Reticular dystrophy of retina

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This rare fundus dystrophy was first described by Sjögren (1950) in a Swedish family. Since then two more families have been described from Holland and Argentina by Deutman and Rumke (1969) and Alezzandrini (1971) respectively. This article describes a case in an Irish family.

Case report

The patient, a 7-year-old boy, was first seen at the Western Ophthalmic Hospital after a school medical examination showed he had poor visual acuity. He had no previous history of any systemic or ocular illnesses. His parents and grandparents were unrelated.

The best corrected visual acuity was 6/12 with +2.75 D cyl., axis 105° in the right eye and 6/9 with +2.50 D cyl. axis 80° in the left eye. External slit-lamp examination and ocular motility were normal. The fundi showed a small group of fine pigment clumps at each macula tightly enclosed within a small, light-

coloured, ring-shaped area. The posterior part of the fundus showed extensive marking with thin pigmented lines forming a net-like pattern of polygons varying in size from half to one disc in diameter. The polygonal pattern was often incomplete. On slit-lamp examination with the contact lens it was clear that the pigment lay at the level of the retinal pigment epithelium. There was no pigment dispersion on the retinal surface except at the maculae. It was found that each pigmented line consisted of fine pigment granules of apparently uniform size. The junctional ends of these lines showed a slight increase in pigmentation. The retina beyond the dystrophy zone was normal. There was no evidence of optic atrophy or narrowing of the retinal blood vessels (Fig. 1*a, b*).

Fluorescein angiography showed marked choroidal background fluorescence in a typical fish-net pattern during the early part of the transit of the dye. The angiography also showed a wider area of involvement than was clinically apparent, extending slightly beyond the nasal border of the optic disc and the temporal vessel arcade. The dark central pigment clump at the macula was surrounded by a fluorescent ring corres-

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FIG. 2 (a) *Fluorescein angiography of left fundus during arterio-venous phase showing black markings on fundus in typical fish-net pattern masking choroidal background fluorescence*

FIG. 1 (a) Ophthalmoscopic view of left fundus showing typical reticular pigmentation at posterior pole. (b) Ophthalmoscopic view of central area of left fundus showing pigment clumping at the macula surrounded by pale ring

FIG. 2 (b) *Fluorescein angiography of right fundus during venous phase showing similar reticular pattern*

ponding to the pale area seen clinically. This was due to retinal pigment epithelial atrophy, allowing an easy transmission of the background fluorescence. There was no evidence of any leakage of dye at any stage of the angiography (Fig. 2a, b).

The patient's sister had no ocular abnormality. The father was myopic, one aunt suffered from myopia and squint, and three other aunts had refractive errors the nature of which was unknown. There was no evidence of consanguinity in the three generations of this family.

Two-and-a-half years later the patient's vision, refraction, and the fundus picture were unaltered.

RETINAL FUNCTION TESTS

Perimetry using a 3 mm white target did not show any field defect. Colour vision testing with pseudoisochromatic plates was normal in both eyes. The electroretinogram was normal in each eye with normal flicker (skin electrode), but the electro-oculogram was low at about 160 per cent in each eye.

Discussion

In this case of reticular dystrophy of the retina

no other members of the family in three generations seemed to have been affected, although there were cases of squint and myopia. It is doubtful if these could be claimed as associated abnormalities. This is in contrast to the three other families reported, in which not only were other cases of reticular dystrophy found but there were also associated abnormalities such as spherophakia, intercalary staphyloma, shallow anterior chamber, and deaf-muteness (Sjögren, 1950). Deaf-mutism has been extensively studied by Holmsgren (1950) in the same family reported by Sjögren.

Summary

A case of an isolated occurrence of reticular dystrophy of the retina is described. The patient had no affected relatives.

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References

- ALEZZANDRINI, A. A. (1971) *Arch. Otol. (B. Aires)*, **46**, 13
- DEUTMAN, A. F., and RUMKE, A. M. L. (1969) *Arch. Ophthalmol.*, **82**, 4
- HOLMSGREN, S. (1950) *Acta ophthalm. (Kbh.)*, **28**, 297
- SJÖGREN, H. (1950) *Ibid.*, **28**, 279