### Title

A microstructural retinal analysis of membrano-proliferative glomerulonephritis type II




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## LETTERS

# A microstructural retinal analysis of membrano-proliferative glomerulonephritis type II

Membranoproliferative glomerulonephritis type II (MPGN type II)/dense deposit disease (DDD) is characterised by electron deposits within the lamina densa of the glomerular basement membrane. Systemic manifestation may include lipodystrophy and a retinopathy characterised by drusenoid deposits.[1–3] MPGN type II is caused by overactivation of the alternative complement pathway (C3 Nephritic Factor, Factor H deficiency or functional defects) in the majority of cases.[4] Recently, Factor H H402Y has been identified as a major-risk haplotype within the alternative complement pathway.[5] In vivo microstructural identification and localisation of these retinal deposits in humans are shown for the first time.

## CASE REPORT

A 54-year-old woman who presented with a history of blurred vision in her right eye, nyctalopia and photophobia for 5 years, was referred with the possible diagnosis of fundus flavimaculatus/RP. The patient's past medical history revealed lipodystrophy (most remarkable in the face, fig 1A) diagnosed at age 11. She was diagnosed as having glomerulonephritis at age 21, for which she had renal transplants at ages 27 and 41. The patient also required insertion of a pacemaker at the age of 52 years. The diagnosis of MPGN type II was confirmed at the time of her first renal transplant. At the age of 54 years, her visual function assessment showed reduced visual acuity (right eye: 0.20 logMAR; left eye: 0.22 logMAR), reduced contrast sensitivity (either eye: 1.05 log) but normal colour vision when tested with HRR plates. Rod- and cone-mediated function was reduced significantly in the recorded

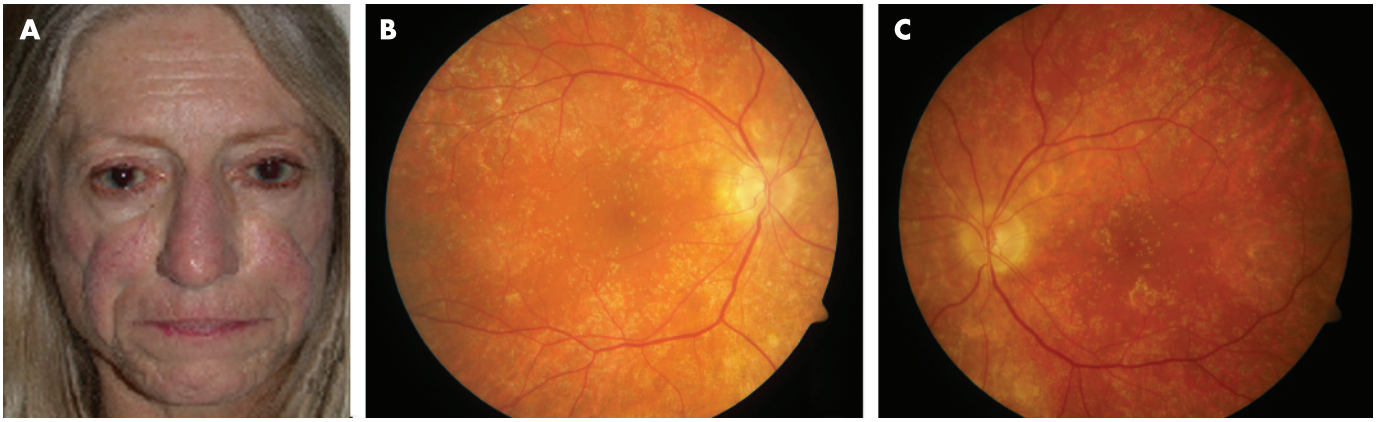


**Figure 1** Face (A) photograph illustrates extended lipodystrophy typical of MPGN type II. Fundus photographs of the right (B) and left eye (C) show drusenoid deposits involving the posterior pole and the peripapillary area.

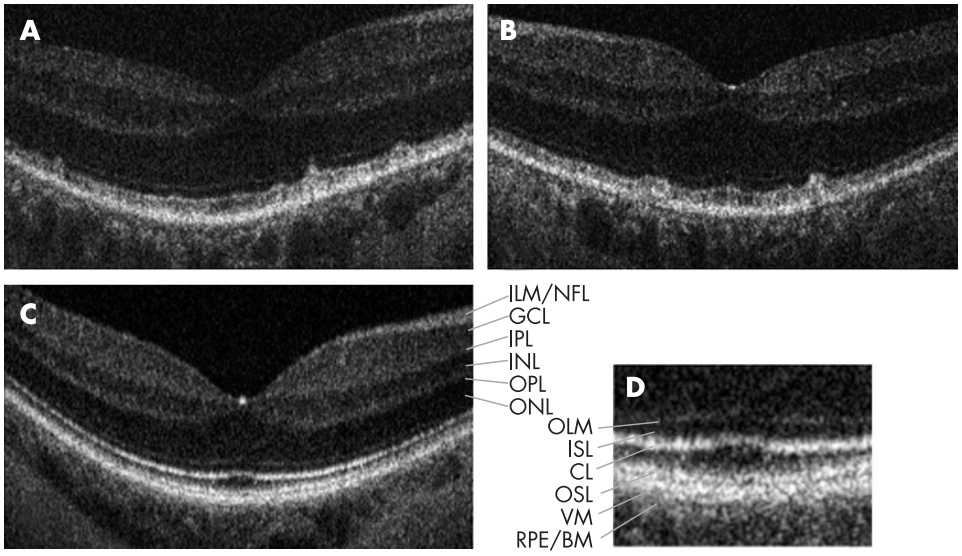


**Figure 2** Horizontal Fourier-domain OCT 5 mm scans through the macula from the patient's right (A) and left eye (B): well-demarcated deposits within the RPE/Bruchs membrane extending through the outer and inner segment layer up to the outer limiting membrane. Retinal lamination is preserved. Retinal lamination depicted in the macular scan of an age-matched control subject shown for comparison (C; foveal magnification: D). CL, connecting cilia; GCL, ganglion cell layer; ILM/NFL, internal limiting membrane/nerve fibre layer; INL, inner nuclear layer; IPL, inner plexiform layer; ISL, inner segment layer; OLM, outer limiting membrane; ONL, outer nuclear layer; OPL, outer plexiform layer; OSL, outer segment layer; RPE/BM, retinal pigment epithelium/Bruch membrane; VM, Verhoeff membrane.

full-field electroretinogram (ISCEV standard[6]) (rod-isolated b-wave: 20%; rod–cone a- and b-wave: 23% and 26%; cone a- and b-wave: 23% and 44%; 30 Hz flicker response: 34% of the mean control) when compared with age-matched control data. Fundus examination of both eyes demonstrated small yellowish partially confluent drusenoid deposits, which were irregularly distributed within the macular area and the posterior pole involving the peripapillary area (fig 1B,C).

High-speed (1000 A-scans/frame; 9 frames/s) high-resolution (axial 4 μm; lateral: 10–15 μm) Fourier-domain optical coherence tomography (Fd-OCT)[7] constructed at UC Davis, combined with a hand-held scanner (Bioptigen, Triangle Park, NC), was used for the acquisition of retinal layer images. Microstructural Fd-OCT analysis revealed an intact inner retinal structure compared with an age-matched control subject.[8] Deposits were observed within the RPE/Bruchs membrane complex, extending through the outer and inner segment layer up to the outer limiting membrane (fig 2).

## Comment

Microstructural in vivo retinal high-resolution imaging identified the localisation and the extent of retinal deposits affecting the RPE/Bruch membrane complex. The mild impact on vision function, characteristic of MPGN type II, is supported by the integrity of retinal lamination including inner and outer segments, which is in contrast to other hereditary macular dystrophy where the photoreceptor structure is disrupted.[9]

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## Doubling of optic disc

Doubling of optic disc is rarely seen and must be differentiated from a pseudo-double disc.[1 2] A 54-year-old woman referred with poor vision. The patient had more than 30-year myopia. The best corrected visual acuity was 0.8 OD with −7.00DS/−1.50DC×10° and 1.0 OS with −4.50DS/−1.50DC×150°. Examination of the anterior segment was normal. Funduscopy examination revealed myopia fundus in both eyes. Appearance simulating a second optic disc in the left eye was located about 1 disc diameter below the actual optic disc, the two being connected by a retinal vessel. The vessels emerging from below the optic disc supplied blood to the inferior retinal. The retinal surrounding the simulating optic disc was hypopigmented. The parents of this patient' family did not have a history of this disorder. FFA (fig 1) revealed that the two discs had separate

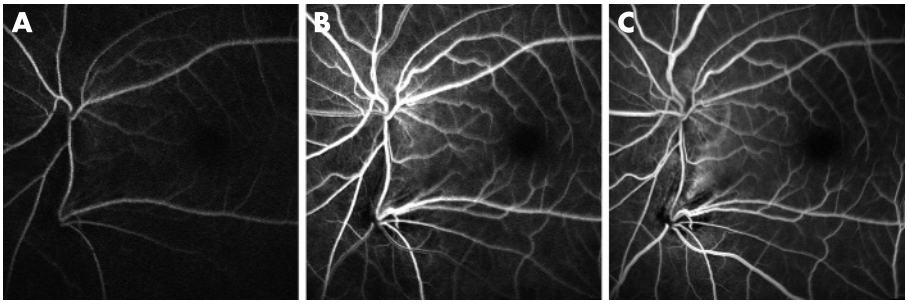


**Figure 1** Early filling of the arterioles coming from the upper disc while the arterioles emerging from the pseudopapilla reveal delayed fluorescence (A). The vessel connecting the two optic discs was also filled at the arterial phase (A). The venous phase shows that the venous drainage was present at the two optical discs (B, C).

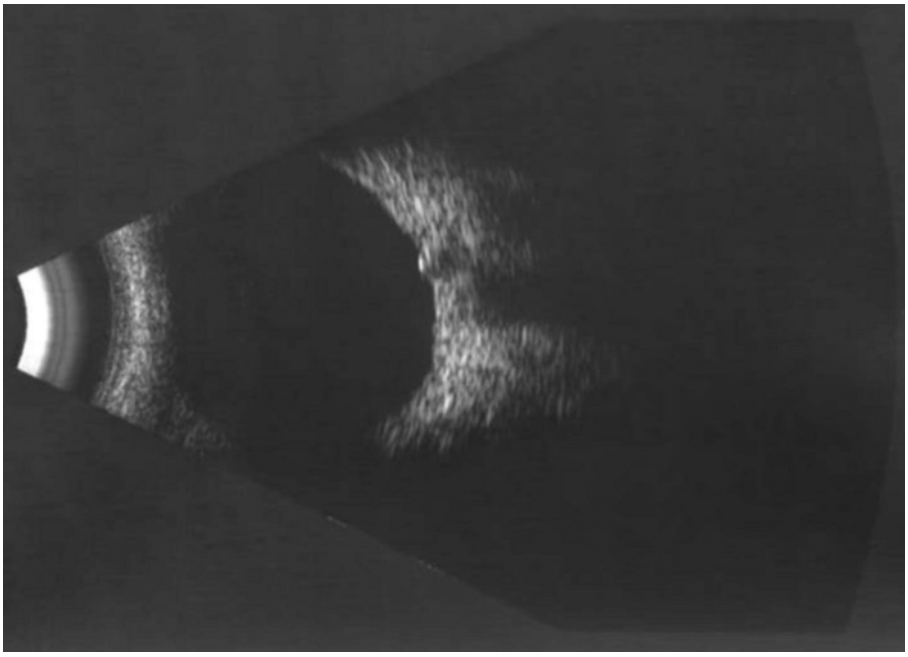

**Figure 2** B-ultrasonagraphy suggested two areas without echo-like optic disc manifestation.

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## Notes