the retinal pigment epithelium (RPE), enables the selective treatment of the choroidal lesions along with sparing of the overlying neurosensory retina. TTT that also uses the diode laser has been used successsfully to treat CNV in age-related macular degeneration (AMD).[4] It has been suggested that TTT causes closure of the choroidal vasculature by hyperthermia-induced endothelial damage.[5] We thought we could use this property of TTT and the longer wavelength of diode laser to close selectively the choroidal vascular network of the leaking juxtapapillary PCV in this case. We did not do PDT in this case since the patient could not afford it. We did not treat the hyperfluorescent lesions superior and superonasal to the fovea since they were not clearly defined in the ICGFA.

The disappearance of the leaking polypoidal lesions following treatment has been seen following laser photocoagulation and PDT as well.[1–3] Our case illustrates the efficacy of TTT in the management of PCV. We do concede the need for future prospective randomised controlled studies to establish clearly the safety and efficacy of TTT in the management of PCV.

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We have no proprietary interest in any of the materials or methods used in this study.

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Sir,

Helicoidal peripapillary degeneration

This exceptionally rare bilateral fundus disorder was first described as choroiditis areata, in Iceland.[1,2] Later, it was renamed helicoidal peripapillary chorioretinal degeneration because of the lesion's resemblance to the propeller (helix) of an aeroplane.[3] Today, in the light of modern genetics, we believe that it is more precise to use the term dystrophy than degeneration, because it is obviously a dominantly inherited disorder.[4] A dominant form was recently mapped to chromosome 11p15.[5] Finally, a novel TEAD1 mutation is recognized as the causative allele in helicoidal peripapillary degeneration.[6]

We have had an opportunity to study three cases of inherited helicoidal peripapillary chorioretinal degeneration: a father, daughter, and son. Although this particular disorder has mostly been observed in Scandinavia, this is the first known case presented in Serbia.

Case reports

Case one

A 20-year-old female patient experienced some visual disturbances as a child and was referred to an ophthalmologist 10 years ago. Unusual atrophic peripapillary change was noticed and helicoidal peripapillary atrophy was diagnosed. Visual acuity was 6/6 for the right eye and 6/12 for the left eye.

At a routine check-up in July 2003, visual acuity had decreased to RE 6/9 with −1.75 D cyl axis 15° and LE 6/12 with −2.25 D cyl axis 170°. Intraocular pressure was normal. Discrete cataracts were observed (an unusual distribution in the anterior lens cortex) with very suggestive helicoidal peripapillary chorioretinal atrophy with tongues next to the fovea on both eyes. A fluorescein angiography was performed (Figure 1a, b).

Case two

Her 15-year-old brother had the same disorder, confirmed 7 years ago. His visual acuity was 6/9 with −0.75 D cyl axis 180° for both eyes, but the intraocular

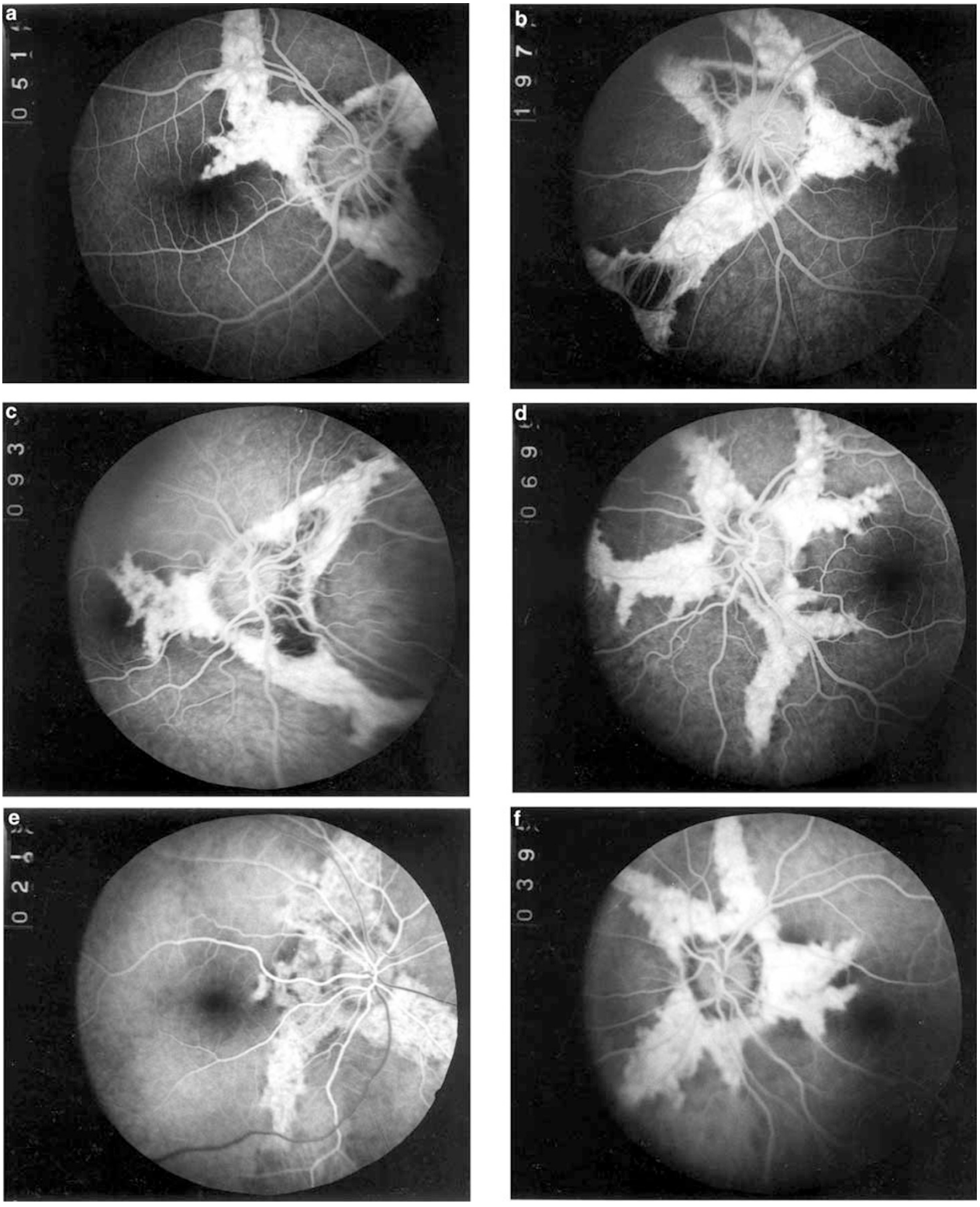

Figure 1 Case one: (a, b) Fluorescein angiograms show bilateral dystrophic peripapillar helicoidal lesions of retinal pigment epithelium and choriocapillaris, extending from the optic disc towards the periphery. On both fundi, atrophic tongues are close to the fovea. Case two: (c) A fluorescein angiogram of the right eye shows a dystrophic peripapillar triangular lesion, extending to the fovea. (d) A fluorescein angiogram of the left eye shows a dystrophic peripapillar lesion, resembling a starfish. Macula is spared. Case three: (e, f) Fluorescein angiograms show bilateral helicoidal peripapillary lesions. Clinical finding is very discrete, but fluorescein angiograms are impressive.

pressure was normal. He had no subjective visual problems. In addition to discrete central cataracts (in the anterior lens cortex), expressive helicoidal peripapillary chorioretinal atrophy was noticed. A fluorescein angiography was performed (Figure 1c, d).

Case three

The father, 45 years old, was also examined. His visual acuity was 6/6 with −0.50 D cyl axis 180° for both eyes. The intraocular pressure was within normal limits. Very discrete cataracts in the anterior portions of the lens were observed. A fundus examination showed only slight irregularity in colour and pigmentation around the disc. A fluorescein angiography was performed and a picture suggesting helicoidal peripapillary chorioretinal atrophy was observed (Figure 1e, f).

Comment

Helicoidal peripapillary chorioretinal degeneration is a rare autosomal dominant disorder characterized by peripapillary tongue-shaped patches of chorioretinal atrophy extending radially from the optic disc, with no inflammatory signs.[1] Choroid and pigment epithelium are completely atrophic,[7] but retinal vessels are not affected. One interesting hypothesis is that the peripapillary fundus lesions are the result of tearing and retraction of the retinal pigment epithelium and Bruch's membrane.[8] We applied Sveinsson's[9] findings, which led us to the conclusion that this disorder probably resulted from developmental anomalies of the choroid, the retinal pigment epithelium, or the short posterior ciliary arteries. A fluorescein angiography helped us to reach the correct diagnosis.

As mentioned earlier, this disorder has been described using various names: choroiditis areata,[2] circumpapillary dysgenesis of the pigment epithelium,[7] helicoidal peripapillary chorioretinal degeneration,[3] or atrophia areata.[4] Since there were no signs of inflammation in our cases, they differ specifically from serpiginous choroidopathy. Other differential diagnoses include angioid streaks, malignant myopia, paravenous retinochoroidal atrophy and radial lattice retinal degeneration.

There has been much confusion and misunderstanding concerning this phenomenon in medical literature to date, but our opinion is that the previously held position (that two variants of the disease exist) is questionable. In the congenital type (our cases), the atrophic peripapillary zone does seem generally to be stationary and probably does not seriously affect the macula during life. However, this is not a rule. The study of a large group of 26 affected individuals with autosomal dominant inheritance, pointed out poor visual acuity in later years.[4] Recently, electrophysiological tests suggest that retinal dysfunction is focal rather than diffuse.[10]

Adult forms, described as separate entities in earlier literature,[3] were probably variants of serpiginous choroidopathy, characterized by the fact that one eye was affected some time before the other, with atrophic tongues that may frequently affect the macula and thereby lead to deteriorated central vision.

Our cases have reasonable visual acuity, with minimal changes over several years. However, in the first case atrophic tongues on both eyes were very close to the fovea, so further careful examination is needed. The third case (father) was rather unusual since diagnosis could only possibly be performed by a fluorescein angiography.

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Sir,

Deep lamellar keratoplasty in corneal dermoid

Corneal dermoids are a rare cause of corneal opacification consisting of abnormal mesoblastic tissue covered by the epithelium. They can involve the deeper layers of cornea leaving an intact descemet's membrane and endothelium. Occasionally, anterior segment structures may be involved by the choristomatous growth and there may occur accompanying ocular malformations. A surgical intervention in the form of lamellar keratoplasty (LK) or penetrating keratoplasty (PK) is warranted on accounts of cosmesis, discomfort, and primarily because they are located in the visual axis. Recently, we treated a child with a corneal dermoid encroaching on to the visual axis. An anterior deep lamellar keratoplasty was performed after the assessment of depth of involvement of the corneal mass by ultrasound biomicroscopy. To our knowledge this is the first report of a successful deep LK in a patient with a corneal dermoid.

Case report

An 8-year-old boy presented with the complaints of pinkish mass and decreased vision in his left eye. This mass was present since birth and had gradually increased in size. The child was the product of normal uncomplicated gestation and had normal development.

Ocular examination disclosed a normal right eye with a visual acuity of 6/6. Left-eye visual acuity was hand movements close to face. Constant divergent squint of approximately 30-prism diopter was present in the left eye. Eyelids were normal in both eyes. Left eye showed a vascularized, moderately elevated, sharply circumscribed pinkish mass $6 \times 6\,mm^2$ in size overlying the cornea superonasally. Also present was a ring of lipid deposit around the mass in the adjacent clear cornea (Figure 1a). The remaining cornea was clear. Anterior segment was otherwise normal. Clinically, a diagnosis of corneal dermoid was made. Ultrasound biomicroscopy (UBM) disclosed a highly echogenic lesion occupying the superficial 60% of the cornea. Deeper part of stroma, descemet's membrane and endothelium were not involved (Figure 2a). An anterior deep LK (DLK) was planned.

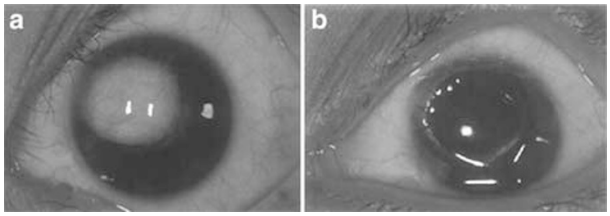

Figure 1 (a) Clinical photograph of left eye showing smooth, elevated, sharply circumscribed mass with an arc of lipid in adjacent cornea. (b) Left eye 6 months after DLK showing clear graft.

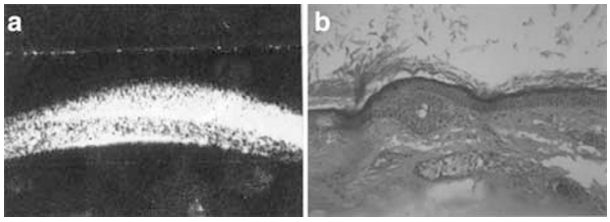

Figure 2 (a) Ultrasound biomicroscopic picture showing highly echogenic lesion occupying the superficial 60% of the cornea. (b) Histopathology slide showing thick keratinized epithelium and a sebaceous gland enmeshed in connective tissue (H&E, × 25).

After a partial thickness corneal trephination with 6 mm trephine, a superficial keratectomy involving the mass was performed. A disposable 30-gauge needle was inserted deeply with bevel down into the paracentral stroma and air was injected. A small opening was made in the air bubble and the remaining stromal layers were removed till the descemet's membrane was bared of the stroma. Donor corneal button 6.5 mm sized, stripped of descemet's membrane was sutured onto the bare descemet's membrane. In all, 16 interrupted 10/0 nylon sutures were given. Postoperatively, the eye was treated with topical prednisolone acetate (1%), tobramycin (0.3%) and a tear substitute four times a day. Histopathology of the corneal dermoid revealed thick keratinized epithelium and sebaceous glands enmeshed in connective tissue (Figure 2b). Patient had an uneventful postoperative course. Visual acuity improved to 3/60 and is currently receiving ambylopia therapy. At 6 months postoperatively the graft has remained transparent with no interface scarring (Figure 1b).