Shabeeba R Hannan, Krishnappa C Madhusudhana, Andrew J Lotery, Richard S B Newsom
Southampton Eye Unit, Southampton, UK

Andrew J Lotery
Human Genetics Division, University of Southampton, Southhampton, UK

Correspondence to: Dr R S B Newsom, Southampton Eye Unit, Tremona Road, Southampton S016 6YD, UK; richard.newsom@suht.swest.nhs.uk

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Choroidal osteoma in association with Stargardt's dystrophy

Choroidal osteoma usually occurs in 20–30 year old, healthy, white women as a well-defined, unilateral (75%), solitary, yellow orange, slow-growing juxtapapillary lesion.[1] This is the first time that a choroidal osteoma has been associated with Stargardt's dystrophy.

Case report

A 16-year-old girl presented with bilateral reduction of distance visual acuity (6/24) since 2 years. Near vision, colour vision, stereoacuity and anterior segments of both eyes were normal. Amsler grid testing in the left eye revealed a blur of the grid superotemporal to the foveal fixation point. Stereoscopic funduscopy revealed bilateral Stargardt's maculopathy (foveolar pigment epithelial changes with parafoveal orange-coloured flecks; figs 1A,B), with an approximately 2.5 mm yellowish subretinal lesion situated 2-disc diameters below the left optic disc (fig 1B). Fundus fluorescein angiography (FFA) revealed macular and perimacular hypofluorescence (dark choroid) with hyperfluorescence and late staining of the lesion inferior to the left optic disc (figs 1C,D). A/B ultrasound echography demonstrated a corresponding 0.9 mm thick, 2.5 mm diameter lesion with high, homogeneous internal reflectivity and marked shadowing of tissue signals posterior to the lesion (fig 1E), characteristic of choroidal osteoma. Serum calcium phosphate and parathyroid hormone levels, electroretinogram, electro-oculogram and visual evoked potential tests were normal. Goldman's perimetry revealed bilateral central scotomas (fig 1F). FFA and B ultrasound scan were used to monitor the patient for subretinal choroidal neovascular membranes (CNVM) or haemorrhage.

Comment

This is the first report of a choroidal osteoma occurring in a patient with Stargardt's maculopathy. The clinical and ultrasound features were characteristic for a choroidal osteoma as described above.

Choroidal osteomas present as unilateral yellow–white to orange–red juxtapapillary choroidal lesions, which can occasionally extend into the macula or involve both choroids.[1] The main differential diagnosis of choroidal osteoma is idiopathic sclerochoroidal calcification.

Sclerochoroidal calcification, common in 50–80 year old men, presents with bilateral (47% are unifocal), geographic, plaque-like or mildly elevated yellow–white fundal lesions along the retinal vascular arcades,[2 3] with rare multiple grey scleral lesions that resemble the choroidal lesion.[3]

Choroidal osteomas, common in women, increase in size in 41% of cases, and can cause a reported secondary poor visual acuity ($\leqslant$6/60) in 45% and 56% of eyes at the 5 and 10 years follow-up, respectively.[4] Spontaneous resorption of the lesion can occur. CNVM, subretinal fluid and haemorrhages complicate 33% of cases. Quantitative CT scans have demonstrated occult lesions in 90% of fellow eyes of clinically

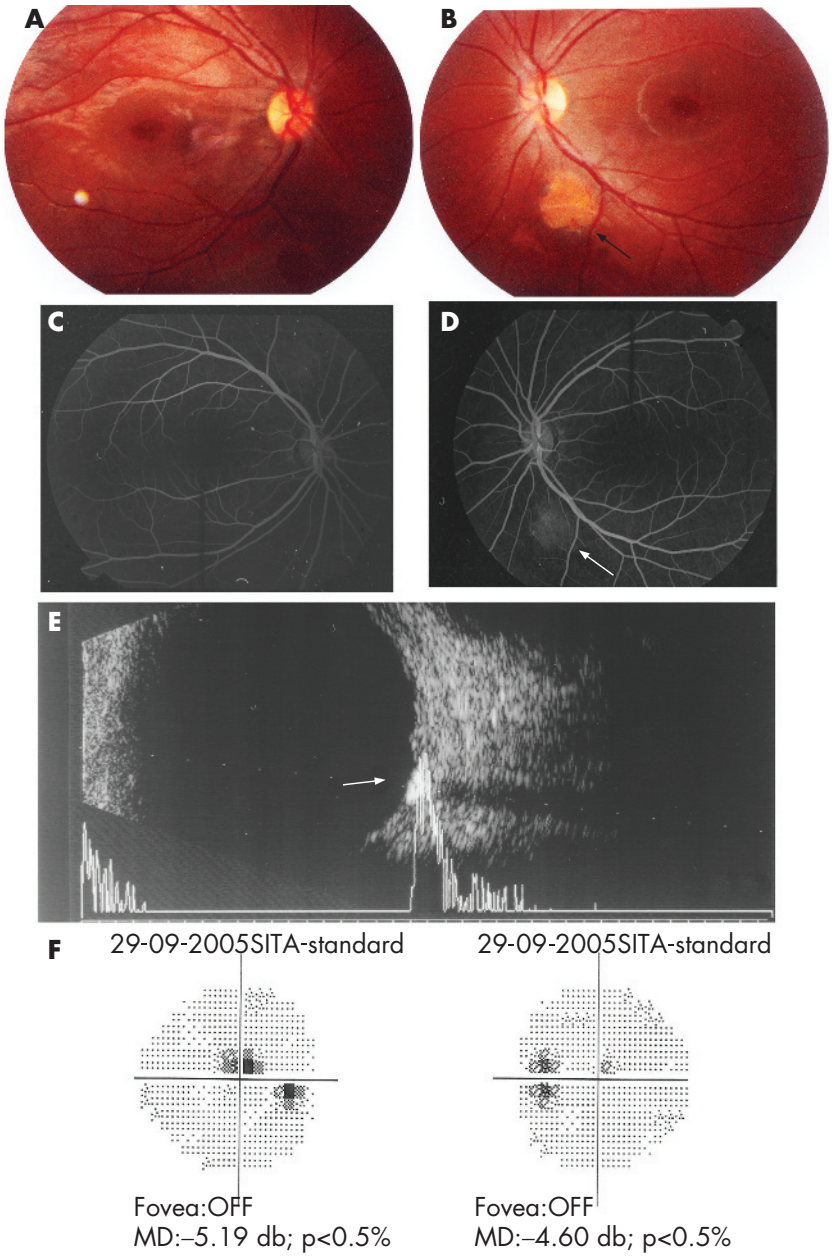

Figure 1 (A,B) Fundus photographs of right and left eyes revealing bilateral bull's eye maculopathy and left juxta papillary calcific lesion (arrow). (C,D) Fundus fluorescein angiography of right and left eyes. Bilateral dark choroids and hyperfluorescent juxtapapillary lesion in left eye. (E) B ultrasound scan reveals shadowing posterior to the highly dense lesion (A ultrasound scan: sound attenuation posterior to the lesion). (F) Perimetric central field defects. MD, mean deviation.

unilateral cases.[1] Oval or round choroidal osetoma lesions with scalloped margins range from 2 to 22 mm in basal breadth and 0.5 to 2 mm in height, with pseudopoidal extensions, surface telangiectases and altered pigmentation.[1] Central serous retinopathy has been associated with choroidal osteomas.[1 5] FFA reveals an early patchy hyperfluorescence and diffuse late staining with vascular tufts on the vitreal surface of the tumour that fill in the early phases.[1] CT scan demonstrates a radiopaque ''bone-like'' density of the lesion. Osteomas seem hyperintense on T1-weighted and hypointense on the T2-weighted MRI images.[1]

Choroidal osteomas must be distinguished by ultrasound/CT scan from adult vitelliform macular dystrophy, serpiginous chorioretinitis, central serous chorioretinopathy, calcified granulomas, trauma, Cogan's plaques and chorioretinal scarring.[1 2]

Idiopathic choroidal calcification has been associated with Rieger anomaly, limbal dermoids, retinitis pigmentosa, spontaneous choroidal haemorrhage, organoid nevus, epidermal nevus, Bartter and Gitelman's syndromes.

The pathogenesis in choroidal osteoma is unknown. Although an association of choroidal osteoma with Stargardt's maculopathy may be coincidental, the association could be related to a dysfunction of the retinal pigment epithelium or choriocapillaries. Complicated extrafoveolar CNVM are treated with photocoagulation/photodynamic/transpupillary thermo therapy. Submacular surgery is indicated in subfoveolar CNVM or haemorrhage.[1]

Edwin C Figueira
Department of Ophthalmology, Prince of Wales Hospital, Sydney, New South Wales, Australia

R Max Conway
Chatswood Grove Eye Clinic, Sydney, New South Wales, Australia; Save Sight Institute, University of Sydney, Sydney, New South Wales, Australia; Sydney Eye Hospital, Sydney, New South Wales, Australia

Ian C Francis
Department of Ophthalmology, Prince of Wales Hospital, Sydney, New South Wales, Australia

Correspondence to: Dr I C Francis, Chatswood Grove Eye Clinic, Suite 12, Chatswood Grove, 12–14 Malvern Avenue, Chatswood, Sydney, 2067, NSW, Australia; ifrancis@student.unsw.edu.au

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Agreement between two non-contact specular microscopes: Topcon SP2000P versus Rhine-Tec

Endothelial cell density (ECD) assessment with the noncontact Topcon SP2000P specular microscope is known to be reliable when the automated mode is followed by manual corrections (touched-up mode). We compared its agreement with Rhine-Tec, a new noncontact specular microscope, in 270 eyes of 160 patients, by comparing the ECD measured in the automated and touched-up modes with that of Topcon touched-up. Good agreement existed between either touched-up modes with a mean difference of only 2 cells/mm^2 95% CI (−27; 23) whereas agreement with the Rhine-Tec automated mode was poor with an overestimation by a mean of 226 cells/mm^2 95% CI (172; 281).

Background

The Topcon SP2000P non-contact specular microscope (Topcon, Tokyo, Japan) is widely used to measure corneal endothelial cell density (ECD). It uses a cell contour recognition algorithm based on contrast differences, and ECD derived from an automated delineation of cell boundaries with manual correction of inaccurately drawn cells (''touched-up'' mode) has been validated.[1 2] A new commercially available non-contact specular microscope Rhine-Tec (Rhine-Tec, Krefeld, Germany) determines ECD by a cell-centre method consisting of identification of centres of contiguous cells either in an automated or a touched-up mode using an operator-defined cell centre. We aimed to determine its agreement with the Topcon touched-up mode.

Methods

The central endothelium of 270 eyes of 160 subjects (healthy subjects and those operated on for cataract or penetrating keratoplasty to explore a wide range of ECDs) was imaged, first with the Topcon microscope and immediately afterwards with the Rhine-Tec specular microscope. On each sharpest image, the ECD and percentage of hexagonal cells were determined on the largest possible area by a trained ophthalmologist blinded to the clinical antecedents and choosing the device (Topcon or Rhine-Tec) randomly. Both automated and touched-up modes were used for Rhine-Tec, and the Topcon touched-up served as the reference.[1, 2] Agreement was determined using the Bland–Altman method.[3] Means were compared using paired t tests (or non-parametric Wilcoxon's test when required) and results considered significant when $p<0.05$.

Results

Mean (standard deviation (SD)) number of cells counted was 129 (46) (range 24–219, median 144) for Topcon and 137 (44) (range 47–279, median 136) for Rhine-Tec ($p<0.001$). Despite a good correlation between the Topcon touched-up and Rhine-Tec automated modes ($y = 0.37x+1497$, $r = 0.8$, $p<0.001$), the agreement was poor, with Rhine-Tec overestimating ECD by a mean of 226 cells/mm^2 95% CI (172 to 281) (fig 1). A continuous bias in ECD determination characterised by overestimation

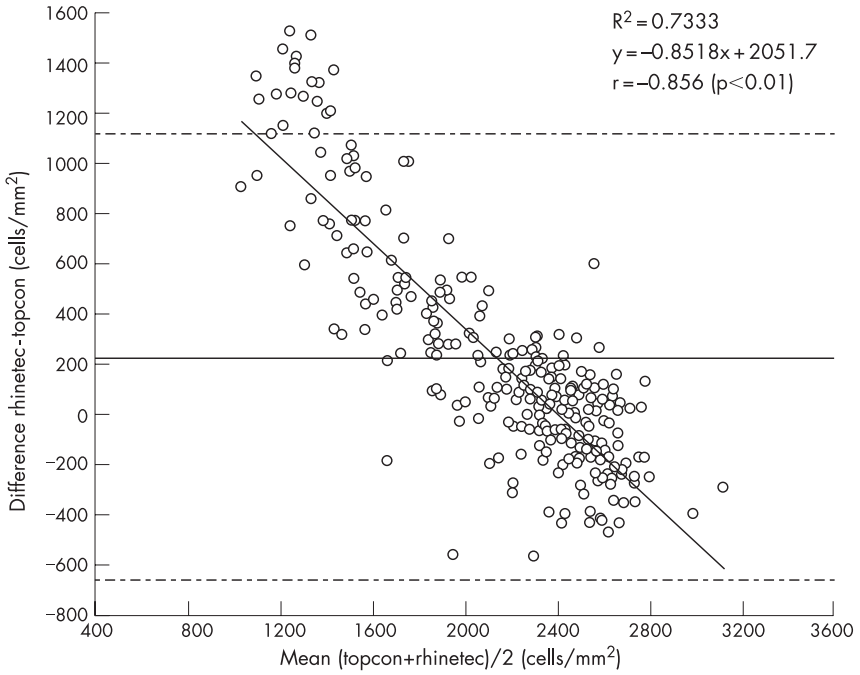

Figure 1 Difference against mean graph showing the agreement between the endothelial cell densities (ECDs) obtained with the Topcon touched-up mode compared with the Rhine-Tec automated mode. ECD measured in Topcon touched-up mode ranged from 417 to 3263 cells/mm^2, with 84 eyes (31%) having <1800 (low ECD), 109 eyes (40%) having between 1800 and 2500 cells/mm^2 and 77 eyes (29%) having >2500 cells/mm^2 (high ECD). The solid horizontal line indicated the mean difference between Rhine-Tec and Topcon (226 cells/mm^2). The agreement was poor with wide 95% limits of agreement (calculated as mean difference (±1.96 SD of the difference) of the difference) set between −663 and +1116 cells/mm^2 (dashed lines). The linear regression line (bold) indicated the significant overestimation of low ECDs and underestimation of high ECDs by Rhine-Tec.