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Gonioscopy-assisted transluminal trabeculotomy for congenital glaucoma secondary to Klippel-Trenaunay-Weber Syndrome: A case report

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ABSTRACT

Purpose: To describe the case of a 9-year-old boy with congenital glaucoma secondary to Klippel – Trenaunay - Weber Syndrome (KTW) with a history of trabeculotomy in both eyes (BE) and further trabeculectomy in the left eye (LE) presented with high intraocular pressure (IOP) and progression in the LE despite maximum tolerated medical therapy.

Observations: GATT surgery was performed firstly in the LE, followed by the right eye (RE) two months apart since the IOP in the RE started to increase later on. First post-operative day the IOP was under 15 mmHg. In the last visit, 6 months after the first surgery, IOPs were 10 and 11 mmHg RE and LE, on one fixed combination; slit lamp examinations were normal with wide open angles and a good view of the Schlemm's Canal (SC) posterior wall.

Conclusions and Importance: GATT surgery can be done after failed incisional surgery in children with glaucoma secondary to KTW syndrome.

1. Introduction

Klippel – Trenaunay - Weber Syndrome represents a congenital neurocutaneous vascular syndrome consisting of cutaneous vascular malformation, venous lymphatic malformations associating soft tissue and skeletal hypertrophy of the affected limb.[1] The most common finding of this syndrome is the cutaneous capillary malformation known as "port-wine spot" which can be seen in 90%–100% of cases.[2] Ocular involvement can be through vascular anomalies of orbit, iris, choroid, optic nerve associated with orbital varices, choroidal angioma, persistent fetal vasculature, and glaucoma. Ocular findings include prominent ocular surface vessels, heterochromia, strabismus, tortuous retinal vessels and choroidal hemangiomas.[3]

Childhood glaucoma associated with this syndrome represents a common feature and may be seen in 30% of the cases.[3] Glaucoma occurs due to angle dysgenesis and/or increased episcleral venous pressure, thereby glaucoma can be refractory and treatment is challenging in some cases.[4]

First treatment option can be medical or surgical therapy depending on the age onset and mechanism of glaucoma.[5]

2. Case report

9 year-old boy with childhood glaucoma due to Klippel – Trenaunay – Weber Syndrome was presented. He underwent trabeculotomy and trabeculectomy in left and right eyes respectively when 2 months-old. Patient was referred for the tube surgery since the intraocular pressure was still high and glaucoma progression was detected in his left eye (LE).

Clinical examination revealed mild facial hemangiomas (port – wine spot) associated with soft tissue and skeletal hypertrophy of the affected left limb (predominantly left) (Fig. 1). Best corrected visual acuities (BCVA) in both eyes were 20/20, IOPs were 17 mmHg and 22 mmHg with 3 medications in RE and LE, respectively. Anterior segment examination revealed mild subconjunctival scar superiorly in the RE but there was no bleb, whereas there was failed bleb with peripheral iridectomy (PI) at 11 o'clock in LE (Fig. 2). Gonioscopy revealed wide-open angles with mild dysgenesis in both eyes and trabecular meshwork was lightly pigmented but visible enough. There was no blood in the Schlemm's canal. In the LE sclerostomy was "anterior to the trabecular meshwork" and there were 2 small peripheral anterior synechiae (PAS) right next to the previous sclerostomy area in the superior quadrant. Fundus examination showed 0.8 cup-to-disc ratios in both

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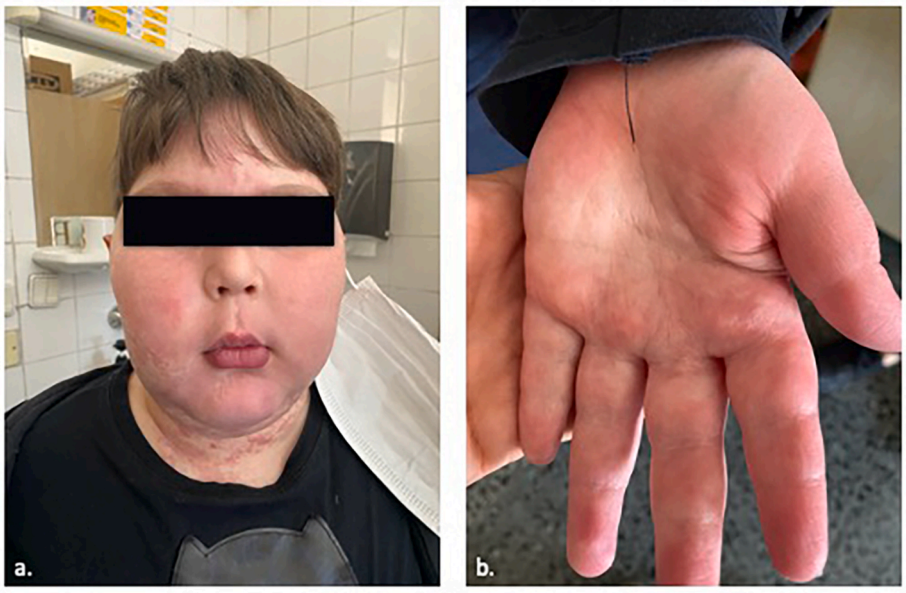

Fig. 1. a-b: Facial hemangioma with bilateral distribution (a); left affected limb with vascular malformations (b).

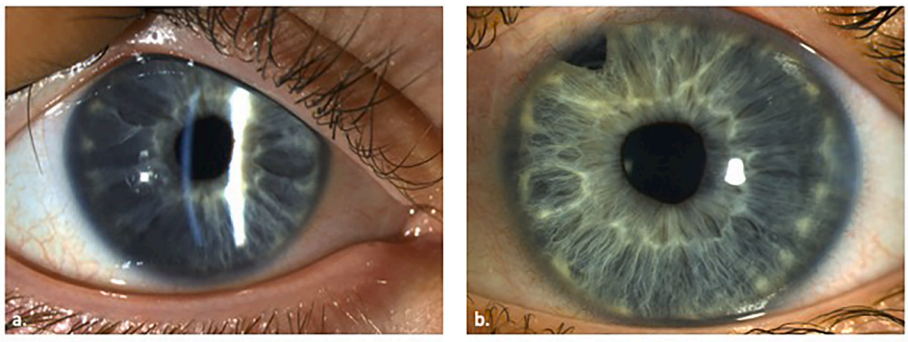

Fig. 2. a-b: Baseline slit lamp examination for RE (a) and LE, non-functional bleb and PI in the superonasal quadrant is seen (b).

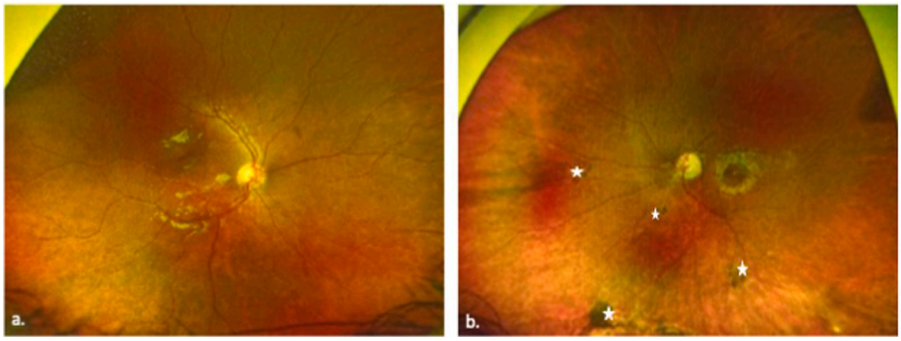

Fig. 3. a-b: Funduscopic images showing cupped optic nerve heads in both eyes (a–b) and retinal pigment epithelial hypertrophy areas (stars) (b).

eyes and retina pigment epithelial hypertrophy. (Fig. 3).

On optical coherence tomography (OCT) glaucoma was stable but there was progressive loss in retinal nerve fiber layer (RNFL) analysis in the LE throughout the patient's follow-up (Fig. 4).

For the surgical treatment of the LE, GATT was recommended.

Surgery was done for the LE under general anesthesia without any complications. After draping the patient. A temporal and superior paracentesis was made with 23-G microvitreoretinal blade along with reformation of the anterior chamber (AC) using cohesive viscoelastic substance after tilting the patient's head and the microscope. Prior to GATT procedure the patient underwent goniosynechiolysis by using a microvitreoretinal forceps through the temporal paracentesis for breaking small PAS next to the sclerostomy site. Microvitreoretinal blade was inserted via temporal paracentesis and a 2-mm goniotomy is performed. Using a hand-held cautery a 5–0 prolene suture was blunted and introduced in the AC using the superior paracentesis. The suture was

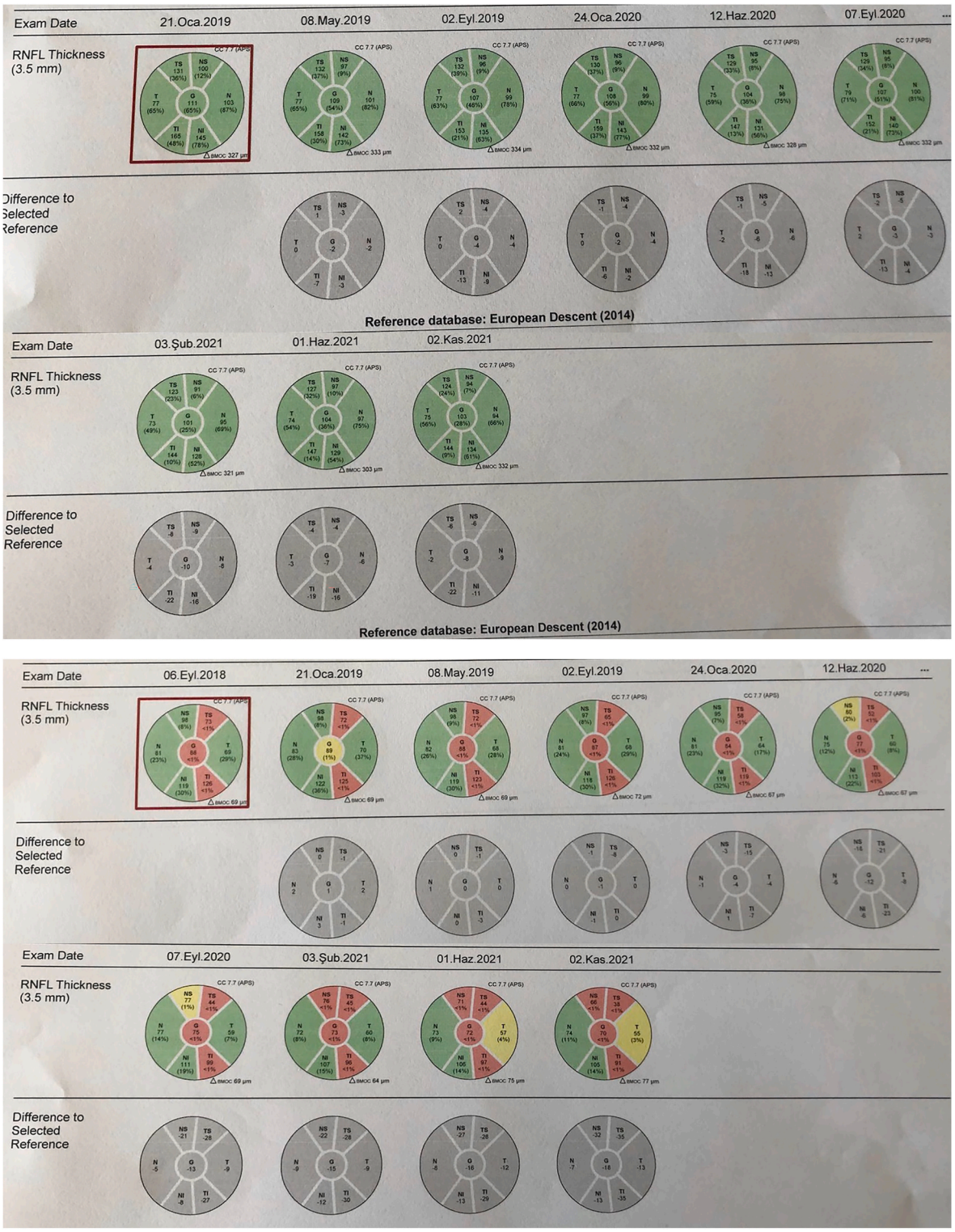

Fig. 4. a-b: SD - OCT Heidelberg Spectralis follow-ups shows glaucoma is stable in the RE (a) however progressing in the LE (b).

entered into the SC through the goniotomy incision and could be advanced circumferentially using a microsurgical forceps from the temporal paracentesis since the previous sclerostomy was done anteriorly and did not involve trabecular meshwork. Then the blunt distal tip was grasped with forceps and the proximal part of the suture was pulled to complete the circumferential ab-interno trabeculotomy. Blood reflux from the SC was seen which was a good prognostic sign revealing patent distal collector channels. The AC was then washed using irrigation/aspiration and viscoelastic substance is used to fill the AC (around 1/5) to prevent blood reflux and hyphema. Patient was put on steroid and antibiotic drops 4 times daily.

On first post-operative day IOP was 11 mmHg. Minimal hyphema was noticed at slit lamp examination which was resolved until next visit, at one week (Fig. 5). IOP was 15 mmHg at 1st week follow-up, and patient was given timolol-dorzolamide fixed combination since cup-to-disc ratio were high. At one month follow-up the IOP for LE was 10

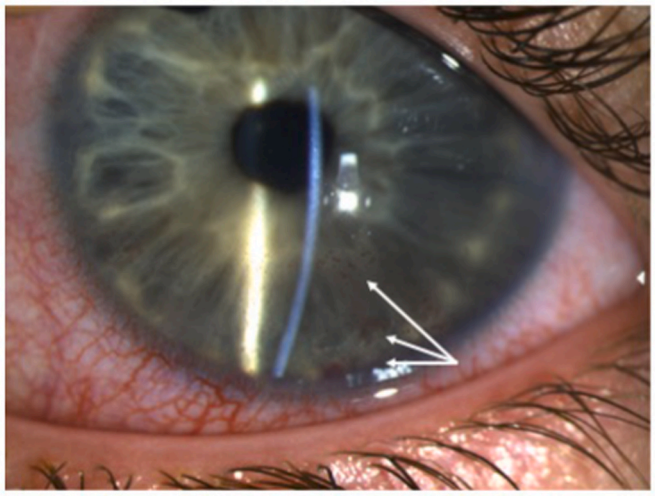

Fig. 5. Slit lamp appearance of the LE at 1 week post-operatively showing minimal hyphema (white arrows).

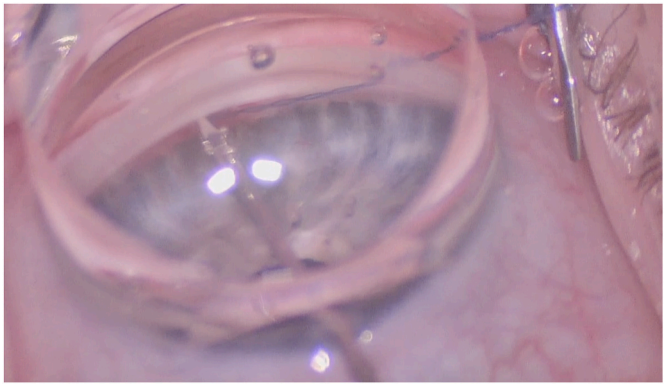

Fig. 6. Dysgenic angle appearance while threading GATT suture in the Schlemm's canal.

mmHg with fixed combination, however IOP in the RE was 28 mmHg with three medications, further on GATT surgery was also performed in the RE.

Surgery was done for the RE under general anesthesia without any complications with the same technique. This time twisted GATT suture (Sharpoint Ophthalmic Sutures) was used (Fig. 6). Circumferential ab-interno goniotomy was done successfully in the RE too.

On first post-operative day again IOP was 12 mmHg with minimal hyphema in the RE, however at the first post-operative week the IOP was 23 mmHg without any hyphema, under the same post-operative protocol treatment. Then pilocarpine 2% and timolol-dorzolamide fixed combination 2 times daily until next visit. At first month IOP was 13 mmHg in the RE again with medication and pilocarpine 2% was stopped.

The final visit occurred 6 months after the first surgery on the LE and IOPs were 10 and 11 mmHg RE and LE, with timolol-dorzolamide fixed combination, respectively; BCVAs in BE were 20/20, anterior segment slit lamp examinations were normal with wide open angles and a good view of the SC posterior wall (Figs. 7 and 8).

3. Discussion

Klippel – Trenaunay Weber Syndrome is a phakomatosis that may cause congenital glaucoma through various mechanisms similar to Sturge-Weber Syndrome. Primary angle dysgenesis with prominent iris processes that in contact with the trabecular meshwork is one of the main mechanisms leading to glaucoma in these patients. Gonioscopic similarities to the primary childhood glaucomas might be seen in these patients. Another mechanism is increased episcleral venous pressure (EVP). Other factors such as peripheral iris synechia, choroidal effusions or retinal pathology associated with neovascular glaucoma can also be incriminated as physiopathological mechanisms in secondary glaucomas secondary to phakomatoses.[3] Treatment options can be stratified according to the type of glaucoma, age of the patient, visual prognosis and associated ocular findings.[6,7]

In recent years minimal invasive glaucoma surgery (MIGS) techniques have had an important role in the surgical management of glaucoma. Goniotomy/trabeculotomy is a generally the first choice technique in childhood glaucoma with spectacular success rates in terms of IOP reduction. Circumferential trabeculotomy or goniotomy techniques have better surgical success rate compared to the conventional trabeculotomy in patients with primary congenital glaucoma.[3,8–12] Congenital glaucoma associated with Klippel – Trenaunay Weber Syndrome, GATT remains a good surgical option for lowering the IOP since it is a modified form of goniotomy and opens the Schlemm's canal circumferentially.

The current case presented with buphthalmos as a congenital glaucoma case due to angle dysgenesis standing for a main pathophysiological mechanism, in contrast to a late presented case linked to the raised EVP. There was no blood in the Schlemm's canal that is commonly seen in the presence of raised EVP. Other than that, facial hemangioma is minimal but bilateral and asymmetric as glaucomatous impairment is also bilateral and asymmetric.

The IOP reduction after GATT surgery depends on the patency of distal collector channels, which can also be affected by the severity of goniodysgenesis.[6]

Before planning GATT surgery after any intraocular intervention; type of the intervention, visibility of trabecular meshwork, position of the sclerostomy (if present) and its relation to the iris should be evaluated in every patient. GATT surgery can be done successfully after

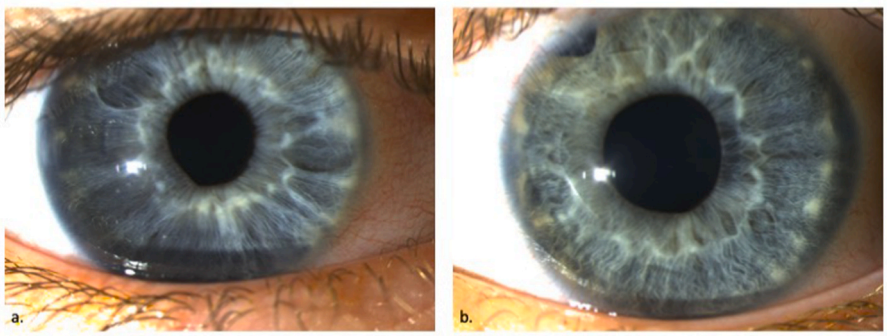

Fig. 7. a-b: Slit lamp examination of the RE (a) at 4 months and the LE (b) at 6 months.

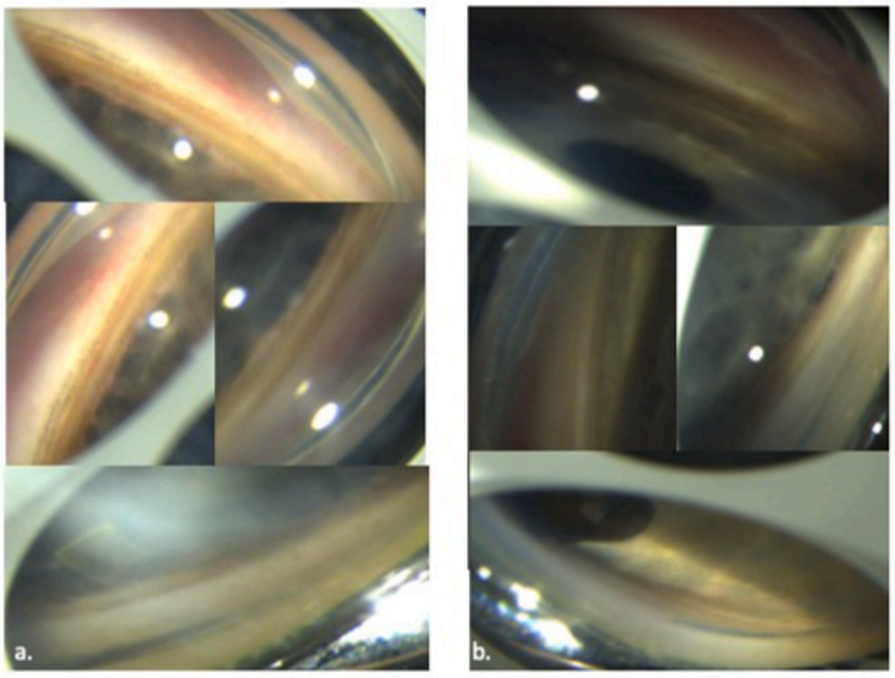

Fig. 8. a-b: Gonioscopy performed after GATT surgery in the last visit demonstrating a wide open angle with normal postoperative appearance and a good view for the posterior wall of the Schlemm's canal in the RE (a) and LE (b).

trabeculectomy in adults.[13,14]

In our case, preoperative gonioscopy revealed mild dysgenesis in both eyes with visible trabecular meshwork circumferentially that looked surgery naïve. In the left eye there was a history of trabeculectomy. However, the sclerostomy site was anterior to the Schlemm's canal so the trabecular meshwork was visible 360-degrees. In the right eye there was a history of previous intervention too. We considered that this was an ab-externo trabeculotomy attempt, since there was no peripheral iridectomy or PAS but only a subconjunctival scar. However, the trabecular meshwork looked normal all the way around. Thus, the authors thought that Schlemm's canal might not have been found during the previous angle/canal surgery.

Since there was mild angle dysgenesis, the trabecular meshwork appeared normal after two interventions, and there was no blood in Schlemm's canal in both eyes, GATT surgery was planned.

Hyphema is one of the most common complications of GATT surgery. This might be a concern in eyes with raised EVP. Thus, patients should be selected carefully and special attention should be given to preoperative findings and the age of presentation as described above. In patients with raised EVP, blood in the Schlemm's canal during ophthalmic examination is very common. In our case, postoperative hyphema was mild and resolved very quickly. If the patient with phakomatosis presents late, a possible underlying mechanism would be the high EVP, and filtering or tube surgeries are recommended in these cases.[3,4] In refractory cases trans-scleral cyclophotocoagulation diode laser (TS – CPC) can also be taken in consideration.[15]

In the current case, glaucoma was bilateral even though it was more advanced in the LE. Angle dysgenesis can be more severe in one eye compared to the fellow eye, resulting in more advanced damage, refractory to previous surgeries.[4,16] However, angle surgery which is the procedure of choice in our case was skipped.

Following GATT surgery early post-operative IOP spikes might be seen, and anti-glaucoma treatment and close follow-up might be required for this reason. The IOP spikes may result from hyphema, clot formation, viscoelastic substance in the anterior chamber, inflammation or steroid response.[6,17] In our case, this complication was noticed shortly after the surgery in the LE and treated with medical treatment easily.

4. Conclusion

Klippel – Trenaunay Weber Syndrome is a rare form of phakomatoses with some important ocular features. Glaucoma is one of them and should be treated as early as possible. Surgical treatment is needed in most of these patients and GATT surgery must be taken into consideration especially in patients with angle dysgenesis since the angle surgery is generally the first option in these patients.

Long term follow-up is very important because of the refractory behavior of glaucoma in this cases. On the other hand, outcomes of angle surgery might not be as successful as seen in patients with primary congenital glaucoma. If the first angle surgery is not circumferential, it can be repeated. Other treatment options such as filtrating surgery, tube surgery or cyclodestructive procedures should be taken into consideration for these patients in a step by step surgical approach if circumferential angle surgery fails.

To the best of our knowledge, this is the first pediatric glaucoma case secondary to KTW syndrome treated with GATT surgery. GATT surgery can also be done after filtering surgery in children if the angle is visible. Thus, it can be applied in these patients with phacomatosis secondary to KTW syndrome even after trabeculectomy, since operative risks of tube surgery are high in these patients. Long term outcomes of randomized controlled trials are still needed to shed light on the place of GATT surgery in the treatment algorithm in patients with glaucoma secondary to KTW syndrome.

Patient consent

The patient's legal guardian consented to publication of the case in writing.

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Authorship

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

Declaration of competing interest

The authors certify that they have NO affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements) or non-financial interest (such as personal or professional relationships, affiliations, knowledge or belifs) in the subject matter or materials discussed in the manuscript.

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