persist almost unchanged until delivery. So perhaps it is wise to report these two cases without explanatory hypothecation.

Summary

Two cases of acute and severe haemorrhagic retinopathy are described in pregnant diabetic women. The condition does not conform either to usual diabetic or to toxic renal retinopathy and the latter was excluded.

I am indebted to Mr. L. H. Savin for close study and reports on the retinal changes.

CRATER-LIKE HOLES IN THE OPTIC DISC*

BY

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ALTHOUGH Duke-Elder¹ has stated that the condition known as "pits" or "crater-like holes in the optic disc" is of relatively frequent occurrence, yet Greear² has been able to collect and tabulate only seventy-two cases including his three cases, the only three he had encountered in eighteen years of active practice. There would seem to be sufficient reason to report any additional cases and possibly in so doing to lend statistical support to the present-day concept of the condition without the necessity of awaiting pathological studies before one can be sure of the nature of this anomaly³.

CASE 1. This 31-year-old patient was examined routinely prior to embarkation. His vision was 20/20 in each eye and his only ocular complaint was difficulty in reading for prolonged periods. There were no abnormal eye findings other than those of the right disc (Fig. 1). This disc was ovoid in shape with a thin, pigmented scleral ring on the temporal side. There was present a small physiological cup about 1/3 D.D. in size located slightly upward and nasally. The cup was about three dioptres in depth and presented a pronounced cribriform plate at its bottom. Just nasal to the physiological cup was a triangular-shaped "pit" with its apex pointing inferiorly. This "pit" did not reach quite to the edge of the disc nor to the nasal edge of the physiological cup. The "pit" appeared to be blue-grey in colour and seven or eight dioptres in depth. A thin connective tissue veil seemed to be present within the cup rendering it difficult to visualize the bottom

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of this "crater." The edges of the "crater" were quite distinct. One small vessel could be seen approaching the nasal side of the disc and then disappearing over the nasal lip of the "pit." This vessel could not be followed very far down into the "pit" for it

FIG. 1.

seemed to disappear completely. There was no further anomaly of structure of this disc. The patient's vision was found to be 20/20 in each eye with correction. Fields and blind spot studies produced the pictures seen in Figs. 2 and 3.

In summary this disc presented a "pit" upon the inferior nasal aspect of the disc triangular in shape and seven or eight dioptries in depth. A temporal crescent was present along with a characteristic field defect. The "pit" was covered over by a connective tissue substance and was blue-grey in colour.

CASE 2. The second case was that of an adult male, aged 51 years, who appeared for a routine refraction and offered the information that he had upon a previous examination been informed that there was present in his right eye a very rare condition known as a "hole in the optic nerve." After learning of this condition the

FIG. 2.

patient had conducted several tests upon himself in which he learned that he had a field defect in his right eye which to him seemed quite marked when compared with the visual field of his left eye.

Examination of the left eye was not remarkable. The right eye too was not unusual, except for the appearance of the right disc (Fig. 4). This showed a pigmented greyish-black scleral crescent upon the nasal third of the disc which was fairly well demarcated in its upper half but not pigmented below. There were three islands of clear area within the crescent. A fairly large physio-

logical cup was present which occupied about one-third the entire area of the disc and was eccentrically displaced nasally. The central retinal vessels emerged in no unusual manner from this cup with the primary arterial and venous bifurcation occurring before emergence. On the temporal portion of the disc a very small round "pit" was present which approached but did not

FIG. 3.

touch either the physiological cup or the disc margin. One small vessel appeared to enter the cup near the upper edge, emerging at approximately "9 o'clock" and running upward over the disc edge. Below the lower lip of the "pit" a small vessel approached the edge of the hole but then curved gracefully away to run off the disc and on to the retina. The "pit" was of pale blue colour and was covered over by a greyish foam-like membrane so that the depth of the hole could not be ascertained. The hole seemed to

take a slightly oblique source temporally. There were no other anomalies of the vessels upon the disc or the retinal structures. A study of the visual field disclosed a defect as pictured in Fig. 5.

FIG. 4.

FIG. 5.

CASE 3. This patient, aged 29 years, was examined at this hospital because of extremely poor vision in his right eye of many years' duration. This eye had always been much poorer than his left eye and although the patient was right-handed he was aware of the fact that he had always used his left eye when sighting through a rifle. About ten years ago the right eye was injured by a small seed brushed from some tall grain. The patient was seen shortly afterwards by a specialist who removed a foreign body from the cornea and then put a patch upon his eye. When this patch was removed the patient noticed that his vision was greatly reduced. He doesn't recall making any effort to determine the cause of this diminished vision subsequently, but knows that at every eye examination thereafter something of interest was discovered in his right eye which caused a good deal of comment by all examiners. Examination disclosed vision in the right eye to be restricted to hand movements. The external ocular examination was negative. The right eye was slightly divergent; no effort was made at fixation. The iris and lens were normal. In the right macula there was a horizontal lesion resembling a tear with the prominent redness of the choroid brightly shining through and with the edge of this cleft strongly pigmented. There was a definite and complete scleral temporal crescent and in the infero-temporal portion of the disc was an obliquely placed slightly triangular

FIG. 6.

"pit" (Fig. 6). This was about $1/8$ D.D. in size and was covered over by a rather dense connective tissue membrane which was somewhat shiny. The "hole" was greyish-blue in colour; its depth could not be measured since one could not focus beyond the range of the connective tissue covering. The temporal edge of the "pit" did not quite reach to the edge of the disc. Its margin temporally was extremely distinct and the oblique nature of the "pit" appeared to be emphasized by this sharp temporal edge. In the inferior portion a vessel seemed to approach the hole and then pull away from it without entering the "pit." There was a pronounced physiological cup with normal vessel arborization. A visual field study done upon this eye failed to illustrate the characteristic sector-shaped defect probably because of the extremely poor fixation resulting from the macular lesion. There was a marked concentric contraction of the peripheral field.

CASE 4. Is that of an adult male soldier 27 years who had been under the care of the eye department at a station hospital for more than a year because of suspected glaucoma. During a routine examination for refraction the peculiar cupped appearance of the patient's left disc was observed (Fig. 7). Following changes noted in the blind spot of that same eye, the patient was put upon an anti-glaucomatous régime. He was observed at very frequent

FIG. 7.

intervals during which time no variation was discovered in the appearance of the blind spots, the peripheral fields or the diurnal tension curve. Vision in each eye was 20/70 but was readily corrected to 20/15. There was no involvement of the peripheral field but studies of the blind spots showed an enlargement (Fig. 8).

FIG. 8.

The fundus of the right eye presented a small colobomatous area just below the disc enclosed by a pigmented crescent and traversed by several retinal vessels which certainly suggested the presence and location of an atypical congenital coloboma (Fig. 9).

CASE 5. A. S., aged 10 years, had been given occlusion therapy because of an amblyopic left eye. This consisted of total occlusion during the summer months and occlusion therapy several hours daily after school. The treatment was continued for a period well

over one year at the end of which time no improvement was noticed. The patient was first seen shortly thereafter. During routine fundus examination a crater-like "pit" in the optic disc

FIG. 9.

of the left eye was noted in the inferior temporal sector of the disc. It was oval in shape, greenish-grey in colour and apparently quite deep, although the depth of the "pit" could not be ascertained both on account of the connective tissue overlay and the obliquity of extension of the hole. Two small vessels approached this crater and then dipped very abruptly over its edge to disappear into its depth. A rather pronounced physiological cup was present in this disc (Fig. 10).

CASE 6. This patient was a 47-year-old female who recently developed symptoms of presbyopia. She had never suffered from any eye condition. There were no irritative phenomena, headaches or blurred vision. Within the past six months difficulty in threading a needle and in reading telephone type had become more and more pronounced. The patient's vision was 20/25 in each eye and was readily corrected to 20/20 in each. The right pupil was smaller than the left. Although each pupil reacted to light and to accommodation, the response of the left pupil was not very prompt to light. There was considerable atrophy of the inner circle of the

FIG. 10.

iris of the left eye with a very mild degree of iris atrophy on the right side. The media were clear. The fundus of the right eye appeared perfectly normal. The left fundus presented the anomaly shown in the fundus photograph (Fig. 11). The fundus in general was albinoid with the choroidal circulation readily showing through to the retina. The disc was slightly oval in shape with the upper four-fifths appearing quite normal in all respects but the lower fifth presented an anomalous depression. This lunar-shaped depression was five dioptres in depth, greyish-green in colour and covered by a thin irregularly-formed veil. On the temporal side two vessels disappeared suddenly just beyond the margin of the cup to become visible again in the depths of the depression. On the nasal side one large vein was seen dipping abruptly into the cup and reappearing just beyond the margin of the disc. The vessel pattern was not particularly abnormal. Because of the irregularity in the size of the pupil, the atrophy of

FIG. 11.

the iris, the appearance of the left disc and because of lack of familiarity of the condition of "pits" a study was made of the patient's intra-ocular pressure, but the diurnal curve failed to show any variation of significance. Provocative tests (*i.e.*, dark room, dilatation, coffee, excess of fluids), caused no increase in the intra-ocular pressure. Studies made upon visual fields and blind spots showed a defect associated with the congenital "pit" (Fig. 12).

CASE 7. This 42-year-old female first sought aid because of persistent diplopia, headaches, nausea and vomiting. For many years she had been able to read only if one eye was kept closed. She also noticed that the left lid had a tendency to droop, the condition becoming quite pronounced when she gazed into the mirror. The patient was found to have a paresis of the left superior rectus muscle along with a pseudoptosis. The fundus of the right eye appeared normal. The fundus of the left eye showed a peculiar cupping of the nerve head. In the lower outer quadrant there was a small partial coloboma about $1/8$ D.D in size and slightly bluish

FIG. 12.

in colour. It was covered over by a connective tissue membrane so completely that no depth could be determined for the hole formation.

Discussion

Most case reports of this condition show the crater to be present in the temporal half of the disc and usually in the lower portion. The "pit" never extends beyond the edge of the disc although that structure may be distorted in a horizontal direction. A. Fuchs² stated that congenital "pits" occupy the sclera adjacent to the lower margin of the papilla and only exceptionally are they seated within the papilla itself. Fuchs presented a very rare case in which a congenital "pit" was within the excavation of a glaucomatous cup. Most "holes" appear to be directed backwards in a straight and not an oblique fashion. Cases 3 and 5 in our series appeared to be somewhat oblique. Also in Case 3 the temporal margin of the disc appeared to be unusually sharp. In three instances the depth of the "pit" could not be ascertained in our cases; in case I the "crater" was seven or eight dioptres in depth. In a case reported by Moffatt⁴ it was noted that the darkest part of the

hole was at its outer edge giving the impression that the deepest portion of the "pit" was behind the overhanging edge of the sclera. This same appearance was also noticed in our third case. In a case reported by Neame⁵ the patient noticed a gap in his visual field which caused him to seek aid much as occurred in our second case. Mann⁶ states "there are no vessels in the pits," which statement is apparently not true, for in a case reported by Nielson⁷ the author states that "vessels descended at the edge of the groove and reappeared upon the other side." Several of our cases also show vessels within the "pits."

A case presented by G. Edmund⁸ showed a groove which occupied the entire temporal half of the disc and reached completely to its margin. This groove was darker in colour but only three dioptres in depth. A second case reported by this author showed an inferior conus with a horizontally oval hole lying between the disc and the crescent. This second case had iridodonesis which was pointed out as a factor in suggesting that the condition is one of congenital abnormality. Other anomalies are said to be associated with this condition—i.e., microphthalmia, ectopia lentis, coloboma, persistent hyaloid artery, etc—but none was observed in our case reports.

Most authors believe the "holes" are colobomata. They may be the result of defective foetal cleft closure, or a remnant of the hollow which connects the optic cup with the cavity of the optic stalk. Wessely⁹ believes the "pits" are remains through which have passed cilio-retinal or optico-ciliary vessels whose atrophy had been completed rather late in intra-uterine life. Edmund⁸ thinks the "pits" are the results of abnormal vascular grooves. Alabaster¹⁰ expressed the opinion that a vascular accident caused a collapse of the small capillaries in the nerve head with a defect subsequently remaining. Mann⁶ states that in all probability these defects *are not holes*, believing that their nature is not always clear since they seldom come to microscopic examination. She furthermore states they may be cysts or pockets filled with transparent tissue, but there is no clinical evidence that these structures are cystic. Greear² has emphasized that the condition is not a true coloboma which would be secondary to disturbance of closure of the foetal fissure. He states that these "pits" are atypical colobomata by which is meant that the "pit" resembles a coloboma but has its origin in an entirely different manner.

SUMMARY. Crater-like holes may be erroneously regarded as evidence of glaucomatous changes, particularly when there are pupillary inequalities, iris atrophy and field defects. They may also be present in amblyopic eyes in which instance, recognition

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*

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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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