

Optimizing conservative management on recurrent hypotony maculopathy following glaucoma drainage devices implantation in steroid responder

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ABSTRACT

This report describes a case of managing recurrent hypotony maculopathy following glaucoma drainage devices (GDD) implantation with steroids. A 29-year-old female presented with blurred vision and a sudden decrease in visual acuity (VA) and intraocular pressure (IOP) 5 months following the GDD implantation. Assuming hypotony maculopathy as the cause, we started conservative treatment with oral and topical steroids, resulting in functional and structural improvement. However, the same symptoms and findings recurred. The steroid medications were then re-established with tapering down and maintenance of dose, the patient's IOP and VA remained stable with conservative treatments. The most common cause of hypotony is glaucoma filtering surgery. Conservative management using steroids is frequently effective in treating hypotony maculopathy, especially in steroid responders.

Key words: Glaucoma, Glaucoma drainage device, Hypotony maculopathy, Steroid, Steroid responder

Glaucoma is a progressive optic neuropathy and is the second most common cause of blindness, after cataract [1,2]. The number of people with glaucoma worldwide is expected to rise from 76 million in 2020 to 11 million in 2040, with Africa and Asia being affected more heavily [3].

The most effective way to control glaucoma is by lowering intraocular pressure (IOP) to prevent progressive degeneration of retinal ganglion cells [3]. Glaucoma drainage devices (GDDs) have a high safety profile and less need for post-operative follow-up, thus the number of GDDs performed is increasing. Despite their proven efficacy, GDDs are associated with a higher complication rate, one of them is post-operative hypotony, which may lead to choroidal effusion, choroidal hemorrhage, and hypotony maculopathy, which can potentially cause permanent visual loss [2-5]. Hypotony maculopathy is characterized by chorioretinal folds, increased vascular tortuosity, and disc edema and is treated with procedures that elevate the IOP, which depends on its cause. Conservative management of hypotony includes topical corticosteroids and oral steroids [6].

We report a patient with juvenile open-angle glaucoma (JOAG) who developed recurrent hypotony maculopathy after non-valved GDD and IOP was controlled by steroids. As

patients with hypotony are at risk of vision loss, it is important to understand how to manage hypotony maculopathy. Given the limited literature on this topic, this case adds important insights into the understanding and treatment of this challenging condition.

CASE REPORT

A 29-year-old female had a history of JOAG diagnosed 2 years earlier. She had undergone trabeculotomy with mitomycin C (MMC) in both eyes but continued to experience uncontrolled IOP. Due to progressive disease, she underwent non-valved GDD implantation in her right eye. Her medical history also included systemic hypertension, but she had no family history of glaucoma or other systemic diseases. Vital signs were normal, and the general examination revealed no abnormalities.

She had an intracameral hydroxypropyl methylcellulose (HPMC) viscoelastic injected on the 1st week after GDD implantation due to low IOP and very shallow anterior chamber (AC), resulting a normalized IOP of 16 mmHg, improved visual acuity (VA) of 6/20 and deep AC a week after. However, her IOP and VA remained unstable and fluctuating during the entire follow-up (Fig. 1).

On her 5 months post-surgery, she complained of blurry vision in her right eye. Examination revealed a reduced IOP of 6 mmHg and VA on the right eye was 6/24. Fundus photography

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showed irregular retinal folds radiating outward from the fovea and tortuous retinal vessels (Fig. 2a). Optical coherence tomography (OCT) also showed significant retinal folds (Fig 3a). Assuming hypotony as the cause, conservative management of topical and oral steroid medication was initiated, which resulted in improvement of IOP, VA, and OCT findings (Figs. 2b and 3b). Considering the hypotony was resolved, we continued her steroid medication and also started topical glaucoma medications to control her IOP on her right eye. On the next follow-up, the patient had no complaint, the VA was 6/6 and the IOP was 33 mmHg. We recommended her to stop her steroid medication. She continued her glaucoma medications and her IOP remained stable with no visual complaints in subsequent visits.

However, a month after discontinuing steroids, the patient returned with once again decreased IOP of 6 mmHg and VA dropped to 6/15. Recurrent retinal folds were found on OCT examination (Fig. 3c) and her central corneal thickness was 405 μ m. To address this recurrence, glaucoma medications were stopped, and oral steroids were reintroduced. Within a month, her IOP increased to 22 mmHg, and her VA improved to 6/7.5. Fundus photography and OCT demonstrated significant improvement in retinal folds and reduced vascular tortuosity (Figs. 2c and 3d).

Due to her response to steroids, the steroid dosage was gradually tapered while closely monitoring the patient's IOP and VA, rather than performing surgical interventions. The patient's IOP remained stable, and her vision did not deteriorate during subsequent follow-ups.

DISCUSSION

Hypotony is the most common complication of glaucoma filtering surgery. The advanced glaucoma intervention study showed

that surgical intervention is necessary when IOP remains high despite maximum medical therapy, which has led to an increase in glaucoma surgeries and, consequently, more complications like ocular hypotony and hypotony maculopathy [1]. When trabeculectomy fails, implantation of GDDs is the procedure of choice [1], such as the Virna glaucoma implant a non-valved GDD product from Indonesia, with affordable price and similar effectiveness and safety profile as other implants [2].

A low IOP with shallow AC was found in our patient on the 1st day after surgery, suggesting overfiltration. It is consistent with the theory that in non-valved devices, hypotony may occur due to improper placement of the ligature or damage to the tube during placement of fenestrations. If the tube is allowed to flow before the formation of a capsule, there is no mechanism to restrict the flow of aqueous and hypotony can ensue [7]. Our patient was treated with intracameral HPMC viscoelastic injection. A recent study reported that 29% of patients with post-operative hypotony resolved with intracameral viscoelastic injection [4]. However, hypotony recurred 2 months later. This condition was explained in a theory that although the technique might effectively control post-operative hypotony, they do so often at the price of multiple revisions and do not offer an adjustable patient-based solution, which consequently results in periods of volatile IOPs [4].

Some risk factors may play a major role in the development of post-operative hypotony maculopathy. The use of non-valved GDD increased the risk of complications [6]. In this case, we used a non-valved implant, which has a greater risk of post-operative hypotony maculopathy. A 5-year data analysis of a comparison study between Ahmed valved implants and Baerveldt non-valved implants concluded that Baerveldt implant offered a greater reduction in IOP and in medication burden at the expense of a greater risk of hypotony. The Ahmed valved implant was reported

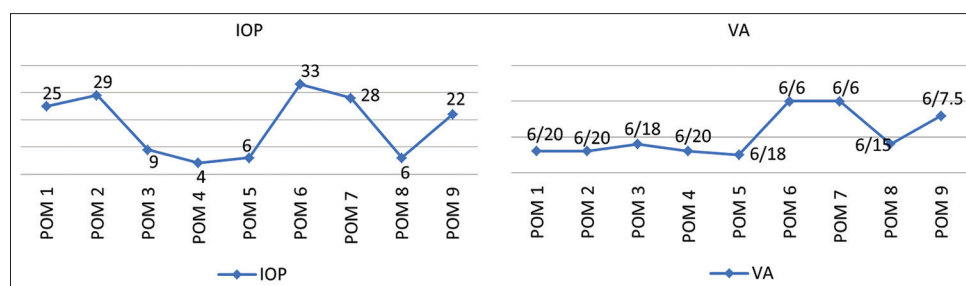


Figure 1: Post-operative right eye intraocular pressure and visual acuity

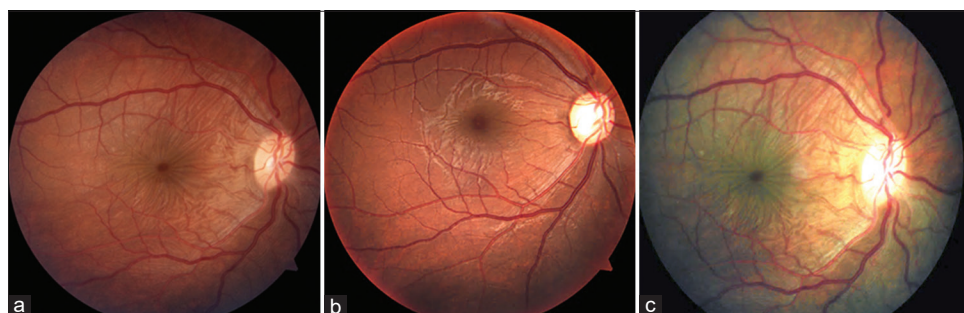


Figure 2: Fundus photography during follow-ups. (a) 5 months post-operative reveals retinal folds and retinal vascular tortuosity. (b) 6 months post-operative reveals a resolution (c) 9 months post-operative showed some folds in the macula and mild retinal vascular tortuosity

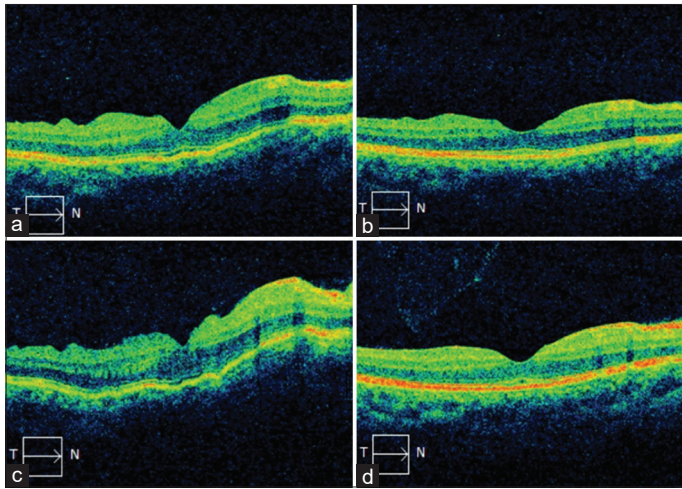


Figure 3: Optical coherence tomography during follow-ups. (a and c) shows significant retinal folds on 5 and 8 months post-operative respectively. Meanwhile, (b and d) show improvement of retinal folds of 7 and 9 months post-operative

to have a 0.4% failure from persistent hypotony while the Baerveldt non-valved implant had 4.5% [8]. In addition, younger patients like ours are at higher risk. It is correlated with low scleral rigidity, the sclera of young patients is thought to be more elastic and flexible than that of older patients and may shrink more in hypotonic conditions. On top of that, hypertension, which the patient also has, has been linked to a higher risk of hypotony maculopathy [1].

Treatment for hypotony depends on its cause. We used conservative management with topical and oral steroids for this case. Steroids can help raise IOP by reducing inflammation and promoting changes in the trabecular meshwork, which helps drain fluid [8]. Reports on recurrent hypotony treated with conservative management are very limited. There is a case similar to our case; however, in that case, a non-penetrating deep sclerectomy with MMC was performed. The patient was also treated with steroids for the recurring hypotony maculopathy, which resulted in the same outcome, the therapy effectively stabilized the patient's IOP [9]. Another case report also describes a patient with JOAG who developed hypotony, however after undergoing trabeculectomy, experienced improvement both in VA and IOP following administration of topical steroid, without any additional surgeries [10]. These cases highlight that conservative management with steroids can be an effective treatment for managing hypotony, regardless of the initial surgeries performed on each patient.

We believe our patient is a steroid responder, as her IOP increased significantly while on steroids, matching the criteria used to define steroid responders in previous studies [11]. This conservative therapy in our patient achieved a resolution of the hypotony. We utilized the ability of the patient to respond

to steroids to control her IOP and prevent vision-threatening complications of hypotony maculopathy.

CONCLUSION

As the number of GDDs performed is increasing due to their safety profile and versatile nature, the possibilities of complications like hypotony maculopathy still remain possible. As patients with hypotony maculopathy are at high risk for vision loss, it is essential to understand the potential complications and the most suitable management for each patient. Treatment options include surgical and conservative approaches. Closely monitored conservative therapy may also give favorable outcomes similar to surgical approach.

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