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Childhood idiopathic bilateral macular dystrophy (New entity)

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Abstract

Macular dystrophies correspond to hereditary diseases affecting the macular region and associating primary abnormalities of the pigment epithelium and the sensory retina, which appear in children or young adults, the aim of this work is to report an atypical case of idiopathic bilateral macular dystrophy observed in a 9-year-old child. It is a new clinical entity of macular dystrophies in children, which remains rare and can be complicated by choroidal neovascularization.

Keywords: idiopathic bilateral, macular dystrophy, choroidal neovascularization

Introduction

Macular dystrophies correspond to hereditary diseases affecting the macular region and associating primary abnormalities of the pigment epithelium and the sensory retina ^[1]. Which appear in children or young adults ^[2].

The examination of the fundus will direct the complementary examinations necessary to confirm the diagnosis whether it is fluorescein angiography, optical coherence tomography, electroretinogram and / or electrooculogram.

The aim of this work is to report an atypical case of idiopathic bilateral macular dystrophy observed in a 9-year-old child.

Observation

One child, born in 2009, presented at the ophthalmology clinic in October 2018 for a bilateral decrease in visual acuity.

On examination, the visual acuity of the right eye was 4/10 not improvable while that of the left eye was 2/10 not improvable.

In addition, the anterior segment was normal and a clear vitreous.

Examination of the fundus revealed a slightly yellowish-white pre-retinal veil with a few perilesional pigmentary lumps and a lower juxtafoveal retrohyaloid hemorrhage in the right eye (Figure 1), while on the left side there was - slightly yellowish retinal with some perilesional pigmented mounds without visible retrohyaloid hemorrhage (Figure 2).



Fig 1: Macular dystrophy with retrohyaloidal hemorrhage

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Fig 2: Slightly yellowish retina with some perilesional pigmented mounds

Fluorescein angiography showed a perimacular area of chorioretinal atrophy in both eyes and with a lower perifoveal subtraction image in the right eye while the retinal vessels were normal in appearance (Figure 3-4).

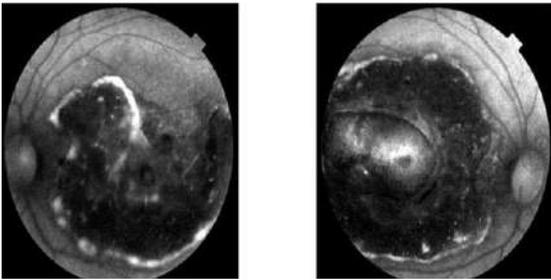


Fig 3 & 4: Fluorescein angiography showed a perimacular area of chorioretinal atrophy in both eyes

In OCT, the lesion's architecture is anarchic and delimits choroidal zones, humped, repelling the retina and responsible for subretinal exudation (Figure 5).



Fig 5: OCT, the retina's architecture is anarchic and subretinal exudation

The young patient subsequently received an injection of bevacizumab in the right eye, and pre-retinal hemorrhage was observed to disappear within a month (Figure 6).

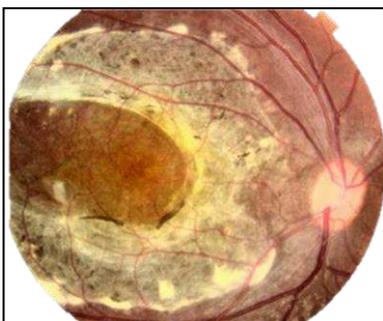


Fig 6: Macular dystrophy without retrohyaloidal hemorrhage

The evolution is marked by the stabilization of uncomplicated lesions after six months of control.

Discussion

Macular dystrophies are often responsible for a decrease in visual acuity and are a major cause of profound visual impairment and affect all ages of life [3].

The examination of the fundus is the key element of the diagnosis and the clinical interview and family history will guide the complementary examinations that must be requested.

Macular dystrophies can be classified into several entities, such as Stargardt's disease, or fluorescein angiography with retinal periphery radiographs for choroidal silence or dystrophy. cones in which the key examination remains the electroretinogram, associated with fluorescein angiography, whereas in case of X-linked macular retinoschisis, OCT is without doubt the most contributory and sufficient examination at diagnosis [4].

There are many other macular dystrophies of genetic origin most often syndromic, forming part of a particular general context already known [5].

In these cases, the involvement of the macula is not isolated and most often occurs in the context of syndromic retinal dystrophy whereas this case reported in this work constitutes an isolated attack of the macular region and does not synonymous with the already known clinical entities hence its name: childhood idiopathic bilateral macular dystrophy and which can be complicated by choroidal neovascularization.

Conclusion

Childhood Idiopathic bilateral macular dystrophy is a new clinical entity of macular dystrophies in children, which remains rare and can be complicated by choroidal neovascularization.

Conflict of interest

No

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