mesenchymal origin. We assume, as do other authors, that the degenerative corneal changes can derive from a primary ectodermal defect.1 * 9 10

To our knowledge, this is the first report of successful penetrating keratoplasty associated with Werner’s syndrome and incomplete Leopard syndrome.

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CASE REPORT

An 8-year-old otherwise healthy boy was first admitted to our hospital in July 1992 with a 1 year history of decreasing visual acuity in his right eye. We saw a greyish, slightly elevated tumour in the area of the temporal vascular arcade including the macula. The left fundus appeared normal at this time.

At the age of 6 full visual acuity (10/10) and normal fundus appearance had been documented for both eyes by the referring ophthalmologist.

On a second admission to our hospital, 18 months later, visual acuity in the left eye was found to have decreased to 6/10 and now a similar process could be seen in the left fundus.

During 2½ years of follow up visual acuity finally decreased to 1/10 in the right eye and remained almost unchanged in the left eye.

Anterior segment findings were normal.

There was no strabismus, and ocular motility was free. Family history was unrevealing.

The greyish tumour surface showed fine wrinkling caused by a superficial gliosis with traction of the great vessels towards the macular area (Fig 1). There was no retinal detachment.

B-scan echography revealed a flat thickening of the retina in the area of the tumour but no extraretinal extension.

Fluorescein angiography showed normal filling times and no exudation in the late phase (Fig 2).

Computerised perimetry (Octopus 1-2-3, G1X-program) showed a diffuse sensitivity loss in the central visual field with a small absolute scotoma in the centre of the right eye.

COMMENT

This case is uncommon for two reasons. Firstly, CHR-RPE is believed to be a unilateral eye disease, leading to monocular (pseudo) strabismus in some cases. Schachat et al12 reviewing 60 cases of CHR-RPE, stated in 1984 ‘neither bilaterality nor...have been seen in combined hamartomas ...’. To the best of our knowledge our patient is the second published case of a bilateral CHR-RPE. In the first case, described by Laqua and Wesing in 1975,13 the tumours were located closer to the optic disc and both lesions were seen on first examination. Angiography was not performed in their patient, probably because the child was only 4 years old.

In both eyes of our patient the clinical appearance was typical for CHR-RPE,1 3 as it met the main criteria: slight elevation, pigmentation, mixture of retinal tissue and pigment epithelial cells, gliosis with superficial traction, and distortion of the vessels.

Our patient had a macular (peripheral) type of the disease with tumours not involving the optic disc and an early age of onset of symptoms.

Differential diagnosis must consider hypertrophy of the retinal pigment epithelium, melanocytoma of the optic disc, choroidal mela-
noma, and other pigmented tumours of the
fundus, but in all these the clinical picture dif-
ers markedly from that of the combined
hamartoma.

The second unusual feature in our case is
that the lesion of the left eye started growing
after the age of 8. This is in contrast with
the views of Schachat et al,2 who believe that
combined hamartomas are always congenital
lesions. The referring ophthalmologist of our
patient found full visual acuity and a normal
fundus on both eyes at age 6. On first admis-
sion to our clinic we still saw a normal
fundus of the left eye while a hamartoma had
developed in the right eye. (No fundus photo-
graphs were taken at that time because the
child was not cooperative.) We believe that the
CHR-RPE need not necessarily be present at
birth (that is, congenital) but may develop
later in childhood.

We agree with others2 that clinical symp-
toms may show up as early as the hamartoma
itself or many years later, when secondary
changes such as contraction of the superficial
gliosis with retinal traction and exudation
occur.

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