Abstract 109

EXTRAFOVEAL RETINAL REMODELLING AS AN EARLY BIOMARKER OF ABCA4-RELATED RETINAL DEGENERATION

Poster

Coppé A.M.*[1], Iarossi G.[1], Falsini B.[1], Lapucci G.[2], Buzzonetti L.[1]

[1]Ospedale Pediatrico Bambino Gesù ~ Rome ~ Italy, [2]Studio Oculistico Coppé ~ Rome ~ Italy

Purpose:

Retinal remodelling (RR) is a common process in inherited retinal dystrophies (IRDs). Studies on animal models (Denilinger B, 2020) indicate that this process is triggered by photoreceptor loss and/or dysfunction. The aim of this study was to evaluate RR in ABCA4-related IRD by using spectral domain optical coherence tomography (SD-OCT).

Methods:

A cohort of children or adolescents affected by IRD, with a clinical Stargardt type I phenotype and an ABCA4 genotype, were examined by SD-OCT. Volumes and thicknesses at the 9 ETDRS retinal fields were measured by a semi-automated segmentation by two different observers. Foveal results were excluded. Relative thicknesses and volumes, expressed by the quantitative ratios between layers were determined. Results were compared with those obtained from an age-matched control group.

Results:

Average thickness and volume of the outer nuclear and retinal ganglion cell layers were reduced ($p < 0.01$) in patients compared to controls. This reduction was independent of genotype severity and age of patients. RR showed a columnar aspect, showing a topographical correspondence between outer and inner pathological changes.

Conclusions:

Extrafoveal RR, as expressed by thinning of outer nuclear and retinal ganglion cell layers with a topographic correspondence, may be an early anatomic biomarker of ABCA4-related IRD. Further studies will clarify if RR with the observed features can be found associated with other genotypes causing juvenile macular degeneration.