

Bilateral Chorioretinal Scars in a Child – Case Report

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ABSTRACT

The eye manifestations of intrauterine infections are multiple, but chorioretinal scars and/or active chorioretinitis are the most frequently reported. When associated with other systemic manifestations of the infection, the diagnosis becomes more obvious, but when eye involvement is the only abnormality, etiology often remains uncertain. We are presenting the case of an 8-year-old female patient whose fundus lesions revealed an unusual choroidoretinopathy, associated with cataract in one eye. Her general examination and her medical history were unremarkable. Blood test for multiple pathogenic agents (HIV, hepatitis Band C, Toxoplasma, Toxocara, Borrelia, CMV, Epstein-Barr virus) were negative. Complete blood count, ESR, fibrinogen and CRP are all within normal limits. Plasma ornithine levels were in normal range (ruling out a diagnosis of gyrate atrophy). The chorioretinal lesions discovered in this 8 years old child may be caused by an acquired or a congenital infection. As in most similar cases, the clinical aspect, the medical history and the blood work did not help too much in establishing the moment of the infection or the causative agent. A complete clinical, structural and functional base-line evaluation is however mandatory when facing such a case. Periodic follow-up is recommended in order to assess the evolution of the disease.

Keywords: chorioretinal scars, cataract, children, intrauterine infection

INTRODUCTION

Ocular manifestations of intrauterine infections are multiple, but chorioretinal scars and/or active chorioretinitis are the most frequently reported. Congenital cataract also suggests a possible intrauterine infectious etiology and is usually discovered with other intraocular findings. Chorioretinal scarring is in a high percentage of cases due to congenital infection with *Toxoplasma gondii*, Herpes simplex virus, Lymphocytic choriomeningitis virus and West-Nile virus. When associated with other systemic manifestations of the infection, the diagnosis

becomes more obvious, but when eye involvement is the only abnormality, etiology often remains uncertain. □

CASE REPORT

We present the case of an 8-year-old girl whose parents noticed a moderate decrease in her visual acuity (VA) (she sits closer to the TV screen) in the last 3 weeks.

Her medical history is unremarkable.

The general clinical examination reveals a weight of 30 kg, 120 cm of height, good general status and a normal physical and mental development, according to her age.

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Her uncorrected distance VA was 0.3 in the right eye (OD) and 0.6 in the left (OS); her best corrected visual acuity was 0.4 (OD) and 0.8 (OS).

Refraction: OD:-0.5Dsf ^ -4.25Dcyl 6 degrees; OS: -0.25Dsf ^ -1.75Dcyl 168 degrees.

The slit lamp examination of the anterior pole reveals posterior subcapsular lens opacities in the OD without any other pathologic findings. There are no signs of inflammation of the anterior pole.

Fundus examination shows in both eyes (OU) a round, well contoured optic nerve head; multiple patchy atrophic chorioretinal lesions, some with colobomatous like aspect and pigmentary disturbances in the peripheral retina following a circumferential pattern and also well demarked chorioretinal scars situated in the paramacular region (Figure 1, 2).

At this moment our diagnosis leads to: Sequelae of chorioretinitis; myopic astigmatism. Blood tests, functional and morphological visual tests are recommended.

Blood samples for HIV, hepatitis Band C, Toxoplasma, Toxocara, Borrelia, CMV, Epstein-Barr virus and ASLO are negative. The result for Herpes simplex IgM was inconclusive C.O.=0.42 and D.O.=0.33. Complete blood count, ESR, fibrinogen and CRP are all within normal limits. Plasma ornithine levels were in

normal range (ruling out a diagnosis of gyrate atrophy).

Goldman perimetry reveals defects that correspond to the chorioretinal scars.

OD: the point of fixation is slightly blurry; central relative scotoma, predominantly temporal up to 23 degrees; the visual field for colors is concentrically reduced; no relative scotoma for colors.

OS: visual field is concentrically reduced, with wedge-shaped defect nasal superiorly and nasal inferiorly; visual field for colors is also reduced, for green more than for red, with no relative scotoma.

The optical coherence tomography (OCT) shows flattening of the foveal region and macular scarring, without revealing any subretinal fluid.

Flash and Pattern VEP show normal values, while the electrophysiologic tests (ERG) reveal pathologically decreased values in both eyes. The maximal registered amplitude is for OD=145 nanoV and for OS=146 nanoV, estimating the extent of the damaged retina.

Considering the clinical aspect of the lesions, and the absence of inflammatory signs and exudation we did not perform a fluorescence angiogram.

The diagnosis in this case was OU: Chorioretinal scars with unknown etiology (possible

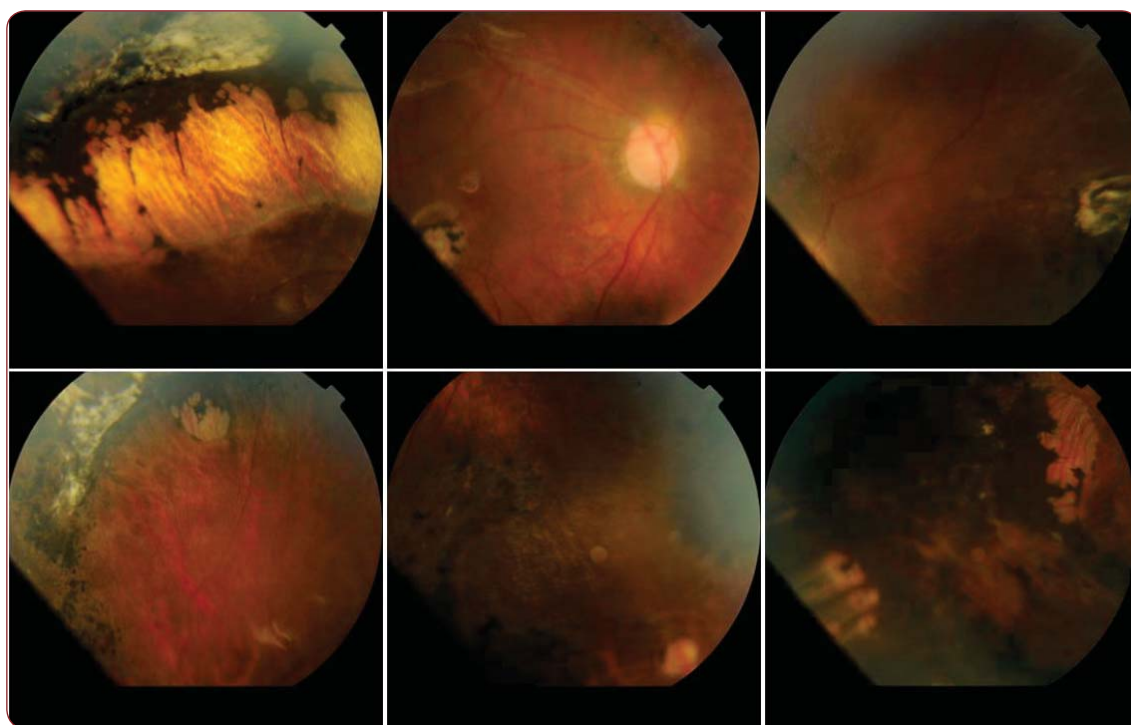


FIGURE 1. Fundus photography of right eye.

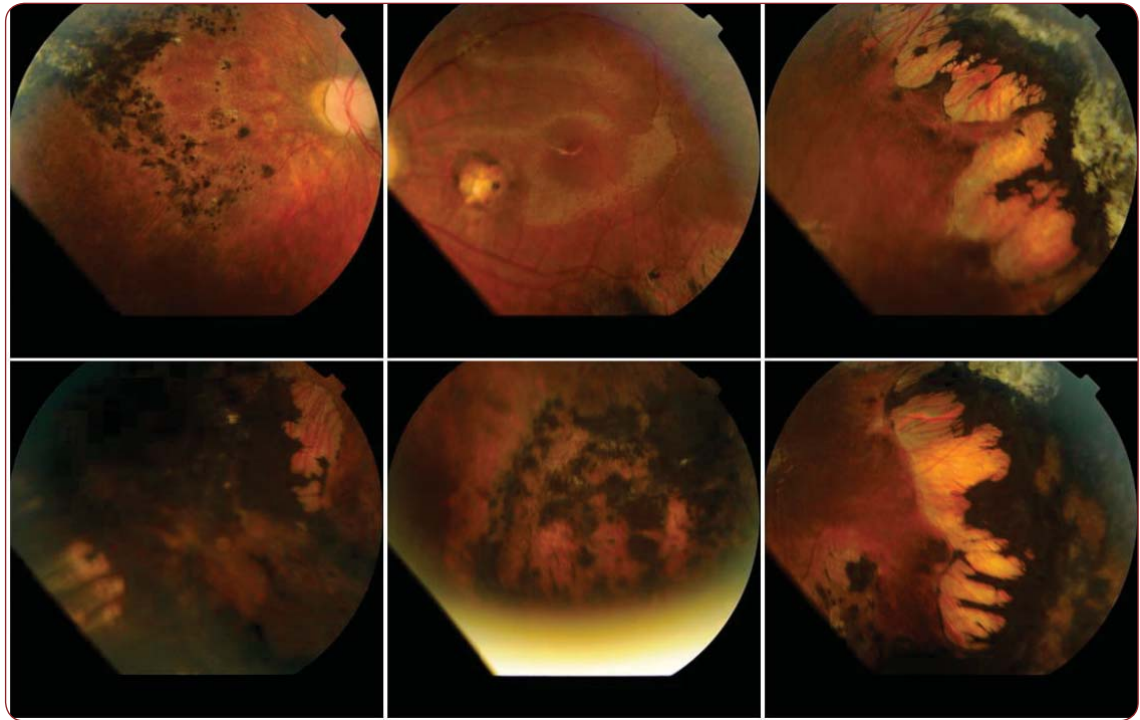


FIGURE 2. Fundus photography of left eye.

intrauterine infection). Myopic astigmatismus. OD: Cataract. $\square$

TREATMENT

An etiological treatment does not come in question since we have not established the cause of these chorioretinal lesions, that ap-

pear to be non-active. Regarding the refractive error, glasses were prescribed OD: $-0.5\text{Dsf} \wedge -2.00\text{Dcyl } 5 \text{ degrees}$ and OS: $-1.00\text{Dcyl } 0 \text{ degrees}$ and a one month check-up was established. We explained the necessity of following up the lens opacities and the chorioretinal lesions. $\square$

Agent	Eye manifestations of congenital infection
Toxoplasma	Chorioretinal scars, microcornea, cataract, optic atrophy, microphthalmia, retinitis, retinal detachment, vitritis, strabismus, nystagmus, phthisis
Rubella	Dacryostenosis, endotheliopathy, glaucoma, keratoconus, iris hypoplasia, chronic granulomatous iridocyclitis, iris coloboma, anisocoria, posterior synechiae, persistent papillary membrane, mesodermal dysgenesis, cataract, salt and pepper retinopathy, primary optic atrophy, microphthalmos, microcornea
Cytomegalovirus	Corneal opacity, chorioretinitis, optic nerve hypoplasia, optic nerve coloboma, optic atrophy, cyclopia, anophthalmia
Herpes simplex virus	Conjunctivitis, keratitis, iridocyclitis, iris atrophy, posterior synechiae, cataract, retinitis, chorioretinitis, chorioretinal scars, optic neuritis, optic atrophy optica, microcornea, microphthalmia
Lymphocytic chorioretinitis Virus	Chorioretinal scars, optic atrophy, nystagmus, optic nerve dysplasia, eso or exotropia, microphthalmos, cataract, retinitis
West-Nile Virus	Chorioretinal scarring
Treponema pallidum	Corneal opacity, uveitis, cataract, glaucoma, pigmentary retinopathy, optic atrophy, Argyll-Robinson pupil
Varicella-Zoster Virus	Chorioretinitis, hypoplasia and atrophy of the optic nerve, congenital cataract, Horner's syndrome
HIV	CMV retinitis, toxoplasmic chorioretinitis
Epstein-Barr virus	Congenital cataract

TABLE 1. Eye manifestations of Intrauterine Infections (MB Mets and MS Chhabra).

DISCUSSION

The difficulty in establishing an etiological diagnosis is on the one hand because of the many possible causative agents and on the other hand because of the low sensitivity and specificity of the blood tests. Sometimes the absence of the IgG antibodies for one specific agent makes the diagnosis of infection improbable without being able to completely exclude it. Tests that determine the specific antigen in the serum or tissues could clarify the diagnosis, but they are more difficult to perform.

Although in our case the blood tests for Toxoplasma were negative, we could not exclude this causative agent (1,2). Other agents that can produce similar chorioretinal lesions are the lymphocytic choriomeningitis virus (3) or the West-Nile virus, but serological testing was not performed.

The clinical aspect of the fundus with vast chorioretinal atrophic lesions, massive pigmentary changes and with no sign of local or general inflammation, associated with subcapsular cataract in OD lead to the hypothesis of a sequelar chorioretinitis, that occurred most probably during the intrauterine life (4). The elements that support this theory are:

- young age (child, 8-year-old)
- bilateral lesions
- absence of inflammatory episode (adenopathy) in the medical history
- association with cataract

Symptoms related to the decrease in VA are difficult to assess and report in very young age groups.

Intrauterine infections are often summarized by the mnemonic TORCH (Toxoplasma gondii, "others", rubella, CMV and herpes sim-

plex virus). "Others" includes Treponema pallidum, Varicella-Zoster Virus, Epstein-Barr virus, HIV, West-Nile virus and the Lymphocytic choriomeningitis virus. These agents produce a mild form of illness in the mother, in comparison to the major impact that they can have on the developing fetus. In the majority of cases the risk of fetus contamination and the severity of the lesions are dependent on the moment of infection, the week of gestation. The risk of transmitting the infection transplacentally grows together with the age of gestation, but the impact on the fetus decreases towards the end of the pregnancy.

Chorioretinal scarring has been reported as sequelae in a variety of congenital infections (Table 1) (5), most often a Toxoplasma infection (6,7), but also Lymphocytic choriomeningitis virus, Cytomegalovirus or West-Nile virus infections. These last ones are often underestimated as causative agents (8,9). □

CONCLUSION

The chorioretinal lesions discovered in this 8 years old child may be caused by an acquired or a congenital infection. As in most similar cases, the clinical aspect, the medical history and the blood work do not help much in establishing the exact moment of the infection or the causative agent. A complete clinical, structural and functional base-line evaluation is however mandatory when facing such a case. Periodic follow-up is recommended in order to assess the evolution of the disease (reactivation possible).

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