CASE REPORT

Steroid-induced protracted severe ocular hypertension in a 14-year-old girl

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SUMMARY

Steroid-induced ocular hypertension (SIOH) is a challenging entity in paediatric age, with many being refractory to medical therapy. Literature is scarce about surgical options in these cases. A 14-year-old girl with bilateral uveitis and macular oedema had received an intravitreal and subconjunctival triamcinolone injection in the right (OD) and left (OS) eye, respectively. While the steroid was effective in resolving the oedema, intraocular pressure (IOP) increased to about 40 mm Hg OD and 34 mm Hg OS, despite being under maximal IOP-lowering therapy. An initial conservative approach was preferred due to the young patient age and given that most cases of SIOH are transient. However, progressive structural changes were documented, and bilateral sequential minimally invasive glaucoma surgery (MIGS: XEN gel stent) was taken. With a follow-up of 6 months, the patient is drug-free with IOP around 14 mm Hg. This report discusses the role and efficacy of MIGS in a paediatric case of SIOH.

BACKGROUND

Steroid-induced ocular hypertension (SIOH) is a challenging entity in paediatric age, with many being refractory to medical therapy. Literature is scarce about surgical options in these cases. In this manuscript, we report a case of bilateral SIOH in a 14-year-old girl, which was successfully managed with minimally invasive glaucoma surgery (MIGS).

CASE PRESENTATION

A 14-year-old girl with a diagnosis of an idiopathic bilateral anterior and intermediate uveitis with macular oedema was referred from another institution with a protracted, refractory steroid-induced ocular hypertension. Four months earlier, she had received an intravitreal triamcinolone injection in the right eye, followed by subconjunctival triamcinolone delivery in the left eye due to uveitic macular oedema. While the steroid proved effective in resolving the uveitic macular oedema and restoring both the normal foveal contour and a near normal visual acuity, intraocular pressure (IOP) significantly increased despite medical therapy. At presentation in our department, she had been fully investigated for infectious and non-infectious causes of uveitis and was on oral ciclosporin A 100 mg twice daily. Her clinical observation was unremarkable, with visual acuity at 20/25 OU, no signs of active inflammation in neither eye, no posterior or anterior synechiae

and an open-angle OU (Schaffer IV 360°) on gonioscopy. IOP was 40 mm Hg in the right eye (OD) and 34 mm Hg in the left eye (OS) (corneal central thickness 546 µm OD and 543 µm OS) despite being under quadruple IOP-lowering therapy — topical prostaglandin analogue, beta-blocker, alpha agonists and oral acetazolamide.

INVESTIGATIONS

Funduscopy examination revealed a bilaterally enlarged cupping (figure 1A, B) with the triamcinolone crystals still floating on the posterior segment OD. Optical coherence tomography (OCT) and visual field revealed a normal thickness of the peripapillary retinal nerve fibre layer and no abnormal retinal sensitivity on both eyes, respectively.

TREATMENT

An initial ‘wait-and-see’ conservative approach was preferred due to the young patient age and given the fact that in most cases the steroid-induced ocular hypertension (SIOH) is transient. A close follow-up scheme in clinic that included a monthly OCT was pursued. After 4 months, since progressive structural changes (higher than 5%) were documented in three consecutive OCT examinations under maximal medical therapy, we considered proceeding to a surgical approach. After explaining the risks and benefits and having the patient and family informed consent, a bilateral sequential MIGS (XEN gel stent implant) was performed. The implant was inserted ab interno as usual, keeping about 3 mm of it in the subconjunctival space, 2 mm intrasclerally, and 1 mm in the anterior chamber. Both procedures were performed under general anaesthesia and were uneventful.

OUTCOME AND FOLLOW-UP

After nearly 6 months of XEN implantation, IOP was well controlled (OD: 14–16 mm Hg; OS: 13–14 mm Hg, with no topical therapy), and the optic cup was noted to reverse, most strikingly in the left eye (figure 1C, D). During follow-up, ocular inflammation was under control with no flare-ups under systemic conventional immunosuppression (subcutaneous methotrexate 20 mg weekly), with no need for topical or systemic steroids. A longer follow-up report would be of uttermost importance to better elucidate the role of MIGS in paediatric age.

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Figure 1 Right and left optic nerve head cup demarcation before (A and C) and after (B and D) glaucoma minimally invasive surgical procedure. A diminished cup:disk ratio after the procedure is striking.

DISCUSSION

SIOH depends on dose, duration and route of administration—intravitreal (IV), periocular (PO) and topical (TP). Reported incidences of SIOH range from 40% and 5%–25% following IV and PO/TP routes, respectively.^{1–3} Literature regarding SIOH in the paediatric age is scarce, but it is assumed a potentially more severe and rapid IOP elevation following ocular steroid therapy than in adults.⁴ A recent study on the outcomes of SIOH in children found that 44% of the cases resolved with steroid withdrawal only, 37% required ocular hypotensive drugs and 19% were performed a filtering surgery.⁵ All the patients who required surgery had documented glaucomatous disc damage.

In our case, an initial ‘wait-and-see’ conservative approach was preferred, but a MIGS procedure was considered due to documented anatomical progression. As a note, and since SIOH is a presumed diagnosis, one may not underestimate the importance of ruling out all the potential infectious aetiologies for the intraocular inflammation, before performing invasive procedures, especially in uveitic eyes.

This option allowed to effectively decreasing the IOP, while avoiding the complications associated with filtering surgery or other more invasive procedure in a teenage patient with SIOH. As such, conjunctiva iatrogenic damage, the potential for cataract formation and intraocular ocular inflammation exacerbation associated with more aggressive surgical procedures were all circumvented (causal factor removal with vitrectomy and subconjunctival excision, as well as filtering surgery with trabeculectomy or a tube). Given the patient’s long life expectancy, the decrease in dependence on glaucoma medications and the good

safety profile of MIGS surgery, this option is certainly associated with better quality of life among other surgical possibilities.⁶

As an implication of this case, it should be noted that SIOH is iatrogenic in nature, which can be prevented by seriously considering the prolonged topical use of steroids or their intraocular or periocular injection. Since the increased IOP is asymptomatic and the use of steroids is associated with intraocular inflammation-related symptom relief, glaucomatous damage may silently develop. This brief case raises awareness for the concept of *primum non nocere*, when considering the use of steroids in the paediatric age group. Conventional systemic immunosuppression (with an antimetabolite, for example) would have been a preferred initial approach to control bilateral ocular inflammation even with macular oedema, instead of bilateral local steroid treatment in the first place.

Learning points

- Steroid-induced ocular hypertension (SIOH) in the paediatric age is uncommon but usually more severe than in adults, with almost 20% requiring glaucoma surgery.
- A minimally invasive glaucoma surgery (MIGS) may effectively decrease the intraocular pressure, while avoiding the complications associated with a more invasive filtering surgery in a teenage patient with SIOH.
- Given the patient’s age, the decrease in dependence on glaucoma medications and the good safety profile of MIGS, this option also allows for better quality of life among other surgical options.
- A *primum non nocere* attitude should be in our minds when considering the use of steroids in the paediatric age group.

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