Intraretinal changes in the Groenblad-Strandberg syndrome

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Systemic elastorrhexis is a widespread hereditary degeneration of mesodermal tissues affecting many structures in the body, particularly the dermis, the media of the arteries, and Bruch's membrane in the eye.

Pseudoxanthoma elasticum is one form of elastorrhexis mainly affecting the skin. Degeneration of the elastic lamina of Bruch's membrane results in the formation of angioid streaks in the retina. The association of angioid streaks and the cutaneous lesions is called the Groenblad-Strandberg syndrome. The other recognized lesions in the fundus are macular haemorrhagic and pigmentary changes (Scholz, 1941); mottling by pigmented deposits (Smith, Gass, and Justice, 1964; Gills and Paton, 1965); choroidal atrophy and choroidal sclerosis (Groenblad, 1958); and atrophy of the pigmentary epithelium (Groenblad, 1958; Carlborg, Ejrup, Groenblad, and Lund, 1959; Goodman, Smith, Paton, Bergman, Siegel, Ottesen, Shelley, Pusch, and McKusick, 1963).

The purpose of this paper is to describe a case of Groenblad-Strandberg syndrome which showed intraretinal bands and peripheral retinal degeneration leading to bilateral retinal detachment. The angioid streaks were not visible ophthalmoscopically but were vividly demonstrated by fluorescein studies.

Case report

A 20-year-old Indian first attended the Ophthalmic Department of the General Infirmary at Leeds in June, 1968, complaining of gradual progressive bilateral deterioration of the visual acuity. He had been aware of a slight visual defect since childhood which had become much more marked during the previous 12 months.

Examination

The visual acuity was 6/24 in the right eye and 6/36 in the left with a small myopic correction. The media were clear and the only ocular abnormalities detected were in the fundi of both eyes. There were bilateral macular degenerative changes associated with pale-coloured bands radiating from the optic discs. On a subsequent visit he was found to have developed bilateral retinal detachments with multiple peripheral round holes.

Treatment

An encircling procedure was carried out on the right eye followed by a similar procedure on the left. Cryoapplication was used to produce chorioretinal adhesions. Although the retinae remained flat, the visual acuity did not improve to more than 6/24 in each eye.

General examination

Linear atrophic striae were noted, mainly on the anterior and posterior axillary folds and inner surfaces of the arms. Initially, these skin changes were suspected to be related to weight loss,
but subsequent histological examination showed these to be consistent with pseudoxanthoma elasticum.

Laboratory investigations
A full haematological investigation showed nothing abnormal. Electrophoresis revealed a slight diffuse increase in the $\gamma$-globulin fraction, particularly affecting $I_\gamma G$ which was 1,330 mg./100 ml. (Normal range in adults 700–1,500 mg./100 ml.: Average 1,000 mg./100 ml.) (Allansmith, McClellan, Butterworth, and Maloney, 1968).

Skin biopsy revealed changes commensurate with the diagnosis of pseudoxanthoma elasticum.

Fluorescein studies
Fluorescein angiography displayed the pattern of the pigment epithelium very well in the early phases of circulation. Definite angioid streaks could be seen radiating from the optic disc especially in the left fundus (Fig. 1). The intraretinal bands of tissue were nonfluorescent in the early phase and were seen silhouetted against the background fluorescence (Fig. 1). The defects in the pigment epithelium were punctate and widespread.

FIG. 1 Fluorescein angiogram of left fundus taken in the retinal vascular phase of dye circulation. The generally mottled background fluorescence indicates widespread disorder in the pigment epithelium. There are radiating streaks of hyperfluorescence at 3 and 9 o'clock around the disc (vertical arrows). The wide area of pigmentary mottling is characteristic of the peau d'orange appearance seen in pseudoxanthoma elasticum (Smith and others, 1964). Radiating hyperfluorescence is typical of angioid streaks. The nonfluorescent radiating intraretinal bands at 10 o'clock in relation to the disc (horizontal arrow) are seen silhouetted against the background fluorescence and are crossed by the supranasal retinal vessels. The appearance of these bands in vivo, unlike the angiogram, was vividly white against the darker background of the fundus. On the other hand, the angioid streaks and the more diffuse pigmentary changes dramatically demonstrated in the angiogram were not visible ophthalmoscopically.

In the left upper temporal fundus there was a large area of coalescent angioid streaks (Fig. 2). In the late phase the intraretinal bands of tissue generally became fluorescent (Figs 3 and 4).
Discussion

The Groenblad-Strandberg syndrome is well recognized and documented (Artuner and Erdinc, 1968; Shevrov, 1968; Percival, 1968; Castel, Renard, Masse, Mollaret, Roche, Chartier, Matieu, and Velly, 1968; Mehta, 1968). This case is of interest for the following reasons.
The angioid streaks were not manifest on ophthalmoscopic examination but could be demonstrated by fluorescein angiography. The intense initial fluorescence of the streaks was due to choroidal intravascular fluorescence seen through gaps in the retinal pigment epithelium induced by changes in the underlying Bruch’s membrane (Rosen, 1968). The streaks usually show persistence of fluorescence (Smith and others, 1964). This has been suggested by Rosen (1968) to be due to scleral fluorescence and staining of collagen in Bruch’s membrane.

The presence of intraretinal bands radiating from the optic discs was very striking. The delayed fluorescence of the bands was also noteworthy. What was the nature of these bands? Retinal striae are white lines which indicate the limits of a receding area of retinal separation (von Jager, 1869). Histologically, Rönne (1910) found these striae to consist of fibrous tissue. In this case the bands were noted before the development of retinal detachments. The distribution of the bands was similar to that of angioid streaks. Seepage of exudate through the degenerate Bruch’s membrane into the retina probably excited localized inflammatory reaction and caused the intraretinal fibrous bands to develop. The delayed fluorescence suggested the fibrovascular nature of the bands.

It is unusual for a case of Groenblad-Strandberg syndrome to be associated with retinal detachment. In systemic elastorrhexis, widespread degeneration of mesodermal tissue involves the media and the elastic lamina of the blood vessels (Carlborg and others, 1959). Such changes, when extended to the retinal and choroidal vessels, could produce retinal degeneration resulting in retinal tears.

Finally, the immunoglobulin levels were found to be on the high side of normal. It has been previously suggested that their level is elevated in pseudoxanthoma elasticum (Johnson and Bloch, 1969).

FIG. 4 Right fundus in the late phase of dye circulation, showing fluorescent bands. Note the partially fluorescent lower band on the temporal side of the disc (arrow).
Summary

A case of Groenblad-Strandberg syndrome is described, in which widespread radiating intraretinal bands and bilateral retinal detachment formation were observed. Occult angioid streaks were demonstrated by fluorescein retinal angiography. The possible aetiology of the retinal changes is discussed with particular reference to widespread vascular degenerative changes which are known to occur in this disorder.

References

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