I. Introduction

In a previous paper\(^1\) I dealt with some anomalies of the optic disc, which by an inexperienced examiner might be mistaken for a glaucomatous excavation. I mentioned in this respect an unusually oblique insertion of the optic nerve, which may be present even in cases of slight myopia and may simulate a glaucomatous cupping by the apparent bending of the vessels at the margin of the papilla (Fig. 9). I also mentioned an unusually large physiological cupping (Fig. 3) of congenital origin, the pale colour of the floor of the papilla making it still more like a glaucomatous excavation. Another anomaly is the branching of the central vessels behind the lamina cribrosa so that the individual branches emerge only at the margin of the papilla (Fig. 10). Finally, a glaucomatous cupping may be simulated by a congenital coloboma of the optic disc, if a circumscribed depression of its surface has produced a pit-like cupping. The congenital pits occupy, as a rule, the sclera adjacent to the lower margin of the papilla. Exceptionally they are seated within the papilla itself, mostly directly downwards or downwards and nasally or temporally. Such pits may be fairly deep so that their floor appears dark because it is not illuminated by the
ophthalmoscope, and may be mistaken for pigmentation. In most of the cases of coloboma, however, the depression of the optic nerve head extends beyond the limits of the papilla.

As a very rare occurrence a congenital pit in the optic disc may coexist with a glaucomatous excavation as in the following case, which I examined in the first Vienna Eye Clinic.

Case 1.—A woman, aged 64 years, discovered by chance, ten years ago, the poor vision of her right eye. An oculist whom she consulted eight weeks ago for a slight inflammation of her eye diagnosed glaucoma. The patient had never noticed blurred or iridescent vision.

Each eye shows no injection, nor precipitates on the cornea, with shallow anterior chamber, normal iris and pupil. The papilla of the right eye is grey with a tinge of red and with a complete excavation up to 4D. In the nasal half of the floor of the cupping there exists in the tissue of the optic nerve head a sharply defined pit in the floor of which one sees a short length of a vessel, without any visible connection with other vessels. At the nasal side the margin of the excavation is overlapping with a sharp bend of the vessels. Apart from reduced size of the retinal vessels the fundus is normal. In the lens some radiating opacities are visible.

Vision 6/18, with -2D. cyl. 160° V. 6/12? with +4D. sph. Jäger No. 2. The upper half of the field of vision is gone.

Tension (Schiotz) 22 mm. Hg, the same after homatropine and after adaptation to darkness.

Left eye normal.

In spite of the low tension I had no doubt, that I had to deal with glaucoma simplex, because of the total excavation. On account of the low tension a decompression operation was not required, but, as there existed also an incipient cataract, an iridectomy was performed to prevent later increase of tension and to be preliminary to the operation of cataract extraction.

The pit within the excavated papilla admits of a twofold interpretation. It may be a deeper part of the general glaucomatous excavation ("double excavation" Schmidt-Rimpler) a less resistant part of the lamina cribrosa yielding to the intra-ocular tension more than the rest, so that this place becomes the seat of a particularly deep, pit-like depression. This occurs only in glaucoma of long standing and with high hypertony, the pit occupying as a rule the vicinity of the entrance of the retinal vessels. In the present case the tension was found to be low and had probably never been higher, so that hypertony could not be the cause of this groove.

On the other hand there may be a simple coincidence of glaucoma and congenital pit; accounts of congenital pits in the papilla were
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published repeatedly, especially in 1908 and 1909, while later the publications have become scarcer, probably because this anomaly was considered to be sufficiently known. The presence of a vessel in the floor of the pit in my case is in accordance with similar findings in congenital pits and pleads in favour of its congenital origin. The reduction of the field of vision was due to the glaucoma which supervened later in life and caused the total excavation.

II. Colobomatous excavation simulating glaucoma

The two following cases simulated glaucoma so closely that they were mistaken for such by some experienced oculists.

Case 2.—The patient, a woman, aged 43 years, had noticed repeatedly three months ago the appearance of a "black wheel" before the eyes, which always disappeared after a few minutes. This symptom did not recur. No blurred nor iridescent vision.

The right eye looks normal externally with an anterior chamber of medium size as in the other eye. The optic disc is deeply excavated, with a sharp bend of the retinal vessels at its margin. The floor of the excavation is yellowish-white and covered by a semi-opaque tissue so that the vessels are not defined sharply enough to determine their refraction; but that the cupping is deep is shown by the strong parallactic displacement. The papilla is somewhat larger than in the other eye and is encircled by a narrow white halo; at its temporal side lie a few irregular patches of pigment.


The visual field examined with Bjerrum’s screen exhibits an increased size of the blind spot, which is a horizontal ellipse with a horizontal extension from 4° to 25° (Fig. 1). The left eye is normal. Vision 6/4 with +2D. sph. 6/4.

Tension, right eye: 20 mm.; left eye 18 mm. (Schiötz).

In the present case the history, the reduced vision, and the aspect of the optic disc pleaded for glaucoma which in fact had been diagnosed by an oculist. Considering the low tension the glaucoma ought to have been a simple one. But the fact that the right papilla was larger than the left proves the case to be not glaucoma but a congenital colobomatous excavation.* My father, to whom I showed the case, confirmed my diagnosis. A year later the picture was the same.

*In anatomical specimens of old glaucoma the scleral canal is found somewhat dilated, but this holds good only of cases of very long standing.
The diagnosis of a congenital malformation of the optic disc is in accordance with the enlargement of the blind spot, which with regard to its unusual size as well as to its extension along the horizontal meridian does not correspond with the findings in cases of glaucoma. The vessels at the floor of the excavation were seen but indistinctly, because they were covered by a layer of neuroglia, as has been recorded also in other cases of coloboma of the optic disc.

Similar to this case is

Case 3.—A girl, aged 12½ years, who came to consult me for conjunctivitis and for spectacles; said to have seen worse always with her right eye.

The papilla of the right eye is deeply excavated, with some of the vessels passing the margin of the papilla with a sharp bend, some disappearing there behind the scleral ring. The floor of the papilla is whitish and blurred, but the vessels are sufficiently distinct to determine their refraction, which is −8D, at the floor of the cupping. The papilla is surrounded by a sort of halo of about one-third of the diameter of the papilla. The halo stands out against the adjacent fundus by its lighter and more mottled colour; its peripheral limit is irregular with some pigmented spots. The halo exhibits a slight declivity, having a refraction of 1D. to 1.5D. higher than the adjacent fundus.

Vision with −1.5D. sph. 5/18, Jäger No. 1.
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Tension normal; corneal astigmatism 0.5D. cyl., according to the rule. Field of vision normal. It was not possible to trace the blind spot.

In the left eye the papilla is normal and very slightly smaller than in the right.

A sister of the patient has in the right eye a conus inferior with markedly less pigmentation of the lower part of the fundus. The mother has normal eyes.

In the present case the papilla simulated a glaucoma by the deep boiler-like cupping, the white colour, and the bending of the vessels at the margin. On the other hand the age of the patient, the normal tension, and the absence of other symptoms of glaucoma pleaded against this diagnosis. It was no doubt a case of congenital colobomatous excavation; although there was no enlargement of the papilla as in the preceding case, the congenital anomaly manifested itself here by the depressed level of the halo, as shown by its higher refraction. Also the irregular pigment spots at the margin of the halo would be very unusual in a case of glaucomatous halo.

Similar cases of pseudo-glaucomatous cupping are on record, the earliest being that of Stood\(^2\). In this case the excavation was not complete, some nervous tissue persisting on the nasal half of the disc, wherefore Stood did not insist on the simulation of glaucoma. Szily, sen.\(^3\) gives the diagram of the papilla of a case in which an operation was advised for glaucoma, but during four years of observation the findings did not change. The anomaly was the same in both eyes and proved to be congenital by the fact that the entrance of the central vessels and their branching at the floor of the papilla were invisible.

Rönne\(^4\) recently published a similar case, without increased tension, and observed for four years.

Nowadays the differentiation of congenital anomalies from glaucomatous excavation is easy, if in the fellow eye exact examination of the blind spot be possible. Notwithstanding, there will always be cases in which the diagnosis will at first remain doubtful, and to be made only after prolonged observation.

The ophthalmoscopic distinction between colobomatous and glaucomatous excavation is easy, if the fellow eye also presents congenital malformations, as have occurred in most of the reported cases. Very defective vision is also in favour of a congenital anomaly if it had been noticed by the patient for a very long time, especially since childhood.

The exact diagnosis is more difficult in some rare cases, as in mine, if the malformation is limited to only one eye, the fellow eye being normal. If then, as in my Case 2, the excavated papilla
is larger than that of the other eye, glaucoma may be excluded, since in this condition both papillae are of the same size.*

Another distinctive feature is the veiling of the vessels at the floor of the excavation, which I do not remember to have seen in glaucoma cases in which it is nearly always possible to focus exactly the vessels and determine their refraction. This veiling of the vessels has been recorded in several of the published cases and is probably due to the filling of the cup with semi-transparent neuroglia, compare the case of Roemer (left amaurotic eye), and of Zade.(6)

Decisive for the diagnosis may also be the aspect of the halo, present in my cases as well as in some of those recorded in the literature. In contradistinction to the glaucomatous the colobomatous halo is irregular and often pigmented. On the other hand the glaucomatous halo which, as a rule, is present only in very advanced cases of glaucoma, is narrower, whitish, and more uniform. The colour of the excavation is not decisive; it may be more red or on the contrary quite white, resembling in the latter case the colour of the glaucomatous cupping, but there are exceptional cases of incipient glaucoma, with a marked red colour of the papilla; in these cases, however, the cupping would be insignificant.

III. Angiectasia of the eyeball

The above recorded cases simulated glaucoma simplex; whilst the angiectasia of the eyeball resembles inflammatory glaucoma. Such cases are very rare, and, therefore, I beg to relate one observed by me.

A boy, aged 14 years, from Cairo, had his eyes injected for five years, and he thought that his vision was decreasing. After reading for some length of time the characters began to dance. He consulted a well-known French oculist, who was passing through Cairo, and who advised an operation on both eyes for glaucoma.

The boy does not open his eyes to the normal extent. The conjunctiva of the lids and the transitional fold is redder, exhibiting between the large vessels very numerous small ones with minute varicosities in places. Besides there are, as a sequela of old

*There exist eyes with papillae of unequal size. Such was a case published by Cords (Klin. Monatsbl. f. Augenheilk., T. LXXI, p. 414, 1922) where one papilla of a girl, aged 11 years, was 65 per cent. smaller than the other. The smaller disc was the malformed one and was of a whitish colour on the temporal side. The vessels appeared here on the lower edge.

I had once the opportunity to observe a case where one papilla was markedly larger than the other. The vision and the refraction were normal in each eye. The smaller disc was apparently the normal one, while the other was larger than normal.
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trachoma, very delicate cicatrices in the conjunctiva, both of the lids and of the transitional folds.

On the eyeball one sees very numerous vessels of unusual breadth and tortuosity, running from the limbus backwards. The dark colour of these vessels shows them to be veins.

In both eyes the cornea is bright, the anterior chamber deep, the pupils of medium size, and the iris blue—quite unusual in an Egyptian. The refracting media are transparent, the optic discs are unusually red with a conus inferior in each eye. The discs are sharply defined without an excavation. In the temporal half of the papilla of the left eye a minute macular vessel begins with numerous, very marked corkscrew-like windings, continuing a more rectilinear course in the fundus. The fundus of each eye is but poorly pigmented and everywhere the choroidal vessels, without any appreciable changes, are visible.

Vision R.E. 5/6 H.M. 1D. sph.; corneal astigmatism (ophthalmometer) 0.75D. cyl., according to the rule.
L.E. 5/18 with +2D. sph. +2D. cyl. 5/8? corneal astigmatism, 1.5D. cyl., according to the rule. Tension normal.

The boy's skin is poorly pigmented; he has brown hair and blue eyes like his father. In the fossa naso-labialis there are fine venous ectasias, whilst there is no change visible in the vessels in the region of the mouth and pharynx.

This case presents a particular condition of the vascular system of the eyeball, viz., a very marked ectasia and tortuosity of the anterior ciliary and the conjunctival vessels, similar to that seen in an eye, which has just had an attack of acute glaucoma, with the difference, however, that other symptoms are absent. During the next months I saw the boy repeatedly and always found the ocular tension low.

In a minor degree an analogous change existed also in the vascular system of the retina, a macular vessel being enlarged and tortuous within the papilla. On the other hand the posterior ciliary arteries were probably normal, as there were no changes visible in the choroidal vessels.

My father, to whom I showed the case, confirmed the diagnosis of vascular change in contradistinction to glaucoma. He remembered to have observed a similar case for many years. It was a woman, aged 43 years, who consulted him for the first time on January 5, 1887. Her right eye had divergent squint since infancy, with high myopia, circumpapillary atrophy of the choroid and central choroiditis. She counted fingers at 50 cm. and read Snellen 1 at a very short distance.

In the left eye the cornea and anterior chamber were normal the reaction of the pupil good, and tension normal. The optic disc, as expressly noted, was not cupped.
Vision 5/9, with -0.75D. sph. 5/6. Snellen 0.5.

In both eyes the anterior part was of a light violet colour because the anterior ciliary vessels, probably arteries as well as veins, were greatly dilated, as in a case of chronic glaucoma of long standing ("annulus arthriticus") so that in spite of the normal tension and absence of excavation a glaucoma was thought to be imminent and the use of pilocarpine was advised.

One year later the condition of the eyes was unchanged. It came out that the patient had had for seventeen years some enlarged glands in the neck, which increased in size every winter and diminished during summer. After prolonged use of iodine the glands had somewhat decreased in size. In May, 1887, two glands, a small and a larger one, were discovered at the anterior margin of the sterno-mastoid muscle.

The patient was seen at intervals during the following twenty years. At her last visit, 1907, the congestion of the vessels was just as striking as when seen for the first time. In the meantime the patient had developed cortical opacities of the lenses, so that the fundi could no more be seen satisfactorily, but there existed no symptoms of glaucoma whatever.

This case is analogous to mine, except for the age of the patient, being aged 14 years in my case and aged 43 years in my father's case in which the beginning of the vascular trouble had not been recorded. At any rate, this particular instance shows that this trouble may persist unchanged for many years.

In the literature I found but two similar cases, recorded in 1880, in a paper by Leber (6).

Case 3 (Leber).—The patient, a man, aged 35 years, stated he had noticed the dilatation of the vessels for six years. Besides opacities of the lens and of the vitreous body there existed enormously dilated "conjunctival veins," suggestive of the dilatation of the veins in the last stage of glaucoma or in an intra-ocular tumour. The capillary loops in the limbus were normal, but the ciliary veins were enormously dilated. Some of these were seen running as far as the transitional fold, whilst the majority disappeared in the recti muscles. The papilla appeared normal to Leber, whilst the oculist of New York, who had treated the patient previously had found the papilla covered by a dense network of fine vessels.

Case 4 (Leber).—Another patient from America, aged 19 years, had developed during the last years a circumscribed hyperaemia on the temporal side of the cornea, which two years later had become worse. In the temporal half of the conjunctiva the veins were greatly dilated and could be followed up to the transitional fold. In contradistinction to congestion associated with inflammation, as for instance scleritis, the dilatation concerned
mainly the large vessels, the capillary network being nearly normal.

In the above four cases the vessels of the anterior part of the eyeball were so highly dilated that the eyes presented the aspect of inflammatory glaucoma. In fact in my case this diagnosis had been made by a distinguished oculist, and my father suspected glaucoma for a while in his case. Leber published his cases on account of their unusual aspect and because, as he specially noted, of their resemblance to glaucoma.

In three of the cases the patients stated that the vascular trouble existed only for some years, so that it was not a congenital condition. The colour of the dilated vessels is bluish red, and I think, that in my case at least not only the veins, but also the arteries participated in the dilatation.

Leber held that the dilatation of the vessels was due to a chronic inflammation of the ciliary body, which had also caused the vitreous opacities in his first case. He furthermore admitted as an unusual fact, that the cyclitis must have come to a standstill, whilst the external dilatation of the vessels persisted, wherefore he supposed that there had been an independent alteration of the vessels.

I also believe that the vascular trouble in the above cases was independent of any intra-ocular inflammation, of which there existed no symptoms whatever in my own case and also in my father's, no such symptoms occurred during twenty years of observation. Furthermore, there existed in my case dilated vessels also in the region of the naso-labial fossa which could in no way be brought into relationship with an intra-ocular inflammation.

The condition seems to be a stationary one, as in my father's case it persisted unchanged for twenty years.

The clinical picture of the above cases is very rare and is important to recognize because of its resemblance to inflammatory glaucoma, and the possibility of an error of diagnosis, which, however, can be easily avoided by the use of the tonometer.

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