

## UNUSUAL TRAUMATIC RETINAL HAEMORRHAGES ASSOCIATED WITH ANGIOID STREAKS\*

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THIS report concerns the effects of apparently slight trauma upon the eyes of a young male patient with angioid streaks and other signs of pseudoxanthoma elasticum.

### Case Report

An apparently healthy 16-year-old youth, whose sight had never been in question, was admitted to hospital complaining that the vision in each eye had deteriorated immediately following a fight. In spite of having been punched about the eyes the only outward sign of trauma was a slight bruise on the bridge of the nose, and both eyes had brisk pupillary reflexes and appeared superficially normal; the visual acuity, however, was reduced to 6/60 in each eye. There were no field defects.

A bizarre appearance of both fundi resulted from widespread chorio-retinal haemorrhages, which seemed to extend through otherwise typical angioid streaks. In each eye the macular area was displaced forwards by a mass of blood, and a tessellated outline to the upper border of this haemorrhage suggested that the nerve-fibre layer had been reached in this part of the retina. Elsewhere the extravasated blood spread evenly and symmetrically in the plane of the retina, and nowhere had it penetrated as far as the subhyaloid space (Fig. 1*a*, opposite).

In addition, in the mid-periphery of each fundus were scattered approximately six round and "punched-out" looking spots, which, apart from an associated clump of dark pigment, were a startlingly white colour; each was about a tenth of the size of the optic disc. These spots appeared to be similar to the "distinctive mosaic" of spots which Duke-Elder (1940) described as being sometimes associated with angioid streaks. There were no crescentic choroidal tears, and no signs of commotio retinae.

The blood absorbed first along the line of the angioid streaks, which, because of associated choroidal atrophy then became a white line passing through the centre of each haemorrhage (Fig. 1*b*). Except in the region of the maculae, where organization and fibrosis produced the typical picture of disciform degeneration, the remainder of the haemorrhages finally disappeared completely (Fig. 1*c*); the usually prolonged sequence of occasional spontaneous haemorrhages and choroidal atrophy, leading ultimately to macular degeneration, was therefore telescoped into the space of a few weeks. Scarring of the left macula was more marked than the right, and the visual acuity 6 months after the injury was 6/12 in the right eye and less than 6/60 in the left.

*General examination.*—Other manifestations of pseudoxanthoma elasticum were confined to typical striae of the skin of the axillae, and to nearly absent radial and ulnar pulses. The results of the following investigations were all normal: bleeding time, clotting time, prothrombin test, serum protein, serum cholesterol, and full blood count (platelets abundant). The Wassermann reaction was negative and the blood pressure was 130/80.

### Discussion

Goodman, Smith, Paton, and others (1963) agreed with the widely held view that angioid streaks are caused by cracks in Bruch's membrane resulting from changes in the elastic fibres; they also noted the tendency towards ocular haemorrhages in

\* Received for publication May 3, 1965.

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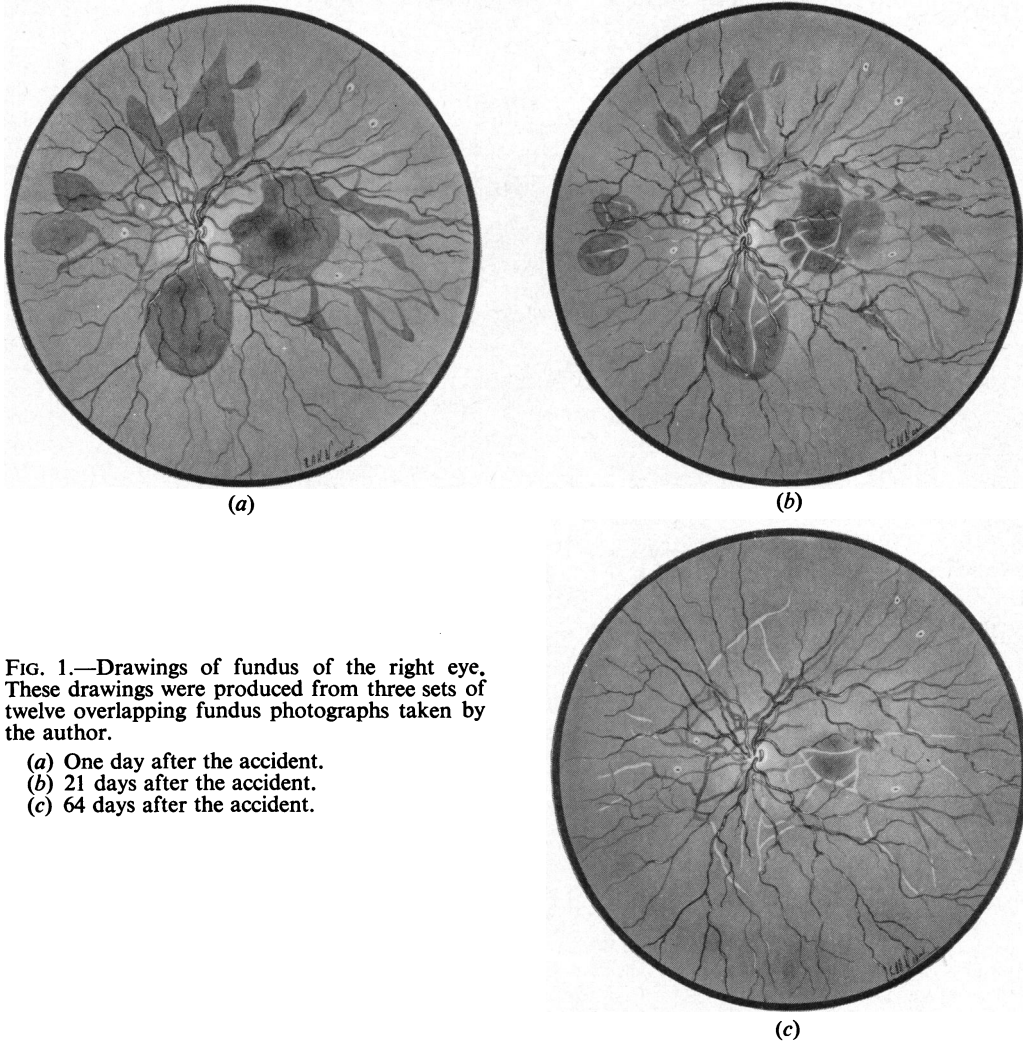


FIG. 1.—Drawings of fundus of the right eye. These drawings were produced from three sets of twelve overlapping fundus photographs taken by the author.

- (a) One day after the accident.
- (b) 21 days after the accident.
- (c) 64 days after the accident.

patients suffering from pseudoxanthoma elasticum, and in addition demonstrated asymptomatic arterial occlusive changes in the limbs of nine out of twelve patients with this complaint. This latter change was due to proliferation of the media of the vessel wall, and not, as in the eyes and skin, to changes in the elastic fibres.

The possibility that relatively slight trauma could cause multiple choroidal haemorrhages, and that the blood might track through the gaps in Bruch's membrane, can easily be envisaged, and this is exactly what appears to have happened; the precise plane where the extravasated blood collected, however, can only be conjectured. In the macular areas the depressed function, the marked swellings, and the shape of the upper parts of the haemorrhages suggest that the nerve fibre layers of the retina have been reached. Elsewhere the blood appears to have spread evenly sideways from the angioid streaks without impedance from adhesions between tissue planes; the absence of field defects corresponding to the sites of these haemorrhages, together with their complete absorption without residual scarring, suggests that the

blood lay behind the photoreceptor cell layer and yet in front of the angioid streaks. It seems highly probable that it collected in the potential space between the retinal pigment layer and the photoreceptor cell layer, access for the blood to reach this normally avascular zone being afforded by the ruptures in Bruch's membrane.

Although Doyne (1889) and Ballantyne and Michaelson (1962) each described a case of angioid streaks thought to be due to blunt trauma, the same is very unlikely to be true of this case; for not only were both eyes symmetrically involved in the absence of severe direct trauma, but there were other signs of pseudoxanthoma elasticum. Similarly, had the angioid streaks and the haemorrhages occurred together as a result of relatively slight trauma to eyes with a weakened Bruch's membrane, it is unlikely that the streaks would have been so extensive and symmetrical; this, however, remains a possible explanation.

The white fundus spots appear to be discrete areas of choroidal atrophy, sometimes occurring in the line of angioid streaks (Batten, 1931) and sometimes, as in this case, in apparently healthy areas of the fundus. Recently attention has been drawn to fundus changes other than angioid streaks occurring in patients with pseudoxanthoma elasticum, and believed to be part of the clinical picture (Shimizu, 1961; Goodman and others 1963; Smith, Gass, and Justice, 1964). These changes occur alone or in addition to angioid streaks, and the one most frequently reported is a diffuse yellowish mottling of part of the fundus, variously described as *peau d'orange* or mottled fundus.

This was not apparent in this case, but it is possible that the white fundus spots represent the final stage of another change noted by Smith and others (1964). These authors described and illustrated yellowish-white spots in the mid-periphery of the fundus similar in size, shape, and site to the spots noted in the eyes reported in this communication; the only difference was in their colour, and it is possible that the ones which they described represented an earlier stage of choroidal atrophy than the startlingly black and white spots described in this paper. Being smaller, rounder, and more discrete than an old focus of choroiditis their appearance was striking, and their presence should at least arouse suspicion of the systemic disorders associated with angioid streaks, even if the latter are absent.

I wish to thank Mr. R. Dagleish for permission to report a case under his care, and for helpful suggestions on many points. I am also especially grateful to Mr. R. A. H. Neave, medical artist to the University of Manchester, for his painstaking work in producing drawings from the fundus photographs.

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