should lead to avoidance of occlusion therapy. The field defect may be quite constant and may be recognised by an observant patient. Case 4 would tend to strengthen the belief that the condition is a congenital one. The possibility of such a "pit" extending back into the orbit and uniting with a cyst, thus producing unilateral exophthalmos, should also be borne in mind¹².

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A CASE OF A GRÖNBLAD-STRANDBERG SYNDROME, WITH DISCIFORM DEGENERATION OF THE MACULAE*

BY

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IN March, 1946, C. R., male, married, aged 38 years, a motor salesman, noticed distortion of objects to his right side, most noticeable when he was car driving, and absent when he closed his right eye. At that time, his visual acuity was 6/5 6/5 (Snellen) with no error of refraction and at six metres, there was $\Delta 1/4$ left hyperphoria, and at 20 cms. esophoria 3°. The media were clear, but each fundus showed several, small, scattered, ovoid patches of yellow-white exudate with discrete, faintly pigmented margins, two of which were in the right macular area. Over these, the macular capillaries were without distortion or interruption.

During one year, the case was repeatedly examined, and several, new, small patches of exudate were seen to form in the mid-periphery of each eye.

In March, 1947, several new patches were seen in the right

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macular area, and subjective testing on Bjerrum screen at two metres revealed a scotoma in the right central area of the field of vision, with a dense nucleus and sloping edge which overlapped the fixation point. The vision of this eye was reduced to 6/24 by April, 1947.

The case was observed closely, and in September, 1947, the patient reported rapid failure of central vision for all purposes, including close work during one week. He was unable to read newsprint, and objects seen with the left eye appeared distorted and blurred. Visual acuity was then O.D. less than 6/60; O.S. 6/36 partly.

The scotoma, recorded before as present in the right central area, was found on Bjerrum screen at two metres, to be more extensive, with a larger, dense nucleus and steeper margins. The scotoma was dense to targets of less size than 10/2000 white (artificial light 100 watt 6 feet above patient). The left field of vision showed a small, central scotoma, relative in quality.

The right fundus, at this time, showed a roughly circular, white patch of exudate in the macular area, approximately twice the size of the optic disc, over which the macular capillaries ran without distortion, though standing out well against the brilliant white background of exudate. This patch gave the impression of being slightly raised above the level of the surrounding normal retina. To the nasal side of the disc, a brown-black angiod streak was

PHOTOGRAPHS 1-3.

DIAGRAM 1.

present with indistinct powdery margins, equal in width to an artery of first division of the central retinal artery, and curved concentrically with the disc margin, separated from this by a narrow interval. Three angeoid streaks ran radially from the convex border of this out towards the nasal mid-periphery of the fundus. (See photographs 1-3 and diagram 1.)

DIAGRAM 2.

The left fundus showed a "haemorrhagic" patch of exudate, roughly circular, in the macular area, and one half the size of the optic disc. This patch increased in size from day to day, becoming constant at the side seen in photographs 4-5 and acquiring a grey-black centre and brown-black margin, as is well seen in the photographs as two separate densities.

PHOTOGRAPHS 4-5.

An angeoid streak was present in the left eye, similar in appearance to that in the right fundus, but placed nearer to the disc margin on its nasal side.

On each fundus, the retinal vessels were normal in appearance, while the patches of exudate in the mid periphery of each fundus were not related to the radial angeoid streaks. The retinal vessels crossed the angeoid streaks without deformation or distortion.

Throughout the period of observation, no signs of uveitis have

appeared, with absence of "K.P." aqueous or vitreous haze, and ciliary injection, while the fields of vision remained of normal, relative extent.

A discrete patch of pigmentation on the anterior surface of the right iris has remained unaltered.

During the period of observation, it was noticed that the patient had altered skin of the left side of the neck. Similar changes were found on extensive areas over the epigastrium, lower chest anteriorly, the anterior fold of the left axilla, and the skin of each antecubital fossa and axilla. In all these areas, the skin was soft, thin when picked up between finger and thumb, excessively mobile on the underlying tissues, and possessed a fine mosaic of yellow ridges, less than a millimetre in width, easily palpable to the pulp of the examining middle finger. The effect of voluntary contraction of platysma myoides on the skin of the neck was less on the left side than on the right. One café-au-lait stain, the size of a British penny postage stamp, was present over the left temporal region.

Investigations

March, 1946.

	$\frac{1}{1,000}$	negative
Mantoux intradermal skin test	$\frac{1}{10,000}$	negative
	$\frac{1}{100,000}$	negative

Wassermann (venous blood), negative. B.P. 160/70. Chest X-ray.—No abnormality seen. X-ray teeth.—Apical infection of two teeth. These were extracted.

September, 1947.

X-ray paranasal sinuses.—"Loculated type of antrum on the right side, with diminished translucency of part of the antrum. ? Congenital anomaly ? after infection." There are no clinical signs or symptoms of antrum disease.

October, 1947.

X-ray skull and pelvis.—"There is no evidence of Paget's disease of bone."

B.S.R. 3mm. in one hour. Coagulation time, 2 mins. 40 secs. Bleeding time, 7 mins. 30 secs. Hb. 92 per cent. R.B.C.'s, 5.06 million. C.I. 0.92. W.B.C.'s 12,600. Polymorphs 62 per cent. Eosinophils 1 per cent. Basoph 1 per cent. Lymphocytes 30 per cent. Mononuclears 6 per cent. The red cells were of normal shape, size and staining. Capillary fragility test.—Slight increase.

Urine.—No abnormal constituents.

V.—O.D. less than 6/60. O.S. 6/60, with -0.50 D.Sph. added—part of 6/36; eccentric fixation—part of 6/24.

The patient readily and suddenly learned to appreciate the value of fixing slightly to one side of the object of regard.

Telescopic lens systems. (Hamblin's). O.D. 6/60. O.S. 6/36 with -1.00 D.Sph. placed behind the system.
part of 6/24 direct fixation.
6/18 eccentric fixation.

With a 12 D.Sph. small lens added to the system. O.D. J.16 read badly. O.S. J. 10 read rapidly and well.

General physical examination showed no abnormal signs, radial pulses equal and normal, 72 per min. There is no history and no other sign of peripheral vascular disease except a history of pain in both calves after walking about a mile or so. This pain subsides after resting, but is inconstant in incidence.

Family History.—The father and mother died aged 75 years and 77 years respectively, and one brother died as a youth of tuberculous peritonitis.

One sister has normal vision, and the patient knows of three cousins with no history of eye disease, though none was available for examination.

Diagnosis.—When the case was first seen, the diagnosis was disseminated choroiditis, using the term in a descriptive sense, rather than the narrower sense of implying the common picture of syphilitic choroiditis.

The significance of the aneoid streaks was clearly recognised and a bad prognosis given.

The skin condition was not discovered until September, 1947. The patient gives a long history of unusual skin texture in the areas described, with no recent change.

The changes in each macular area have been labelled disciform degeneration, and the photographs show perhaps two stages in a common process, the right eye a late stage, the left an early "haemorrhagic" state.

Treatment.—Focal sepsis was searched for, but, apart from the teeth mentioned, none was found.

Sulpha drug, penicillin, mixed vitamins, iodides, salicylates, coagulin ciba, and other drugs have been given a trial, but the case has steadily progressed to the present state.

Photographs and diagrams

Comments on Photographs.—These photographs stress the severe limitation of fundus photography as so far evolved. In them, the size of the areas of macular exudate is shown, and

the difference in appearance on the two sides is also shown well. The angeoid streaks are recorded but faintly and appear similar in the one side on photographs 1 and 2—as opposed to obvious defects and artefacts which have moved in position on the two photographs.

The arteries and veins of the same fundus differ in different photographs in apparent diameter and appearance. This illustrates my personal opinion that fundus photography cannot be held to record accurately such factors as the nature of the light streak, arterio-venous crossings, venal diameter, though the degree of tortuosity (or “straightness”), especially of macular capillaries, can be recorded thus.

This opinion I have reached after photographing a series of 150 fundi in healthy persons of greater age than forty years.

The Grönblad-Strandberg syndrome consists of angioid streaks of the fundi with pseudo-xanthoma of the skin. Originally described by Grönblad, with Strandberg as the authority on the skin condition, this syndrome has been repeatedly found and cases published.

The existence of other abnormalities has been found in the presence of the syndrome, or of the angeoid streaks.

The syndrome, plus exudation at the macula resembling disciform degeneration, was described by Goedbloed² who quotes Wildi, 1926, as describing three stages of development of the lesion in the macular area.

1. Streaks with normal central fundus.
2. Detachment of the retina by exudate in the macular area.
3. Organisation of the exudate to form a grey disc.

The case described falls broadly into this description.

Angeoid streaks and senile elastosis of the skin are associated in two cases by Goedbloed².

Angeoid streaks and Paget's osteitis deformans of bone have been described by Terry³, Lambert⁴ and others⁸. The syndrome, however, has not been described in the presence of Paget's disease.

A case of moderately advanced Paget's disease was observed by me over a period of twelve months, and developed scattered small areas of yellow exudate in both fundi, some in the central areas. There were no skin changes in this case, and no streaks. The familial tendency of streaks has been established by Law⁹, Wassenaar¹⁰, Goedbloed², who quotes Franceschetti, and Roulet, 1936, and the factor appears to be recessive.

Pathological studies by Hagedoorn⁵ suggested degeneration of elastic tissue of Bruch's membrane, with perhaps similar alteration of the elastic structure of blood vessels. Law⁹ found folding of the

retina and accumulation of pigmented debris to coincide with the streaks.

Recently, attention has been given to the blood vessels of the extremities in cases of angeoid streaks by Guenther⁶ and Scheie⁷ with Freeman⁷. Guenther investigated pulse wave velocity and form in the peripheral musculo-elastic arteries (Muskelelastische Arterien) in nine cases of the syndrome, eight of arteriosclerosis, and nineteen "normals." The velocity was lower in the syndrome cases, while the proportion of the height of the base of the incisure, compared with the amplitude, expressed as a percentage, was higher than normal.

The alterations found were considered significant, and indicative of degeneration of the elastic tissue elements of the vessel walls. There was no confirmation by biopsy of the vessels. Symptoms attributed to vascular changes were weakness of extremities, coldness of hands and feet, dyspnoea. Scheie⁷ and Freeman⁷ supply some confirmation in describing three cases of streaks. In two cases, the whole syndrome was present, with severe vascular disturbances in the limbs. Absence or diminution of pulsation of peripheral arteries was present. Oscillometric records of pulsation showed a generally diminished amplitude in these vessels in both cases. In one case, biopsy of an ulnar artery was carried out. The diminished pulsations were confirmed during the dissection.

Histologically, the intima and internal elastic lamina were normal. The media showed fragmentation and vacuolation of elastic tissue by Weigert's stain to a marked degree. There was hypertrophy of muscle fibres.

In both cases showing vascular abnormalities, calcification of major limb vessels was evident radiographically. These writers suggest routine ophthalmoscopic examination of cases of unexplained peripheral vascular disease.

The association of Paget's disease with the angeoid streaks, angeoid streaks with changes in the dermal elastic tissue, the streaks and skin changes with peripheral vascular disease, and with abnormal calcification of peripheral major vessels, point to a probable common factor. Perhaps more systematic investigation of each type of case may be productive of further knowledge, but the tendency for each writer to be confined to a small collection of each manifestation operates against this.

Permission to publish the account of this case by Mr. J. J. McCann, F.R.C.S., of Liverpool, under whose care the case was observed, is acknowledged. Use of the fundus camera at the Liverpool Medical Research Institute was granted by the Director, Dr. I. Harris, Liverpool.

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A MYXO-HAEMANGIOMA SIMPLEX OF THE CONJUNCTIVA BULBI*

BY

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GRONINGEN

A WOMAN, aged 35 years, came into my consulting-room, complaining that she had the sensation of a foreign body in her right eye. For six weeks she was aware of the presence of a small tumour in the inner angle. Nothing was known about any trauma. The tumour was growing rapidly and at the time of my investigation it was about 4 mm. high and had a diameter of 3 mm. It was of a fungus like shape and had a broad and short pedicle. It was movable on the underlying tissue. Its base was partly covered by the semilunar plica. The tumour had a cherry-red colour and a smooth surface. At its base a tortuous and dilated vessel was visible.

Because of its disfiguring effect and its rapid growth I decided to remove the tumour. This was performed under cocaine anaesthesia. The tumour was excised by the single snip of a pair of scissors. There was little or no haemorrhage. Fixation with Bouin's liquid.

The microscopical examination gave the following picture: The whole tumour is completely covered with normal epithelium, containing many goblet cells (Fig. 1). The main substance of the tumour consists of compactly arranged small blood-vessels (dilated capillaries). The endothelial cells are extremely swollen. Here and there we find more than one layer, so that the lumen is obliterated by them. Some larger vessels intersect the tumour. In various sections we see large oedematous areas and some smaller

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