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SHORT COMMUNICATION

Indocyanine green angiography in congenital hypertrophy of the retinal pigment epithelium

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PURPOSE. *To report a case of congenital hypertrophy of the retinal pigment epithelium followed up for 7 years showing features not previously reported.*
METHODS. *A complete fundus examination including fluorescein angiography was performed at first examination and at follow-up.*
RESULTS. *The area of congenital hypertrophy of the retinal pigment epithelium presented morphologic changes, showing enlargement of the lesion on one side associated with partial regression on another side. In addition, the indocyanine green angiography findings revealed that this test was more useful than fluorescein angiography to delineate the real boundaries of the lesion and disclosed hypofluorescent areas inside the main lesion that were not appreciated at ophthalmoscopy or fluorescein angiography.*
CONCLUSIONS. *Indocyanine green angiography is an useful test to understand the findings and evolution of congenital hypertrophy of the retinal pigment epithelium. (Eur J Ophthalmol 2005; 15: 162-4)*

KEY WORDS. *Congenital hypertrophy of the retinal pigment epithelium, Fluorescein angography, Indocyanine green angiography*

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INTRODUCTION

Congenital hypertrophy of the retinal pigment epithelium (CHRPE) is a flat and black benign lesion of the retinal pigment epithelium (RPE) (1, 2). In most cases the plaques of CHRPE show some enlargement over years (3, 4). Fluorescein angiographic features of CHRPE are well known and help to differentiate this lesion from benign and malignant tumors of the choroid (5, 6); however, indocyanine green (ICG) angiographic findings of this lesion are much less studied.

We describe a case of CHRPE that showed an unusual partial regression and in which the ICG angiography gave more information about several aspects of the lesion than fluorescein angiography.

Case report

A 12-year-old girl was referred to the service of fluorescein angiography of the University Eye Clinic of Palermo because of a pigmented lesion in the right fundus that was found during a routine examination of the eye. She did not have visual symptoms and her visual acuity was 20/20 in both eyes. Nothing remarkable was seen at the ophthalmologic examination, except in the fundus of right eye: inferiorly to the temporal retinal vascular arcade, a round, black, and flat plaque, measuring about 1.5 disc diameters, was seen (Fig.1A).

Two small round white atrophic lesions were evidenced inside the black plaque and an incomplete subtle white annulus was recognized parallel and inside the nasal border of the plaque.

Fluorescein angiography outlined the border of the whole lesion that appeared uniformly hypofluorescent, except for choroidal vessels that were seen through the two atrophic spots and the incomplete annulus that showed barely visible transmission hyperfluorescence (Fig. 1B). No abnormality of the retinal vascular bed, as rarefaction of capillaries or microaneurysms, was found upon the lesion. A diagnosis of CHRPE was made and no other diagnostic test was performed.

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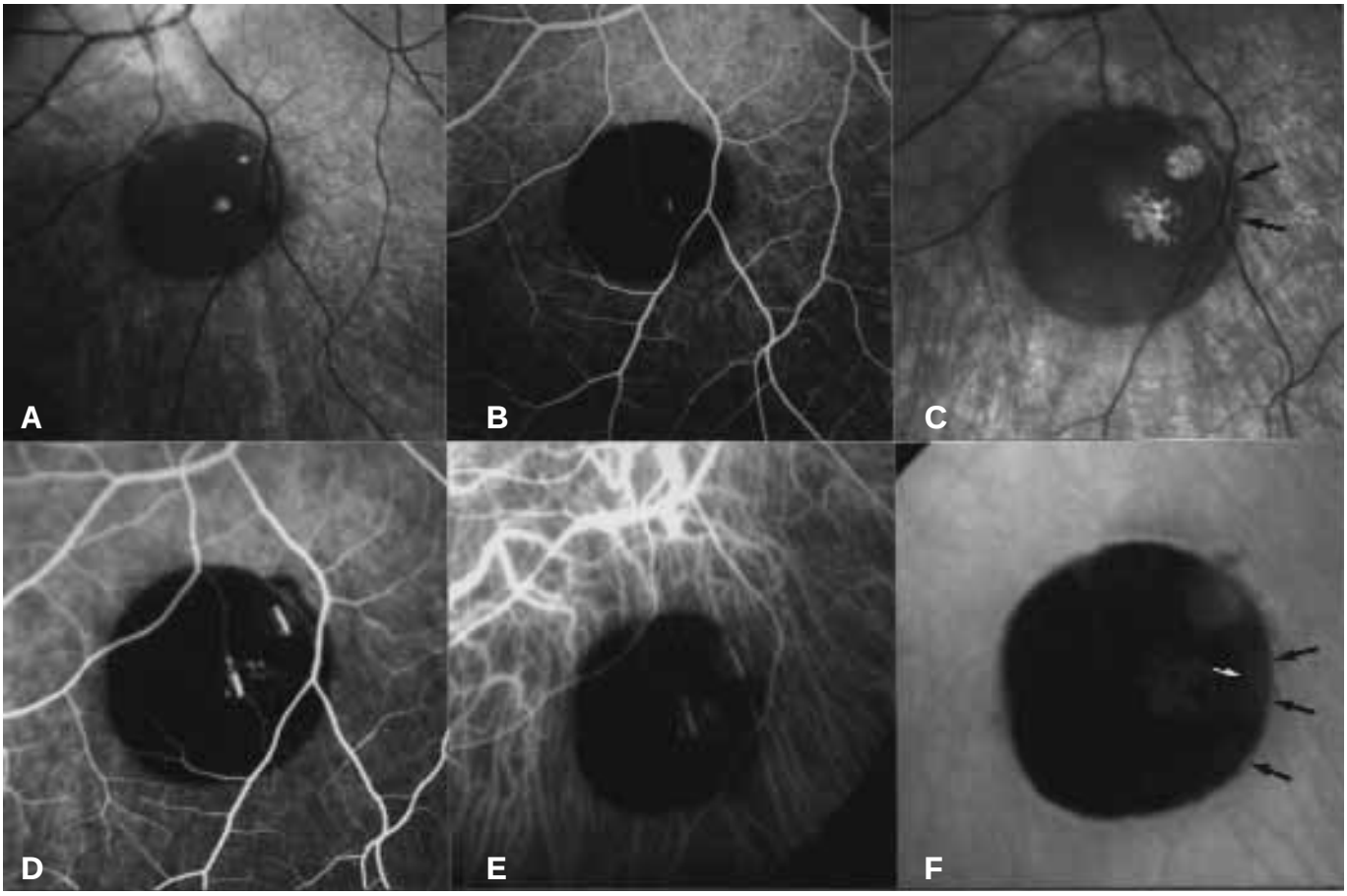

Fig 1 - (A) *Red-free photograph showing a plaque of congenital hypertrophy of the retinal pigment epithelium (CHRPE) with a depigmented halo and two lacunae.* **(B)** *In fluorescein angiography the lesion is hypofluorescent. The halo is barely seen.* **(C)** *Seven years later the lesion enlarged toward the temporal side (on the left). On the nasal side, however, the depigmented halo as well as the hyperpigmented external ring were withdrawn (arrows).* **(D)** *Fluorescein angiography showed masking hypofluorescence of both the main lesion and the external pigmented ring, and transmission hyperfluorescence of the depigmented halo.* **(E)** *Indocyanine green (ICG) angiography during an early phase shows a hypofluorescent area corresponding to the CHRPE; the depigmented halo and the hyperpigmented external ring are invisible. Choroidal vessels are seen through the lacunae.* **(F)** *ICG angiography during a late phase revealed hyperfluorescence of the lacunae as well as of areas not appreciated in ophthalmoscopy or fluorescein angiography (arrows).*

The patient was seen again 7 years later. The lesion had a somewhat different appearance as it enlarged toward the temporal side, while it seemed to be a little less expanded in the nasal side, where the white annulus was withdrawn as well as the external ring of hyperpigmentation (Fig. 1C). The two lacunae enlarged. Fluorescein angiography showed that the borders of the hypofluorescent lesion exactly corresponded to the external pigmented border seen at ophthalmoscopy; the retinal vessels on the plaque had a normal appearance (Fig. 1D). The ICG angiography in an early phase showed a hypofluorescent lesion that coincided to the lesion area located inside the white annulus; the annulus itself and the external hyperpigmented ring were not appreciated (Fig. 1E).

The late phase of ICG angiography better delineated the lesion and the lacunae were clearly seen as zones of transmission hyperfluorescence; in addition, other hyperfluorescent areas similar to those corresponding to the lacunae, that were not evident at ophthalmoscopy or fluorescein angiography, were seen inside the hypofluorescent plaque (Fig. 1F).

DISCUSSION

CHRPE is a unifocal, plaque-like, black thickening of the RPE (1, 2, 4). Generally CHRPE does not cause any visual problem and is casually found during routine fun-

dus examination. The differential diagnosis with choroidal nevus and melanoma is easy on clinical grounds alone. Even if CHRPE is probably present at birth (7), it tends to grow and as much as 83% of cases show some enlargement in a 3-year period (4). Typically, areas of depigmentation (lacunae) appear within the CHRPE with the enlargement of the lesion.

CHRPE cells are full of round granules of melanin (8, 9). Around the border of the CHRPE a more or less complete annulus of depigmentation is often found (10). Histologic study shows that it is formed by elongated cells with longitudinal axes oriented parallel to the lesion's circumference; the depigmented halo was interpreted as contact inhibition between two cell populations: the cells of the CHRPE and the cells of the nearby healthy RPE (9). The annulus of depigmentation is a sign of growth (hyperplasia) of the CHRPE. In addition, a second border, which is hyperpigmented, is seen external to the previous: it is composed of cells of RPE in which a greater amount of lipofuscin is present (9). As the lesion enlarges, the hypopigmented halo advances, changing its position.

The patient we described presented a reduction of the CHRPE on one side, associated with an enlargement on another side, with a general moderate change of the shape of the lesion. Reduction of CHRPE was not previously described, although Chamot et al (3) reported a case in which a depigmented area vanished and then reappeared. This occurrence shows that even if the more frequent case is the progression of the CHRPE toward the normal RPE (4), the opposite, i.e., the progression of normal RPE toward the CHRPE with withdrawal of the hypopigmented halo, is possible.

ICG angiography performed in our patient evidenced some not previously described findings. First, the borders of the CHRPE appear hypofluorescent as can be expected by the masking effect of the melanin present in this lesion, but the halo, as well as the hyperpigmented external border, are invisible in ICG angiography: probably the lipofuscin that accumulates in RPE around the CHRPE is not sufficient to cause a masking effect. These ICG angiographic findings are different from the features we found in fluorescein angiography where CHRPE and hyperpigmented nearby RPE are all completely hypofluorescent. Thus ICG angiography can delineate the borders of the CHRPE more carefully than fluorescein angiography, and especially in cases in which the hypopigmented halo is lacking it shows the exact limits of the CHRPE. In addition, ICG angiography showed the presence of hypofluorescent areas similar to lacunae that were not apparent in ophthalmoscopy or fluorescein angiography. Probably these are zones in which a smaller amount of melanin is present that is not enough to determine a masking effect; thus it is likely that they are areas that are developing lacunae.

In conclusion, ICG angiography is a useful diagnostic test to better understand the various findings and the evolution of CHRPE.

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REFERENCES

1. Reese AB, Jones IS. Benign melanomas of the retinal pigment epithelium. Am J Ophthalmol 1956; 42: 207-12.
2. Buettner H. Congenital hypertrophy of the retinal pigment epithelium. Am J Ophthalmol 1975; 79: 177-89.
3. Chamot L, Zografos L, Klainguti G. Fundus changes associated with congenital hypertrophy of the retinal pigment epithelium. Am J Ophthalmol 1993; 115: 154-61.
4. Shields C, Mashayekhi A, Ho T, Cater J, Shields JA. Solitary congenital hypertrophy of the retinal pigment epithelium. Clinical features and frequency of enlargement in 330 patients. Ophthalmology 2003; 110: 1968-76.
5. Claery PE, Gregor Z, Bird AC. Retinal vascular changes in congenital hypertrophy of the retinal pigment epithelium. Br J Ophthalmol 1976; 60: 499-503.
6. Cohen SY, Quentel G, Guiberteau B, Coscas GJ. Retinal vascular changes in congenital hypertrophy of the retinal pigment epithelium. Ophthalmology 1993; 100: 471-4.
7. Champion R, Daicker BC. Congenital hypertrophy of the pigment epithelium. Light microscopic and ultrastructural findings in young children. Retina 1989; 9: 44-8.
8. Wirz K, Lee WR, Coaker T. Progressive changes in congenital hypertrophy of the retinal pigment epithelium. An electron microscopic study. Graefes Arch Clin Exp Ophthalmol 1982; 219: 214-21.
9. Lloyd WC III, Eagle RC Jr, Shields JA, Kwa DM, Arbizo VV. Congenital hypertrophy of the retinal pigment epithelium. Electron microscopic and morphometric observations. Ophthalmology 1990; 97: 1052-60.
10. Kurz GH, Zimmerman LE. Vagaries of the retinal pigment epithelium. Int Ophthalmol Clin 1962; 2: 441-64.