

Presence of a Triple Concentric Autofluorescence Ring in *NR2E3*-p.G56R-Linked Autosomal Dominant Retinitis Pigmentosa (ADRP)

Advances in multimodal retinal imaging with widefield lenses have dramatically improved patient examination. In a paper entitled “Double Concentric Autofluorescence Ring in *NR2E3*-p.G56R-Linked Autosomal Dominant Retinitis Pigmentosa” and published in this journal in 2012,¹ we reported the presence of a double concentric hyperautofluorescent ring of fundus autofluorescence (FAF) as a highly prevalent and possibly pathognomonic early phenotypic marker for this dominant retinitis pigmentosa (RP). These observations were based on the use of a 55° lens for fundus autofluorescence imaging (Heidelberg Retina Angiograph 2; Heidelberg Engineering, Heidelberg, Germany). At present, ultra-widefield scanning laser ophthalmoscopy allows us to acquire from 102° (Heidelberg Retina Angiograph 2; Heidelberg Engineering) to 200° views (ultra-widefield Daytona Optomap; Optos, Dumferline, UK) of the patient’s retina in routine examination. When we recently re-examined the patient affected by *NR2E3*-p.G56R-linked ADRP described in Figure 7C of the 2012 paper with a 200° ultra-widefield imaging technology (Optos), we observed a third ring of hyperautofluorescence in the periphery of the retina, in addition to the previously observed double hyperautofluorescent rings (Fig. 1). Therefore, a triple ring of hyperautofluorescence is present in this patient.

In 2014, the patient was then 24 years old; imaging of fundus autofluorescence with a 55° lens on a confocal scanning laser revealed no changes in the concentric double ring of autofluorescence we observed with a 30° lens in 2011.¹ The inner hyperautofluorescent ring was still located in the perimacular region and the outer ring around the optic disk and along the vascular arcades (Fig. 1A). The recent re-examination at the age of 26 years with the 200° retinal

imaging system revealed now this third ring of hyperautofluorescence in the periphery (Fig. 1B). We hypothesize that the rim of this hyperautofluorescent ring progresses centripetally toward the midperiphery. Indeed, we observe numerous pigment deposits in the far periphery, suggestive of retinal degeneration having already occurred there. Remarkably, in the midperiphery where the previously reported double hyperautofluorescent ring is present, the outer ring has completely separated from the inner ring in the inferior perimacular region, consistent with our hypothesis that the outer ring progresses centrifugally toward the periphery.

To our best knowledge, this is the first report of a triple concentric ring of hyperautofluorescence in RP patients. With a growing number of clinics using widefield retinal imaging systems and given the relatively high incidence of the *NR2E3*-p.G56R mutation among ADRP patients, we anticipate this triple ring to be observed more frequently.

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References

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FIGURE 1. Fundus autofluorescence examination of a previously reported *NR2E3*-p.G56R-linked ADRP patient. (A) Examination of FAF of the patient at age 24 years with a 55° lens (left eye). The outer hyperautofluorescent ring is located along the vascular arcades, except for the inferior perimacular region, where the inner and outer rings are barely separated. (B) Examination of FAF of the patient at age 26 years with a widefield 200° lens (left eye). A third hyperautofluorescent ring is present in the periphery. Numerous small pigment deposits are present in the far periphery, beyond the third hyperautofluorescent ring. In the midperiphery, where the previously reported double hyperautofluorescent ring is present, the outer centrifugal hyperautofluorescent ring has completely separated from the inner ring in the inferior perimacular region. (C) Schematic representation of the FAF findings in *NR2E3*-p.G56R-linked ADRP patients. The innermost perimacular hyperautofluorescent ring (red dotted line) progresses centripetally (red line) toward the fovea (filled grey circle). The outer hyperautofluorescent ring (blue dotted line) located around the optic disk (black circle) and along the vascular arcades (black lines) progresses centrifugally (blue line) toward the midperiphery. The third hyperautofluorescent ring (dotted green line) progresses centripetally (green line) and delimits the far peripheral region where pigment deposits are present (black stars).



