

Sir,
A case of idiopathic perifoveal Telangiectasia preceded by features of cone dystrophy
 Idiopathic Perifoveal Telangiectasia (IPT), or type 2 macular telangiectasia, is an uncommon, nonexudative, bilateral condition of obscure etiology.^{1–3} Cone dystrophy is another uncommon macular disease.⁴ Here, we describe a patient who developed clear signs of IPT years after being diagnosed with cone dystrophy. It seems unlikely that two conditions arose independently.

Case report

A 57-year-old male presented with gradual impairment of reading vision and photophobia in 2004, 20/50 OU. Fundus examination showed subtle, relatively discrete retinal pigment epithelial (RPE) abnormalities in the

central maculae. Central-foveal reflexes were attenuated (Figure 1a and b) without loss of macular transparency. Fluorescein angiography revealed a central window defect corresponding to RPE abnormalities in both eyes without discernible vascular changes (Figure 1c–f). Some mild, diffuse hyperfluorescence in the temporal maculae was present (Figure 1e and f). Both eyes had normal rod-driven responses; cone-driven responses to both single-flash and 30 Hz flicker showed significantly reduced amplitudes in Ganzfeld–electroretinography⁵ (Figure 1i–l). Interestingly, OCT examination showed prominent inner–lamellar holes (Figure 1g and h). A diagnosis of a progressive cone dystrophy was made.

By 2007, visual acuity had dropped to 20/200 OU. Funduscopy revealed vascular telangiectasis and right-angled vessels in the temporal perifoveal region of both

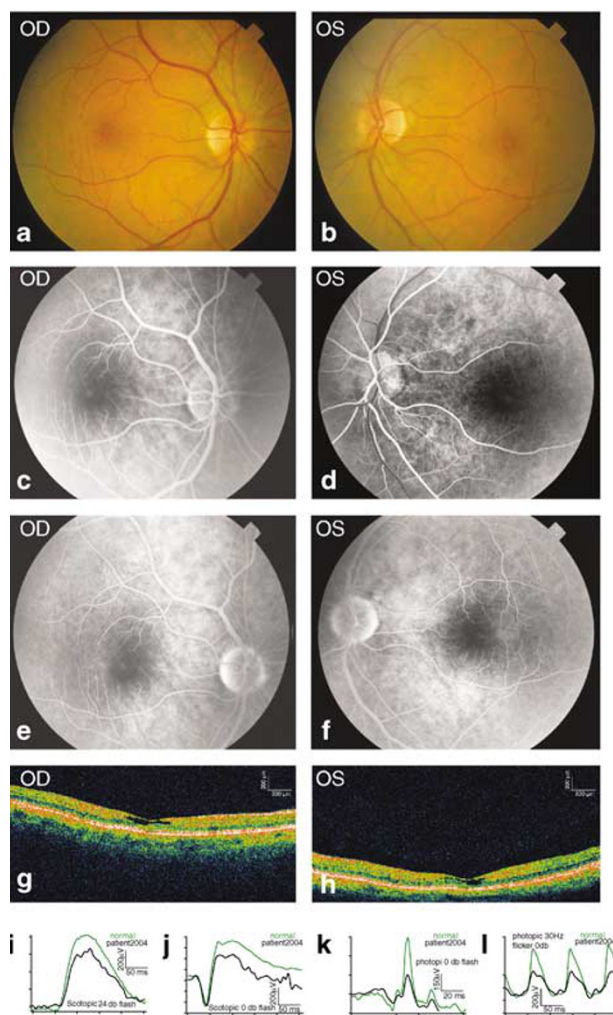


Figure 1 Right (a) and left eye (b) fundus photographs in 2004 show pigment changes in the central macula. Early fluorescein angiographs (c and d) reveal a central window defect. Late fluorescein angiography images (e and f) show irregularities in background fluorescence. No telangiectasis or vessel staining is visible. OCT images (g and h) exhibit foveal inner-lamellar holes. Ganzfeld–ERG recordings demonstrate normal rod-driven responses (i and j) and reduced amplitudes in cone-driven responses (k and l).

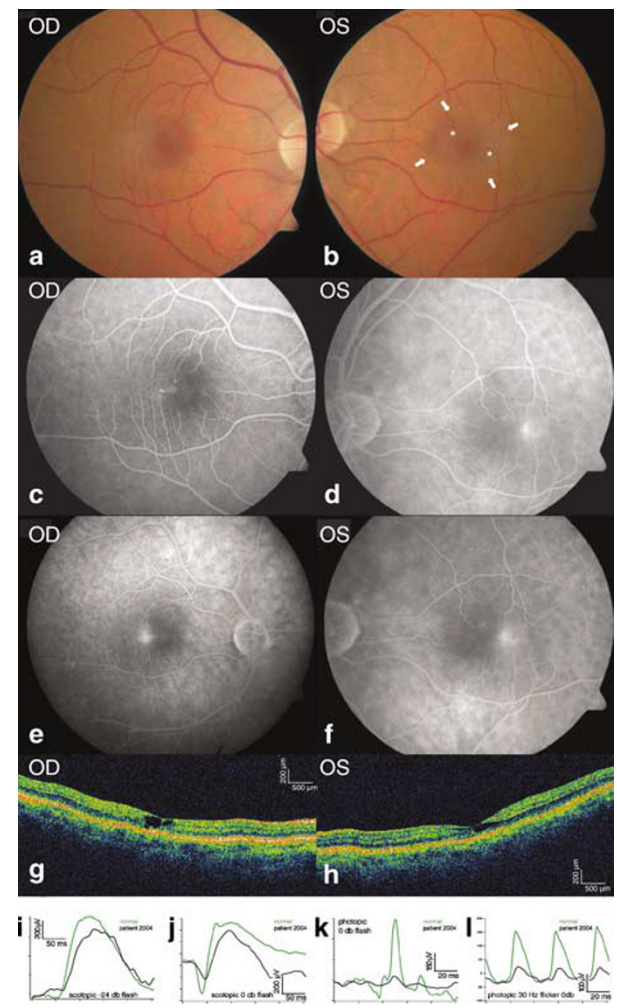


Figure 2 Fundus photographs taken in 2007 (a and b) show a loss of transparency (arrows) of the central macula and pigment abnormalities with right-angled vessels (*) and crystalline deposits in the temporal perifovea. Early fluorescein angiography images (c and d) show telangiectatic vessels in the temporal perifoveal zone, while late angiographs (e and f) show more generalised hyperfluorescence. OCT images (g and h) still show slightly larger inner-lamellar holes. Rod-driven responses in Ganzfeld–ERG (i and j) are still normal while cone-driven responses have decreased further (k and l).

eyes (Figure 2b). Features typical of IPT, such as reduced macular transparency and superficial white crystals, had developed since the previous visit (Figure 2b). Changes were similar in both eyes. Fluorescein angiography showed telangiectatic changes in the early phase as well as diffuse-late hyperfluorescence (Figure 2c-f). Inner-lamellar holes seen in OCT in 2004 were still present and somewhat larger (Figures 2g and h).

Comment

The clinical findings in IPT are most obvious in the retinal blood vessels, however, it is possible that the primary defect lies in other retinal components, such as the retinal pigment epithelium, neural cells (including photoreceptors) or Muller/glia. The inner-lamellar holes may result from the neuroretinal decay rather than being of exudative origin since they were already present before vascular changes and are not visible during fluorescein angiography. The precession of typical vascular changes of IPT by the less well-recognised neuronal changes, as demonstrated in this case, suggests that photoreceptor damage may contribute to the early pathogenesis of the disease.

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Conflict of interest

Dr Mark Gillies is Executive Scientific Manager of the MacTel Research Project.

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Sir, Recreational use of 3,4-methylenedioxymethamphetamine ('ecstasy') relieving symptoms of posterior scleritis

Posterior scleritis is an uncommon form of scleral inflammation. While the prevailing consensus is that scleritis is an immune-mediated disease,¹ its precise pathogenesis remains enigmatic.

We describe a 23-year-old man with intermittent painful right eye and vision loss since 3 years due to posterior scleritis, which resolved when taking recreationally 3,4-methylenedioxymethamphetamines (MDMA, 'ecstasy').

Case report

A healthy 23-year-old man was referred to our hospital with pain and severe loss of visual acuity in the right eye (20/400) since 3 months. History revealed similar episodes during the last 3 years, which resolved partially when using ecstasy on a recreational basis. Since the patient had stopped ecstasy abuse 3 months ago, symptoms worsened.

Echographic evaluation showed choroidal thickening and oedema in Tenon's space in the right eye (Figure 1a). Clinical examination revealed a swollen optic disc surrounded with chorioretinal folds (Figure 1b). Pupillary reflexes and ocular motility were normal.

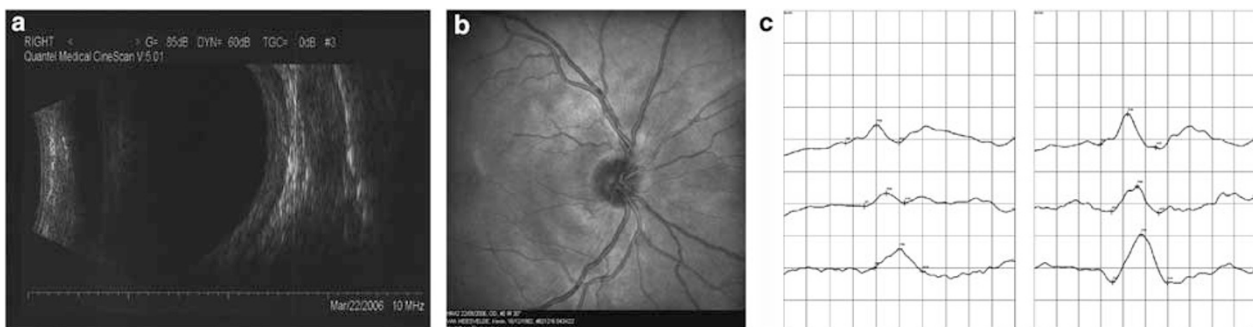


Figure 1 (a) B-scan echographic examination of the right eye showing choroidal thickening and oedema in Tenon's space at presentation; (b) red-free image of the right optic nerve surrounded by chorioretinal folds; and (c) pattern visual evoked potential (pVEP) showing half of the height of the amplitudes in the right eye compared to the left eye, and normal latencies.