on fluorescein angiography (*B*) where a well-defined CNV is evident. In the early ICG angiogram (*C*) the CNV appears poorly hyperfluorescent and is encircled by an hypofluorescent rim. In the late phases (*D*) the CNV is still hyperfluorescent.

[3–5]. All the patients we studied had well defined CNV on FA. ICGA enabled us to detect CNV in all the cases. However CNV were well visible in 41% with a late staining, while in the remaining 59% appeared hypofluorescent and less detectable than FA. ICGA therefore, considering the absence of occult CNV in this disease, did not seem particularly useful for detecting CNV in patients with JHC.

In 11 of the eyes we studied (65%) ICGA detected choroidal areas with varying degrees of hyperfluorescence, visible as blurred patches at the posterior pole or in the peripapillary region. In 5 of these patients the fellow eye was found to be involved, though asymptomatic clinically and on FA. Such hyperfluorescent areas are frequently seen in central serous chorioretinopathy. It has been suggested that they are the manifestations of hyperpermeability of the choriocapillaris in the affected and fellow eye (9–10). This finding in patients with JHC suggests there may be a more diffuse choroidal alteration in this pathology too, with

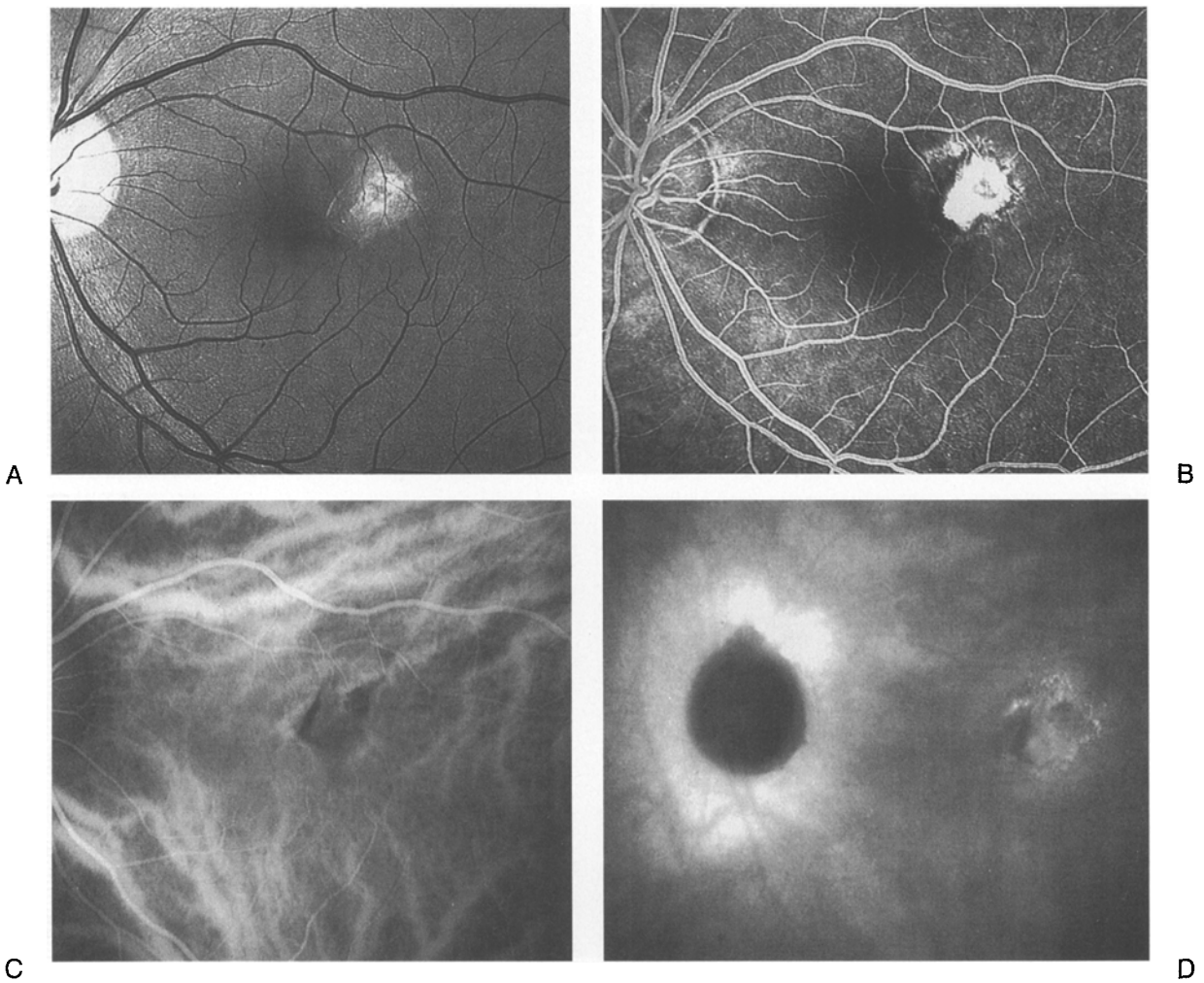

Fig. 2. A macular CNV, located temporally to the fovea, is evidenced by red-free retinography (*A*) and fluorescein angiography (*B*). It is surrounded by an atrophic area. The CNV appears hypofluorescent in the early ICG phases (*C*) with a slight staining in the late ICG phases (*D*). A large hyperfluorescent area is visible around the optic disk, in the late phases.

vasomotor derangement and focal choroidal vascular hyperpermeability.

In 2 eyes (12%) we observed multiple clear-cut areas of widespread hypofluorescence, dotted around in the macular region, in the peripapillary region and even beyond the vascular arcades. These areas became visible in the intermediate phases of the examination and were clearer in the late phases. Such findings have been reported in pathologies suspected of having an inflammatory etiology. In acute posterior multifocal epitheliopathy, large areas of hypofluorescence are described which have been interpreted as areas of capillary non perfusion, or with masking by reactive edema [4]. Areas of hypoperfusion are also seen in serpiginous choroiditis and in multifocal choriocapillaritis. In the former, it has been suggested that there is inflammation of the large choroidal vessels, and in the latter the lesions appears to be more superficial, the medial choroidal capillary network being preserved [3].

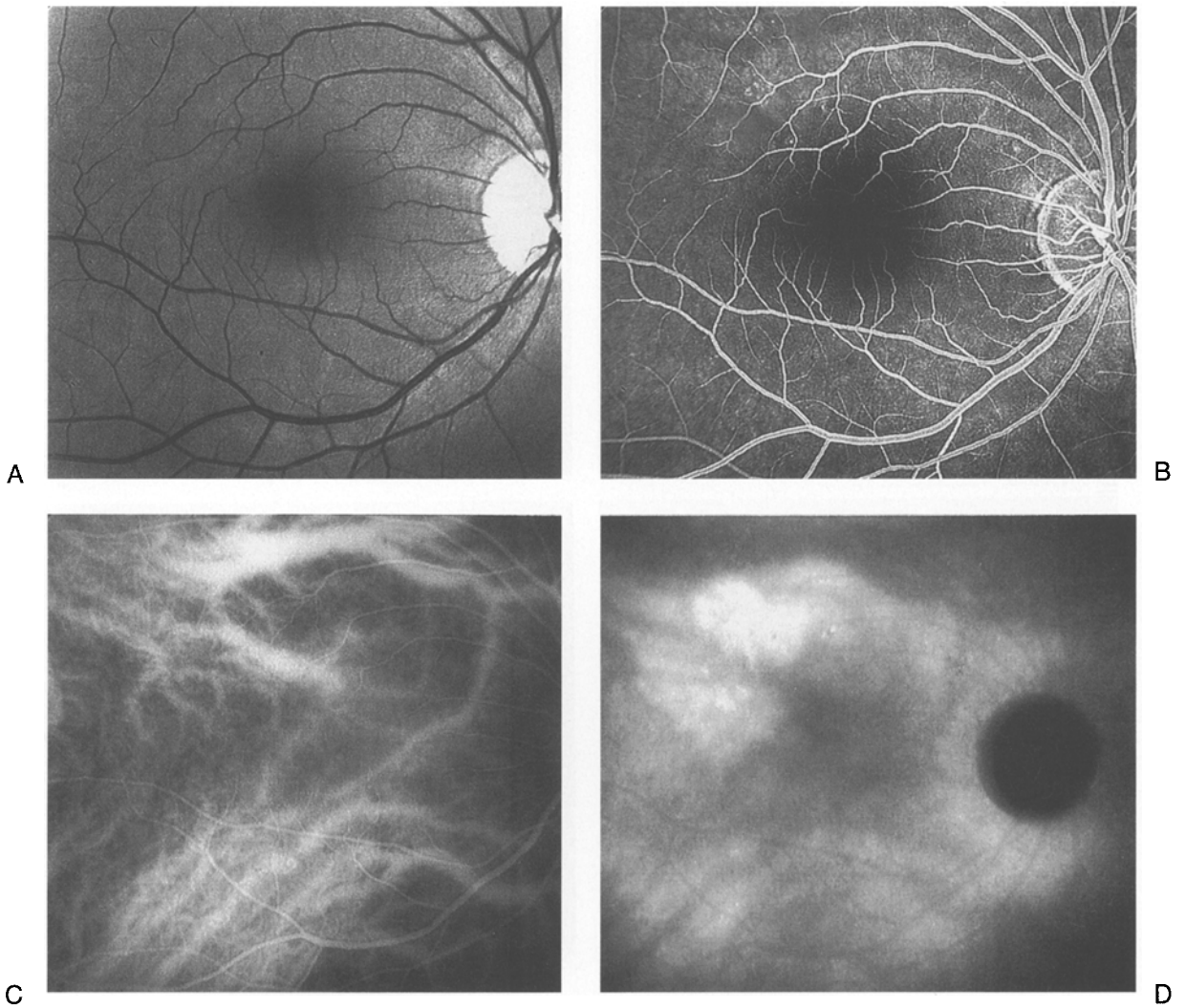

Fig. 3. Fellow eye of patient presented in Fig.2. Red-free retinography (*A*) and fluorescein angiography (*B*) are quite normal. ICG angiography shows in the late phases (*D*) widespread hyperfluorescent areas located at the posterior pole.

In our experience hypofluorescent spots are often observed in multifocal choroiditis. In this disease, multiple areas of choroidal hypofluorescence may be present which could be correlated with a wide choroidal involvement.

In this study, both of the patients with choroidal hypofluorescence showed hyperfluorescence of the optic disk on FA. As shown in Fig.4, we can assess that though the FA pattern is typical of JHC, ICG showed an aspect very similar to multifocal choroiditis, supporting an inflammatory pathogenesis.

In conclusion, the present study showed that ICGA is not indispensable for showing up choroidal new vessels in JHC as these are quite easily detectable with standard retinal FA. ICGA is however, worth using for assessing choroidal function. The case in which FA showed hyperfluorescence of the optic disk appeared to have more serious choroidal involvement, with alterations suspected as being of inflammatory origin. In such cases it could be useful to prescribe adequate general medical theraphy to control the inflammation.

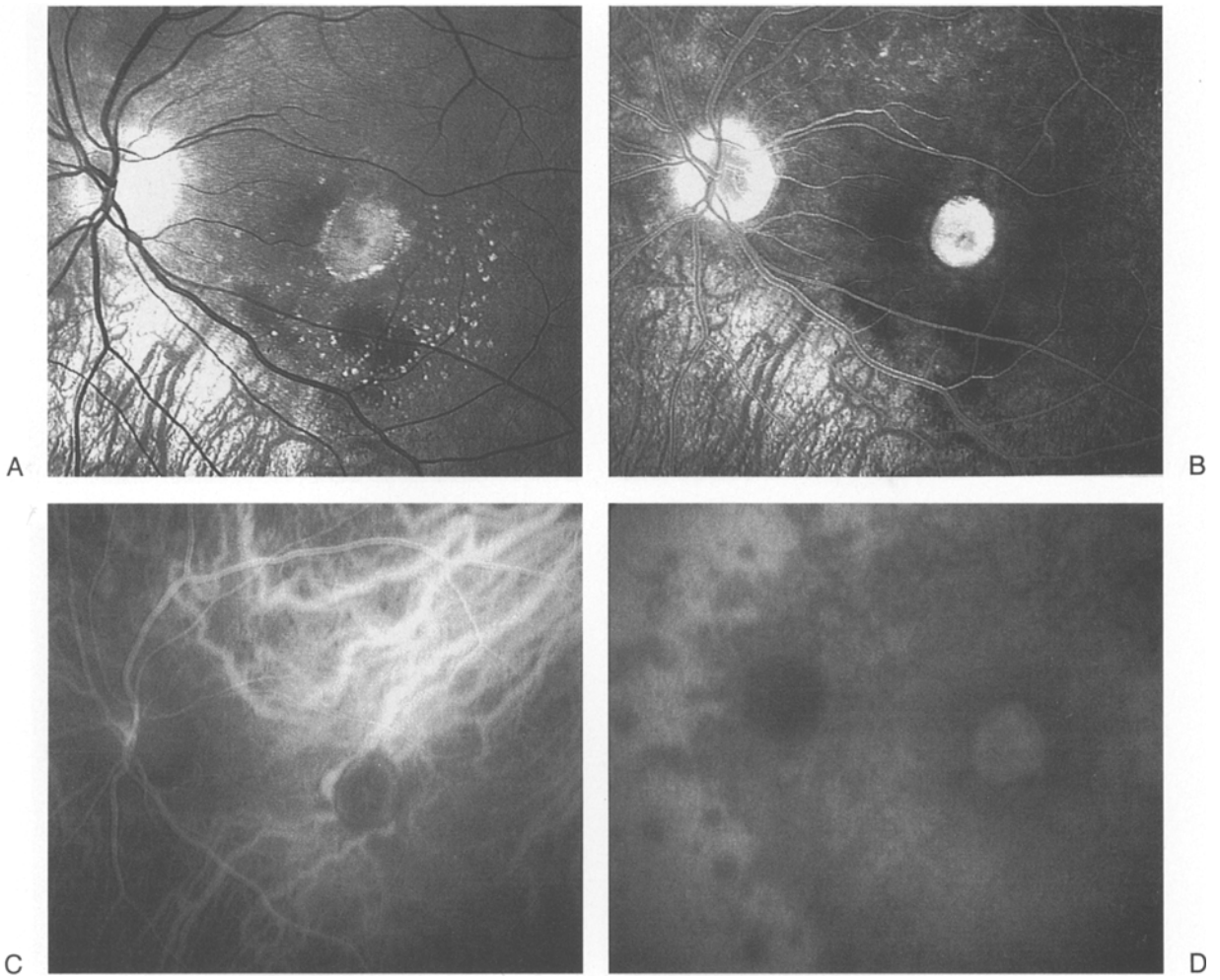

Fig. 4. Red-free retinography (*A*) evidences a macular CNV associated with a deep haemorrhage and numerous hard exudates. Fluorescein angiography (*B*) shows a well-defined macular CNV. A late hyperfluorescence of the optic disk is present. The posterior pole appears hypofluorescent in the late phases (d). Hypofluorescent dots are present outside the vascular arcades. CNV is hypofluorescent in the early ICG phases (*C*) while in the late phases appears as fluorescent as the remaining uninvolved choroid.

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