COMMUNICATIONS

FLUORESCEIN FUNDUS PHOTOGRAPHY OF ANGIOID STREAKS*†

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THE purpose of this paper is to present the first reported fluorescein fundus photographs of angioid streaks, and to document another ophthalmoscopic finding which may occur in patients with pseudoxanthoma elasticum, the *peau d'orange* fundus.

Case Report

A 37-year-old white female, seen through the courtesy of Dr. Victor H. Witten and Dr. Harvey Blank, came to the Dermatology Department because of dissatisfaction with the healing process following a cut to her right hand by a piece of broken glass. At that examination, she was found to have severe and widespread pseudoxanthoma elasticum (Fig. 1), which was verified by skin biopsies. She stated that the changes in her skin had begun at about 5 years of age and had since been slowly progressive. But for wearing glasses most of her life, she had enjoyed good health and otherwise had no complaints. She knew of no other persons with similar problems in her family.

FIG. 1.—Left lateral view of patient's neck, showing pseudoxanthoma elasticum.

Examination.—The corrected visual acuity was 20/25 in the right eye and 20/15 in the left. The external and biomicroscopical appearances were normal. Applanation tonometry showed that

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the ocular tension was 20 mm. Hg in each eye. The ophthalmoscope showed classic angioid streaks in both eyes (Figs 2 and 3, opposite). An early macular lesion in the right eye appeared consistent with the visual acuity (Fig. 4, opposite). The peripheral fundi showed interesting areas temporal to the macula, which were symmetrically located in each eye and had a stippled appearance which resembled that of the cutaneous changes on the neck. The term *peau d'orange* fundus appeared appropriate in describing this lesion (Figs 4, 5, 6).

Several small round yellowish-white spots were also seen in the right lower fundus (Fig. 7, opposite). Fig. 8 shows an angioid streak in the left upper nasal quadrant.

The impression was that of a case of the Groenblad-Strandberg syndrome, with pseudoxanthoma elasticum and angioid streaks.

FIG. 5.—Typical *peau d'orange* fundus appearance as seen temporal to the macula in both eyes.

FIG. 6.—Macula of left eye, showing *peau d'orange* appearance temporal to fovea (symmetrical appearance in other eye).

FIG. 8.—Angioid streak in upper nasal quadrant of left fundus.

FLUORESCEIN FUNDUS PHOTOGRAPHY

FIG. 2.—Right eye, showing angiod streaks.

FIG. 3.—Left eye, showing angiod streaks.

FIG. 4.—Macula of right eye, showing angiod streaks (to the right) and *peau d'orange* appearance temporal to the fovea (to the left).

FIG. 7.—Right eye, showing "salmon spots" in lower fundus. One angiod streak is seen above.

Discussion

Controversy formerly existed in the literature concerning the exact histological nature of angioid streaks. This varied from breaks in Bruch's membrane, abnormal choroidal vessels, breaks in Bruch's membrane allowing normal choriocapillaris to be seen, and breaks in Bruch's membrane allowing abnormal choroidal vessels to be seen, to breaks in Bruch's membrane with ingrowth of scar tissue. It was thought that it would be interesting to study such a case by fluorescein fundus photography (Smith, David, Hart, Levenson, and Tillett, 1963), because this technique allows a physiological interpretation of fundus morphology, so that one can study independently the arterial, arteriovenous, and venous phases of retinal circulation. As is seen in Figs 9 and 10, in this case the angioid streaks definitely fluoresced in the arterial phase. Furthermore, with passage of time, the streaks became more prominent, and indeed they stained with fluorescein for some time after the retinal vessels had become clear of the dye.

FIG. 9.—Fluorescein fundus photograph of right eye (arterial phase). Note that the angioid streaks light up in the arterial phase.

FIG. 10.—Fluorescein fundus photograph of same right eye (late venous phase). Note that angioid streaks still fluoresce after retinal vessels are nearly empty of dye.

There are several possible interpretations of these photographs. It would seem, from their appearance, that angioid streaks are not blood vessels. Further, as is seen in the histological study of another case (Fig. 11), there is no doubt at present that disruptions in the continuity of Bruch's membrane occur in this disorder.

FIG. 11.—Photomicrograph of another case with angioid streaks. Note break in continuity of Bruch's membrane.

Loss of pigment epithelium with resulting baring of the underlying choriocapillaris might account for more prominent fluorescence in these areas. On the other hand, ingrowth of glial tissue and capillaries into the break in Bruch's membrane might occur to such a degree that early fluorescence might be noted. Altered tissue permeability might allow extravascular retention of fluorescein for a longer period than elsewhere in the retinal tissues. Such a finding may be seen, for instance, in old disciform degenerative lesions of the macula (Norton, Smith, and Justice, 1964). Which of these or other factors best explains the combination of *early appearance* and *delayed transit* of fluorescein from the angioid streaks is unknown at this time.

The term *peau d'orange* was proposed to describe the stippled appearance noted in both eyes in this patient temporal to the macula. The similarity of its texture to that of the skin changes of pseudoxanthoma elasticum on the neck was striking. One of us (J.D.M.G.) noted similar ophthalmoscopical changes in both eyes of a child who also had pseudoxanthoma elasticum.

It would appear that more careful study of the peripheral fundus with binocular indirect ophthalmoscopy is warranted in patients with angioid streaks, to assess the frequency of associated ophthalmoscopic findings as well as their clinical significance. The histological picture of scar tissue growing into dehiscences of Bruch's membrane accounts for the not infrequent distressing loss of central vision in some of these patients. The tendency to the involvement of blood vessels elsewhere explains the calcification of peripheral vessels noted in lateral roentgenograms of the legs in this disorder. For the same reason, a search for angioid streaks should be made in persons with repeated gastrointestinal tract haemorrhage despite repeatedly negative radiological studies. These points are helpful in diagnosing cases of the Groenblad-Strandberg syndrome when the fundus lesions may be more subtle than those in the patient here described.

Summary

The first reported fluorescein fundus photographs of angioid streaks are presented. Fluorescence appeared in the streaks in the arterial phase, increased in the venous phase, and tended to persist for a time even after the retinal vessels had cleared themselves of the dye. A fundus change seen in this patient somewhat resembled the cutaneous changes of pseudoxanthoma elasticum, and for this the term *peau d'orange* fundus is proposed.

Grateful acknowledgement is made to Dr. Lorenz Zimmerman for use of Fig. 11.

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ADDENDUM

Since this article was submitted, our attention has been drawn by Dr. David Paton to a previous report of the mottled or *peau d'orange* fundus in association with angioid streaks. This paper by K. Shimizu is entitled "Mottled Fundus in Association with Pseudoxanthoma Elasticum" (*Jap. J. Ophthalm.* 5(1): 1-12, Jan. Mar. 1961); it is written in English and has excellent illustrations, and Fig. 3 is strikingly similar to Fig. 6 in the present paper. More attention should be devoted to the "salmon spots" also seen with angioid streaks.