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TREATMENT OF INCONTINENTIA PIGMENTI RELATED RETINAL PATHOLOGIES

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Introduction:

Purpose: To evaluate the ocular clinical features of Incontinentia pigmenti (IP) related retinal pathologies and determine the surgical and laser treatment outcomes.

Materials and methods:

The clinical records of IP patients with at least 12 months of follow up in our clinic between the years 2016-2024 were retrospectively analysed. Fourteen eyes of 7 female patients were included in the study. Indirect ophthalmoscopic findings, color fundus photographs, B-scan ultrasonography and FA with RetCam were analysed. Demographic features, family histories, associated systemic findings, genetic mutation analysis in addition to ophthalmological findings, symmetry of the disease between the eyes, applied treatments, and results of the treatments during follow up were evaluated. Laser photocoagulation (LPC) was applied to avascular peripheral retina and vitrectomy was performed in eyes with tractional retinal detachment (TRD).

Results:

The mean age of patients was 4.5 months (0.5 -14months). Mothers of the 3 babies had the same disease. Skin lesions were detected in 100% of the patients, within postnatal first months. Alopecia and dental anomalies were recorded in 3 patients. IP was confirmed with molecular analysis in 4 patients with different gene mutations. Retinal findings on application were; TRD in 7 eyes, one of which was accompanied with vitreous hemorrhage, 1 had epiretinal membrane. Peripheral avascular retina was detected in 6 eyes, The median duration of follow-up was 3 years (range, 1-8years). Two of the TRD eyes were evaluated to be inoperable. Of the 7 patients with TRD, 6 of the fellow eyes had only peripheral avascular retina which points out the asymmetric nature of the disease. 6 of the eyes with TRD and 1 eye with epiretinal membrane (ERM) underwent vitrectomy with or without lensectomy. Six eyes underwent LPC for the peripheral avascular retina which were all stable, with good visual outcomes (Fixation and following- 1.0 Snellen). 4 out of 6 eyes undergoing LFC, needed a second laser session. Anatomical success could be achieved in 71% of the operated cases. However, visual acuity varied between LP and HM in eyes operated for TRD

Conclusions:

IP is usually an asymmetric disease, causing tent like TRD in one eye and relatively silent with only avascular peripheral retina in the other eye which can be controlled very effectively with LPC. TRD carries a guarded prognosis in terms of functional outcomes, despite the better rates of anatomical success of vitrectomy. This may be attributed to the asymmetrical nature of the disease, and the deep, inevitable amblyopia.