WATERSHED ZONE DEGENERATION, A CLINICAL SYNDROME?

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INTRODUCTION

The purpose of this study is to interpret the clinical, angiographic and perimetric features of a group of six male patients with decrease of vision and metamorphopsia. Sometimes we found pigment mottling and oedema in the macula and around the optic disc. Angiography showed in some patients hyperfluorescent leaking spots in the macular area and in a vertical zone temporal to the optic disc. In others, only hyperfluorescence in a zone temporal to the optic disc was seen. Perimetry revealed corresponding deep scotomas.

CASE I

This healthy 41-year old man suffered from decreased vision for a few months and metamorphopsia in both eyes. Examination showed a visual acuity correctable to 6/18 in the right eye and finger counting in the left eye. In both macular regions we found pigment epithelium degeneration (P.E.D.) with oedema. The optic discs were normal. No signs of inflammation were found. In the right eye a triangular area of P.E.D. was seen in the periphery. Fluorescein angiography showed besides a normal arm-retina time, band- and sector-shaped hyperfluorescent leaking areas in a vertical plane temporal to the optic disc. In the macular region hyperfluorescent spots were seen with leakage on later pictures. There was a marked symmetry between the two eyes. Kinetic perimetry showed corresponding absolute scotomas. Clinical, biochemical and neurological examination revealed no abnormalities. The situation of both eyes took an unfavourable course, despite high doses of Prednisone. Also bilateral retinal detachment occurred, which disappeared later on.

CASE II

This 46-year old healthy man noticed decreased vision with metamorphopsia in his left eye. On examination, corrected visual acuity in the right eye was 6/6 and in the left 6/24. In the left macula there was subretinal oedema. On fluorescein angiography we found bilateral lesions in the posterior pole, almost equal to those seen in the first patient. The leakage, however, was doubtful. Perimetry showed corresponding absolute scotomas.

Fig. 1a. The configuration of this degenerative area is the same as that of early filling defects in normal patients.

CASE III

A 39-year old Eurasian healthy man was seen because of a central serous choroidopathy in his right eye. His corrected vision in both eyes was 6/6. In the right macula we noticed oedema; the left eye appeared normal. Fluorescein angiography revealed in both eyes large wedge-shaped areas of hyperfluorescence inferior to the optic disc with leakage and irregular patches in the macular region. Corresponding absolute scotomas were found. Up till now, the central visual function remains good.

CASE IV

This 51-year old man noticed decreased vision in his left eye. Corrected vision was 6/6 in both eyes. On the temporal side of the — normal — optic disc of the left eye pigment degeneration was noticed. Fluorescein angiography showed a prolonged arm-retina time, narrow retinal arteries and an irregularly

Fig. 1b. The visual field shows a corresponding scotoma.

bordered area of P.E.D. temporal to the optic disc without leakage (see Fig. 1a). This degenerative area corresponded with an absolute scotoma (Fig. 1b).

CASE V

This 42-year old man was seen because of amaurosis fugax. He had vision 6/6 in both eyes. Around both optic discs, but mainly temporal, areas of P.E.D., suggesting clusters of Elschnig spots, were seen. Fluorescein angiography showed a prolonged arm-retina time. Ophthalmodynamography was suggestive of bilateral carotid artery stenosis.

CASE VI

This 46-year old man was referred because of decreased vision in the right eye. His history revealed a myocardial infarct in 1971. Corrected vision in the right eye was 6/12, in the left 6/6. Examination of the right eye showed P.E.D. between macula and optic disc; the left eye revealed yellow spots in the macula and so-called Siegrist streaks (1899). The affected area in the right eye was wedge-shaped without leakage on fluorescein angiography. Visual field analysis showed a relative centrocoecal scotoma.

Fig. 2a. These pictures show pigment epithelium degeneration in the temporal zone of the optic disc, adjacent to a not yet filled area (watershed zone). A few seconds later this area also shows pigment epithelium degeneration (Fig. 2b).

DISCUSSION

Ophthalmoscopy suggested in cases II and III a central serous choroidopathy as the cause for the decline of vision, but when we saw the angiograms we were at a loss to put all signs under a single heading. Features that tally with local choroidal circulation disturbances were visible in three patients: a triangular syndrome (AMALRIC, 1971, 1973; case I), Siegrist streaks (SIEGRIST, 1899; case VI) and Elschnig spots (KLIEN, 1968; case V). The myocardial infarction (case VI), bilateral carotid artery stenosis (case V) and prolonged arm-retinal time (cases IV and V) hinted at a more general vascular disorder. We were struck by the fact that the areas of P.E.D. coincided with early choroidal filling defects (see Fig. 2). Recent publications on choroid circulation (HAYREH, 1972, 1975) have shown that the arterial blood supply is sectorial. The temporal part of the choroid is usually filled by the lateral posterior ciliary artery (P.C.A.), the nasal part by the

Fig. 2b. For legend see Fig. 2a.

medial P.C.A. These two parts are separated by a so-called watershed zone (HAYREH, 1974). In man this zone is often recognizable as an early filling defect in the temporal peripapillary area (SHIMIZU, 1974). Also in the macular region these filling defects can be seen where they represent watershed zones of short P.C.A.'s.

Comparison of these data with the angiograms of our patients suggested to us the existence of a watershed zone syndrome. Also Hayreh's hypothesis that the watershed zones are the most vulnerable parts of the choroid in case of circulation disturbance, is in accordance with the general and local vascular affections in four of our patients. The visual fields point to lesions of the retinal outer layers overlying the P.E.D. areas and thus the watershed zones. On angiography the discs were normal and as we expected there were no fibre bundle defects.

SUMMARY

Six patients with pigment epithelium degeneration in the posterior pole have been discussed. The typical findings in these middle-aged men are:

1. Decrease of visual acuity and metamorphopsia.

2. Widespread patches of hyperfluorescence, wedged shaped and geographically distributed, mainly in a vertical zone temporal to the disc, but sometimes in the macular area. A few cases show leakage.
3. In most patients the lesions are bilateral and symmetric and
4. are accompanied by large and intense scotomas.
5. They run a chronic and in one case a very unfavourable course.
6. The angiographic localization of the lesions correlates with the watershed zones.
7. Symptoms are mentioned that are suggestive for vascular disorders as a cause for this disease.

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DISCUSSION

Dr Gass: I want to comment briefly on Dr Bos' paper. We have seen about 12 patients very similar to the one he presented, with the exception that every one of our patients had had at least in one eye a serous detachment of the retina involving the macula. And I think these cases represent a severe form of central serous retinopathy. The large areas of pigment epithelium atrophy that often have a teardrop shape may extend all the way to the periphery of the fundus and they are related to chronic, recurrent, long standing retinal detachment. Often it is outside the macular area and the patient does not know that he has it. Very often there is demonstrable leak in the upper portion of this zone. So I think it is the same disease. It is of interest that all these patients were males and middle-aged. I think these patients should be watched extremely carefully, given an Amsler grid, be-

cause these are the patients who can lose significantly pericentral as well as central vision over a period of years and should be photocoagulated if you can catch them with a detachment.

Dr Bos: We considered also the diagnosis of central serous retinopathy. However, the peripapillary degenerative areas did not present localized leaking points but did diffuse over their entire surface. These areas correspond to watershed areas. It is not proved if there is an essential difference in pathogenesis between the two diseases. Theoretically an analogous etiology (watershed zone) could be postulated for central serous retinopathy.

The incidence of chorioretinal arteries in 1000 consecutive patients was determined by review of intra-ocular fundus photographs and fluorescein angiograms. One or more chorioretinal arteries was present in 4.5 per cent of all patients or in 32.1 per cent of the 1000 eyes of this series. They occurred bilaterally in 14.6 per cent and contributed to some portion of the macular circulation in 18.7 per cent of the patients. A great deal of variability of size, number and distribution of chorioretinal arteries was observed.

Chorioretinal arteries were found to be particularly prominent in patients with central serous retinopathy in 7 per cent of the 1000 eyes of this series.

The chorioretinal arteries were observed in the early phase of the disease and in the late phase of the disease. The chorioretinal arteries were observed in the late phase of the disease in 14.6 per cent of the 1000 eyes of this series. The chorioretinal arteries were observed in the late phase of the disease in 14.6 per cent of the 1000 eyes of this series.

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