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Management of Ectopic Lens Cataract by Lensectomy, Vitrectomy and Iridial Fixation Implantation (Artisan): About 02 Eyes of a Child Admitted to CHU IOTA

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ABSTRACT

Objective: To report two cases of ectopic cataracts in the context of Marfan's syndrome diagnosed and treated by vitrectomy, lensectomy associated with iridial fixation implantation with good functional results. Case report: This 16-year-old patient was admitted to the African Tropical Ophthalmology University Hospital Center (CHU-IOTA) for a decline in visual acuity since childhood, and was diagnosed as having Marfan syndrome with bilateral ectopic cataract, taking into account the Berlin anatomical criteria. Apart from ocular and musculoskeletal involvement, cardiac, pulmonary, biological and biochemical examinations were normal. After surgical management with vitrectomy, lensectomy and iridofixation implantation (artisan), initial visual acuity counts fingers at 1 meter and at 2 meters in the right and left eyes respectively, improved to 5/10 without correction in both eyes and to 10/10 with optical correction made after postoperative follow-up. Conclusion: Iris-fixation implantation after vitrectomy and lensectomy is an effective surgical technique that can guarantee good functional results in children with ectopic cataracts due to Marfan syndrome.

Keywords: Ectopy, lens, Marfan, vitrectomy, lensectomy, Iris implantation, visual acuity, child.

INTRODUCTION

Lens ectopia is a congenital displacement of the lens, bilaterally and symmetrically, due to a zonular anomaly. It is often associated with syndromic diseases such as Marfan syndrome. The latter is an autosomal dominant disorder with an estimated frequency of 1/5,000 individuals [1], diagnosis of which is based on certain criteria, and management is multidisciplinary. We report here two cases of ectopic lens cataracts of Marfan syndrome diagnosed and treated by vitrectomy, lensectomy associated with scleral fixation implantation with good postoperative functional results.

CLINICAL CASE

This was a 16-year-old student wearing corrective lenses for myopia, with a family history of his mother's long-sighted morphotype. He was seen in ophthalmology consultation at CHU-IOTA for a progressive drop in visual acuity since childhood. On external examination, ophthalmology revealed moderate leukocoria with decreased red reflex of the fundus in the bruckner test, and good oculomotricity. On physical examination, the right eye showed uncorrected visual acuity at 1 metre and 4/10 with correction, ectopia with lens opacity (opaque spherophakia, displaced superiorly, abnormally visible zonular fibres elongated inferiorly, showing a bright red crescent on retro-illumination), intraocular pressure at 10 mmHg and normal fundus. Examination of the left eye revealed uncorrected acuity at 2 metres

and 5/10 with corrected glasses, with the same signs in the anterior segment and fundus (Figure 1).

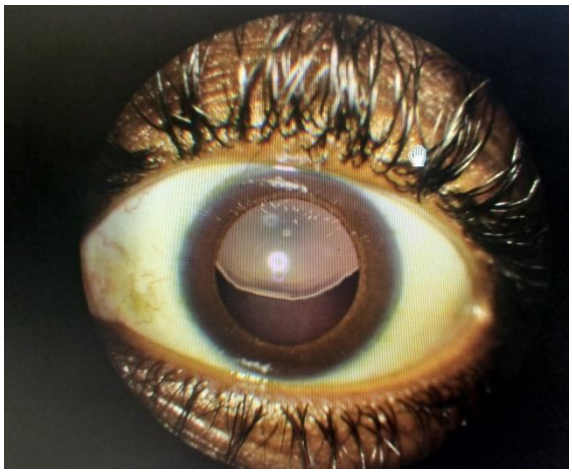

Figure 1: Ectopia of the lens of the left eye.

General examination revealed musculoskeletal damage (dolichostenomelia with a height of 185 cm, arachnodactyly with walker Murdoch's thumb sign, scoliosis and anteroposterior dolichocephaly). Cardiovascular examination was normal, with no mitral insufficiency or aortic dilatation. Pulmonary examination was normal. Biological and biochemical tests (CBC, ESR, cholesterol, blood glucose, creatinine, azotemia, uricemia, ALT/ASAT, TSHus, blood ionogram) were also normal.

The diagnosis of Marfan syndrome with bilateral cataractous lens ectopia was made according to the 1986 Berlin criteria (involvement of two systems + family history of Marfan) and surgical management by vitrectomy, lensectomy and iridial fixation implantation (artisan) was proposed. The right eye was operated on first, and the left eye after post-operative follow-up of the first eye. In the operating room, after the 03 approaches had been set up, an anterior vitrectomy, a vitreotome lensectomy and an iridium-fixing implant (18.5 dioptres in the right eye and 20 dioptres in the left eye) were performed (Fig. 2).

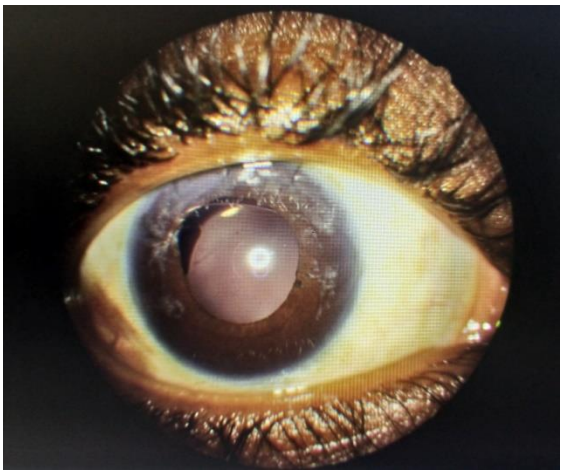

Figure 2: Iris fixation implant in the right eye.

The immediate post-operative course was marked by moderate diffuse conjunctival hyperemia and mild corneal edema on Day 1. After treatment with eye drops (dexamethasone phosphate-Neomycin sulfate, indomethacin), ophthalmic ointment (dexamethasone-oxytetracycline) and tablets (fucidic acid and prednisone), ocular examination was normal on D17, with uncorrected visual acuity of 5/10 in both eyes and corrected acuity of 10/10 on D37 postoperatively in each eye.

DISCUSSION

Our patient with Marfan syndrome presented with bilateral ectopia of the lens. Indeed, nearly 6 out of 10 people with Marfan syndrome present with lens displacement in one or both eyes [2]. According to a Paris study by The Parc et al, lens ectopia is the major sign of ocular damage in 60-80% of cases [1]. Other signs of ocular damage may include axial myopia with high values above -7 dioptres, as in the case of our patient wearing myopia lenses, central corneal flattening, accommodation disorders and retinal detachment, a classic complication of severe myopia.

In our case, there was also musculoskeletal involvement. According to the literature, this is the main feature of Marfan syndrome, with dolichostenomelia, arachnodactyly, joint hypermobility, scoliotic deformity, acetabular protrusion, pectus carinatum or pectus excavatum thorax deformity, anteroposterior axis dolichocephaly, micrognatism and malar hypoplasia. Our patient had a suggestive dolichostenomelic morphotype, with a height of 185 cm. The Parc et al. had found in their study a height of 185+/- 10 cm, and this large size concerns 90% of subjects with Marfan's disease. [1]

Preoperative uncorrected visual acuity was less than 1/10 in both eyes of our patient. According to a study conducted at the ophthalmology department of CHU Mohammed VI in Marrakech from 2012 to 2018 on 48 eyes, this acuity was less than or equal to 1/10 [3]. Our preoperative corrected visual acuity was 0.5 (5/10). This finding is similar to the study by The Quoy et al. Who found a mean preoperative visual acuity of 0.5 [4].

The diagnosis of Marfan syndrome was made on the basis of our patient's suggestive morphotype and the 1986 Berlin diagnostic criteria, which stipulate that in the case of involvement of two systems in a person with a direct relative with Marfan, the diagnosis of this syndrome can be confirmed. It should be noted that in the absence of a direct relative with Marfan syndrome, skeletal involvement and involvement of two other systems are required before a diagnosis can be made [1]. These criteria were revised in 1996 by De Paepe et al., and other authors are now talking about the contribution of molecular biology to the diagnosis of Marfan syndrome. These include the protein test (possible by skin biopsy, with study of fibroblasts for identification of modified fibrillin) and the molecular test itself for the FBN1 gene of this fibrillinopathy. However, these two tests alone cannot confirm or rule out Marfan syndrome, as other fibrillinopathies may give the same results. Their indirect contribution is only important when there is a strong clinical suspicion of Marfan syndrome. [1]

The indication for treatment was surgical in our patient, with the technique of vitrectomy, lensectomy (phacophagy) and iridial implantation. In the study by Abou El Houdou et al. In Morocco, the surgical indication was retained in all their patients, and the technique consisted of phacophagy with anterior vitrectomy in 64% of cases [3]. The Quoy et al. Also recommended

surgical treatment for their patients with crystalline ectopia, but the technique was posterior vitrectomy combined with scleral fixation implantation [4]

Postoperative functional results were good in our patient, with uncorrected visual acuity of 5/10 in both eyes and corrected visual acuity of 10/10 with glasses. Our results are comparable to those of Abou El Houdou et al. Who also had satisfactory functional results after surgery in 40% of cases, with a better corrected visual acuity of 5/10 [3].

In the study by The Quoy et al. The mean post-operative visual acuity was 0.8 (8/10) at 6 months follow-up [4].

CONCLUSION

The management of Marfan syndrome is multidisciplinary. Iris-fixed implantation after vitrectomy and lensectomy is an effective surgical technique that can guarantee good functional results in children with ectopic cataracts of Marfan syndrome.

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