Inflammatory pigmented paravenous retinochoroidal atrophy

KATSUHIRO YAMAGUCHI,¹ SATOSHI HARA,¹ YASUHIRO TANIFUJI,²
AND MAKOTO TAMAI¹

From the ¹Department of Ophthalmology, School of Medicine, Tohoku University, Sendai, and

²Tanifuji Eye Clinic, Morioka

SUMMARY A 47-year-old Japanese man had a progressive degeneration of the retina and choroid along the retinal veins associated with uveitis of two years' duration. The lesion was characteristic of paravenous retinochoroidal atrophy: a contiguous atrophy of the retinal pigment epithelium and choroid of one-half to one disc diameter in size was present along most of the veins from the posterior pole to the far periphery. Fluorescein angiography showed a window defect in the retinal pigment epithelium, with hyperfluorescence representative of retinal pigment epithelium and choriocapillaris degeneration. Good visual acuity was attained after extracapsular cataract extraction for complicated cataract and vitrectomy for severe vitreous opacity had been performed in both eyes. The cause of this new inflammatory disease was unknown.

Pigmented paravenous retinochoroidal atrophy¹⁻⁴ is a rare condition in which the atrophic areas extend one-half to one disc diameter on either side of the vein and follow the vein's course, even when it branches. The cause of the condition is still unknown.⁵⁻⁹

We examined a patient who had gradually progressive paravenous retinochoroidal atrophy accompanied by an active panuveitis of more than two years' duration. We believe ours is the first report to describe active inflammatory causes of paravenous retinochoroidal atrophy.

Case report

A 45-year-old Japanese man was examined by one of us (YT) on 3 March 1985 and found to have had bilaterally blurred vision for six months. His family and past history were uneventful. Ophthalmic examination showed his best corrected visual acuity to be 20/25 in both eyes. Slit-lamp examination disclosed keratic precipitates, cells in the anterior chamber, and floaters in the anterior vitreous. Fundus examination showed small, patchy, multifocal cobblestone-like areas of retinal pigment epithelial

degeneration along the retinal veins in both eyes and round the optic disc (Fig. 1)

Although the patient was treated with systemic and locally administered corticosteroids, the uveitis persisted without improvement. With the development of iridic posterior synechiae, complicated cataract of both lenses and the opacity of the vitreous increased, and the patchy retinal pigment epithelial degeneration enlarged to form contiguous lesions.

The patient was referred to Tohoku University Hospital on 6 April 1987. Ophthalmic examination disclosed a best corrected visual acuity of 20/200 in both eyes. Slit-lamp examination showed posterior synechiae of the iris, a few cells in the anterior chamber, and complicated cataract in both eyes. The fundi were obscured by severe vitreous opacity. A scotopic electroretinogram with single white-flash stimulation showed reduced a wave and b wave amplitudes (Fig. 2). Electro-oculography showed low standing potential and no response to light bilaterally.

The patient was admitted to hospital on 16 April 1987. Extracapsular cataract extraction and simple vitrectomy were performed in the right eye on 18 May and 5 June, and in the left eye on 23 June and 7 July. Systemic and local corticosteroids were administered postoperatively. The corrected visual acuity in both eyes was 20/20. The Humphrey field analyser

Correspondence to Katsuhiko Yamaguchi, Department of Ophthalmology, School of Medicine, Tohoku University, 1-1 Seiryō-cho Sendai, Miyagi, 980 Japan.

Fig. 1A

Fig. 1B

Fig. 1. Fundus photograph from 3 March 1985. A: Right fundus shows retinochoroidal atrophy round the optic disc, along the superonasal vein, and in the retinal veins two disc diameters inferotemporal to the macula. B: Left fundus had retinochoroidal atrophy around the disc and in the vein just temporally to the macula.

Fig. 2 Right scotopic electroretinogram (ERG) with single white flash stimulation of 20 ms after 20 minutes of dark adaptation, pupillary dilatation, and topical anaesthesia (0.5% proparacaine hydrochloride). In both eyes a and b waves were reduced. Control illustrates normal responses on ERG.

Fig. 3 Humphrey field analyser showed geographic scotoma and an enlarged blind spot in both eyes.

Fig. 4A

Fig. 4B

Fig. 4 Fundus photograph from 26 September 1987. (A) Right and (B) left eyes. The degeneration progressed to form a continuous lesion along most of the veins in both eyes.

Fig. 5A

Fig. 5 Fluorescein angiograms from 26 September 1987, of the (A) right and (B) left eyes. The degeneration of the retinal pigment epithelium and choriocapillaris is obvious along the retinal veins and round the optic disc.

Fig. 5B

disclosed geographic scotoma (Fig. 3). A test of colour vision with Ishihara pseudoisochromatic plates was completed without error. Both fundi showed contiguous delimited atrophy of the retinal pigment epithelium along most of the retinal veins and round the optic disc. The atrophic regions were of almost the same size, and there were few clumps of pigment in the peripheral regions (Fig. 4). Fluorescein angiography in the early phase revealed a window defect of retinal pigment epithelium and choriocapillary atrophy that disclosed a large choroidal vasculature. In the late phase the degenerative area along the veins showed homogeneous hyperfluorescence (Fig. 5).

No systemic abnormality was found. Results of laboratory studies included normal complete and differential blood cell counts, serum amino acids, serum electrolytes, serum protein electrophoresis, erythrocyte sedimentation rate, and angiotensin converting enzyme. There was no serological evidence of syphilis, toxoplasmosis, systemic lupus erythematosus, or rheumatoid arthritis. A skin test for tuberculosis was within the normal range, as were serum antibody levels for herpes simplex virus, herpes zoster virus, cytomegalovirus, and measles virus.

Discussion

This patient's fundi and visual function were characteristic of pigmented paravenous retinochoroidal atrophy. The clinical course, however, was unique.

To our knowledge there have been no reports of active inflammation with this disease. It was obvious, however, that inflammation had a major role in the pathogenesis of the retinochoroidal atrophy in our patient. Also unique to this patient was a small patchy area of atrophy that progressed to continuous lesions along the veins over a three-year period; Pearlman and colleagues⁷ have reported the disease as being progressive.

Thus we believe that this case represents a new type of inflammatory disease of unknown origin marked by a progressive degeneration of the retinal pigment epithelium and choroid that occurs along the retinal veins. The main region to be affected was the retinal pigment epithelium. That was clearly demonstrated by fluorescein angiography and supported by an extinct pattern on the electro-oculogram.

The differential diagnosis includes both chorioretinal degeneration and inflammatory disease that cause chorioretinal atrophy, such as gyrate atrophy of the choroid and retina,¹⁰ helicoid peripapillary chorioretinal degeneration,¹ choroiditis proliferans,¹¹ sarcoidosis, syphilis, acute retinal necrosis, cytomegaloviral retinitis, tuberculous disseminated choroiditis, onchocerciasis, toxoplasmosis, and frosted branch angiitis.¹² These clinical features, however, were different from those in our case.

Although steroid therapy cleared up the anterior and postoperative uveitis in our patient, it did not help the inflammatory pigmented paravenous retinochoroidal atrophy.

References

- 1 Franchschetti A. A curious affection of the fundus oculi: helicoid chorioretinal degeneration. Its relation to pigmentary paravenous chorioretinal degeneration. *Doc Ophthalmol* 1962; **16**: 81–110.
- 2 Brognoli C. Spora un case di pigmentazione anomala del fondo oculare (meranosi della retina). *Arch Ottalmol* 1949; **53**: 99–119.
- 3 Morgan OG. Congenital pigmentation of the retina. *Proc R Soc Med* 1948; **41**: 726–7.
- 4 Pearlman JT, Kamin DF, Kopelow SM, Saxton J. Pigmented paravenous retinochoroidal atrophy. *Am J Ophthalmol* 1975; **80**: 630–5.
- 5 Chisholm IA, Dudgeon J. Pigmented paravenous retinochoroidal atrophy. *Br J Ophthalmol* 1973; **57**: 584–7.
- 6 Skalka HW. Hereditary pigmented paravenous retinochoroidal atrophy. *Am J Ophthalmol* 1979; **87**: 286–91.
- 7 Pearlman JT, Heklenlively JR, Bastek JV. Progressive nature of pigmented paravenous retinochoroidal atrophy. *Am J Ophthalmol* 1978; **85**: 215–7.
- 8 Brown TH. Retino-choroiditis radiata. *Br J Ophthalmol* 1937; **21**: 645–8.
- 9 Hsin-Hsiang C. Retinochoroiditis radiata. *Am J Ophthalmol* 1948; **31**: 1485–7.
- 10 Takki K. Gyrate atrophy of the choroid and retina associated with hyperornithinaemia. *Br J Ophthalmol* 1974; **58**: 3–23.
- 11 Fuchs A. Choroiditis proliferans. *Klin Monatsbl Augenheilkd* 1959; **135**: 775–91.
- 12 Watanabe Y, Takeda N, Adachi-Usami E. A case of frosted branch angiitis. *Br J Ophthalmol* 1987; **71**: 553–8.

Accepted for publication 26 September 1988.