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COMMUNICATIONS

CAVERNOUS OPTIC ATROPHY AND ITS RELATION
TO GLAUCOMA*

BY

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EXETER

THE thesis I wish to maintain is that cavernous atrophy is a distinct entity; that its association with glaucoma is casual and not causal; it may exist without glaucoma, presenting symptoms somewhat resembling that disease; or it may exist with it, not causing it, though the two diseases will influence each other.

The cases to be dealt with are seven in number. It will be necessary to deal with them in some detail; it is proposed to do this mainly by charts and diagrams, which will make it easier to follow the points dealt with.

The following premises will enable the descriptions of the cases to be shortened:

- (1) None of the cases had any nerve symptoms apart from the condition of the optic nerves.
- (2) The discs were pale where the lamina cribrosa was exposed, not elsewhere. The vessels were not contracted.
- (3) The vision given is that obtained by using fully correcting glasses where necessary.
- (4) The tension was measured by the Schiötz tonometer. That weight was used which brought the indicating point between 2

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and 4 on the scale, except where the lowest weight (5.5 gms.) gave a higher figure than 4, *i.e.*, a lower tension than 20 mm.

(5) The disc cups are given in percentage of the whole area of the disc surface. The disc diagrams are constructed according to the method used by me in a previous paper (*Brit. Jl. of Ophthalm.*, p. 81, 1923).

(6) Under the conditions used, the white 3/1,000 and red 10/1,000 were nearly equal in a normal field.

(7) The red fields are shown with interrupted lines.

The cases now to be detailed bear a fairly close resemblance to glaucoma in the visual fields, loss of central vision, and cupping

of the discs; but they differ from it in having no rise of tension and in the colour fields conforming to the atrophic type.

Case A. H. T., aged 73 years, male. Onset eleven months ago, under observation three months. Both disc cups are 100 per cent. (*i.e.*, occupy the whole surface) and 4D. deep. Vision, R. 6/18(1), L. 6/24. Tension, R. 25, L. 18.

The main points in the right field are:

- (1) An annular scotoma, about 10° wide, starting from the blind-spot, sweeping above and round the fixation point through $14/24$ of a circle, breaking out above through the 3/1,000 boundary.
- (2) A marked limitation of the 1/1,000 field; and
- (3) A general contraction of the red 10/1,000 field.

The left field shows a concentric limitation, especially marked for white 5/300, 1/1,000, and red 10/1,000.

Except for their normal tension and small colour fields these eyes would easily pass as glaucomatous. Observations on four

dates gave tension varying; Right, 18 to 25; Left, 18 to 23. The tension shows there is no glaucoma, the colour fields show there is optic atrophy. For these reasons and the large cups this is called a case of cavernous optic atrophy.

Case B. Mrs. C.S., aged 63 years. This patient came for spasmodic entropion of the right lower lid, with no complaint as to her vision. The condition to be described was found in the ordinary routine examination.

The right disc cup is 43 per cent.; its depth 3D., the left 100 per

cent., depth 3D. There is an irregular ring of white atrophic choroid around the left disc.

Tension, R. 15 mm., L. 15 mm. Vision, R. 6/4.5(4), L. 6/6(3).

Both fields show great concentric limitation for white and red, with enlarged blind-spots; but this is more marked in the right, especially for red 5/300.

The special points in this case are that the affection of the field is less in the eye with the larger cup; and that the destruction of the colour field in the right eye is particularly great. From this it is deduced that the process is primarily a nerve condition, and that the destruction is greater in the eye in which the disc cup less resembles glaucoma. As it is fair to assume that the same pathological process is attacking both eyes, and the left has all the features of cavernous atrophy, it follows that the right also has cavernous atrophy. The difference in appearance of the two discs is quite in accordance with Schnabel's account of the pathology of cavernous atrophy.

Case C. Mrs. H. H., aged 79 years. Under observation two years. R.E. only affected. Very slight cortical striae in both lenses.

The right disc cup is 50 per cent.; a narrow ring of atrophied choroid round two-thirds of the disc. It has not altered in appearance during the two years. Depth, 1.5D. Corrected V.= 6/60, also unchanged. Tension, 15 mm.

The field for 3/1,000 is slightly and generally contracted, with a large blind-spot. The 2/1,000 field is much contracted, 1/1,000 cannot be seen. The red field, 10/1,000 is much contracted.

In this case the incidence of the disease is on the centre of the field for white. The large blind-spot and the colour field establish

the diagnosis of optic nerve affection, the large cup puts it among the cavernous atrophy group, though the cup is not deep. The absence of progression in two years suggests the possibility of the disease having come to its final stage. It will be seen that the tension is markedly low.

Case D. Mrs. L.H., aged 78 years. L.E. only affected. Under observation three years. Three minute dots in centre of lens. Disc cup 85 per cent., 2D. deep. Tension, 21. Corrected vision, 6/18(3).

The field for white 3/1,000 is generally contracted, about 10° being lost. That for white 1/1,000 is much contracted. The

blind-spot is large, the red field, 10/1,000 from 15° to 5° smaller than white 3/1,000.

It will be seen that this is similar to Case C., differing mainly in having a larger, deeper cup.

Case E. Miss F.B., aged 44 years. Under observation one year five months. A very slight increase in left disc cup, none in right. The annular scotoma spreading from the blind-spot has become larger, especially for 3/1,000. Both eyes affected. Tension has varied, R. 20—28, L. 23—28.

R.E. disc cup 36 per cent., 2D. deep. Tension, 25. Vision, 6/9(5). White field 10/300 slightly contracted, 3/1,000 full, 1/1,000 contracted and irregular. Blind-spot much enlarged to a half ring below, 8° wide. Red not recorded.

L.E. disc cup 83 per cent., 3D. deep. Tension, 27. Vision, 6/12(1). Field for 10/300 slightly contracted, 3/1,000 good,

1/1,000 irregularly contracted in outer and upper quadrants. Blind-spot slightly enlarged.

This case illustrates well the want of relationship between the size of the cup and the alteration in the blind-spot, the left eye with a very large deep cup has much less alteration than the right eye with a cup of quite average size. Like Cases C. and D. the incidence is on the blind-spot and the centre of the field in both eyes.

It must be pointed out that the tension in both eyes is near the margin of safety; opinions might well differ as to which side it is on. But the red field 10/1,000, taken January 1, 1923, but inserted in the chart of March 19, 1924, shows conclusively that there is

optic atrophy, while the 83 per cent. disc cup of the left eye entitles the case to be included in the cavernous atrophy series.

(The red field for Case E. had been tested on July 22, 1924. In both eyes it is very small; in the left smaller than that shown in the field exhibited, in the right conforming very nearly to the 10° circle, while the field for white, 3/1,000 is practically unaltered; which fully confirms the position of this case as one of optic atrophy.)

Case F. Mdlle. B., aged 48 years. Right only affected. Treated as glaucoma with pilocarpin by a distinguished Continental oculist.

Disc cup 60 per cent., 2D. deep. Tension 21 mm. Vision 6/5(6). Field good except for flattening above for 1/1,000 white, and an enlarged blind-spot.

This is included as an example of a slight case.

Case of Chronic Glaucoma

This is described to show the close resemblance to the cases of cavernous atrophy and to illustrate the differences. It will be seen that the larger cup corresponds with the worse central vision and field, but that the better eye is distinctly affected.

Case G. Mrs. A.M., aged 63 years. Recent gradual but indefinite onset. Disc cups; R. 100 per cent., 2D. deep with ring of white atrophy around disc with a crescent of tessellated choroidal pigment on the outer side; L. 26 per cent., 1.5D. deep. Tension, R. 30, L. 30. Vision, R. 6/18(2), L. 6/5(5).

The right white field shows a large annular scotoma, starting from the blind-spot, for 3/1,000, having a very indented border near the fixation point, a greatly contracted field for 1/1,000.

The left white field for 3/1,000 is full except a very localized loss on the nasal side, like a bite out of a slice of bread; that for 1/1,000 is contracted and irregular; the blind-spot is enlarged.

The red field 10/1,000 follows the 3/1,000 remarkably closely, except in the vertical meridian above the right fixation point, where two portions are separated from the main red field.

That this is a case of glaucoma will be granted by most, because of the tension and the general agreement of the white and red colour fields, but there is a great resemblance in the white field to Cases A. and E.

The want of complete coincidence of the red field with the white suggests that there may be a slight atrophic element.

Relative Glaucoma

Case H. Mrs. L., aged 77 years. R.E. disc cup 100 per cent., 2D. deep. Tension 21. Vision 6/6(4). The field shows general contraction for 3/1,000 and 1/1,000; 2/1,000 shows much loss on the nasal side. The blind-spot is enlarged for 3/1,000, and it is prolonged as a relative scotoma for 2/1,000 in an annular direction. The red field for 10/1,000 conforms fairly closely to white 3/1,000, but with a larger blind-spot than the latter.

L.E. Disc cup 72 per cent., 2D. deep. Tension 21. Vision, 6/9(3). The field is generally contracted for 3/1,000 and 1/1,000,

the blind-spot is enlarged. The red field for 10/1,000 is a little smaller than white 3/1,000, but generally conforms to it.

The special features of this case are the low tension and the close resemblance in other respects to glaucoma. But the red field is on the small side, especially in parts. A feasible explanation is that it is a mixed case, in which the incidence of the atrophy is mainly on the supporting tissues of the nerve head rather than on the nerve fibres. This would produce a condition of relative glaucoma; the nerve fibres, unprotected by the connective tissue, would suffer as they do from increased tension, the colour field keeping pace with the white field. But it is not a pure case of relative glaucoma, for the correspondence of the fields is not accurate, especially at the right blind-spot.

Case H. has been seen by Mr. W. Lang from 1904 to 1920, who has kindly given me an abstract of his notes. Vision in 1910 was 5/5 in each eye; tension was full. The field in 1910 for white 20/300 was full for both eyes. In both eyes there was a scotoma above the fixation spot, in the right between 5° and 15° vertically,

from 45° to 135° laterally, in the left from 3° to 8° vertically, 90° to 135° laterally. A comparison with the charts given in this paper will show that the scotoma in the right field has fused with the blind spot; there was no scotoma for 3/1000 in the left eye when examined by me. As 2/1000 was not charted for that eye, a scotoma for that cannot be excluded.

A comparison between these details and those given by me will show that there has been very little progress indeed in the past 14 years, a point of great interest in estimating the prognosis in such cases.

That a physical condition is being dealt with in the cases A. to F. and in H. is shown by the condition of the disc cups. That there is an affection of the optic nerve fibres is shown by the fields. In cases A. to F. this is accompanied by a disproportionate loss of the red fields, as in ordinary optic atrophy. The question at once arises whether these cases are a clinical entity or ordinary primary or secondary atrophy happening to show unusually large deep cups; also whether these are members of the group of optic atrophies which, according to some, would develop general nerve symptoms if the patients lived long enough. My concern is to point out the differences from ordinary atrophy, the ultimate destination of the latter group not being the question at issue in this paper. The differences are: (1) The absence of the dead white colour; (2) the vessels slightly or not at all contracted; (3) the large deep disc cups. The pallor was no greater than is shown by anything which causes the lamina cribrosa to be exposed. The appearance in these cases may be summed up as "typically glaucomatous" in every way. A comparison of a group of all the cases of optic atrophy of known cause observed during the same period will establish the marked difference between disc cups in the two groups.

OPTIC ATROPHY OF KNOWN CAUSE.		CASES OF CAVERNOUS ATROPHY.		
Percentage size of cup.	Depth in Dioptres.	Percentage size of cup.	Depth in Dioptres.	Case letter.
60	Not recorded	100	4	A
54	1.5	100	4	A
32	2	43	3	B
31	2	100	3	B
23	1.5	50	1.5	C
22	1	85	2	D
18	1.5	36	2	E
12	1	83	3	E
12	1	60	2	F
Average		Average		
29	1.5	73	3	

It must finally be insisted on that none of these cases showed any symptoms of nerve disease. To those who would regard them as

cases of optic nerve atrophy, which will later show signs of general nerve lesions, it must be pointed out that this involves the assumptions: (a) That the general signs will develop; (b) that the appearance of the optic disc is different in this group of delayed general nerve lesions from that in which these are developed. With (a) I have no concern except to point out that an appeal to the unknown future is no argument, but (b) stands on a different footing. The appearance of the atrophied optic disc in cases, such as locomotor ataxy, where the atrophy is a precursor of the general signs, or the latter are present in slight degree, differs in no way from the atrophied discs seen in those cases which show the full symptoms of such diseases. It is therefore maintained that the cases A. to F. and H. are essentially different from ordinary simple optic atrophy.

The symptoms which separate this group from chronic glaucoma have now to be considered. The appearance of the disc cups and the fields for white would agree completely with chronic glaucoma; the essential differences are the colour fields and the tension. It is generally accepted that, in a typical case of glaucoma, such as G. in this paper, the colour field is lost in nearly the same proportion as the white field. The meaning of this is that the conduction of the nerve fibres for all light sensations, white or coloured, is equally destroyed by pressure. This is generally accepted, is in accordance with my own experience, and is assumed in this paper. On this assumption there can be no doubt that the cases of the present group differ from glaucoma.

As a further though minor proof that the cases are different from glaucoma, cases B. and E. may be cited. In the right eye of each case, with the smaller disc cup, the alterations in the white field and blind spot are greater than in the left eye with the larger disc cup. This is distinctly contrary to the rule in glaucoma, where the larger cup is almost invariably associated with the greater effect on the field. I have never seen an exception to this rule.

The tension has next to be considered. In no case was over 28mm. (Schiötz) found; in most of the cases it was well below this amount. Cridland has put down 30mm. as being approximately the beginning of abnormally high tension, and most would agree with this standard. There can be no doubt according to this that the cases described have no increase of tension. Now the pressure exerted by the vitreous upon the optic nerve head is that exerted by a viscous fluid upon a more or less solid substance. If the pressure of the fluid remains constant and normal any alteration in the position of the surface of contact of the fluid and solid must be considered as essentially due to alterations in the latter. If the solid surface recedes, as in the present cases, it is because of lessened resistance in the nerve head. To speak of glaucoma in such cases is a misuse of terms, for glaucoma is a condition of

increased pressure, not of diminished resistance in the nerve head. Moreover the use of a wrong term in this relation will prejudice one's attitude towards treatment.

It is quite true that certain cases may be regarded as relative glaucoma, if only the real facts are not lost sight of. Such is case H., where with low tension the red field fairly agrees with that for white. In the published accounts of the pathology of cavernous atrophy it will be noticed that the incidence of the disease on the different tissues of the optic nerve varies. As already suggested, if the cavernous condition affected only the supporting tissue, the unprotected nerves would suffer from pressure and the red field would keep pace with the white field. As far as my experience goes, this relative glaucoma is exceptional in cavernous atrophy; the rule is that the colour field is proportionately less than that for white.

It may be argued that these are cases in which the pressure varies much at different times, that the tension was raised during the times in which no observations were made. It must be admitted that all the tensions were taken between 10 a.m. and 5 p.m. But the colour fields dispose of this contention. Fluid pressure can only act as fluid pressure; how could it cause colour fields of the atrophic type when acting at one time of the day and not when acting during the ordinary hours of observation? It is not denied that intraocular pressure varies at different times; what is denied is that its results are of a different kind.

For these reasons it is asserted that this group of cases constitutes a distinct entity, with differences which separate them from ordinary optic atrophy on one hand and glaucoma on the other. The name cavernous atrophy is given because they conform clinically to the account of that disease as described by Schnabel, saving that there is no glaucoma present.

Association with Glaucoma

It remains to be considered what association is found between this condition and glaucoma. It is obvious that, should they occur together, the combined effect upon the common symptoms of each would be much intensified; the large deep disc cup, the affection of the visual field in general, of groups of nerve fibres in particular, and the enlargement of the blind spot, would all tend to be very marked. To disentangle the symptoms would be impossible; it would only be by weighing against one another the main distinguishing symptoms, the tension and the colour fields, that an estimate could be formed.

The cases of cavernous atrophy detailed in this paper are obviously of different degrees of severity as they stand at present. It is therefore quite possible that this condition may occur in conjunction

with glaucoma in greater or less amount, that there may be different degrees of cavernous atrophy in conjunction with glaucoma. This assumption would explain some cases in which, after a successful operation with consequent lowered tension, the vision fails, and particularly the visual field breaks down. The colour field might be of use in such cases to give an idea of the amount of atrophy present as compared with the destruction by pressure, and so aid in a more accurate prognosis.

The symptoms dealt with in this paper may be summed up as consisting of four variables:—

The size of the optic disc cup.

Tension.

The field for white.

The field for red.

It is the alteration in the disc cup that usually calls attention in the routine examination. Moreover, it is an essential sign, for without it in one or both discs contraction of the optic nerve tissue cannot be inferred.

It will be common ground that different eyes react very differently to alterations of tension, so that one eye may easily withstand a tension that in another will produce glaucoma. Yet as a general rule tensions of 30 mm. and upwards are accompanied by alterations characteristic of glaucoma. With a tension under 30 mm. there must be other good reasons why the case is to be considered as glaucoma, the lower the tension the less likelihood is there that true glaucoma exists.

The field of vision for white is only characteristic for glaucoma because this is the commonest cause for such fields. Affections of the nerve are well recognized as causing similar fields. The present series shows very well that, while certain general characteristics are present, there may be variation in detail.

The final test in cases having tension below 30 mm. after a due consideration of the field for white, must be the colour field. It will have been seen that the cases A. to F. show a field generally recognized to be characteristic of optic atrophy. Case H., on the other hand, does not differ greatly from the colour field of glaucoma, but its low tension and long duration entitles one to assume that it is not glaucoma but a variety of cavernous optic atrophy. In glaucoma the effect upon the nerve fibres is that of a total or partial destruction of conductivity to all light sensations in equal degree. In optic atrophy it is, for some unexplained reason, of unequal degree, but it is not unreasonable to suppose that in the form of optic atrophy now considered it will occasionally happen that the lowering of conductivity should be, in the main, equal for all colours, which will explain the exceptional case H.

Prognosis

Several of these cases have been under observation for some time ; 2 years, 3 years, $1\frac{1}{2}$ years and, in the case of H., 14 years. These are long periods for so little progress to have taken place. It raises a presumption that, in at least a proportion of such cases without increase of tension the outlook is fair ; that the disease arrives at a moderate degree of severity and afterwards makes very little progress. In making this statement I am quite aware of the long intervals of apparent quiescence that may occur in other forms of optic atrophy, but I believe that this generalization is justifiable. Finally I should again like to emphasize the importance of this class of case in that borderland of difficulty which separates the normal eye from the glaucomatous, and the probable usefulness of the colour field in distinguishing an atrophic element in cases of glaucoma.

Addendum

It has been suggested that the cases dealt with in this paper are cases of glaucoma which are either early, intermittent, or in which the pressure has passed away, and that the condition found may be explained under one of these three heads. The question of intermittent tension has already been dealt with. The argument used there applies to all three of the above suppositions, that pressure wherever it is applied should produce the same results, it being generally acknowledged that the colour fields are lost in the same proportion as the white fields. The fields of the atrophic type in the present cases cannot be explained by pressure, however or whenever applied. The suggestion that the pressure has entirely passed away requires further reasoning. Conceivably it might be the case that when pressure is relieved changes could go on in the retinal nerve elements which would lead to an atrophic type of field. Assuming that the field to begin with is not atrophic, and that the relief of pressure continues, the atrophic type could only be reached by the white field enlarging, but not the colour field ; or by the colour field contracting. To be quite frank, I know of no case of completely subsided glaucoma which has been dealt with from this point of view. It is obvious that material from operated cases will not suffice. What is required is as follows:—A case of definitely increased pressure existing for some time, with fields in which the white and red fields are equally affected, with large disc cups, where the pressure subsides without operation. Should the fields then alter to the atrophic type, whether by enlargement of the white field only or by diminution of the red field only, it would prove the possibility of cases such as those detailed in this paper arising from a subsided glaucoma. Until

these conditions are fulfilled, it is justifiable to assume that diminution of pressure, where the tissues are capable of repair, merely reverses the action of increased pressure, so that the white and colour fields increase *pari passu*. Test conditions such as are stated above will not often occur. Really chronic glaucomas which recover without operation to an extent that justifies stopping treatment are rare; few surgeons would risk leaving cases for long without operation on the chance of recovery—though some still leave them because they despair of doing good by operation. Thus the case wanted for proof would be hard to come by.

The cases recorded in this paper are easily explained by assuming an alteration in the optic nerve tissues, they are only explained as glaucomatous by granting two undemonstrated suppositions—increased pressure at an unknown time and an atrophic field as the result of this pressure or its disappearance. It is more logical to take the simpler explanation. Moreover, in the right eye of case E the field is distinctly that of a retrobulbar affection. If the left disc were not cupped, nobody would think of diagnosing the right as a glaucomatous condition; nothing but a diagnosis of retrobulbar neuritis would fit it. The assumption that this inflammatory process can go on to contraction, for which Schnabel has given pathological evidence, completes the reason for assuming that the left eye has the same affection as the right, a conclusion which is supported by the doctrine of the paucity of causes. The other cases can be claimed by a similarity of reasoning not to be glaucomatous.

DERANGEMENTS OF THE ORGANO-VEGETATIVE NERVOUS SYSTEM AND OF THE ENDOCRINIAN SYSTEM IN ESSENTIAL GLAUCOMA

BY

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THE peculiar form of "attacks" generally assumed by essential glaucoma, and particularly obvious in its acute form, and also the intermittency of the attacks noticed in the chronic form, have led many authors to the "neuropathic" conception of glaucoma, and several of them do not even hesitate to call it a diathesis.⁽¹⁾ This makes it necessary to separate, in the hypertension syndrome, the pure glaucoma from the accidents of the same kind, the origin of which is infectious or neoplastic.

As "essential asthma" had to be separated from asthmatic dyspnoea⁽²⁾, so must glaucoma be isolated from the disturbances which succeed in copying it, if its description is to be of any