CASE REPORT

MANAGEMENT OF UVEITIC GLAUCOMA IN CHILDREN WITH BLAU SYNDROME

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ABSTRACT

Introduction: *Glaucoma in children is a condition that potentially cause blindness. Management of uveitic glaucoma is challenging because of the many mechanisms involved in its pathogenesis*
Purpose: *To report clinical characteristic and management of uveitic glaucoma in children with Blau Syndrome.*
Methods: *A 13 years old boy came to the hospital with chief complain blurred vision in both eyes. The patient was diagnosed as Blau Syndrome one year ago. He had history of trabeculectomy on his left eye. Scalling skin and swollen joints was found on physical examination. Visual acuity was hand movement in both eyes, intraocular pressure was 34 (right) and 40 (left). Gonioscopy of the right eye was schwabe line in superior and peripheral anterior synechia in three quadrants. At the left eye, there was scleral spur in all quadrant. At the anterior segment of right eye, there was band keratopathy, posterior synechia 360 o , peripheral anterior synechia, and cataract. While at the left eye, there was bleb, band keratopathy, posterior synechia, peripheral iridotomy, and cataract. Trabeculectomy, 5 fluorouracil, synechiolysis, iris retractor, and membranectomy was performed for the right eye.*
Conclusion: *Uveitic glaucoma in children poses a significant risk of blindness and needs an aggressive treatment to control intraocular pressure and amblyopia therapy to preserve vision.*

Keywords: *uveitic glaucoma, Blau syndrome, pediatric glaucoma, trabeculectomy*

INTRODUCTION

Glaucoma in children is potentially blinding condition. Uveitis causes secondary glaucoma in children. The prevalence of uveitic glaucoma in children ranges from 5 to 13.5%.[1,2]

The management of uveitic glaucoma is difficult and challenging. Inflammatory reactions may reduce the effect of some hypotensive drugs. Refractory uveitic glaucoma can be treated with surgery. Postoperative inflammatory reactions can increase the risk of filtration surgery failure in uveitic glaucoma.[1,4] This case report will discuss further about the management of uveitic glaucoma in a 13-year-old boy with Blau syndrome.

CASE REPORT

A 13-year-old boy was consulted by Pediatric Ophthalmology Clinic, Cicendo National Eye Hospital for glaucoma surgery. The patient is regularly control to Pediatric Ophthalmology Clinic since 2 years ago . The chief complaint was blurry vision in both eyes since 4 years ago.

The symptom is getting worse since 1 year ago. The patient's mother complained there is some white patch in his eyes that obstruct his vision.

The others symptoms were recurrent red eyes, watering, headache, glare, and seeing rainbows when looking at light. The patient have already complained about recurrent red eyes since 2.5 years old but sometimes its disappeared without treatment. The patient also had red spots on his skin that was treated in Dermatology Clinic, Hasan Sadikin General Hospital. He also had swollen knee, wrist joints and fingers. The patient was diagnosed as Blau's syndrome based on histopathological examination from his skin lesions. The patient had done trabeculectomy surgery on his left eye 2 years ago.

On general examination revealed scaling skin, painless swollen joints of the knees, wrists, and fingers. On ophthalmological examination, visual acuity was 1/300 in both eyes, intraocular pressure was 34 mmHg in right eye, 40 mmHg in left eye. On gonioscopy examination of the right eye: schwalbe line superiorly, PAS (peripheral anterior synechia) temporally, nasally, and inferiorly, left eye: scleral spur in all quadrants. Anterior segment of right eye: palpebrae and bulbar conjunctiva: within normal limit, cornea: band keratopathy, anterior chamber: VH gr II, flare/cell -/-, pupil: seclusion, iris: posterior synechiae 360^{o}, PAS > 180^{o}, lens: cloudy. Anterior segment of left eye: bulbar conjunctiva: bleb + cornea: band keratopathy, anterior chamber: VH gr II-III, f/s -/-, pupil: irregular, iris: posterior synechiae, lens: slightly cloudy.

Figure 1. Clinical presentation

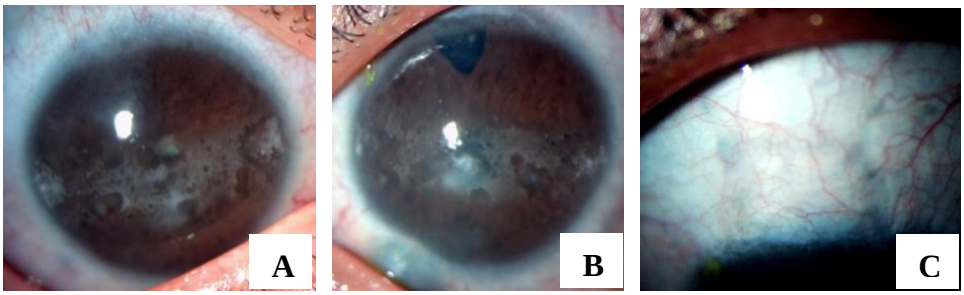

Figure 2. A. Anterior segment of right eye; B,C. Anterior segment of left eye

The patient was diagnosed as uveitic glaucoma, complicated cataract, band keratopathy in both eyes and Blau syndrome. The patient was treated with timolol maleate 0.5% eye drops

2 times a day in both eyes, brinzolamide eye drops 3 times a day in both eyes, methylprednisolone tablets 1x24 mg, methotrexate tablets 1x10 mg per week (from pediatric immunology department of Hasan Sadikin General Hospital), and was planned for trabeculectomy, synechiolysis, lens extraction ± IOL on right eye in general anesthesia.

During surgery, surgeon found that the lens had become membrane. Then manual synechiolysis, using viscoelastic, was performed to free posterior synechiae and peripheral anterior synechiae, insertion of iris retractors, and membranectomy. After membranectomy, There was no posterior lens capsule, so it was decided not to implant an intraocular lens.

On first day postoperative, visual acuity of right eye: cffc (close to face finger counting), intraocular pressure of right eye: 10mmHg, left eye: 44mmHg. Anterior segment of right eye: lid: minimal blepharospasm, bulbar conjunctiva: bleb +, cornea: band keratopathy, minimal edema, anterior chamber: VH gr III, pupil: irregular dilatation, synechiae -, PI (peripheral iridectomy) +, lens: aphakia . The patient was given postoperative therapy of prednisolone acetate 1% eye drops 6 times a day on right eye, levofloxacin eye drops 6 times a day on right eye, chloramphenicol + hydrocortisone eye ointment 3 times a day on right eye, paracetamol tablets 3x250 mg, amoxicillin tablets 3x250 mg. The patient would have a post surgery follow up at Glaucoma Clinic 1 week later.

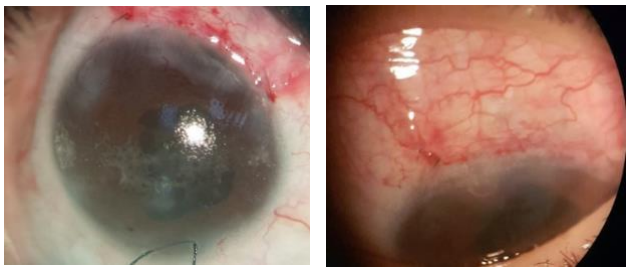

Figure 3. Anterior segment of right eye post surgery

On ophthalmological examination 1 week postoperative visual acuity of right eye: cffc, left eye: 1/300, intraocular pressure of right eye: 7mmHg, right eye: 39mmHg. Anterior segment of right eye: bulbar conjunctiva: bleb +, cornea: band keratopathy, anterior chamber: VH gr III, pupil: irregular dilatation, synechiae -, PI +, lens: aphakia. The patient was treated with tapering off prednisolone acetate 1% eye drops, sodium hyaluronate eye drops on the right eye and timolol maleate 0.5% eye drops, brinzolamide eye drops on the left eye.

On ophthalmological examination 1 month postoperative IOP (intraocular pressure) of right eye: 44mmHg, left eye: 38mmHg. The anterior segments of the right and left eyes were the same as in the previous examination. The patient was given additional treatment with timolol maleate 0.5% eye drops, brinzolamide eye drops, and sodium hyaluronate eye drops on

both eyes. Patients were routinely controlled until 1 year postoperative, intraocular pressure of right eye: 19, left eye: 28 using a combination of timolol maleate and brinzolamide eye drops.

DISCUSSION

Glaucoma is the third leading cause of visual impairment at any ages. Prevalence of blindness in pediatric glaucoma is 1.2% in the UK and reaches 3-7% in India. The incidence of glaucoma in children reaches 2.29 per 100,000 population in the United States with the most common type of glaucoma is secondary glaucoma. The incidence of pediatric uveitic glaucoma in the UK ranges from 5-13.5%. This patient was diagnosed as Blau syndrome. Blau syndrome is a rare autoinflammatory disease caused by mutations in the nucleotide-binding domain of CARD15/NOD2 gene. This patient has triads of clinical characteristics of Blau syndrome, that is arthritis, dermatitis, and uveitis. Uveitis caused secondary glaucoma in this patient.[1,2,5,6]

The intraocular pressure in uveitis was increased due to inflammatory reaction and side effects of corticosteroid therapy. Glaucoma can be either open-angle or closed-angle. In this patient, there was pupillary seclusion which caused pupillary block and anterior peripheral synechiae which caused closed angle and then obstructing the outflow of aqueous humor.[1,3,11]

Management of uveitic glaucoma includes controlling the inflammatory reaction and intraocular pressure. The first step in the management of uveitic glaucoma is administration of anti-inflammatory drugs. Corticosteroid is chosen to control inflammatory reaction aggressively. Patient with uveitis is given immunomodulatory drugs in case the patient does not respond to corticosteroids or to reduce the dose of systemic corticosteroids, related side effects that cannot be tolerated by children. This patient has been given prednisolone acetate eye drops and methotrexate tablets to control the inflammation.[3,4]

Antiglaucoma drugs have varying effectiveness depend on the presence of inflammation and when used in conjunction with steroid therapy. Uveitis with active inflammatory reaction can reduce absorption of antiglaucoma drugs up to 70-80%. Alpha-2 adrenergic agonists and prostaglandin analogues can induce inflammation, so their use is avoided in uveitic glaucoma. This patient had been given timolol maleate 0.5% and brinzolamide eye drops but his intraocular pressure was still high.[4,7]

Refractory uveitic glaucoma can be treated with surgery. There is no gold standard surgical technique in the management of uveitic glaucoma in children. Heinz et al reported a success rate of trabeculectomy is 88%, compared with modified deep sclerectomy is 50%. The success rate of trabeculectomy in children is lower than in adults because of excessive postoperative fibrosis and more frequent complications, such as hypotonia and serous choroidal

detachment. Antifibrotic drugs such as 5 fluorouracil and mitomycin C can reduce risk of postoperative fibrosis. This patient underwent trabeculectomy + 5 fluorouracil, syneciolysis, iris retractor, membranectomy. Cataract surgery in children with uveitis is very complicated. Seth et al reported that 86.7% patients with pseudophakia required postoperative topical steroids and antiglaucoma drugs to control inflammation and intraocular pressure. Fifty percent of patients with pseudophakia require further surgery. Carreno et al reported a better success rate of IOP reduction in trabeculectomy alone in uveitic glaucoma compared with combine phacotrabeculectomy. Iris manipulation and intraocular lens implantation will increase postoperative inflammatory reaction. This patient underwent trabeculectomy, lens extraction and synechiolysis as the synechia caused secondary glaucoma in this patient. But intraocular lens implantation was not performed because absence of posterior capsule and in aphakia eyes, degree of inflammation and intraocular pressure was lower. The risk of trabeculectomy failure in these patients is high. This patient requires long-term monitoring and multidisciplinary therapy to control inflammation and intraocular pressure. [8,9,10]

Prognosis for this patient is quo ad vitam dubia ad bonam. In Blau's syndrome there are other symptoms such as dermatitis, arthritis, and vasculitis that require further evaluation and monitoring. Quo ad function is ad malam because this patient had chronic panuveitis with uveitic glaucoma that intraocular pressure has not been controlled for a long time. Quo ad sanasionam is ad malam because recurrence rate of uveitis and risk of trabeculectomy failure is high.

CONCLUSION

Uveitic glaucoma in children causes blindness and requires aggressive multidisciplinary therapy to control inflammation, intraocular pressure, and amblyopia therapy to maintain vision. Management of uveitic glaucoma in children is challenging and requires more attention and wise decisions to determine surgical technique to control intraocular pressure.

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