X-linked Retinoschisis Discovered after Congenital Cataract Surgery

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ABSTRACT

X-linked retinoschisis (XLRS) is a rare, hereditary, and bilateral vitreoretinal degeneration, associating maculopathy and vitreoretinopathy. We report the case of X-linked retinoschisis diagnosed after congenital cataract surgery in a child.

Keywords: Retinoschisis, Optical Coherence Tomography, vitreoretinal degeneration.

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I. INTRODUCTION

X-linked retinoschisis is a rare, hereditary, and bilateral vitreoretinal degeneration, associating maculopathy and vitreoretinopathy.

II. CASE REPORT

We report the case of X-linked retinoschisis diagnosed in the aftermath of a child operated on for congenital unilateral OD cataract.

Child aged 5 years, followed for unilateral congenital cataract of the operated OD, and implanted in the sac with post rhexis and ant vitrectomy. The evolution was marked by a deep amblyopia of the average OD of the OG despite an OC and a TTT of well-conducted amblyopia.

The anterior segment was normal on the left and on the right: the IOL was in place, no secondary cataract and the intra ocular pressure was normal.

In the ocular fundus:

Right eye: there is an inter-papillo-macular tractional membrane with tortuosity and stretching of the vessels. (Fig. 1).

Left eye: Wheel raillon aspect (Fig. 2).

OCT: more marked and more extensive macular retinoschisis on the right (Fig. 3).

ERG: wave « b » is negative and wave « a » is positive. (Fig. 4).

Fig. 1. Right eye: An inter-papillo-macular tractional membrane with tortuosity and stretching of the vessels.

Fig. 2. Left eye: Wheel raillon aspect

Fig. 3. More marked and more extensive macular retinoschisis on the right eye.

Fig. 4. A negative wave « b » and a positive wave « a ».

III. DISCUSSION

The most common mode of discovery of XLRS is decrease in Visual Acuity [1], it often affects males with rare cases in females. [2,3].

It is characterized by still present bilateral foveal retinoschisis, peripheral retinoschisis, perivascular cuffs, vitreous veils, and pigmento-atrophic changes [4]. Other anomalies can be associated: axile hyperopia, strabismus, and cataract [5].

The electroretinogram is still disturbed and shows a pattern characteristic of the condition [6].

The appearance on OCT of XLRS is very evocative with a superficial cleavage in the form of hyporeflective cavities which are particularly predominant in the macula [7].

The evolution is marked by a stable VA during the first 2 decades [8].

IV. CONCLUSION

X-linked retinoschisis is a rare pathology, its clinical presentation is variable, and the diagnosis is difficult in children, but can be precipitated by the occurrence of complications. The forms diagnosed in children are more often severe and the prognosis remains poor regardless of the age of diagnosis, hence the interest of genetic counseling

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