Sir,

Retinal and choroidal angiopathy and electrophysiological disturbance in a case of amyloid AA

The common ocular manifestations of amyloidosis include deposition in the ocular adnexa, conjunctiva, cornea, glaucoma, and vitreous.[1] Retinal and choroidal involvement is rare.

Case report

A 35-year-old woman complained of progressive nyctalopia. She had previously been diagnosed of secondary AA amyloidosis affecting her kidneys, bone marrow, thyroid gland, and colon (confirmed on biopsy). Despite extensive investigations no underlying disease was identified.

Her vision was 6/6 in each eye. Ocular adnexae, anterior segments, and intraocular pressures were normal. Fundoscopy showed widespread small drusen-like white deposits, diffuse pigmentary changes, and microaneurysms (Figure 1). Autofluorescence imaging showed a diffuse speckled pattern (Figure 2a). Fluorescein angiography (FA) showed multiple microaneurysms (Figure 2b). Indocyanine green (ICG) angiography showed dilated large choroidal vessels with variable calibre, and hyperfluorescence and staining along retinal and choroidal vessels (Figure 2c and d). OCT showed normal foveal thickness. Electrophysiological testing suggested maculopathy (reduced P50 on pattern ERG; increased latency from central and peripapillary rings from both eyes on multifocal ERG) (Figure 3a). Full-field ERG showed decreased and delayed scotopic and photopic responses with borderline maximal responses, suggesting generalized photoreceptor dysfunction (Figure 3b).

Comment

Only a few cases of retinal changes have been described in FAP.[2–4] These described pinpoint white deposits, retinal vascular sheathing, microaneurysms, and scattered retinal haemorrhages. Angiographic changes similar to our patients were seen (vascular closure, focal staining, and microaneurysms on FA; hyperfluorescence and staining along choroidal and retinal vessels on ICG).

Although the fundal changes could potentially be due to an underlying disease causing the secondary amyloid, they are strikingly similar to those described in FAP and amyloid AL. We propose that the changes result from amyloid deposition in the choroidal and retinal vasculature, which causes ischaemia and further photoreceptor and macular dysfunction. The microaneurysms and pigmentary changes are likely the result of retinal ischaemia. The pinpoint white deposits are similar in appearance to drusen and probably reflect amyloid deposition within the retina, resulting in the speckled AF pattern. Indeed Aβ amyloid has been identified in drusen and may have a role in the pathogenesis of age-related macular degeneration.[5] These new findings may help in further understanding

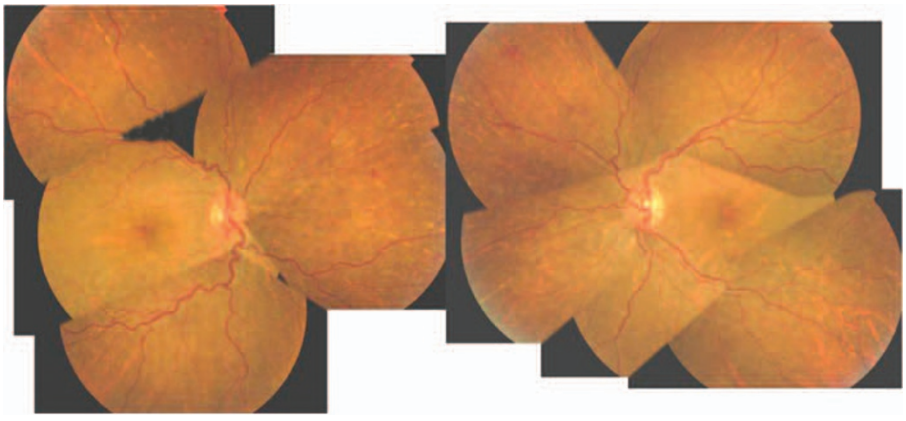

Figure 1 Fundus photography showing widespread small drusen-like white deposits and diffuse pigmentary changes.

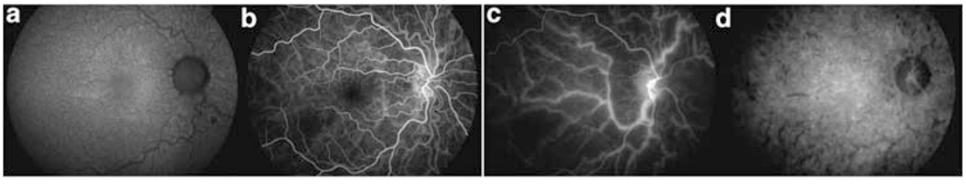

Figure 2 (a) Speckled pattern on autofluorescence imaging. (b) FFA showing multiple microaneurysms. (c) ICG showing variable-calibre choroidal vessels and (d) staining along retinal and choroidal vessels.

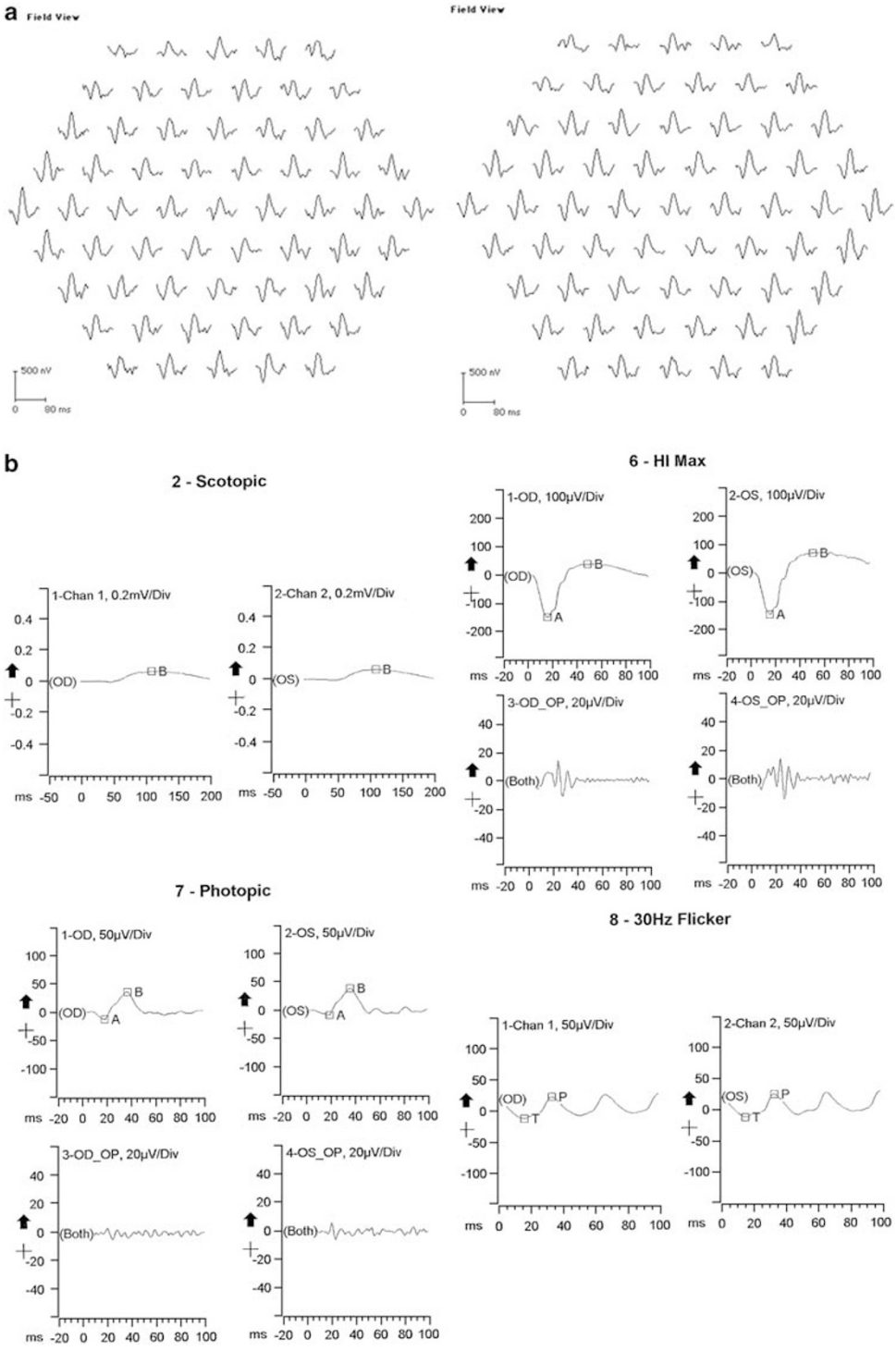

Figure 3 (a) Multifocal ERG tracing showing reduced and delayed responses from the central rings. (b) Full-field ERG tracings showing decreased and delayed scotopic and photopic responses, and borderline maximal responses.

the ocular involvement of amyloidosis in forms other than FAP.

Conflict of interest

The authors declare no conflict of interest.

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Eye (2010) **24,** 1117–1119; doi:10.1038/eye.2009.259; published online 6 November 2009

Sir,

Comment on macular full-thickness and lamellar holes in association with type 2 idiopathic macular telangiectasia

The article by Charbel Issa *et al*[1] is an interesting report on the association of type 2 idiopathic macular telangiectasia (IMT) with both full-thickness and lamellar macular holes (MHs). The aetiology of IMT is unknown, but possibilities include chronic leakage from hyper-permeable capillaries and ischaemia. The authors hypothesise that MHs in IMT are caused by tissue loss consequential to these factors, with secondary draping of an as yet undefined membrane, possibly ILM, over these areas rather than the accepted concept of epiretinal tractional forces that result in idiopathic MHs.

We describe a case of IMT with ERM that showed visual improvement with vitrectomy, which is relevant to the aetiology of IMT and the evolution to a MH. A 66-year-old pseudophakic woman presented with a 3-year history of gradually reducing central vision. Fundoscopically she had the typical signs of IMT, which were confirmed on angiography. In addition, she had very fine surface ERM in both eyes, which was worse in the left eye. Vitrectomy was carried out on the left eye, with separation of an incompletely attached posterior hyaloid face and peeling of the ERM. Postoperatively her vision improved from 6/36 to 6/12. In view of the improvement in the left eye, a similar surgery was carried out on the right eye. Once again, visual acuity improved from 6/60 to 6/18. Her vision remained stable at follow-up 3 years later.

ERM formation can be seen in a number of situations in which chronic retinal vascular leakage and hypoxia occur, most notably in diabetic maculopathy. ERM formation in these situations is likely to be driven partly by hypoxic cytokine-driven tissue repair.[2–4] Perhaps in IMT, this also has a significant role in leading to a detrimental repair process causing ERM, and in some cases, traction. Vitrectomy increases oxygenation in the vitreous cavity, increasing the availability of oxygen to the retina and thus potentially dampening tissue repair processes, as well as improving retinal function.[5]

Vitrectomy may therefore both remove detrimental traction and improve retinal oxygenation. It is possible that patients with early symptomatic IMT, particularly those with epiretinal membranous changes, could actually benefit from vitrectomy and membrane peeling before atrophic MH formation occurs. Further study on this aspect is needed.

Conflict of interest

The authors declare no conflict of interest.

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Eye (2010) **24,** 1119; doi:10.1038/eye.2009.243; published online 2 October 2009

Sir,

Sequential corneal infection with two genotypically distinct *Acanthamoebae* associated with renewed contact lens wear

Acanthamoeba keratitis (AK) is a rare infection that is estimated to occur in between 1–100 cases per million