Tel: +60 1 289 08877;
Fax: +60 1 289 08877.
E-mail: chuaoxford@hotmail.com

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Sir,

Pericentral pigmentary retinopathy: long-term follow-up

Pericentral pigmentary retinopathy is a rare disorder characterized by annular chorioretinal atrophy with varying degrees of bone spicule pigmentation extending temporally from the optic disk along the main retinal vessels in an arcuate fashion. Rarely, macular complications may be observed, such as bull'eye maculopathy and macular hole due to foveal ischemia and vitreomacular traction.[1] The prognosis varies, as the visual acuity and ocular findings appear stable in the autosomal recessive form[2] (OMIM[3] 268060) and progressive in the autosomal dominant form[4] (OMIM 180210).

Case report

A 14-year-old patient born to consanguineous parents was first seen in the Retina Unit of the University of Naples Federico II in 1987, complaining of night blindness. Patient's general health was good, metabolic and enzymatic profiles were within normal ranges, no relevant infectious disease was reported and history of exposure to toxic agents was negative. Visual acuity was 20/20 OU, while at fundus examination a bilateral peripapillary and pericentral chorioretinal atrophy with bone spicule pigmentation that spared the peripheral retina and the macula was noticeable. Fluorescein angiography showed bilaterally areas of pigmented epithelium atrophy along main retinal vessels, with medium to coarse pigment clumping. In both eyes automated perimetry showed an annular scotoma, the ERG was nearly extinct, Ishihara colour test was altered. Examinations of both parents were normal. During 18 years of follow-up the visual acuity of the patient has slowly worsened to 20/200 OD and 20/40 OS. At fundus examination progressive peripheral extension of chorioretinal atrophy and ring-shaped pigment, associated to a mild atrophy of the foveolar pigment epithelium have been observed. Periodic fluorescein angiography has shown wider peripheral atrophy of the retinal pigment epithelium and peripheral retinal nonperfusion. Multiple episodes of chorioretinal neovascularization arising in ischaemic areas have occurred. Thus, repeated argon laser photocoagulation of both ischaemic areas and neovessels has been performed (Figures 1 and 2). The electroretinographic responses have become unrecordable and automated perimetry has shown a progressive enlargement of the annular scotoma.

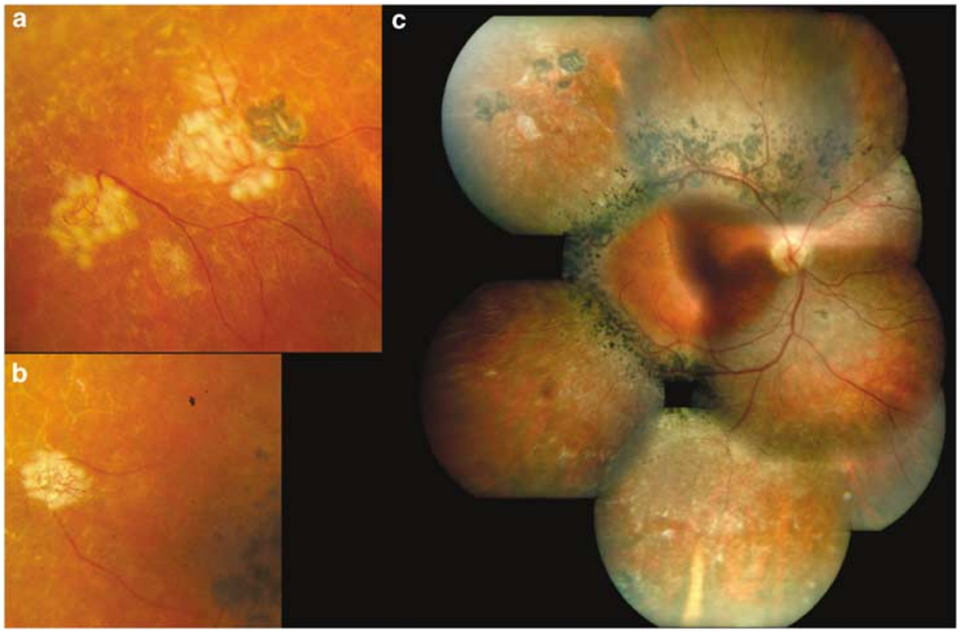

Figure 1 Pericentral pigmentary retinopathy. (a and b) Laser-treated chorioretinal neovessels of the right eye. (c) The colour map of the same eye shows the scars of photocoagulation in the supero-temporal periphery.

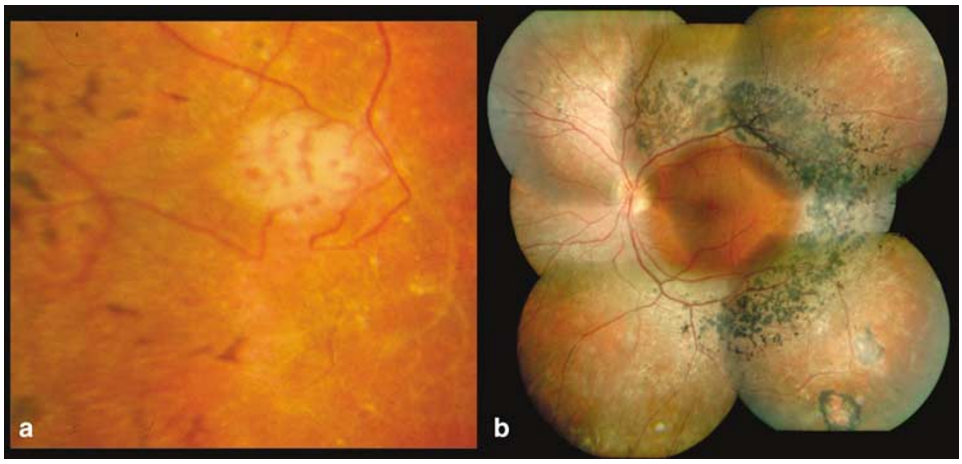

Figure 2 Pericentral pigmentary retinopathy. (a) Laser-treated chorioretinal neovessels of the left eye. (b) Colour map of the left eye. The scar after photocoagulation is noticeable in the infero-temporal periphery.

Comment

In presence of geographic areas of chorioretinal atrophy along retinal arcades, the main entities to differentiate from pericentral pigmentary retinopathy are pigmented paravenous chorioretinal disease, helicoid-serpiginous-geographic dystrophy, hyperornithinaemia with girate atrophy of the choroid and retina, postinflammatory pigmentary retinopathy, angioid streaks, Leber's congenital amaurosis.[2] In pigmented paravenous chorioretinal disease[5] there is no night blindness and most cases are sporadic, although an autosomal dominant or X-linked recessive mode have been described.[5,6] Leber's disease may present with a periarteriolar distribution of well-demarcated yellow lesions,[7,8] and is characterized by severe visual deficiency, with total or nearly total blindness, present at birth or shortly thereafter. In our case, the parents' consanguinity strongly supports the hypothesis of an autosomal recessive gene defect, and a diagnosis of pericentral pigmented retinopathy is suggested by the inherited origin of the phenotype, the young age of the patient, the presence of night blindness, the good visual acuity at presentation, the absence of metabolic, enzymatic or infectious disease or exposition to toxic agents. In 1988 Traboulsi *et al*[2] described pigmentary retinopathy in a pericentral distribution in siblings born to consanguineous parents. In these cases the optic discs, maculae and retinal vessels were normal, and at a follow-up of 13 years the fundus and visual acuity remained unchanged. Our patient developed progressive, severe worsening of visual acuity and findings on fundus, fluorescein angiography, electroretinogram, and visual field examinations. Furthermore, development of chorioretinal neovessels in ischaemic retina has occurred. To our knowledge, neither worsening of vision and clinical findings in autosomal recessive forms nor development of peripheral neovascularization have been described in literature. To explain this latter abnormality we can suppose that large areas of peripheral chorioretinal atrophy could lead to vascular perfusion defects, retinal ischaemia, and subsequent angiogenesis. Owing to the fact that our patient is the only affected member in the family, no linkage analysis can be performed. The only feasible approach to identify the molecular defect underlying this phenotype is a screening of mutation in candidate genes. Although several genes have been identified as responsible of autosomal recessive forms of retinal dystrophies,[9] no one is described as causing this particular form of pericentral pigmentary retinopathy. We could therefore consider that either an unidentified gene, or a particular mutation in a known gene could be involved in causing this phenotype.

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G de Crecchio[1], MC Alfieri[2], G Cennamo[3], F D'Esposito[4] and R Forte[1]

[1]Department of Ophthalmology,
University Federico II,
Naples, Italy

[2]Ophthalmic Division,
Vecchio Pellegrini Hospital,
Naples, Italy

[3]Department of Ophthalmology,
University of Catania, Italy

[4]CEINGE-Biotecnologie Avanzate/Dipartimento di Biochimica e Biotecnologie Mediche,
University Federico II,
Naples, Italy

Correspondence: G de Crecchio,
Dipartimento di Scienze Oftalmologiche,
Università Federico II,
Via Pansini 5,
Naples 80131, Italy
Tel: +39 8174 62293;
Fax: +39 8174 62383.
E-mail: crecchio@unina.it

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Sir,

Photodynamic therapy for solitary retinal metastasis from breast carcinoma

Although metastasis to the uveal tract is relatively common, metastasis to the retina is extremely rare. We report a case of symptomatic, solitary retinal metastasis in a patient with breast carcinoma treated with photodynamic therapy.

Case report

A 55-year-old female was referred to the Ocular Oncology clinic at the Royal Hallamshire Hospital, Sheffield in June 2005 with an unusual lesion affecting her right eye. Her only complaint was of metamorphopsia in that eye and at presentation her Snellen acuity was 6/4 bilaterally. Anterior segment examination and intraocular pressures were normal. The left fundus was entirely normal. Fundus examination of the right eye showed a discrete, white retinal lesion temporal to the fovea (Figure 1a). The lesion appeared vascular and this was confirmed on fluorescein angiography (Figure 1b). Ultrasonography revealed the lesion to have a maximum thickness of 1.6 mm. An accurate assessment of the internal reflectivity was not possible owing to the small height of the lesion.

The patient's past medical history was significant for breast carcinoma treated by lumpectomy in 1996 followed by mastectomy 2 years later. She had received adjuvant Tamoxifen for 3 years, however, this had been stopped for the last two years. In May 2005 CT scanning had revealed multiple pulmonary lesions consistent with metastases and she had been commenced on Anostrozole (Arimidex, AstraZeneca) 1 mg daily.

A diagnosis of solitary retinal metastasis was made and various treatment options were discussed. These included observation, fractionated radiotherapy, ruthenium plaque brachytherapy, transpupillary thermotherapy (TTT), or photodynamic therapy (PDT). The patient was concerned about the use of radiotherapy and the need for regional anaesthesia with TTT and wished to consider PDT. The rationale behind offering PDT was that the lesion was vascular, well-circumscribed and solitary. The risks and benefits of this novel treatment were explained and the patient consented to treatment with PDT using the standard TAP protocol.[1] A single 83 s application was used of 5.2 mm spot-size, delivering 600 mW/cm^2. There were no immediate complications.

At review, 8 weeks later, the patient reported that her symptoms had resolved and visual acuity was recorded as 6/5. Fundscopy showed the lesion to have regressed with minimal surface scarring and resolution of the associated subretinal fluid (Figure 1c). No other new metastases were detectable.

Comment

Breast carcinoma is the commonest primary tumour giving rise to ocular metastases.[2] Metastases to the ocular tissues are relatively common but almost always affect the choroid. Retinal metastases have been described but are extremely rare and few reports deal with more than