Xen® Gel Stent as a surgical option for glaucoma secondary to iridocorneal endothelial syndrome refractory to medical treatment

Xen® Gel Stent como opção cirúrgica para o glaucoma secundário à síndrome endotelial iridocorneana refratária ao tratamento médico

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ABSTRACT

Our objective was to report the case of the use of Xen® Gel Stent in the surgical treatment of glaucoma secondary to iridocorneal endothelial syndrome. Female patient, 38 years old, with unilateral glaucoma secondary to iridocorneal endothelial syndrome in the right eye, using maximum tolerable medications (three medications), with medicated intraocular pressure oscillating between 16 and 18 mmHg, demonstrating structural and functional progression, underwent minimally invasive bleb surgery with ab interno closed-conjunctival implantation of a Xen® Gel Stent. The surgery occurred without complications and the postoperative evolution has been favorable for up to 12 months, with intraocular pressure varying from 8 to 12 mmHg, without hypotensive medications and without complications or reinterventions. The Xen® Gel Stent implant provided good intraocular pressure control in the intermediate term with an excellent safety profile. It opens a perspective for new studies of the use of these devices in cases of glaucoma secondary to iridocorneal endothelial syndrome refractory to medical treatment.

RESUMO

O objetivo foi relatar o caso do uso do Xen® Gel Stent no tratamento cirúrgico do glaucoma secundário à síndrome endotelial iridocorneana. Paciente do sexo feminino, 38 anos de idade, portadora de glaucoma unilateral secundário à síndrome endotelial iridocorneana no olho direito, que usa de medicação máxima tolerável (três medicações), com pressão intraocular medicamentosa oscilando entre 16 e 18 mmHg, demonstrando progressão estrutural e funcional, foi submetida à cirurgia minimamente invasiva de bolha com implante ab interno fechado-conjuntival de um Xen® Gel Stent. A cirurgia ocorreu sem complicações e a evolução pós-operatória foi favorável por até 12 meses, com pressão intraocular variando de 8 a 12 mmHg, sem medicamentos anti-hipertensivos e sem complicações ou reintervenções. O implante do Xen® Gel Stent proporcionou um bom controle da pressão intraocular em prazo intermediário com um excelente perfil de segurança. Isso abre uma perspectiva para novos estudos sobre o uso desses dispositivos em casos de glaucoma secundário à síndrome endotelial iridocorneana refratária ao tratamento médico.

INTRODUCTION

In a group of pathologies collectively known as iridocorneal endothelial (ICE) syndrome, there are three main changes in the affected eye: corneal endothelial degeneration, changes in the anterior chamber angle, and structural changes in the iris.[1]

There are three variations of ICE syndrome: Chandler syndrome, essential iris atrophy, and Cogan-Reese syndrome. Each variation may also be associated with secondary glaucoma, and in general, glaucoma occurs in about 50% of the cases of ICE syndrome.[1]

Iridocorneal endothelial syndrome is a primary abnormality of the corneal endothelium.[2] The endothelial layer typically has a hammered silver appearance with thin guttate cornea-like lesions on slit-lamp biomicroscopy. Specular microscopy often reveals pleomorphism and polymegatism of the endothelial mosaic. Endothelial cells characteristically present as dark cells with light borders, showing a reversal of the normally expected pattern.[1-3]

Abnormal endothelial cells can proliferate and migrate through the angle of the camera to the surface of the iris. Subsequent contraction of the migrating endothelial cell layer causes the formation of peripheral anterior synechiae (SAP), iridocorneal adhesions, and iris defects (polycoria or multiple pedunculated lesions).[1-3]

Iridocorneal endothelial syndrome is usually unilateral and affects women in the third or fourth decade.[2] Peripheral anterior synechia (PAS) are common, but they are not directly related to the increase of intraocular pressure (IOP), since the outflow of the aqueous is primarily obstructed by the endothelium that covers the angle.[1-2]

In Chandler's syndrome, the primary clinical finding is corneal changes with minimal iris abnormalities. Stalk lesions of the iris are the main feature of Cogan-Reese syndrome. The iris takes on a velvety, spiral-shaped surface with loss of crypts and ectropion uveae. Atrophic iris defects are the defining feature of essential iris atrophy. The defects result from a progressive distortion of the pupil on the side opposite to the PAS.[1-3]

Corneal edema can be treated with hypertonic saline, although more severe cases may benefit from corneal surgery, such as endothelial keratoplasty with Descemet removal.[4]

Glaucoma secondary to ICE syndrome is usually difficult to control clinically.[1] Prostaglandin analogues may be useful in certain cases if the surface of the iris is relatively uninvolved. Laser trabeculoplasty or trabecular microinvasive surgeries are unlikely to be beneficial.

Glaucoma incisional surgery is often required to control elevated IOP. Trabeculectomy with mitomycin C may be effective initially, but these eyes are prone to failure as the endothelial membrane migrates through the internal ostium.[5] Membrane goniopuncture with Nd:YAG laser can often restore flow through the ostium, but the effect is usually temporary as the membrane recovers.

The tube should be kept long and inserted away from the cornea and iris surface to decrease the risk of blockage by the proliferating membrane. Placement of the tube in the groove or pars plana may decrease the likelihood of its occlusion and decrease potential damage to the cornea.[6,7]

Recently, minimally invasive surgeries with subconjunctival drainage, such as Xen® Gel Stent and Preserflo®, have an interesting therapeutic potential. Both are drainage tubes that provide a new drainage pathway through direct communication between the anterior chamber and the subconjunctival space. The interest of implants is that they make it more difficult for endothelial cells to obstruct by migration, while at the same time, because they have small diameters, they tend to interfere less with the corneal endothelium and the iris surface.

The objective of the present study was to report the case of the use of Xen® Gel Stent implant in the surgical treatment of glaucoma secondary to iCE syndrome.

CASE REPORT

Initial exam

In April 2022, DSDC, a 38-year-old woman, was referred for evaluation of difficult-to-control unilateral glaucoma.

In this first examination, she had visual acuity (and respective optical correction) in the right eye of 1.0 (-0.75 sph. -2.00 cyl at 170°) and in the left eye of 1.0 (-0.75 sph. -1.00 cyl at 5°). Biomicroscopy showed an inferior nasal corectopia in the right eye. Her left eye did not show any abnormality. Applanation tonometry was 18 mmHg in the right eye (with the following medication: 0.5% timolol maleate + 0.2% brimonidine tartrate + 0.005% latanoprost) and 10 mmHg in the left eye (without hypotensive medication).

Gonioscopy showed the presence of PAS in the lower nasal region, high iris insertion, as well as difficulty in identifying angular structures due to the presence of 360° pectineal remains. In the left eye, the angle was open with good visualization of the angular structures.

Fundoscopy showed an increased cup-to-disc ratio in the right eye (0.8), with superior thinning of the neuroretinal ring. The left eye showed a physiological cup-to-disc ratio (0.2).

The complementary exams at the time showed the following results:

- White-on-white automated perimetry: normal in both eyes
- Optical coherence tomography: reduction of the layer of peripapillary nerve fibers in the upper region of the right eye.
- Central corneal thickness: right eye: 583 µ and left eye: 573 µ.
- Specular microscopy: typical ICE syndrome cells in the right eye. Left eye showed normal endothelial cell appearances and count (Figure 1).

The diagnosis of ICE syndrome in the right eye (probable Chandler) was confirmed. We decided to maintain the medical therapy and follow up with exams every 4 months.

Follow up

From April 2022 to November 2023, the patient attended for periodic reviews every 4 to 6 months. During this period, IOP (right eye/left eye) and complementary tests varied as follows:

- August 2022: 18/11 mmHg. Right eye using the same medication. Stable functional and structural examinations.
- March 2023: 16/10 mmHg. Right eye using the same medication. Stable functional and structural examinations.
- November 2023: 21/11 mmHg. Right eye using the same medication. Significant worsening of right eye exams. Appearance of scotomas in the lower arcuate region on automated perimetry of the right eye and diffuse reduction in the thickness of the peripapillary retinal nerve fiber layer in the right eye (Figure 2). On this occasion, anti-glaucomatous surgery with Xen® 45 gel stent implantation in the right eye was indicated, and a target IOP < 15 mmHg was established.

Surgical procedure

The surgery on the right eye was performed without any complications in January 2024. The implantation of the Xen® 45 gel stent was performed using the *ab interno* technique, without opening the conjunctiva, with topical anesthesia associated with intravenous sedation with propofol. An intraoperative subconjunctival application of Mitomycin C 0.2 mg/mL was also performed in the area of the filtration bleb, followed by verification of the absence of Tenon limiting the movement of the device (primary needling).

The patient was discharged without occlusion of the right eye, with guidance to avoid physical activities and compression of the operated eye and was using a fixed combination of moxifloxacin and topical dexamethasone every 3 hours, in addition to 1% atropine eye drops every 24 hours.

Postoperative follow-up

Day 1

Calm right eye, with the presence of discrete stretch marks on the cornea, deep anterior chamber, dilated pupil, diffuse filtering bubble. IOP: 05 mmHg in the right eye and 12 mmHg in the left eye. Medication in the right eye was maintained.

Day 15

Calm right eye, no corneal stretch marks, deep anterior chamber, dilated pupil, diffuse filtering bleb. IOP: 07 mmHg in the right eye and 12 mmHg in the left eye. The eye drops

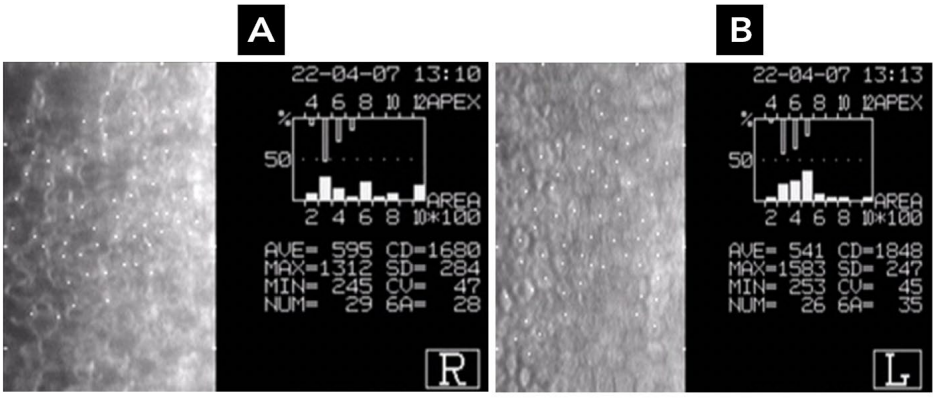

Figure 1. Endothelial mosaic in the right eye (A) and left eye (B). The right eye has features of irido-corneal-endothelial syndrome (pleomorphism, polymegatism, dark cells with light borders).

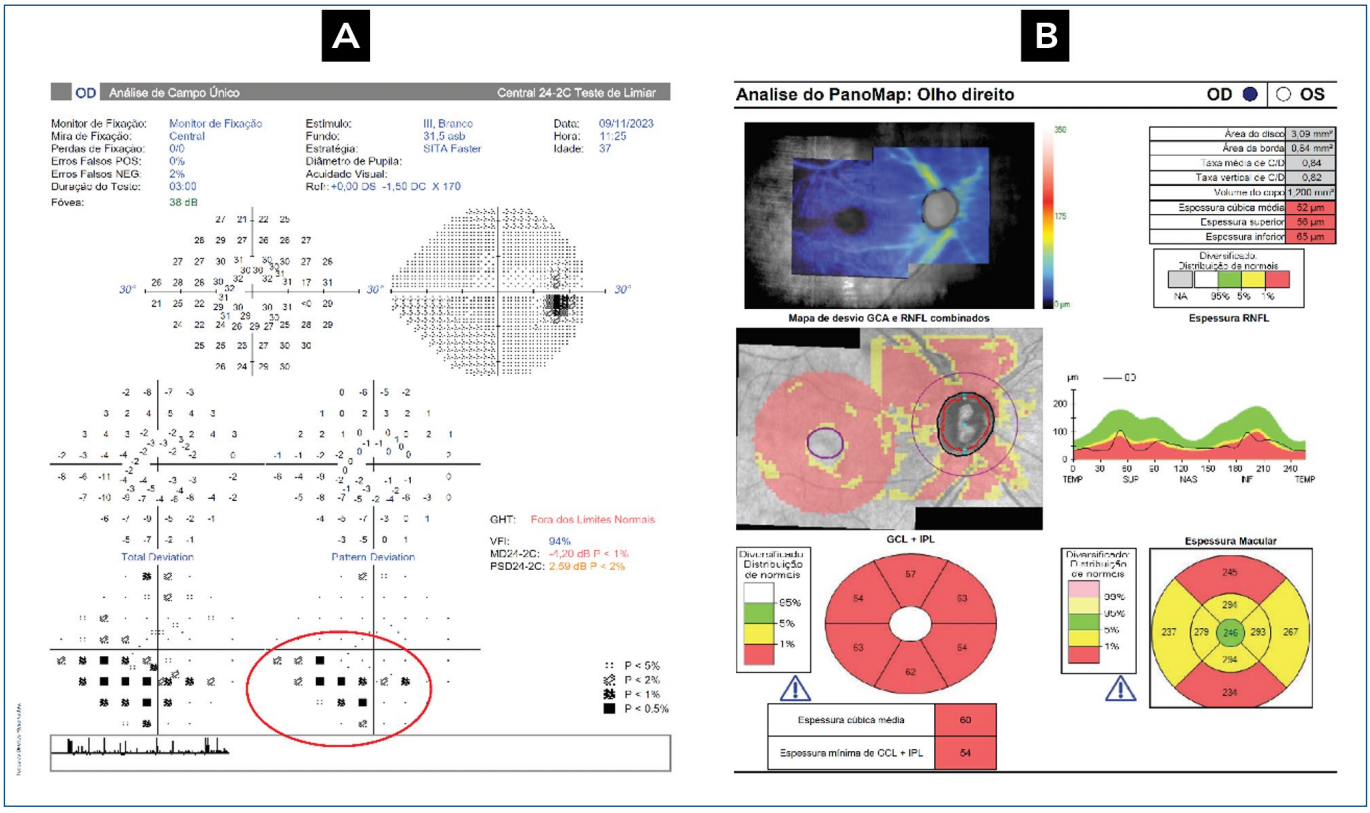

Figure 2. Functional – automated perimetry; (A) and structural examination with optical coherence tomography (B) of the right eye. An inferior arcuate defect (red circle) can be noticed in the visual field exam (A), which appeared during follow up. Optical coherence tomography (B) shows a large disc, a large cup-to-disc ratio and a diffuse reduction in the nerve fiber layer thickness. Both functional and structural progressions justified the surgical indication.

of fixed association of moxifloxacin and dexamethasone were replaced by an eye drop of dexamethasone alone, every 3 hours. Atropine 1% eye drops were discontinued.

Day 30

Calm right eye, deep anterior chamber, photo-reactive pupil, diffuse filtering bleb, no signs of excessive fibrosis. IOP: 08 mmHg in the right eye and 12 mmHg in the left eye. Gradual reduction of corticosteroid eye drops was scheduled every 7 days until complete discontinuation.

Third month

Patient already without use of corticosteroid eye drops. The right eye was calm, the anterior chamber was deep, the pupil was photoreactive, and a diffuse filtering bleb was present (Figure 3). Corrected visual acuity of 1.0 in both eyes. IOP: 08 mmHg in the right eye and 12 mmHg in the left eye. The patient was instructed to maintain frequent follow-up every 3 months.

Sixth month

Calm right eye, deep anterior chamber, photo-reactive pupil, diffuse filtering bleb (same appearance as month 3). Corrected visual acuity of 1.0 in both eyes. IOP: 12 mmHg in the right eye and 12 mmHg in the left eye. The patient was instructed to maintain frequent follow-up every 3 months.

Twelfth month

Calm right eye, deep anterior chamber, photo-reactive pupil, diffuse filtering bleb (same appearance as month 3). Corrected visual acuity of 1.0 in both eyes. IOP: 12 mmHg in the right eye and 12 mmHg in the left eye. The patient was instructed to maintain frequent follow-up every 4 months.

DISCUSSION

The present case report shows the possibility of controlling IOP in the short term with the Xen® Gel Stent device in this case of refractory glaucoma secondary to ICE syndrome.

The treatment of glaucoma secondary to ICE syndrome is a challenge, as the progressive nature of the disease leads to high rates of failure, both in medical therapy and in traditional incisional surgery (trabeculectomy).[1] The best results are found with the use of long drainage

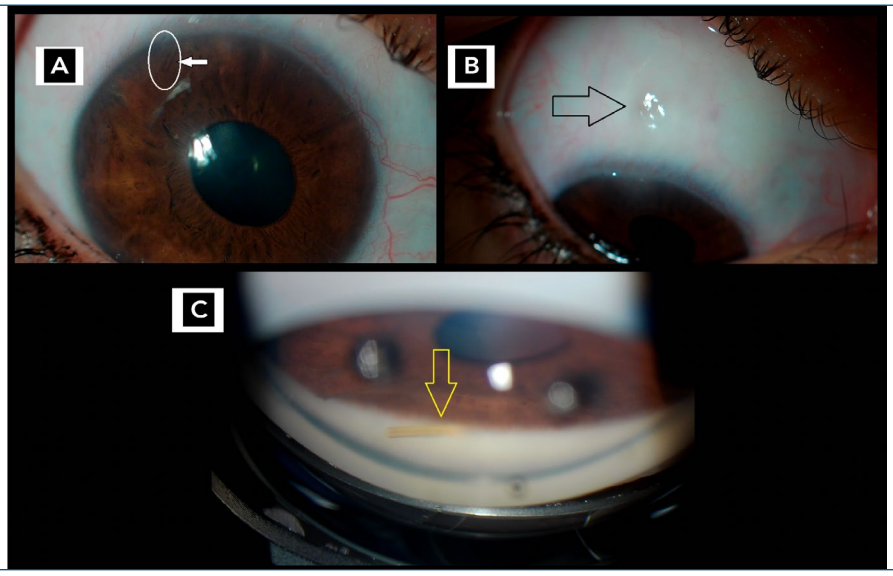

Figure 3. Clinical assessment after 3 months postoperatively. In (A), white arrow and white circle point the intraocular portion of the Xen® 45 gel implant in the anterior segment biomicroscopic view. (B) Presence of a superior, diffuse, and avascular subconjunctival bleb formation (black arrow). (C) The gonioscopic view of the anterior chamber angle and the intraocular portion of the Xen® 45 gel implant (yellow arrow).

implants (tubes and valves).[1,6,7] These, in turn, can lead to the worsening of corneal endothelial disease, with acceleration of endothelial loss, due to the presence of the tube in the anterior chamber.[1,6,7]

The potential advantages of using the Xen® Gel Stent are numerous: decreased surgical trauma; smaller diameter implantation compared to traditional drainage implants; less impact on the corneal endothelium; effectiveness in reducing IOP; and superior intraoperative and postoperative safety.[8]

To our knowledge, there are no reports in the literature of the use of new minimally invasive glaucoma implants, such as the Xen® Gel Stent in glaucoma secondary to ICE syndrome cases. A possible explanation is the low prevalence of this syndrome and the relatively recent availability of these devices.

The outcome described in this report is still very incipient, as it is a single case, with an intermediate follow-up time (12 months). However, it opens a perspective for new studies of the use of these devices in cases of ICE syndrome refractory to medical treatment.

The Xen® Gel Stent implant provided good intermediate IOP control in this case of ICE syndrome, refractory to maximum tolerable treatment with eye drops. The positive evolution of the case points to a less invasive possibility of surgical treatment for this type of glaucoma.

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