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Sir,

Concentric annular macular dystrophy

Concentric annular macular dystrophy is a very rare condition which was first described by Deutman in 1974.[1] The main features are bull's eye maculopathy, perifoveal circular pigment epithelial atrophy and dyschromatopsia. As far as we are aware this is the first case reported in a British journal.

Case report

A 49-year-old Asian woman was referred to the Eye Department following a routine visit to an ophthalmic optician who noted an abnormal macular appearance. At presentation the patient had no visual complaints but on direct questioning did admit to having slight difficulty seeing under dark conditions. There was no history of chloroquine ingestion. Her father was under treatment for primary open angle glaucoma. No other family members had visual problems.

The corrected visual acuity by Snellen chart was right eye 6/9 and left eye 6/6. Anterior segment examination and intraocular pressure were normal. There was a bilateral bull's eye macular appearance (Fig. 1). The optic disc and retinal blood vessels were normal, as were the perifoveal and peripheral fundus. Colour vision by Ishihara testing was normal. Visual fields by computerised perimetry (Humphrey Full Field 120 Point Program) were full.

Fluorescein angiography revealed bilateral circular perifoveal areas of mottled hyperfluorescence with a few areas of hypofluorescence (Fig. 2). There was no leakage but late staining was observed (Fig. 3). The macular area itself did not have any remarkable features on angiography.

Electrodiagnostic testing found normal photopic and scotopic electroretinograms (ERG). The electro-oculogram (EOG) was slightly subnormal. Arden ratios were RE 1.40 and LE 1.37. Flicker fusion frequency was greater than 30 Hz in both eyes.

Comment

In 1974, Deutman reported four cases of a macular dystrophy with a bull's eye appearance clinically, but with angiographic and electrodiagnostic features differing from other macular dystrophies.[1] He named this condition benign concentric annular macular dystrophy. In each case there was a characteristic macular appearance consisting of central hyperpigmentation surrounded by a hypopigmented ring which itself was surrounded by a darker pigmented halo. All patients had normal or near normal vision with mild defects in colour vision.

In Deutman's series, all patients belonged to the same family and a pattern of autosomal dominant inheritance with variable expressivity was seen. In those patients who were affected at a younger age there were more

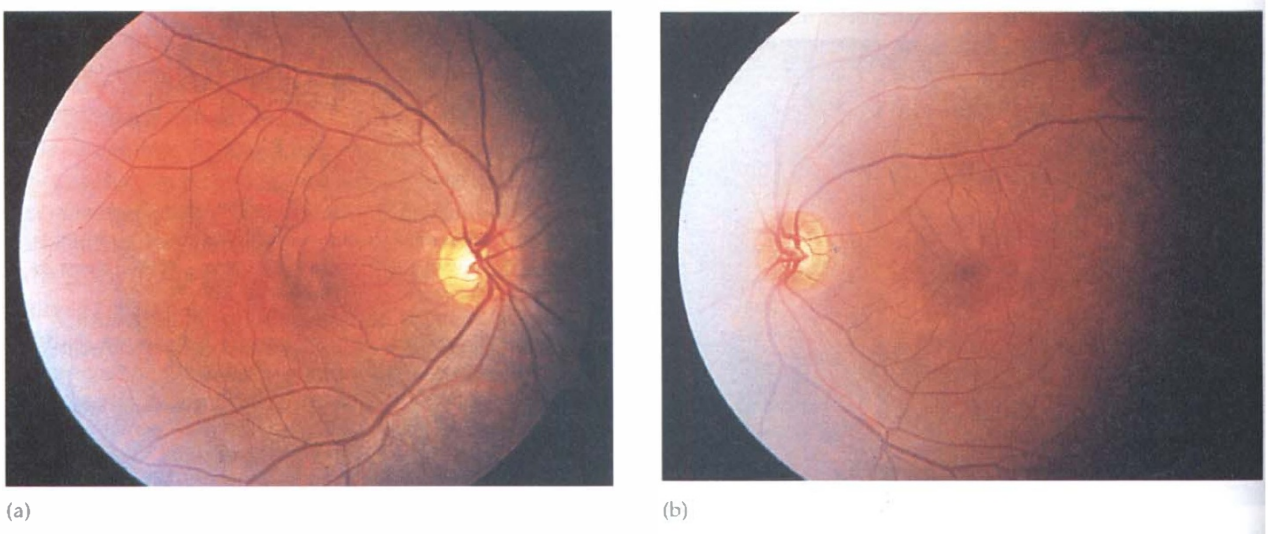

Fig. 1. *(a) Right fundus. (b) Left fundus.*

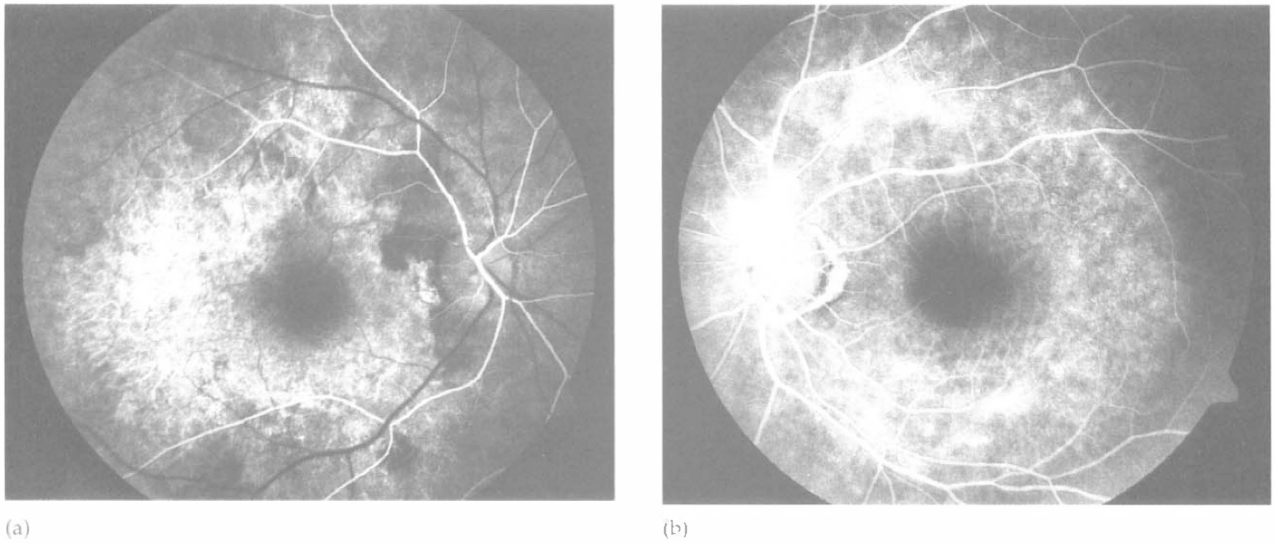

Fig. 2. *Fluorescein angiography: early phase.*

severe defects in visual acuity and colour vision with corresponding abnormalities in the ERG and EOG. The older patients in the series had normal or near normal visual acuity and the ERG and EOG were normal or only slightly subnormal. It therefore appears that high penetrance of the abnormal gene leads to earlier presentation and more severe effects on visual function as opposed to those cases which present later with only minimal effects on visual function.

Of particular interest was the finding on fluorescein angiography of perifoveal circular areas of stippled hyperfluorescence with diffuse hypofluorescence. These did not leak but had late staining. These areas did not have any visible corresponding clinical lesions in the affected areas. This pattern of staining is indicative of a widespread dystrophy of the choriocapillaris and retinal pigment epithelium. This would be consistent with the finding of a subnormal EOG as in our case. In fact, van den Biesen *et al.*[2] proposed that this condition could be a primary defect of the retinal pigment epithelium with a secondary non-selective defect in photoreceptor function. The localised defect in the macular region would be in keeping with a normal ERG, again consistent with the findings in our case. The follow-up series of the patients 10 years later showed that in some patients visual acuity, colour vision and night vision became worse, indicating progression. These, however, were the younger patients (second to third decades) with a likely higher expressivity of the abnormal gene. Those patients who presented at an older age (fifth decade) showed very little change in their condition, demonstrating non-progression. This serves to highlight the highly variable nature of this condition, but the most consistent and reliable feature in all cases was the bull's eye macular appearance.

The other causes of a bull's eye maculopathy to consider would be cone and cone–rod dystrophy, fenestrated sheen macular dystrophy, adult vitelliform macular dystrophy and central areolar pigment epithelial dystrophy. Cone dystrophy and cone–rod dystrophy

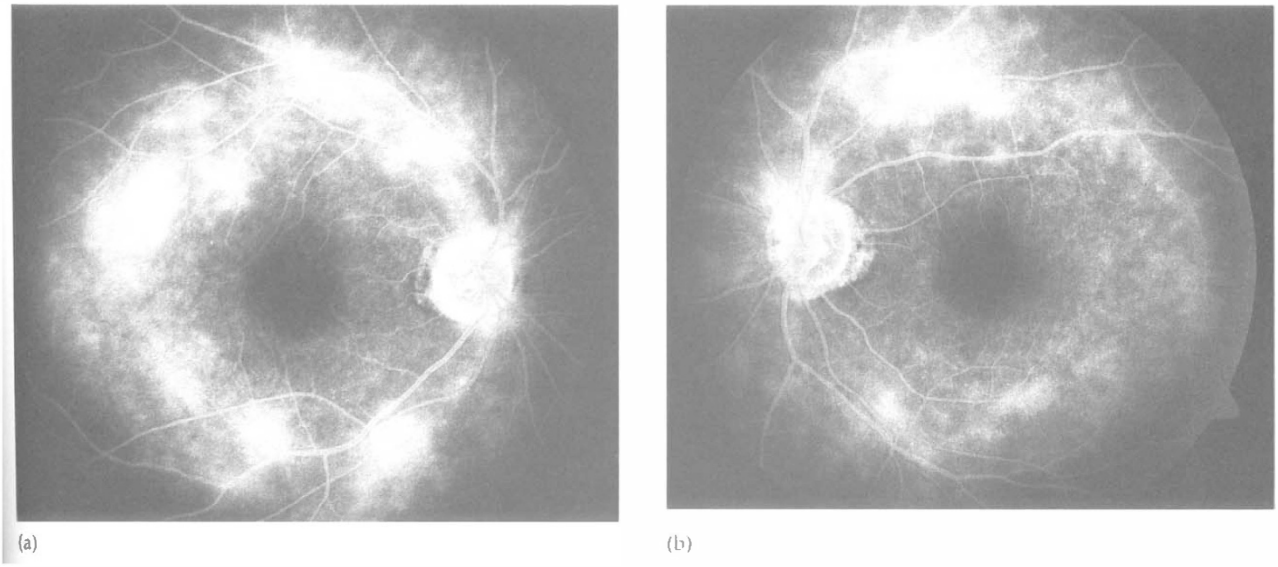

Fig. 3. *Fluorescein angiography: late phase.*

have well-described abnormalities in the ERG which define these conditions.[3] Fenestrated sheen macular dystrophy has a characteristic clinical appearance, as does adult vitelliform macular dystrophy, with raised yellow subretinal circular deposits which leak on fluorescein angiography.[4,5] Central areolar pigment epithelial dystrophy has a characteristic well-demarcated atrophy of the retinal pigment epithelium. None of these features are present in our case.

The finding of a normal ERG and subnormal EOG occurs in pattern dystrophy of the retinal pigment epithelium, but these are not associated with a bull's eye maculopathy and the fluorescein angiogram shows the characteristic pattern of the specific dystrophy. This was not present in our case.

Dyschromatopsia was not a predominant feature in our case whilst it was common in the series by Deutman[1] and also in that of Copetto and Ayazi.[6] The defects were predominantly in the tritan axis and were detected using the Farnsworth–Munsell 100 Hue Test. We tested colour vision by the Ishihara test, which has limitations in detecting tritanopic defects. Even so, there was a high degree of variability in findings in the series by Copetto and Ayazi, highlighting the pleomorphic nature of this condition.

In summary, we have presented the case of a 49-year-old patient with normal visual acuity and bilateral bull's eye maculopathy. Fluorescein angiography revealed perifoveal circular dystrophy of the choriocapillaris and retinal pigment epithelium. The ERG was normal and the EOG was slightly subnormal.

We propose that this is a case of concentric annular macular dystrophy, which is a very rare disorder with a highly variable clinical picture and likely autosomal dominant mode of transmission. At present, this disorder is thought to be a dystrophy of the retinal pigment epithelium with a secondary effect on photoreceptor function. Progression may occur but this is an inconsistent finding.

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Sir,

An unusual cause of orbital apex syndrome

Orbital apex syndrome is a clinical entity consisting of a triad of ophthalmoplegia, visual loss and proptosis.[1] It can be caused by a variety of conditions located at the orbital apex such as infection, inflammation or neoplasm. We present the first reported case of orbital apex syndrome caused by a metastatic prostatic cancer to the pituitary gland.

Case report

A 74-year-old man presented to the ophthalmic casualty with a 2 week history of right painless loss of vision. Otherwise, he had no past medical history of note. The best-corrected visual acuity was 6/24 in the right eye and 6/6 in the left. There was a right relative afferent pupillary defect and fundoscopy revealed a swollen optic disc. Left fundal examination was normal. The ESR was normal. An initial working diagnosis of right non-arteritic anterior ischaemic optic neuropathy was made.

When the patient was reviewed in the outpatient clinic 2 weeks later there was a marked deterioration in the right vision to counting fingers. In addition he had developed a right proptosis and restriction of eye movements. The measurements with Hertel's exophthalmometer were 24 mm in the right eye and 21 mm in the left. A brain and orbital MRI scan (Fig. 1) showed a mass arising from the pituitary gland extending into the right orbital apex. The patient was referred to the physician for further management.

Haematological and biochemical profiles revealed normochromic normocytic anaemia and a significantly raised prostate-specific antigen (PSA) level. On direct questioning the patient admitted to urinary symptoms consistent with prostatic disease. The rectal examination revealed a large craggy prostate gland. Subsequent bone scan revealed widespread bony metastases including the cranium. The oncologists were confident these lesions represented secondary prostatic malignancy and the patient was treated with systemic steroid, radiotherapy and hormonal manipulation. Following treatment the

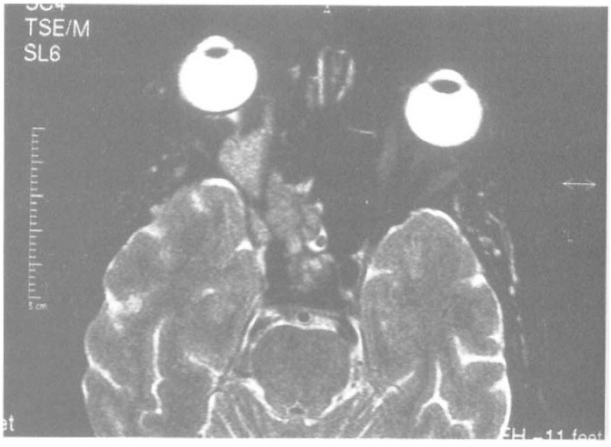

Fig. 1. *Brain and orbital MRI scan showing a mass arising from the pituitary gland and extending into the right orbital apex.*