Use of the XEN gel implant in a patient with aniridia-associated glaucoma

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ABSTRACT

Purpose: To report on the implantation of a XEN 45 gel implant in a patient with glaucoma associated with congenital aniridia.

Observations: A 60-year-old man with familial congenital aniridia and glaucoma presented with intraocular pressure of 30 mmHg in his right eye despite maximal topical treatment with four medications. Implantation of a XEN gel stent was performed without complications after subconjunctival injection of mitomycin-C (0.1 mL of 0.02%). At the two-year follow-up, the patient’s intraocular pressure was 12 mmHg with one topical medication. No needling of the filtering bleb was required. No excessive fibrosis of the conjunctiva or worsening of the existing keratopathy was noted.

Conclusions: The XEN 45 gel implant could be an option in the treatment of aniridia-associated glaucoma.

1. Introduction

Congenital aniridia is a panocular malformation that involves the cornea, iris, anterior chamber angle, lens, retina, and the optic nerve. Secondary ocular complications such as keratopathy, cataract, and glaucoma are common. Approximately 50% of patients with aniridia have glaucoma, and it often requires surgical intervention.1

In recent years, minimally invasive glaucoma surgery has become an alternative to traditional glaucoma surgery. The XEN 45 (Allergan, Dublin, Ireland) was recently approved by the US Food and Drug Administration for the treatment of glaucoma. This gelatin stent is placed ab interno, creating a drainage pathway from the anterior chamber via a clear corneal incision and advanced through the sclera slightly anterior to the trabecular meshwork. The implant was placed in the subconjunctival space in the superonasal quadrant. Implantation with XEN 45 for indications such as open-angle glaucoma,2 uveitic glaucoma,3 neovascular glaucoma,4 ICE syndrome,3 and steroid-induced ocular hypertension5 have been reported.

In this report, we describe the implantation of a XEN gel stent in a patient with congenital aniridia and glaucoma.

2. Case report

A 60-year-old man had familial congenital aniridia and related nystagmus, ptosis, keratopathy, cataract, and a 23-year history of bilateral glaucoma. Cataract surgery was performed in his right eye in 2015. In 2018, the intraocular pressure (IOP) in his right eye increased to 30 mmHg despite maximal topical medications with a fixed combination of brinzolamide and brimonidine tartrate (Simbrinz, Alcon, Fort Worth, TX, USA) and timolol and bimatoprost (Ganfort UD, Allergan, Dublin, Ireland). Oral acetazolamide (Diamox, Mercury Pharmaceuticals Ltd, London, UK) and topical preservative-free dexamethasone 1 mg/ml (Monopex, Laboratoires Théa, Clermont-Ferrand, France) 4 times daily was prescribed. Four weeks later, a XEN 45 gel stent was implanted. Subconjunctival injection of mitomycin-C (MMC) (0.1 mL of 0.02%) was performed. Then, the anterior chamber was filled with an ophthalmic viscosurgical device (OVD) (Healon GV; Abbott Medical Optics, Abbott Park, IL, USA). The injector needle entered the anterior chamber via a clear corneal incision and advanced through the sclera slightly anterior to the trabecular meshwork. The implant was placed in the subconjunctival space in the superonasal quadrant. The OVD was evacuated, corneal incisions were hydrated, and bleb formation was confirmed. Finally, intracameral cefturoxime (Aprokam, Laboratoires Théa, Clermont-Ferrand, France) was injected. Topical dexamethasone 6 times daily was continued for the first month then gradually tapered off over a period of 6 months. The IOP was 4 mmHg on the first post-operative day, 4 mmHg at 1 week, 5 mmHg at 1 month, 10 mmHg at 3 months, and 11 mmHg at the 6-month follow-up. A small cystic appearance was noted in the filtering bleb at this time (Fig. 1). At the one-year follow-up, the IOP was 16 mmHg with preservative-free tafluprost (Tafloton, Santen, Osaka, Japan). 17 months after the surgery, tafluprost was discontinued and substituted with preservative-free timolol (Timosan, Santen, Osaka, Japan). At the two-year follow-up, the IOP was 12 mmHg with preservative-free timolol as monotherapy.

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Needling was not performed during the postoperative period. No excessive fibrosis of the conjunctiva or worsening of the keratopathy was noted (Fig. 2).

3. Discussion

The prevalence of glaucoma in congenital aniridia is close to 50%, and often presents during childhood or early adulthood. There is an age-dependent increase in the incidence of glaucoma, and the increased IOP could be a result of both congenital abnormalities in the Schlemm’s canal and progressive closure of the anterior chamber angle due to forward movement of the iris stump. Medical therapy is considered the initial treatment in patients with glaucoma. Congenital aniridia is often associated with severe stem cell failure, and preservative-free eye drops should be chosen. However, local antiglaucomatous treatments often fail, which makes surgical procedures necessary. Surgical treatment of aniridia-related glaucoma is challenging. The effect of goniotomy in established aniridic glaucoma is questionable, but could have a role as a prophylactic treatment in children. Trabeculotomy had an 83% success rate in one study in a young age group. Trabeculectomy, still considered the gold standard in glaucoma surgery, has a high failure rate in relation to congenital aniridia. One study found better results in a younger age group. Studies of glaucoma drainage devices, such as the Ahmed valve implant or Baerveldt implant, have shown promising results. However, complications such as retinal detachment and tube migration may occur. Surgery with cyclocryotherapy is associated with high rates of phthisis bulbi, at least in children and should be avoided if possible. Cyclophotocoagulation is another surgical option. However, the treatment is destructive, and potential complications such as phthisis bulbi are a concern. The XEN stent is made of collagen-based gelatin that is soft and flexible in the eye. It is well tolerated, without foreign body reaction. The implantation is an ab interno procedure, eliminating the need for a subconjunctival incision, and is less time-consuming than trabeculectomy or procedures with glaucoma drainage devices. The implant is 6 mm long, and the inner lumen is 45 μm, which provides approximately 6–8 mmHg of flow resistance. This reduces the risk of postoperative hypotony. Typically, as in our patient, the IOP is low the first week, then gradually inclines due to increased subconjunctival resistance, and stabilizes approximately 1–3 months postoperatively.

An intraoperative subconjunctival injection of MMC is routinely used in XEN implantation. In addition to the antifibrotic effect of the MMC, the tissue is elevated and expanded, which makes the implantation easier. Use of MMC in trabeculectomy surgery may cause severe limbal stem cell deficiency, and efforts to prevent the MMC from reaching the limbus is advisable. This can be achieved by injecting the solution far posterior to the superior limbus. Additional use of a cotton swab placed at the limbus may also help keep the MMC away from the limbal stem

cells. No avascularity of the perilimbal area or worsening of the keratopathy was observed in our patient. Due to the existing limbal stem cell deficiency in aniridic patients, we believe that special care should be taken to prevent the MMC from reaching the limbus or leaking through the conjunctiva and being exposed to the corneal epithelium. A lower dose of MMC than the frequently used 0.1 mL of MMC 0.02% could be considered.

4. Conclusions

This case report shows that the XEN 45 could be an option in the treatment of glaucoma associated with congenital aniridia. In our institution, one case of aniridia-related glaucoma has been treated with the XEN gel implant so far. Due to the promising results over two years follow-up, we will consider this intervention in future cases. Other case reports, case series or studies with longer follow-up are required to assess the efficacy and safety of this treatment. The procedure involves a filtering bleb, and potential complications such as device exposure, infection and IOP elevation require lifelong monitoring.

Patient consent

Written informed consent to publish the case report was obtained from the patient, and a copy is available for the journal.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

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