Flecked retina syndrome with palisade appearance of the periphery
A case study with a family investigation

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Abstract

A 31 year old woman without symptoms presented bilateral, deeply seated retinal flecks in the midperiphery of both eyes. The flecks increased and became more dense towards the ora serrata, where they formed a palisade-like pattern. Fluoresceine angiography and visual acuity were normal, whereas other subjective or objective tests showed slight to moderate deficits. It is concluded that the condition represents a new variant of the Flecked Retina Syndrome. A family study suggested a hereditary basis, but no safe conclusion could be drawn.

Introduction

In 1965 the term 'Flecked Retina Syndrome' (FRS) was proposed as a common denominator for degenerative conditions characterized by 'limited or widespread distribution of deep yellowish or white lesions of various size and configuration without vascular and optic nerve abnormalities or pigment migration.' (7). The authors included the following conditions under this heading: fundus flavimaculatus (FF), fundus albipunctatus (the stationary form of retinitis punctata albescens, the latter being a tapetoretinal degeneration (6)), drusen degenerations of various morphology, the speckled multicolored fundus and the fleck retina of Kandori.

A patient showing retinal lesions conforming to the definition of FRS has been examined. In contrast to earlier reports on FRS the flecks were nearly absent from the posterior pole, whereas they extended beyond the equatorial regions. Just posterior to the ora serrata a 360-degree palisade pattern was present. The study was completed by a family investigation.

Case history

A 31-year old woman was referred from a medical department because of a slight arterial hypertension. General physical examination had disclosed a chronic, moderate pyelonephritis. She had no visual complaints whatsoever.

The visual acuity was 6/6 with +1.25 sph. OU. Position, motility and pupillary reactions were normal. No pathological changes were found by slit lamp examination and the IOP was 14 mm in both eyes.

Ophthalmoscopy revealed multiple, deeply seated yellow streaks in the midperiphery of both eyes (Fig. 1). Examination with the Goldmann three-mirror lens showed, that they extended far beyond the equator and that their dimension and concentration increased towards the oral region. Here they ended up by forming a 360-degree, densely packed band of palisade-like configuration (Fig. 2). Some of the deposits showed a fish-tail outline in connection with a small eye-like area finely powdered with pigment.

The optic nerve heads were normal. The foveal regions were well defined with normal reflexes; in particular, no signs of an atrophic macular lesion were present (Fig. 3). The arterioles were marked by a slight hypertensive angiopathy. The venules were normal, and the vessels crossed the degenerative regions without showing affection. The two eyes were essentially alike.
The patient read Ishihara's plates without faults. Mapping of the visual fields with Goldmann's perimeter demonstrated slightly contracted isopters. A test with Goldmann-Weekers' adaptometer showed a decreased, although not absent rod sensitivity. Fluorescein angiography showed a completely normal filling of the retinal vessels without any signs of leakage. Neither were any pigment layer defects present.

The ERG showed slightly reduced amplitudes with normal latencies in both eyes. No EOG light rise was present in the right eye (Arden ratio = 100), whereas the left eye presented a normal EOG (Arden ratio = 220). The VER was recorded with a chequer board stimulus and gave completely normal responses in both eyes.

Family study

The proband's father, three living siblings and all members of the third generation were examined (Fig. 4). Visual acuity, color vision and visual field were checked, and ophthalmoscopy supplemented by Goldmann's three-mirror lens (the latter not in