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Phacoanaphylactic by dislocation of the lens in the Marfan Syndrome

Facoanafilaxia por cristalino mergulhado na Síndrome de Marfan

Breno Barreto Ribeiro[1] https://orcid.org/0000-0002-7330-1440
Grazziella Acácio e Almeida[2] https://orcid.org/0000-0001-9965-6402
Marcela Caetano Bastos[3] https://orcid.org/0000-0003-0663-0460
Luciano Pimenta de Figueiredo[4] https://orcid.org/0000-0003-3539-2844
Luciano Sólia Násser[5] https://orcid.org/0000-0002-0864-6047

Abstract

Marfan syndrome is an autosomal dominant inheritance disease that affects connective tissue with phenotypic manifestations involving the skeletal, cardiovascular and ocular systems. The main ocular manifestations are the subluxation of the lens, myopia and retinal detachment. The aim of this article was to report the clinical and surgical management of a patient with Marfan syndrome with luxated lens for the vitreous cavity and who developed a severe phacoanaphylactic reaction characterized by severe secondary glaucoma and corneal decompensation.

Keywords: *Marfan Syndrome; Glaucoma/complications; Vitrectomy*

Resumo

A síndrome de Marfan é uma doença de herança autossômica dominante e que afeta o tecido conjuntivo com manifestações fenotípicas que envolvem os sistemas esquelético, cardiovascular e ocular. As principais manifestações oculares são a subluxação do cristalino, a miopia e o descolamento da retina. O objetivo deste artigo foi relatar a conduta clínico-cirúrgica de um paciente portador da síndrome de Marfan com cristalino luxado para a cavidade vítrea e que evoluiu com severa reação facoanafilática caracterizada por um glaucoma secundário severo e descompensação corneana.

Descritores: Síndrome de Marfan; Glaucoma/complicações; Vitrectomia

[1] Glaucoma Department, Oftalmos, Montes Claros, MG, Brazil
[2] Department of Retina and Vitreous, Oftalmos, Montes Claros, MG, Brazil
[3] Residency in Ophthalmology, Fundação Hilton Rocha, Belo Horizonte, MG, Brazil
[4] Department of Retina and Vitreous, Clínica Oftalmológica Luciano Násser, Montes Claros, MG, Brazil
[5] Universidade Estadual de Montes Claros – Unimontes, Montes Claros, MG, Brazil

Universidade Estadual de Montes Claros – Unimontes, Montes Claros, MG, Brazil

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Introduction

Marfan syndrome is a disease of autosomal dominant inheritance affecting the connective tissue with phenotypic manifestations involving the skeletal, cardiovascular and ocular systems.[1] The major ocular manifestations are crystalline subluxation, myopia and retinal detachment.[2] Skeletal manifestations include thoracic and spinal deformities, dolichostenomelia, arachnodactyly, and tall height. Displacement of the crystalline to the vitreous cavity is a relatively uncommon but potentially serious complication in these patients.[3,4] The displaced crystalline may trigger an important inflammatory reaction that will produce uveitis, corneal edema, vitreous opacification, and secondary glaucoma, with a corresponding reduction in the visual acuity.[5] Attempts by the surgeon to remove the luxate crystalline without the use of vitrectomy may intensify complications and also produce retinal detachment. Complications increase proportionally to the lenth of stay of the crystalline material in the vitreous.[6] Effective vitrectomy associated with facofragmentation of the crystalline material promotes a significant and even definitive resolution of the phacoanafiltic reaction in these cases.[7] The objective of the present paper is to report the clinical and surgical management of a patient with Marfan syndrome who progressed with a severe phacoanafiltration reaction secondary to the displacement of the crystalline to the vitreous cavity.

Case Report

F.A.M., male, 48 years old, dark skin, and natural from the north of Minas Gerais. Patient attended the emergency room of Hospital Municipal in the city of Montes Claros reporting sudden low vision and severe right eye pain (RE); he denied any trauma or other complications in this eye. In association, he had severe headache and nausea. Upon entering the office, it was observed that the patient had Marfan syndrome due to the skeletal alterations presented: tall height and alterations in the hands and chest (Figure 1).

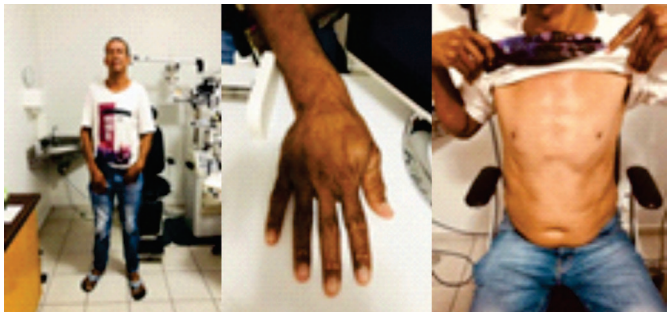
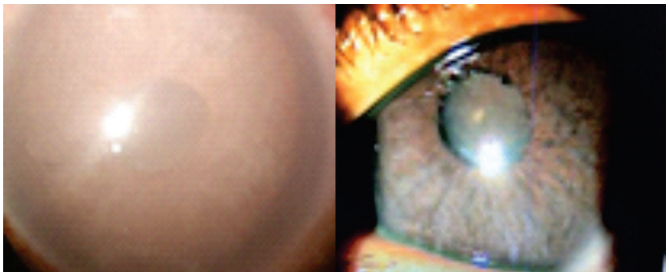


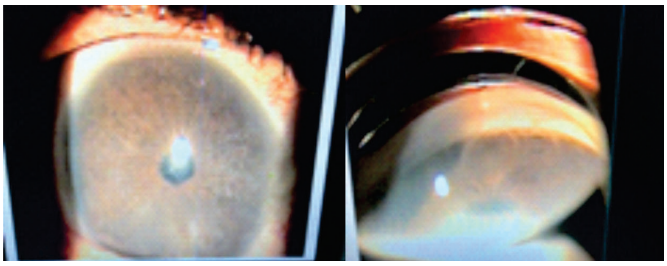
Figure 1: Tall height, dolichostenomelia, arachnodactyly, and chest alterations.

Examination of the RE showed figure visual acuity (VA), conjunctival hyperemia of 3+/4+, corneal edema, iridodonese and aphakia. In the left eye, the presence of subluxated cataract was noted (Figure 2).

Intraocular pressure (Po) in the RE was greater than 50 mmHg. Immediate treatment with ocular hypotension and topical corticosteroids was instituted, as well as oral acetazolamide (maximum dosage with potassium replacement). In the first 24-48 hours there was a substantial improvement in the clinical condition (Figure 3).

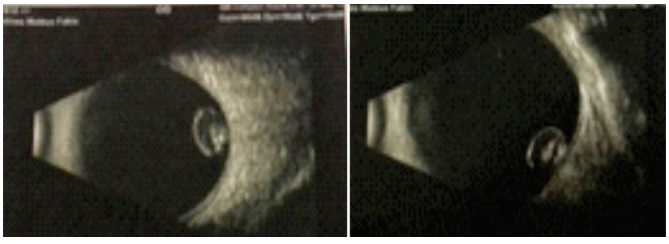


Figures 2: Biomicroscopy of the RE and LE, respectively, showing corneal decompensation in the RE.



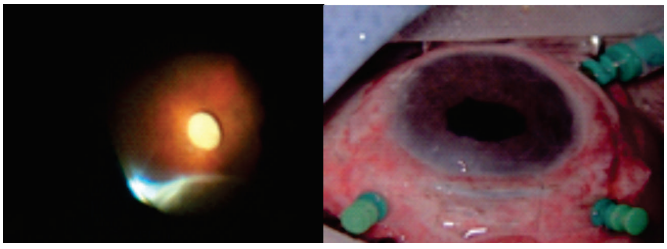
Figures 3: Biomicroscopy and goniscopy of the RE after onset of clinical treatment.

Ultrasonography was performed on this eye, where the presence of the displaced crystalline was detected at the bottom of the vitreous cavity and resting on the optic disc (Figure 4).



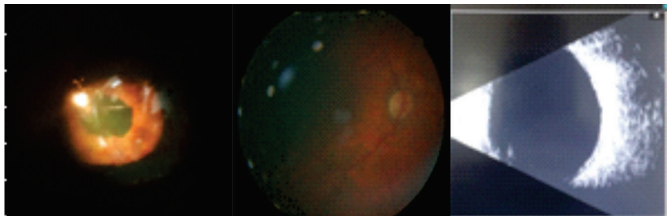
Figures 4: Presence of the luxate crystalline in the vitreous cavity.

After the surgical risk was determined, the patient with the eye stabilized was successfully submitted to posterior vitrectomy surgery with phaco-fragmentation of the dipped nucleus (milky white cataract) and 10-diopter intraocular lens (IOL) implantation by the Net technique, which consists of creating a ciliary sulcus mesh with 9.0 polypropylene yarn with two straight needles (Ethicon®) where Lio is supported (Figures 5).



Figures 5: Peroperative of the RE showing cataract nucleus dipped over the retina.

Postoperatively, the patient progressed uneventfully, and 2 months after surgery he had an eye without inflammatory signs, a VA with flat refraction of 20/40, compensated cornea, Po of 14 mmHg (using 2 topical hypotensors), and preserved eye fundus (Figures 6).



Figures 6: Two months postoperative of the RE showing calm eye, transparent cornea, and attached retina.

Discussion

Crystalline displacement to the vitreous cavity is one of the main mechanisms of phacoanafilaxia, since the crystalline material has antigenic properties.[6] In addition to Marfan syndrome, other causes may be mentioned as homocystinuria, ocular trauma, and of course complications inherent to cataract surgery. Histopathological studies show that the crystalline material has antigenic properties, and this material causes a reaction that can range from an antigen-antibody response to a late hypersensitivity reaction such as phacoallergic endophthalmitis.[8] The inflammatory response begins 24 hours to 14 days after the crystalline is displaced into the vitreous cavity.[9] However, there are cases in which the inflammatory response may take up to 3 months after it has occurred, and there are also nuclei without significant cortical material in which stimulation of this response may occur within 2 years.[6,9]

The corneal decompensation that occurs is usually transient in about 30 to 50% of the cases, and in most cases it is a reflex of the elevated Po and also of the surgical trauma. Only in 10% of these patients does corneal edema remain, resulting in bullous keratopathy whose treatment is corneal transplantation.[6]

In cases where there is good VA without the presence of uveitis or glaucoma, simple observation may be considered, but studies show that over 6 to 12 months there is a risk of ocular complications.[6,10] Due to the high rate of complications inherent to the inflammatory reaction, the best approach is the removal of the luxate crystalline into the vitreous cavity through vitrectomy associated with facofragmentation of all crystalline material.[6,7,11] Surgery should be as early as possible due to the phacoanafiltic response, but some studies show that eyes treated around 3 weeks have a lower incidence of postoperative glaucoma.[9,10] It is recommended that surgery be carried out on stable eyes, although in fact surgeries occur in the presence of elevated Po and with active inflammatory response. After vitrectomy and facofragmentation, IOL implantation was chosen, and both anterior and posterior chamber lenses with scleral fixation could be used.[12] In the case reported, the technique of IOL implantation was performed by the Net technique, as there was no support for IOL, and to avoid the use of an anterior chamber lens and a probable worsening of the cornea state.[13]

Description of the surgical technique for scleral fixation of the IOL: A peritomy was performed in the 4 quadrants of the ocular globe from the limbus, and after cauterization of the scleral surface a superficial groove was created in the sclera 2 mm from the limbus and parallel to it with 4 mm lenth. Polypropylene 9.0 suture with two straight needles (Ethicon®) was used. The straight polypropylene needle was inserted into one end of each scleral groove, and was inserted into another 26 gauge needle (13 x 4.5 mm) inserted into the diametrically opposite sulcus as they were on the visual axis. The polypropylene needle was pulled by the 26-gauge needle until it exited through the sulcus on the other side of the ocular globe. The polypropylene needle returns through the same sulcus as it exited, but at the other end (4 mm in length) being pulled by the 26-gauge needle to exit in the opposite sulcus also 4 mm away from the initial entry point. A U-knot was attached inside the scleral groove so that the thread was protected and did not touch the conjunctiva. This procedure was performed on the horizontal and vertical axes of the ocular globe, forming a square-shaped mesh of 4 mm in length on each side, on the visual axis. A 3-piece hydrophobic folding acrylic IOL (Sensar®-Jhonson & Jhonson®) of 10 diopters was inserted through a 2.75 mm corneal incision over the mesh, remaining stable and centered.

Regarding secondary glaucoma, the persistent immune response may decisively and definitively obstruct the trabecular meshwork causing secondary glaucoma onset.[8] Studies have shown that about 50% of patients remain with a significant increase in Po and thus indicate an additional anti-glaucomatous surgical treatment.[14] It is noteworthy that in some cases cortisonic glaucoma can also occur, with consequent involvement of the patient's final vision, since if surgical intervention is not indicated, the phacoanaphylactic reaction can only be clinically controlled with prolonged use of topical and systemic corticosteroids.[10]

It is concluded that the earlier the removal of crystalline material from the vitreous cavity, the better the progression of the clinical condition, and with less complications in patients with Marfan syndrome.

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Corresponding author:
Luciano Sólia Násser
Rua Valter Ferreira Barreto, 57, Bairro Ibituruna,
ZP Code: 39401-347, Montes Claros-MG.
Phone Nº.: (55)38 99993:8034