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Solitary congenital hypertrophy of the retinal pigment epithelium: A case report

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Abstract
Congenital hypertrophy of the retinal pigment epithelium (CHRPE) is a benign pigmented lesion located at the retinal pigment epithelium (1-4). Hyper pigmented fundus lesions are divided into three different forms: solitary, grouped and multiple (5). The solitary form often appears as a flattened pigment spot, generally considered to be benign, with no potential for malignant growth or transformation (6).

Keywords: congenital hypertrophy of the retinal pigment epithelium, retinal pigment epithelium, malignant transformation

Introduction

Observation

We report a case of a 40-year-old male patient with no specific history. Admitted for loss of visual acuity. Ophthalmologic examination revealed an AVL at 8/10 ODG, which increased to 10/10 after correction. Examination of the anterior segment was normal. Funduscopy is without particularity on the left, on the right eye we found an isolated hypo-pigmented lesion in supero-temporal, of large size of gray-brown color surrounded by a double border, first clear then pigmented scalloped with a central gap, without other associated lesions (Figure 1). Fluorescein angiography shows a hypofluorescent lesion with a window effect next to the central halos (Figure 2). The clinical and angiographic aspect made it possible to eliminate the choroidal melanoma and the other pigmented tumors of the FO, and to retain the diagnosis of solitary HCEPR.

The course of action was to prescribe an orthoptic correction with annual monitoring.

On a 1 year follow-up, the evolution was stationary (Figure 3).

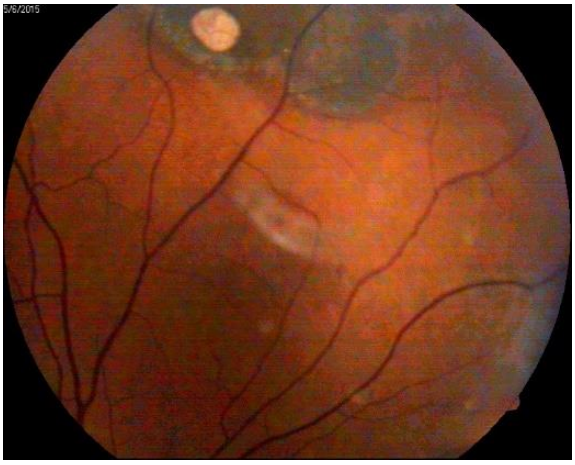

Fig 1: Solitary congenital enlargement of the super-temporal retinal pigment epithelium in retinography.

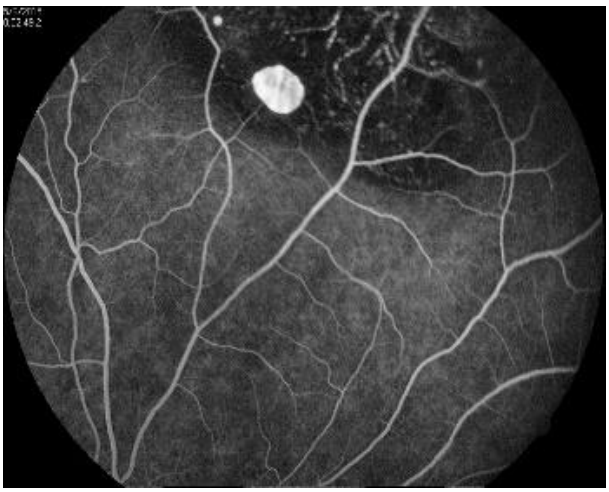

Fig 2: Congenital hypertrophy of the pigment epithelium in fluorescin angiography with the presence of gaps.

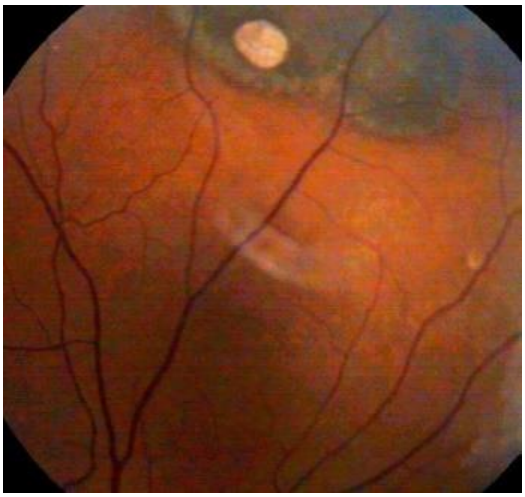

Fig 3: Congenital hypertrophy of the pigment epithelium in retinography after one year of evolution.

Discussion

Congenital enlargement of the retinal pigment epithelium is a benign lesion of the fundus, which often appears as a flat, well-defined pigmented patch [7]. This finding is almost always mild and asymptomatic [8]. Its prevalence is highly variable ranging from 0.3 to 40% of the population [9, 10]. It is more common in women with a 2: 1 ratio [11]. In the solitary form, the pigmented lesion is isolated, more frequently observed in the temporal retina than in the nose [12], their shape is oval or round with smooth and regular margins in 2/3, and geographic with scalloped margins in the 1/3 of the cases).

The lesions were also distributed almost equally between those with (52%) and without (48%) atrophic gaps [13]. Approximately 88% of the lesions are pigmented, brown in about 1/2 of the cases, black in 1/3 of the cases and gray for the rest. Rarely, it can be depigmented.

In angiography, CHRPE has been shown to be hypoauto fluorescent and uniformly dark, explained by the histopathological absence of lipofuscin in strongly and uniformly pigmented RPE cells. Gaps with slightly higher autofluorescence [14]. The variation of iso to hyperautofluorescent vacancies may depend on the thickness of the intermediate choroid, particularly in individuals strongly pigmented with a darker choroid [15].

Drusens and lumps of pigment do not develop on HCEPR lesions and their presence should suggest melanoma [16]. The characteristics suggesting a tumor of the RPE are type of neo retinal vessels feeding the tumor and exudates lipo-protein yellow retinal and under retinal, the drusens and the clods pigmented, the raised and hyper echogenic character of pigmentation with ultrasound, and the association with exudative retinal detachment, hyalitis, vitreo-retinal traction [17].

Bilateral or multiple CHRPE are disseminated over the entire fundus in both eyes and can be associated with the Gardner's syndrome or familial adenomatous polyposis (FAP). [18].

Conclusion

Congenital hypertrophy of the pigment epithelium (HCEPR) or Sutton retinal nevus (halo nevus) is a benign lesion of the eye fundus that often appears as a flattened pigment spot. The risk of malignant transformation, which is certainly exceptional but possible, hence the interest of annual monitoring.

Declaration of interest

The authors declare having no interest links.

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