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Case Report

Steroid-induced Ocular Hypertension in Case of Gyrate Atrophy Treated with Posterior Sub Tenon Triamcinolone Acetonide for Cystoid Macular Edema

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Abstract

Gyrate atrophy is a rare congenital retinal dystrophy that causes cystoid macular edema (CME). The treatment options include intravitreal bevacizumab, intravitreal triamcinolone, topical carbonic anhydrase inhibitors, and subtenon steroid injections. The raised IOP may be controlled with IOP-lowering medication or can be refractory to any medication. Early tenotomy and removing PST deposits may give the patient early rehabilitation. We discuss a 17-year-old male presented with a painless diminution of vision in both eyes. The ocular examination revealed gyrate atrophy with cystoid macular edema. A sub-tenon injection of 40mg triamcinolone was given, for which the patient developed elevated intraocular pressure. The raised IOP was refractory to antiglaucoma medications. The patient was treated with a micropulse transscleral cyclodiode laser (MP3 laser) followed by surgical removal of subtenon triamcinolone deposits to reduce intraocular pressure.

Keywords: Cystoid Macular Edema; Gyrate Atrophy; Intraocular Pressure; Micropulse Transscleral Photocoagulation; Posterior Sub Tenon Triamcinolone

Key Messages

Young patients with gyrate atrophy and cystoid macular edema treated by posterior subtenon injection of Triamcinolone may have an increased risk of elevated intraocular pressures unresponsive to maximum medical treatment. In such cases, surgical removal of subtenon steroid may be considered as the primary treatment option to help decrease intraocular pressure.

Introduction

Gyrate atrophy (GA) is a rare autosomal recessive chorioretinal dystrophy caused by mutations in the ornithine aminotransferase (OAT) gene [1,2]. This gene, located on chromosome 10q26, encodes pyridoxine (vitamin B6)-dependent mitochondrial matrix enzyme ornithine-δ-aminotransferase [3,4]. OAT deficiency causes hyperornithinemia, which results in progressive chorioretinal atrophy. The symptoms include night blindness and peripheral constriction of visual fields. Cystoid macular edema is one common association with gyrate atrophy of the retina and choroid [5].

Among the options available for treating cystoid macular edema (CME), the administration of intravitreal or posterior steroid injection (PST) is an effective option but can significantly elevate

intraocular pressure. The risk factors associated with IOP increase are younger age, lower baseline IOP, and steroid dose [6,7]. While most cases with elevated pressures may be well controlled with IOP-lowering medications, some may require filtering surgeries. The use of micropulse transscleral cyclophotocoagulation in these patients has not been previously reported. Here we report a case of gyrate atrophy with CME, which had a significant elevation of IOP after PST injection, which was treated with micropulse transscleral cyclophotocoagulation (MP3 laser) resulting in a significant reduction of IOP. However, as the desired lowering in IOP was not obtained, a Tenotomy was done to remove the PST deposit. The IOP was normalized to 14 mm Hg in both eyes within two weeks.

Case history: A 17-year-old male patient presented with complaints of decreased vision in both eyes. The associated symptoms include decreased night vision and peripheral vision loss. On examination, his vision was 20/60 in both eyes. The retina analysis showed well-circumscribed regions of chorioretinal atrophy with hyperpigmented margins in the mid periphery suggestive of gyrate atrophy in both eyes (Figures 1a and 1b). Optical coherence tomography confirmed the presence of CME (Figures 1c and 1d). He has advised low protein diet and pyridoxine supplements. To manage CME, the patient has given PST 40mg injections in both eyes a week apart, the right eye followed by the left eye. On follow-up one week after the PST injection the patient presented with pain and severe photophobia. On slit lamp examination, microcystic corneal edema and ciliary congestion were noted in both eyes. The IOP was 48 mm Hg in the right eye and 42 mmHg in the left eye, as recorded with Goldmann applanation tonometry. The patient was started on topical IOP lowering medications Timolol, a beta blocker, Brimonidine, ocular specific alpha agonist, and topical carbonic anhydrase inhibitor Dorzolamide and oral carbonic anhydrase inhibitor acetazolamide 250 mg TID. Even though the intraocular pressure decreased initially, it reached 48 mmHg in the right eye and 36 mmHg in the left eye within a week. Since the pressure was not controlled with maximum medical therapy and oral acetazolamide, intravenous mannitol 1 mg/kg body weight was given over half an hour. The right eye was treated 360 degrees with a 2500 MW micropulse diode laser for 160 sec.

One week after micropulse transscleral cyclophotocoagulation, the pressure in the right eye was reduced to 30 mmHg. As the pressure was still high, we planned for surgical removal of sub tenon triamcinolone depot. A conjunctival and tenon opening was made at the site of the triamcinolone depot. After careful dissection, it was separated from the surrounding adhesions, and the steroid depot was removed. Two weeks after surgical removal, the IOP returned to 14 mmHg. Similarly, we carried out the surgical removal of depot steroid in the left eye, after which the left eye pressure normalized to 14 mmHg within a week.

Figure 1

Discussion: Gyrate atrophy (GA) is a rare autosomal recessive disease characterized by a deficiency of ornithine-δ-aminotransferase (OAT). OAT deficiency causes hyperornithinemia, which results in progressive chorioretinal atrophy [8]. The typical symptoms of GA resemble those of retinitis pigmentosa (RP), beginning with decreased night vision and loss of peripheral visual field, usually during the 1st decade of life. However, the retinal appearance is distinct, manifesting as chorioretinal atrophy, which resembles brain gyri in shape and gives the condition its name. The macula and central vision remain frequently preserved into midlife, although posterior subcapsular cataracts, epiretinal membrane, and CME often occur [6]. In our case, the patient presented in the second decade with the typical clinical picture of gyrate atrophy in the retina with CME and had symptoms of night blindness.

The treatment options for cystoid macular edema include intravitreal bevacizumab, topical carbonic anhydrase inhibitors, and intravitreal or sub-tenons triamcinolone injection [9]. A few case series of uveitis patients showed IOP peaking 2 months after the injection and returned to normal after 12 months [6]. Younger age and higher baseline IOP were risk factors for steroid-induced IOP elevation. In electron microscopy-based anatomical analyses of the trabecular meshwork in steroid-induced glaucoma, the trabecular meshwork exhibited morphological changes secondary to fibrillar elements and extracellular material; thus, obstructing the aqueous humor drainage. Reduced phagocytosis activity in trabecular meshwork cells led to increased aqueous humor flow resistance [7]. In our case report, the patient was young, and the IOP increased within one week. The right eye was first treated with micropulse transscleral cyclophotocoagulation, which was followed by sub-tenon triamcinolone removal. The mechanisms by which micropulse trans scleral cyclophotocoagulation reduces Intra ocular pressure include sub-threshold cell damage to the ciliary body, increasing uveoscleral outflow, and pilocarpine like effect [10]. As there was good IOP reduction after the removal of sub-tenon depot steroid in the right eye, the left eye was directly considered for sub-tenon triamcinolone removal. After the removal, the pressure was gradually reduced to 14 mmHg in both eyes within two weeks.

Conclusions

Gyrate atrophy of the retina and choroid is a rare autosomal recessive disease with ornithine aminotransferase deficiency and is sometimes associated with cystoid macular edema. Steroid injections, though reported to be effective in the treatment of CME can sometimes cause elevated intraocular pressure in steroid responders. Initial management of choice is conservative with topical or oral glaucoma medications. If the pressure is refractory to medications, transscleral micropulse laser may prevent the need for removal of steroid depots; however, the removal of sub tenon injection depot may be considered in refractive cases of elevated intraocular pressure.

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