

The Use of Intravitreal Steroid Implant (Ozurdex) for Treatment of Refractory Cystoid Macular Edema Secondary to Retinitis Pigmentosa

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Abstract

Introduction: Retinitis pigmentosa is a clinically & genetically diverse group of diffuse retinal dystrophies that initially affecting rod with subsequent degeneration of cones, Maculopathy is one of the complications of RP that affecting vision and usually takes different types 1. (Atrophic), 2. (Cystoid Macular Edema) 3.(Macular retraction), The prevalence of CME was 52% in AD RP , 39 % in AR RP and 35% in XL.

Method: We retrospectively review a 24y old male patient that presented with bilateral refractory Cystoid macular edema secondary to RP which is not responding to topical carbonic anhydrase inhibitor and multiple anti VEGF injections of Lucentis in both eyes, the patient underwent bilateral 0.7 mg Intravitreal dexamethasone (OZURDEX) implant of 1 week apart, the spectral domain OCT was performed before and one month after OZURDEX injection and to assess the efficacy of Intravitreal steroid implants in treatment of refractory CMO in RP.

Results: The Spectral Domain OCT after 1 month of injection showed a significant anatomical and functional improvements, CMT improved from 314 um to 233 um in the right eye and from 590 um to 207 um in the left eye. The BCVA improved from 6/18 to 6/9 in the right eye and from CF 1 meter to 6/60 in the left eye.

Conclusion: It seems that the refractory CMO in RP is mostly inflammatory driven, for this reason it shows a dramatic response to Intravitreal steroids, in our case, the patient has received multiple anti VEGF injections of Lucentis with no improvement, but he responded very well anatomically and functionally to Intravitreal steroid implant of OZURDEX, the results in our case was matching other small case series results. EX: In retrospective study of Ozdemir et al reported the efficacy of Intravitreal steroid in a group of five eye of five patients. Further preferably randomized trials may establish the place of Intravitreal dexamethasone in the treatment of refractory CME related to RP [1,2].

Keywords: Dexamethasone; Ozurdex® treatment; Retinitis pigmentosa; Cystoid macular edema

Abbreviations: CMO: Cystoid Macular Edema; RP: Retinitis Pigmentosa; CMT: Central Macular Thickness; VEGF: Vascular Endothelial Growth Factor; BCVA: Best Corrected Visual Acuity

Discussion

It seems that the refractory CMO in RP is mostly inflammatory driven, for this reason it shows a dramatic response to Intravitreal steroids, The pathogenesis of RP-related macular edema is still not known. Spalton et al. [3] investigated 25 patients with RP and angiographic retinal leakage. Edema was present in dominant and X-linked inherited disease and was likely to be present in recessive disease as well. The authors suggested that this might be a general inflammatory response seen in many types of tapetoretinal degeneration against actively degenerating photoreceptors and retina pigment epithelium (RPE). Kuchle et al. [4] investigated blood-aqueous barrier in patients with RP with a laser cell photometer in 56

eyes of 29 patients and found that aqueous flare values were higher in patients with RP than in controls and patients with RP-

related CME had even higher values with a mean of 14.66 photon counts per millisecond compared with 9.65 for patients with RP without CME. Furthermore, Heckenlively et al. [5] looked for the presence of antirational antibodies in a group of 30 consecutive patients with RP and CME, 30 consecutive patients with RP but without CME and 50 normal subjects. Twenty-seven (90%) of patients with RP and CME had antirational protein antibody activity compared with three (6%) of 50 normal subjects and only four (13%) of 30 control patients with RP with no CME (Figures 1 & 2).

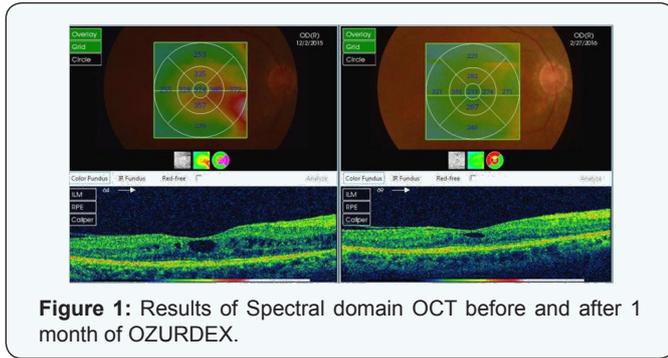


Figure 1: Results of Spectral domain OCT before and after 1 month of OZURDEX.

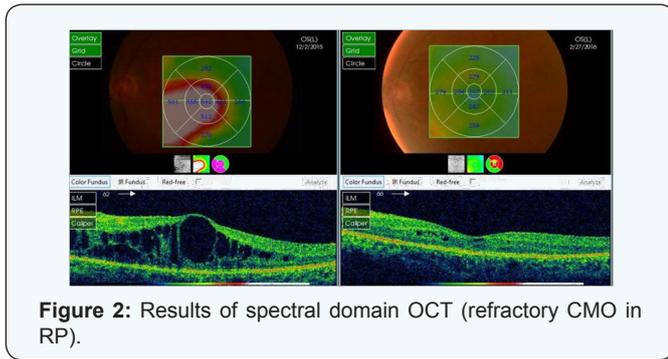


Figure 2: Results of spectral domain OCT (refractory CMO in RP).

Conclusion

This study suggests that patients with RP and CME may have an underlying autoimmune process that is contributing to the macular edema formation in RP. Based on these findings, Intravitreal steroids were administered in small case series with RP-related CME to modulate this autoimmune process and the inflammatory mediators, in our case, the patient has received multiple anti VEGF injections of Lucentis with no improvement, but he responded very well anatomically and functionally to Intravitreal steroid implant of OZURDEX, the results in our case was matching other small case series results, EX: In retrospective study of Ozdemir et al reported the efficacy of Intravitreal steroid in a group of five eye of five patients. Further preferably randomized trials may establish the place of Intravitreal dexamethasone in the treatment of refractory CME related to RP [6-14].

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