and cite the EUCLID substudy as corroborative evidence.[4] In that particular study, the researchers measured *plasma* as apposed to *serum* VEGF levels,[4] making a comparison difficult. Indeed, the use of serum to measure systemic VEGF levels has to be interpreted with caution, because activated platelets (during blood clotting) release VEGF into serum, and thus, results based on serum samples may be inaccurate in view of the possible artifact relating to the source of VEGF levels.[5] Also, the EUCLID study did report an upward trend in plasma VEGF levels, with increasing retinopathy (no retinopathy *vs* nonproliferative *vs* proliferative) (mean levels pg/ml: 11.5 *vs* 12.9 *vs* 16.1, respectively), an association that approached statistical significance ($P=0.06$) but was severely confounded by the disproportionate numbers in the groups (eg for nonproliferative $n=167$, proliferative $n=8$). In our own study, plasma levels of VEGF were elevated in diabetic patients, with the highest levels seen in more severe retinopathy (grade 2 and 3) ($P<0.05$).[3]

Furthermore, the title of their article[1] states circulating 'endothelial progenitor cells' but in fact, the study concentrates on stem cells (c-Kit+) and haematopoietic progenitor (CD34+) mononuclear cells. Increasingly, evidence is emerging that endothelial precursor cells are a heterogenous population with potentially different parent cell lines, but with some overriding phenotypic similarities in markers such as CD34, CD133, and VEGF receptor-2 (KDR). Furthermore, to prove endothelial lineage of these cells, uptake of acetylated low-density lipoprotein and Ulex Europaeus lectin should be demonstrated.[6] Thus, Lee *et al*[1] do not positively confirm that the increase in c-Kit+ and CD34+ cells in their study is due to a concurrent increase in the endothelial progenitor cell fraction, and we suggest that they should be more circumspect in their conclusions.

Lastly, the study by Lee *et al*[1] suffers from the obvious limitations of a cross-sectional design, and the many associated co-morbidities frequently seen with diabetic patients (hypertension, obesity, cardiovascular disease, etc) and drug treatments have to be fully taken into consideration, especially in a relatively small study. Many comorbidities in diabetics (as well as drugs such as angiotensin converting enzyme inhibitors and statins) could have a large effect on the various indices measured, making their results difficult to interpret.

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Sir,

Peripapillary choroidal neovascularization in Bietti crystalline retinopathy

Tapetoretinal degeneration with crystals at the corneal limbus, numerous yellow, glistening retinal deposits, and progressive atrophy of the retinal pigment epithelium (RPE) and choroid was reported by Bietti in 1937.[1] The corneal deposits are detected in only about one-third of patients, and when corneal changes are absent, the term 'Bietti crystalline retinopathy' is used.[2,3] The inheritance

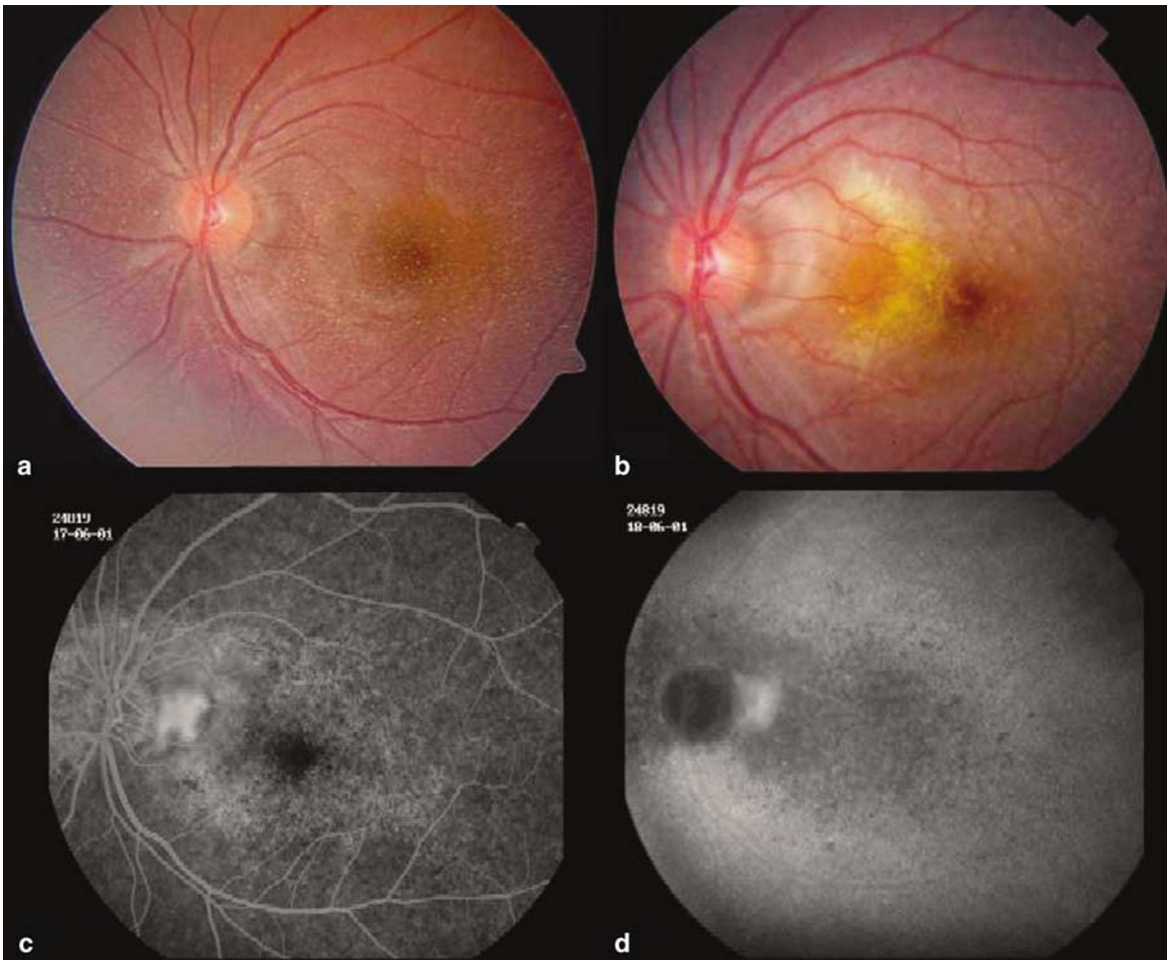

Figure 1 (a) Fundus photography of left eye at first examination revealing intraretinal crystals scattered throughout posterior pole. (b) Fundus photography of left eye 14 months after first examination. Exudation with localized retinal detachment at temporal side of optic disc extending to fovea is seen. (c) Late stage fluorescein angiogram demonstrating poorly defined hyperfluorescent peripapillary membrane with leakage and hypo- and hyperfluorescent spots at posterior pole. (d) ICGA showing hyperfluorescent peripapillary membrane with hypo- and hyperfluorescent spots.

is autosomal recessive and mutations in *CYP4V2* gene on chromosome 4q35 has been associated with Bietti crystalline retinal dystrophy.[4]

Central visual loss usually occurs in the later stages of the disease due to RPE and choroidal atrophy.

Choroidal neovascularization (CNV) may be seen with any pathological process disturbing the RPE and Bruch's membrane. However, to the best of our knowledge, CNV has not been reported in Bietti crystalline retinal dystrophy. Here, we report a case of peripapillary CNV with serous retinal detachment in Bietti crystalline retinal dystrophy.

Case report

A 13-year-old girl was referred to our institution for further evaluation of a presumed retinal dystrophy. Her past medical and family histories were unremarkable. She did not have any visual complaints. Visual acuity was 20/20 and anterior segment examination was normal in both eyes. However, fundus examination revealed bilateral numerous yellowish, glistening intraretinal crystals distributed throughout the posterior pole and peripapillary RPE atrophy (Figure 1a). Visual field examination was normal with Humphrey 30-2 standard automated perimetry.[5] Electrophysiological tests were performed within the limits of ISCEV standards[6,7] and electroretinography showed normal photopic and scotopic responses. Electro-oculography was also normal, with an Arden ratio of 2.08 OD and 2.06 OS. The patient was diagnosed with Bietti crystalline retinal dystrophy on a clinical basis and it was not possible to confirm the diagnosis by molecular analysis. After 14 months, she came for urgent re-evaluation because of a sudden decrease in vision to 20/50 in her

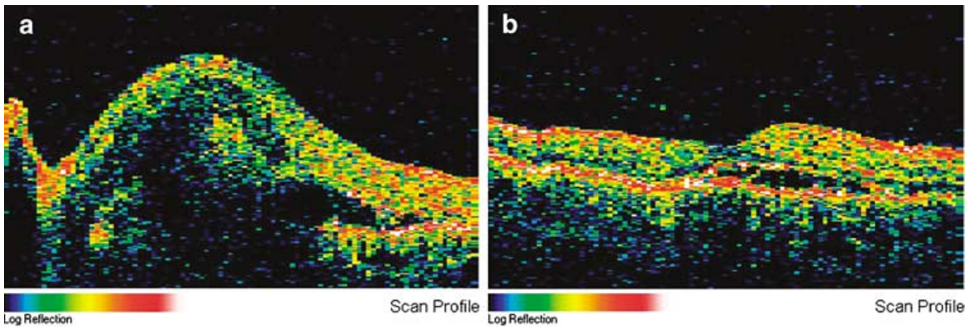

Figure 2 The optical coherence tomography revealed sensory retinal detachment and serous retinal pigment epithelial detachment extending from optic disc to fovea. (a) Nasal-to-temporal, (b) superior-to-inferior.

left eye. Fundus examination revealed exudation with localized retinal detachment at the temporal side of the optic disc extending to the fovea (Figure 1b). There was no autofluorescence of the disc, and fluorescein angiogram demonstrated a rather poorly defined hyperfluorescent peripapillary membrane with a hypo- and hyperfluorescent spots at the posterior pole in the early stages and leakage from the peripapillary membrane in the late stages (Figure 1c). The indocyanine green angiogram (ICGA) showed a hypofluorescent peripapillary membrane with hypofluorescent spots at the posterior pole in the early stages and a hyperfluorescent peripapillary membrane with hypo- and hyperfluorescent spots at the posterior pole in the late stages (Figure 1d). Optical coherence tomography revealed a sensory retinal detachment and serous retinal pigment epithelial detachment extending from the optic disc to the fovea (Figure 2a and b).

Comment

It has been reported that visual acuity is relatively good during the early stages of Bietti crystalline retinopathy with subsequent progressive but rather slow visual loss over decades due to chorioretinal atrophy.[8] In our case, central vision deteriorated rapidly due to extension of exudation of peripapillary CNV to the fovea. The mechanism of CNV formation in our case is unclear; however, RPE and choroidal atrophy may have played a role.

In this case, ICGA showed hypofluorescent spots at the posterior pole in the early stages and hypo- and hyperfluorescent spots in the late stages. These findings could be explained by the study of Salati *et al*,[9] where early phase ICGA revealed focal lobular areas of choriocapillary atrophy at the equator, with concomitant RPE changes at the posterior pole on FA in a patient with Bietti crystalline retinopathy. Progressive sclerosis of ciliary and choroidal arteries was also noted on ICGA.

In conclusion, CNV can occur in Bietti crystalline retinal dystrophy. Therefore, in patients with Bietti crystalline retinal dystrophy and rapid visual loss, CNV should be suspected.

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Sir,

Exudative retinal detachment following deep sclerectomy in Sturge–Weber syndrome

Trabeculectomy for Sturge–Weber (SW) syndrome is associated with a risk of choroidal effusions and haemorrhage.[1] To minimise these complications, a two-staged Baervedlt implant combined with prophylactic posterior sclerotomies[2] has been suggested. Deep sclerectomy (DS) has been reported as a successful treatment modality in one case of glaucoma associated with SW syndrome.[3] We present two patients with SW syndrome and glaucoma who underwent non-penetrating DS with mitomycin-C (MMC) augmentation by an experienced surgeon (NA) and were complicated with exudative retinal detachments (RD).

Case report 1

A 17-year-old male with SW syndrome, diffuse choroidal haemangioma, and intraocular pressures (IOP) in the 40 mm Hg range on maximal treatment, including oral acetazolamide, underwent DS and intraoperative MMC application. The anterior chamber (AC) was noticed to shallow as soon as the Schlemm's canal was de-roofed. Attempts to deepen the AC by injecting air and balanced salt solution through a paracentesis were unsuccessful. Two inferior V-shaped sclerotomies beginning 7 mm from the limbus were performed in the inferior quadrants and a moderate amount of straw-coloured fluid was drained. The AC deepened and a large air bubble was left to prevent contact between the iris and the trabeculo-descemet's membrane (TDM). The next day, the IOP was 34 mm Hg and a superior exudative RD involving the macula was observed even though no RD had been found on screening intraoperatively. Three days later, the detachment had resolved (the patient had been prescribed oral prednisolone 30 mg daily) and the visual acuity improved to 6/12 unaided (preoperatively 6/5). IOP remained high as the TDM was completely obstructed by iris tissue. A trabeculectomy with adjunctive MMC was performed 3 months later with prophylactic choroidotomies. Once again the eye developed a superior exudative RD. This resolved after photodynamic laser therapy (PDT) of the choroidal feeder vessels. On the last visit, the IOP was 18 mm Hg, but the visual acuity was 6/36 due to sub-retinal pigment deposition under the fovea following PDT (Figure 1).

Case report 2

DS with intraoperative MMC was performed on a 24-year-old male with SW syndrome and two previous failed trabeculectomies. The preoperative visual acuity was 6/12 and IOP was 29 mm Hg on Timolol and dorzolamide combination (Cosopt) eye drops. Three days later, a circumferential exudative RD was confirmed by our vitreo-retinal surgeon, who observed anterior proliferative vitreoretinopathy and early funnelling. B scan showed no choroidal detachment and total RD. Phacoemulsification and internal repair of RD was performed. No retinal break was found. Silicone oil was left for internal tamponade. Six months later, the silicone oil was removed and repeat trabeculectomy with intraoperative MMC was performed. Postoperatively he developed a posterior RD, which did not extend peripherally. It resolved spontaneously within a month. Final VA was 6/12. The trabeculectomy failed shortly after and the IOP was 22 mm Hg on maximal topical medical therapy on last follow-up.

Comment

Rebolleda *et al*[3] proposed 'non-perforation' of the AC in DS as a potential advantage over trabeculectomy as there

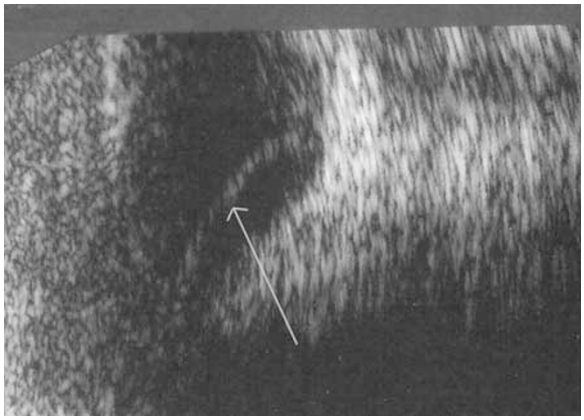

Figure 1 Low-frequency B scan ultrasonographic appearance of patient 1 showing an exudative retinal detachment (white arrow).