



Bilateral Lens Dislocation In Congenital Aniridia: A Case Report

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Abstract: The uncommon disease aniridia, caused by a mutation in the gene PAX6, affects the whole ocular globe. We present the case of a 37-year-old woman who has bilateral eye pain and is malvoyant, known to have an aniridia, and hypoplasie of the fovea.

Index Terms - Congenital aniridia, Lens dislocation, Foveal hypoplasia, Cataract.

I. INTRODUCTION

Congenital aniridia is a rare genetic disorder associated with mutations in the PAX6 gene, the major gene for eye development [1]. Lens changes [1] in aniridia can include changes in size, shape, location and transparency [2]. The purpose of this presentation is to demonstrate how the lens changes during congenital aniridia and the therapeutic management.

II. CLINICAL CASE

We report the case of a 34 year old woman, visually impaired, consulted for a bilateral ocular pain installed since 3 days, the ophthalmological examination reveals a visual acuity reduced to a luminous perception, a nystagmus, a diffuse conjunctival hyperaemia, a transparent cornea and a crystalline dislocation with cataract and an aniridia at the level of the 2 eyes (figure 1), the IOP is 24 mmhg at the right eye and 27 at the left eye, the fundus of the eye is inaccessible. The patient benefited from an ocular ultrasound in B mode and found a flat retina in both eyes. The patient benefited from a local hypotonizing treatment and a manual extraction of the lens with an anterior vitrectomy first on the left eye and then on the right eye 1 month after the 1st surgery. The evolution was marked by an improvement of the visual acuity with finger movements.

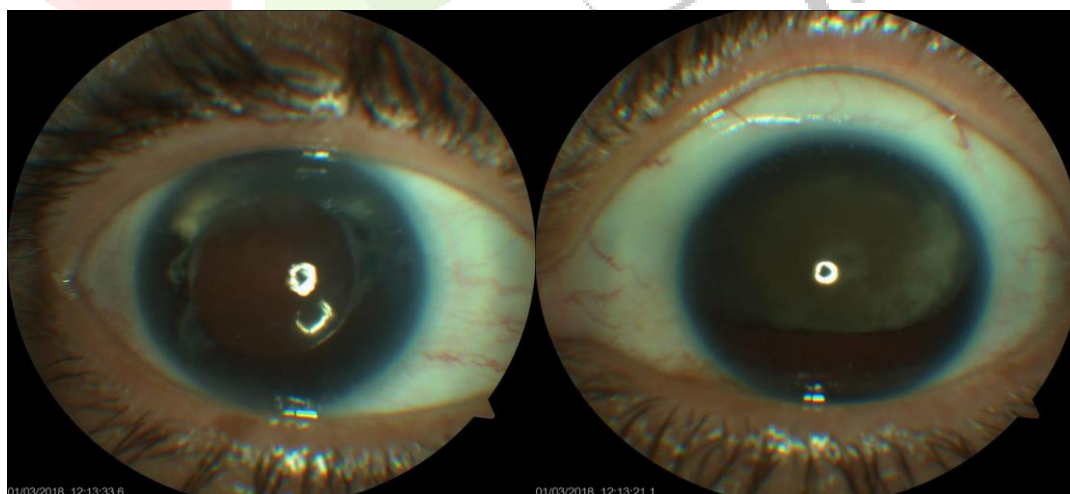


Figure 1: dislocation of the cataractous lens during congenital aniridia.

III. DISCUSSION

Congenital aniridia is a genetic eye condition that affects the eyeball and is defined in part by the partial or complete absence of the iris [1]. The prevalence ranges from 1 in 40,000 to 1 in 96,000 births, but it can be underreported [3]. Additionally, a syndromic aniridia might be formed when it is connected to a number of systemic signs.

This condition links a number of ocular disorders: glaucoma, cataract, Neovascularization with corneal opacities, Nystagmus in addition to foveal hypoplasia, Hypoplasia of the papilla and optic nerve.

The symptoms of the disease can differ from one person to the next, even within the same family. The most reliable signs are iris-like abnormalities and foveolar hypoplasia. The adult patients with severe visual impairment are frequently regarded as having legal adult-age cécité and have very compromised visual prognoses [4].

The removal of the cataracted crystal is not a consistent process during congenital aniridia but rather depends on visual acuity and the appearance of a luxation [2].

IV. CONCLUSION

In the majority of cases of aniridia, the lens becomes progressively opaque over time and develops into a cataract. Other complications may occur such as subluxation of the lens, glaucoma and others.

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