Section of Ophthalmology

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Optic Nerve Compression and its Differential Diagnosis

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OPTIC nerve compression is of interest to the ophthalmologist, neurologist, and neurosurgeon. Like many such problems it lies on the borderland between ophthalmology and neurology. Nor is the subject one of clinical interest only, for we are by no means certain of the cause of the pallor of the optic disc which ensues after prolonged intracranial compression of the optic nerve. It is said to be due to disappearance of the capillaries in the optic disc, and may be of vascular origin consequent on compression of the nerve. The term optic atrophy infers degeneration of the optic nerve, and yet we use it as a clinical term to indicate the appearance of the optic disc, when we should perhaps speak of optic disc pallor.

I wish to refer to the clinical aspects of optic nerve compression, and to a group of cases in which unilateral compression occurs as the initial manifestation. The literature on this subject is scanty. Cushing (1938), in describing meningiomas of the inner portion of the sphenoidal ridge, refers to the unilateral failure of vision in association with a "primary" optic atrophy, which may occur, and progress to near blindness, before the other eye becomes involved. McKendree and Doshay (1936) describe 6 cases of optic nerve compression, 3 meningiomas and 3 aneurysms, in several of which the compression was predominantly unilateral. Schlezinger, Alpers and Weiss (1946) describe 4 cases of suprasellar meningioma in which the initial compression was of the optic nerves, instead of the more usual chiasmal involvement. The authors refer to the expanding scotomatous or sector defect in the visual fields which is so typical of optic nerve compression, though in none of their cases was the compression predominantly unilateral. Foster Moore (1925) describes a mycotic aneurysm of the ophthalmic artery which presented with monocular blindness of sudden onset. Jefferson (1937) refers to compression of the optic nerves by aneurysms, but from his recorded cases it appears that aneurysms rarely present with unilateral optic nerve compression alone. Other structures, such as the optic chiasm or tract, or the oculomotor nerves, are usually involved as well.

When pressure is exerted initially on the optic chiasm, as in cases of pituitary adenoma or suprasellar meningioma, the early bitemporal character of the field defect usually gives a clue to the localization of the lesion, although there may be later evidence of optic nerve compression as well. When the initial compression involves the optic nerves, however, as in anteriorly situated basal meningiomas, bilateral visual failure and optic atrophy

may be the only presenting clinical features, and diagnosis may be by no means easy, especially when other signs are absent. Anosmia would, of course, assist in the diagnosis of an olfactory groove meningioma, and the presence of unilateral exophthalmos would indicate a space-occupying lesion, and occurs, for instance, in cases of optic nerve tumours and meningiomas involving the sphenoidal ridge. The presence of unilateral visual failure with a pale disc, in association with papillœdema in the other eye, is also a well-established clinical syndrome in frontal tumours (Foster Kennedy syndrome). I mention these well-known accompaniments of optic nerve compression, as they were little in evidence in the present series of cases.

The group of cases I propose to discuss consists of those in which optic nerve compression occurs as the early and presenting feature, and in which the compression is predominantly or entirely unilateral. From the clinical aspect it will be realized that these cases offer considerable difficulty in diagnosis, and it is this aspect to which I wish to refer in particular.

The 11 cases on which this communication is based consist of 7 basal meningiomas, 3 aneurysms or vascular anomalies, and 1 glioma of the optic nerves and chiasm. It is, of course, not unknown for a pituitary adenoma to present with unilateral optic nerve compression, instead of the usual chiasmal features; for a nasopharyngeal neoplasm on rare occasions to appear with a similar clinical picture, instead of the more usual oculomotor and trigeminal involvement; and for an arteriosclerotic and tortuous internal carotid or ophthalmic artery to mimic a tumour or aneurysm by compressing the optic nerve. How often this last occurs is conjectural, for if such a condition is suspected, intracranial exploration is usually avoided, especially in the older age-group. I have seen a tortuous carotid artery result in compression of the optic tract, and mimic a tumour to a nicety, and it may well be that optic nerve compression by a pathological artery, without aneurysmal formation, is more frequent than we realize. Sunderland (1948) refers to this subject when discussing neurovascular relations at the base of the brain, and shows a photograph of a tortuous and atheromatous carotid artery compressing one optic nerve. I have avoided mentioning cases of intracranial tumour in which optic nerve compression occurs as a late phenomenon, as such cases present less difficulty from a diagnostic aspect.

It is of interest that all the cases reported here are basal meningiomas, or aneurysms, with one glioma of the chiasm and optic nerves. This is perhaps to be expected, since basal meningiomas and aneurysms are often so placed as to be likely to come in contact with the optic nerve early in their development. An inferior frontal or temporal glioma can compress or invade the optic nerves, and this subject has been fully discussed by Jefferson (1945) in his Doyne lecture. Involvement of one optic nerve, however, is probably rare as an initial or presenting feature in gliomas. Jefferson (1945) states that the Foster Kennedy syndrome, other things being equal, is itself evidence in favour of a meningioma, as opposed to a glioma, though not exclusively so, but that when one disc is atrophic and the other normal, the probabilities of meningioma are greatly increased.

Clinical Features

The first effect of optic nerve compression is unilateral visual failure. This is usually of gradual onset, but is occasionally more sudden, particularly in aneurysmal compression. Further, an early peripheral loss of field may pass unnoticed, and the patient may state that the loss of vision was sudden when the macular fibres become affected later on. I mentioned this apparent abrupt onset which occasionally occurs, as it may lead to a diagnosis of retrobulbar neuritis, as will be described later.

The macular fibres form a large proportion of the optic nerve, and appear to be particularly vulnerable in optic nerve compression. The visual field defect in early cases may consist of a central scotoma, but by the time the patient first consults his medical adviser there is often a gross *central scotomatous defect breaking through to the periphery at one point;* so that all that may remain of the visual field is a peripheral crescentic area of relatively intact vision. This was of particularly common occurrence in the present series of cases (figs. 1, 4, 5, 7, 11). Later, of course, the eye becomes blind, and the patient then presents himself with monocular blindness and optic atrophy.

Traquair (1946) states that central vision is nearly always reduced in cases of optic nerve compression, and that a central scotoma is common. He also refers to the frequency of a wide sector depression of the field, of irregular quadrantic or hemianopic shape.

In a recent paper (published after the present communication was given) Mooney and McConnell (1949) state that a central scotoma is particularly liable to occur when the optic nerve is elevated or depressed in the region of the optic foramen, and refer to the possible mechanisms, such as pressure of the nerve against the margin of the optic foramen, or pressure on, or kinking of, the ophthalmic artery. Case IV in their paper is similar to the cases in the present series, in presenting with unilateral central scotoma and optic atrophy, due to a probable aneurysm lifting up the optic nerve. They also refer to the rarity of a central scotoma in cases of pituitary adenoma, though the latter frequently lies between the optic nerves and presumably exerts pressure on these structures, as well as on the chiasm.

Visual hallucinations in the affected eye were present in one patient at the onset (Case X), in association with fairly rapid unilateral visual failure. This patient, an elderly woman, noticed "sudden lights in front of the right eye, like blue stars with a red rim". Some hours later a grey mist appeared in the temporal field of the right eye, and within a short time the eye was almost blind, but a small area of vision remained in the peripheral field above. The visual hallucinations lasted a day or so. Some months previously she had noticed a sudden beating noise in her head, which persisted, being more marked on exertion. The aneurysmal or vascular nature of the optic nerve compression seemed almost certain, as a loud systolic bruit was heard on auscultation of the right eye and head, which was abolished by pressure on the right common carotid artery in the neck.

The *optic fundus* may show no abnormality if the patient is seen early. Later, it is usual to find a progressive pallor of the disc, though it may be several months before this is evident. Papillœdema was observed in one patient as a temporary phenomenon early in the course of the compression (Case X). At this stage the patient was almost blind in the eye, though with a localized sparing of a part of the peripheral field, thus mimicking a case of retrobulbar neuritis very closely, were it not that an ocular bruit was heard with a stethoscope. What appears to be a very rare mode of presentation—and I have not seen it described in the literature—is that of a central retinal venous thrombosis. This was the first clinical evidence of optic nerve compression in one patient (Case XI), and was followed by thrombotic glaucoma and enucleation of the eye. The patient was referred to me with a temporal hemianopia in the remaining eye, and proved to have a glioma involving both optic nerves and chiasm.

In addition to involvement of the vision of the ipsilateral eye, pressure on the posterior or chiasmal extremity of the optic nerve may involve the *crossed nasal fibres from the other eye,* which pass forwards into the posterior extremity of the optic nerve before passing back into the tract. From clinical observations

it seems likely that the lower crossed nasal fibres are particularly prone to be affected by pressure at this situation (Diagram A), with consequent defect in the periphery of the opposite upper temporal field. In one of the present series of cases, the appearance of this upper temporal defect in the other eye was the first almost certain proof that a unilateral optic atrophy was due to pressure, and exploration revealed a large meningioma arising from the tuberculum sellæ (Case III, fig. 6).

Further, a *partial crossed homonymous defect* in the contralateral fields, which may be confined to the upper or lower quadrants, can occur in association with a severe central loss of vision in the ipsilateral eye, and is probably suggestive of aneurysm. This feature was present in two of this series of cases (figs. 13, 15), both probably aneurysms, and has been mentioned by Jefferson (1937). This clinical picture infers that the aneurysm is large enough to affect both optic nerve and optic tract on one side.

I have described the effect of optic nerve compression on the vision, fields, and optic fundus, and I would now like to refer to the question of pain. In several cases of proved meningioma with unilateral optic nerve compression, the ***absence of pain and headache*** has been a striking feature (Cases I and III). In Case I, a male aged 27, unilateral visual loss of fairly sudden onset had appeared ten years

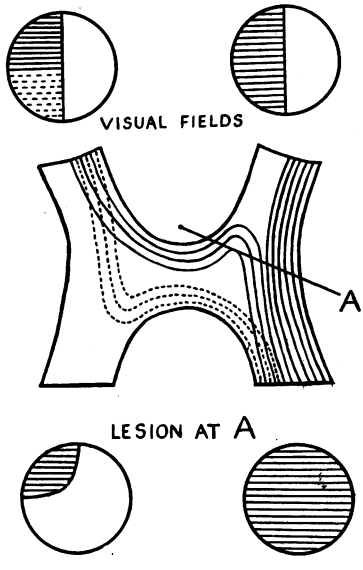

DIAGRAM A.—To show probable course of crossed nasal fibres in optic chiasm, and field defect due to a lesion at A, as in Case III. (Continuous line represents lower nasal fibres, and interrupted line represents upper nasal fibres.)

previously at the age of 17, and a central scotomatous defect had persisted over this period, without any pain. The original diagnosis had been that of acute retrobulbar neuritis. After ten years, headaches and vomiting appeared fairly suddenly, and exploration revealed a large meningioma in the suprasellar region. In Case III, a male aged 49, there had been progressive failure of the vision of the right eye over a period of no less than five years, with no history of headache or pain. The difficulty in diagnosis in this case is, perhaps, emphasized by the fact that the following diagnoses had been suggested at different times during the five years: chronic glaucoma, chiasmal arachnoiditis, an unusual retrobulbar neuritis, hæmorrhage into the optic nerve, and pituitary tumour. X-rays of the skull and optic foramina were normal. The appearance of a slight defect in the upper temporal field of the normal eye five years after the onset of symptoms gave a clue to the diagnosis, and exploration revealed a large suprasellar menin-

gioma arising from the tuberculum sellæ. The absence of pain needs emphasis, perhaps, as it may direct one's thoughts away from a tumour as the cause of monocular blindness. On the other hand, cases of optic nerve compression due to aneurysm often have a history of severe pain, sometimes early in the history and temporary only, and the significance of this early episode may be missed when the patient appears later with a painless blind eye and optic atrophy (Case VIII). Yet even in aneurysmal compression of the optic nerve pain may be entirely absent.

A systolic cephalic bruit, best heard by auscultation over the eye, is occasionally heard in aneurysmal cases; it may be diminished or abolished by compression of the ipsilateral carotid artery in the neck (Case X). The absence of a bruit does not exclude aneurysm. It is more commonly absent than present.

The clinical course of many of these cases is of interest. It will be seen from the case histories that the length of history during which the only sign is unilateral loss of vision may extend to a period of several years, particularly in cases of meningioma (ten years in Case I, and five years in Case III). This fact, perhaps, needs emphasis, especially as the early stages may be entirely painless. In some cases, too, the clinical state may appear to be static over a prolonged period, and can be misleading (Case I). The appearance of a slight defect in the peripheral temporal field of the sound eye is of great diagnostic importance, and is an almost certain proof of compression as the cause of the unilateral visual failure (Case III).

In aneurysmal compression there may be an improvement in the vision, quite apart from the effect of surgical treatment. In one patient (Case VIII) who had an aneurysm pressing on one optic nerve, there was a central scotoma, and vision was reduced to "counts fingers" in this eye. About a year later, without any radical treatment, though there had been an intracranial exploration, the vision had improved to 6/9 and the central scotoma had disappeared.

Accessory Methods of Investigation

These include radiographic examination of the skull and optic foramina, examination of the cerebrospinal fluid, arteriography and ventriculography. Radiography of the skull may show evidence of hyperostosis, as in meningiomas arising from the sphenoidal ridge, tuberculum sellæ or olfactory groove, or there may be evidence of localized bone erosion, and even calcification within the meningioma. An aneurysm may show evidence of calcification in its wall. Enlargement of the optic foramen occurs when an optic nerve tumour, glioma or meningioma, has an intra-orbital and an intracranial portion. Arteriography by the percutaneous method is indicated when an aneurysm is suspected, and is a relatively safe procedure. It is, however, of considerable use in tumour cases, as the arteries may be displaced by a tumour, or the meningioma itself may take up the radiopaque substance, outlining itself clearly by the so-called "blush" (Case VI, fig. 10). Ventriculography is usually indicated in tumour cases, when displacement or deformation of the anterior horns or of the third ventricle may be seen in cases of large basal meningiomas.

One would expect that further investigation on these lines would clear up any doubts as to the diagnosis. But this is by no means always the case, especially when investigations are undertaken early in the course of the condition. Ventriculography may be normal when a tumour is small enough not to deform the ventricles, and, of course, it is in precisely such a case that one would expect operative results to be best (Case V). Arteriography may also be quite negative in the case of an undoubted aneurysm, either because of clotting in the wall of the aneurysm, or when the aneurysm is not in direct continuity with one of the main

vessels (Case VIII). The corollary seems to be that if optic nerve compression is suspected and all investigations are negative, then intracranial exploration of the optic nerve is justifiable. This happened in 3 of the present series of cases. In one case a small meningioma was found arising from the anterior clinoid process, and was completely removed, with recovery of vision from hand movements to 6/9 within three weeks (Case V). In another (Case IV), there was a plaque meningioma surrounding one optic nerve. A small aneurysm was found in a third case, which did not show on arteriography (Case IX).

Differential Diagnoses

The most likely condition which is apt to be confused with unilateral optic nerve compression is unilateral retrobulbar neuritis. In this condition the onset is usually abrupt, a central or paracentral scotoma is present, and the fundus may be normal or the disc slightly swollen. Pain is frequently present, and tenderness of the globe common. Recovery of vision during the following few weeks is the rule. I should recall, however, that pain is sometimes absent, and that occasionally there may be little or no recovery in the vision, so that the patient is left with a permanent defect in the central field, usually associated with a pale disc. It is this type of case in which diagnosis from optic nerve compression may be one of extreme difficulty. One must remember, too, that optic nerve compression by an aneurysm may be painful and abrupt, and even associated with swelling of the optic disc in the early stage (Case X), and the defect in vision may even improve (Case VIII), thus closely mimicking retrobulbar neuritis. The point of practical importance is to remember the possibility of optic nerve compression in the case of a supposed retrobulbar neuritis of unusual type, where the vision does not clear. This difficulty does not often arise, but, as will be realized, early operation in optic nerve compression may lead to complete recovery of vision (Case V), whereas if we leave a doubtful case and observe masterly inactivity, it may prove almost impossible, at a later date, to remove a large meningioma (Case I). In 2 of the present series, both cases of meningioma, the early presence of a central scotoma of fairly abrupt onset, with optic atrophy, led to a provisional diagnosis of retrobulbar neuritis. In one of these patients (Case II), further observation showed that the scotoma was gradually extending to the periphery, and exploration was undertaken. In the other patient (Case I), a youth of 17, the vision and scotomatous defect appeared to have changed little, if at all, over a period of ten years, when headaches and vomiting appeared, by which time the tumour was very large indeed.

The only other conditions likely to be confused with optic nerve compression are vascular lesions of the retina and optic nerve, and chronic glaucoma. The vascular groups include narrowing of the retinal arterial tree, with sudden occlusion or more gradual ischæmia, in addition to vascular lesions of the nerve, about which our knowledge is scanty. As mentioned previously, a tortuous and sclerotic carotid artery may also involve the optic nerve. It would be unwise for a physician to refer to glaucoma, except to state that such a condition has been known to appear in the neurological or neurosurgical wards as a suspected intracranial tumour. In one recent case, not included in this series, which I suspected of having a meningioma or aneurysm compressing the optic nerve, surgical exploration revealed evidence of arachnoiditis of the chiasm and optic nerve. We know little about this condition, and unless it has been proved by inspection, such a diagnosis should be viewed with suspicion.

In conclusion I would emphasize that unilateral visual failure with a central scotomatous or sector defect may be the presenting feature of a basal meningioma or aneurysm, that the onset may be gradual or fairly sudden, and that there may

be a complete absence of headache and pain. The history may go back many years without any abnormality other than unilateral blindness and optic atrophy, and negative investigations should not deter us from considering intracranial exploration.

CASE RECORDS

CASE 1.—*Unilateral visual failure and optic atrophy of ten years' duration caused by meningioma arising from diaphragma sellæ.*

W. C., male, aged 27. Early in 1938 he noticed a fairly rapid loss of vision of the left eye, especially in the centre of the field. He had no headaches or pain. Examination seven months after the onset revealed left optic atrophy, with vision reduced to 6/60. There was a central scotoma with full peripheral fields. The left pupil reacted fairly briskly to light. The nervous system appeared otherwise normal.

The condition appeared to remain almost stationary till 1948, i.e. for ten years, during which period his general health was excellent. Even in 1944, six years after the onset, visual acuity in the left eye was still 3/60, with a left central scotoma. In August 1948 the patient developed generalized headache and vomiting, occurring several times weekly. Examination in October 1948 showed primary optic atrophy in the left eye and slight pallor of the right optic disc. In the left eye vision was reduced to hand movements in the temporal field only, the right visual acuity being 6/6. The left visual field was reduced to a crescentic area in the periphery of the temporal field, while there was a slight defect in the periphery of the right upper temporal field (fig. 1). The left pupil reacted poorly to direct light, but well consensually. The nervous system was otherwise normal.

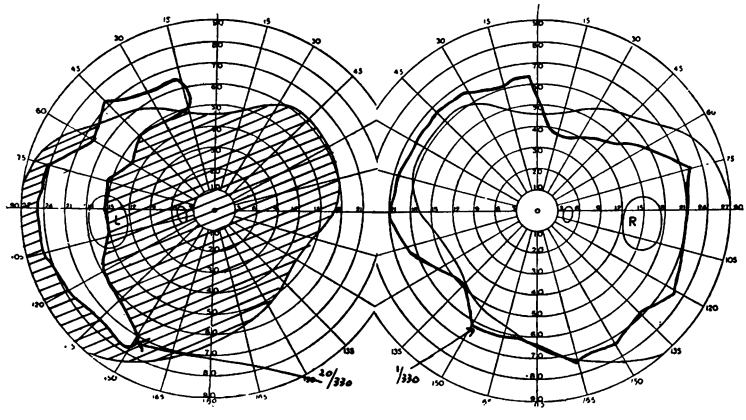

FIG. 1—Visual fields of Case I, ten years after onset of unilateral visual failure, showing early defect in upper temporal field of contralateral eye.

Examination of the cerebrospinal fluid in 1948 showed a slightly raised pressure (170 mm.), a total protein of 110 mg.% and a positive globulin test, but the cell count was normal and the Wassermann reaction negative. Radiography of the skull revealed destruction of the dorsum sellæ and posterior clinoid processes, with normal anterior clinoid processes and optic foramina. Ventriculography revealed gross symmetrical hydrocephalus of the lateral ventricles, with lack of filling of the third ventricle.

On November 19, 1948, operation by Mr. Valentine Logue revealed a large hard tumour extending forwards and laterally from the region of the sella turcica, and so distorting the left optic nerve and chiasm that they could not be identified. The site of the origin of the tumour could not be identified for certain, but was probably the diaphragma sellæ (figs. 2 and 3). No radical treatment was possible. On microscopy the tumour proved to be a slowly growing meningioma.

After a course of radiotherapy, a second surgical exploration was undertaken in April 1949, when the left frontal lobe was amputated. The tumour was seen to consist of three large lobes. One occupied the whole of the sella turcica, and had displaced the pituitary gland and infundibulum posteriorly; a second lobe projected under the left frontal lobe; and a third lobe projected backwards and upwards into the third ventricle. The left optic nerve was a thin band 0·75 cm. across and about 2·5 cm. in length, firmly adherent and incorporated in the tumour capsule, from the optic foramen to the chiasm. The latter had been pushed backwards and was closely applied to the posterior extremity of the tumour. The right optic nerve was stretched round the right side of the tumour. The tumour was removed entirely, after dividing the left optic nerve at the optic foramen and just in front of the chiasm. The patient died forty hours after operation from hypothalamic disturbance.

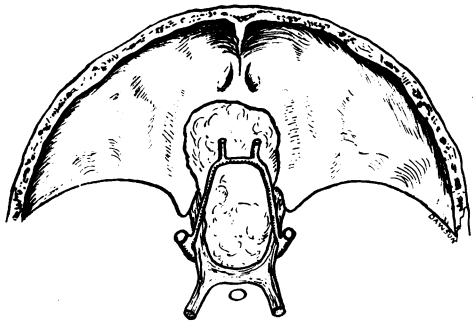

FIG. 2.

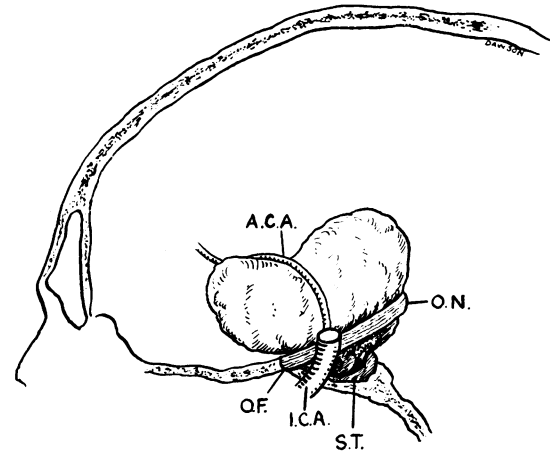

FIG. 3.

A.C.A. Anterior cerebral artery. O.N. Optic nervs
O.F. Optic foramen. I.C.A. Internal carotid arter
S.T. Sella turcica.

FIGS. 2 and 3.—Diagrams of operative findings in Case I, showing tumour lying between optic nerves, and displacing the optic chiasm backwards.

Comment.—This patient was first seen before one had become wise to the clinical picture. The extraordinary feature was the relatively static clinical condition for a period of over ten years, with visual loss confined to one eye. The late surgical intervention showed how large and difficult these meningiomas may become, and stresses the need for early exploration at a stage when the tumour is operable.

CASE II.—*Meningioma of anterior end of falx cerebri with unilateral optic atrophy and central scotoma.*

M. C., female, aged 46. About Christmas 1943 the patient noticed a black spot just below the centre of vision of the right eye, which slowly enlarged in size to involve most of the lower part of the right visual field. There had been no pain associated with the loss of vision, but the patient had had periodic headaches for many years, though she had been free from them for a long period till quite recently, when they recurred. Her general health had remained excellent.

Examination in April 1944, four months after the onset, showed a fairly dense scotoma below fixation in the right visual field, which did not quite reach the periphery of the field below. The right optic disc was moderately pale, and the right visual acuity 6/36 uncorrected. The rest of the examination of the nervous system was normal. Radiographs of the skull and optic foramina were normal.

During the succeeding six months the visual field defect in the right eye spread upwards to involve fixation, and downwards to the periphery of the field, i.e. a large central scotoma

spreading to the periphery in the lower nasal field (fig. 4). Visual acuity fell to 6/60 in the right eye. The left visual field was full, though the left optic disc appeared slightly pale.

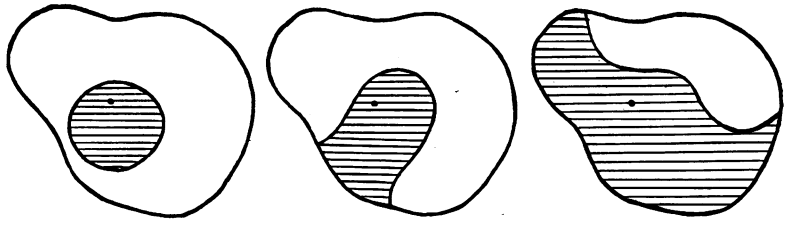

FIG. 4.—Diagram of progress of visual field defect in Case II.

Lumbar puncture was performed in December 1944, when the pressure was raised to over 300 mm. The cerebrospinal fluid was quite normal, the total protein being 15 mg.%. Radiographic examination of the skull at this time showed evidence of erosion of the base of the skull in front of the anterior clinoid processes, with rarefaction of the posterior clinoid processes.

Ventriculography performed on January 22, 1945, showed the presence of a large left frontal tumour, extending across the mid-line. Operation on the same day, by Mr. Wylie McKissock (left frontal approach), revealed an enormous left frontal meningioma, which appeared to arise from the anterior end of the falx cerebri, and one large lobule of tumour extended beneath the falx. The latter was the obvious cause of the loss of vision in the right eye. The tumour was completely removed. Convalescence was uninterrupted.

The patient was last seen in February 1949, four years after operation, when she was fit and well. There was right-sided optic atrophy, with visual acuity less than "counts fingers". The right visual field showed little change from its pre-operative state (fig. 5). The left visual acuity was 6/6.

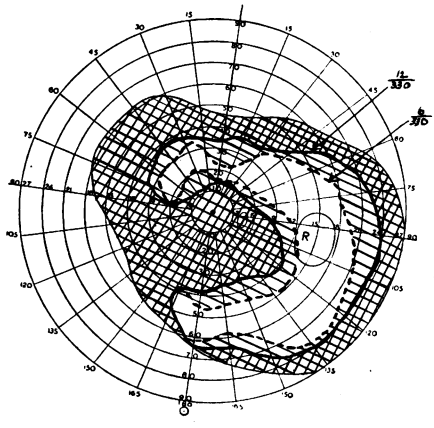

FIG. 5.—Visual field of Case II, three and a half years after removal of meningioma compressing the right optic nerve.

CASE III.—*Meningioma of tuberculum sellæ, with monocular blindness.*

P. R., male, aged 49, complained of progressive failure in the vision of the right eye over a period of about five years, since 1943. In December 1943, it was reported that there was a defect in the upper temporal field, and that visual acuity was reduced to 6/60. At one time he was reported to show a defect in the upper nasal quadrant suggestive of chronic glaucoma. There had been no headaches or pain, and his general health had remained good.

The following diagnoses had been suggested during this progressive visual failure: chronic glaucoma, chiasmal arachnoiditis, an unusual retrobulbar neuritis, hæmorrhage into the optic nerve, and pituitary tumour.

Examination in December 1946 showed well-marked primary optic atrophy on the right side, with visual acuity reduced to counting figures in the lower nasal field. There was no vision in the temporal or upper nasal field or at fixation point. The right pupil reacted poorly to direct light, but well consensually. Sense of smell was normal, and there were no other abnormal signs in the nervous system.

Skiagrams of the skull and optic foramina in January 1947 showed no abnormality of the optic foramina, clinoid processes, or sella.

In June 1948 he was examined again. The right eye was quite blind. There was now a slight defect in the upper temporal field of the left eye (fig. 6), and the left optic disc had become slightly pale. The left visual acuity was 6/5.

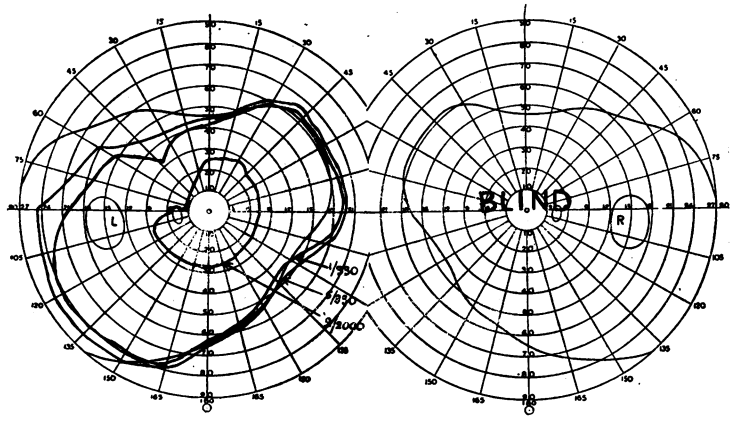

FIG. 6.—Visual fields of Case III, showing monocular blindness and early defect in upper temporal field of contralateral eye.

Investigations in hospital (July 1948) showed that the cerebrospinal fluid was under slightly increased pressure (170 mm.) and the content was normal except for a slight rise in protein (50 mg.%). Skiagrams of the skull showed ? hyperostosis of the tuberculum sellæ.

Ventriculography (July 17, 1948) showed normal lateral ventricles, and a very narrow third ventricle. Myodil ventriculography was therefore performed, and 0·75 c.c. of myodil was injected into the lateral ventricle and manipulated under the screen into the third ventricle. This showed a filling defect in the optic recess, strongly suggestive of a meningioma growing from the tuberculum sellæ.

On July 19, 1948, Mr. McKissock explored the region of the tuberculum sellæ by a right frontal approach. Elevation of the frontal pole revealed a typical suprasellar meningioma lying in the mid-line, and measuring 2½ inches in each direction. The tumour arose from a circular area of the tuberculum sellæ extending from one optic foramen to the other, and backwards into the sella. Amputation of the frontal pole of the brain was necessary before the tumour could be removed. The tumour was pressing on both optic nerves, the right being splayed out and destroyed by the tumour. Histological examination showed the tumour to be a meningioma.

He developed temporary diabetes insipidus after operation. There was no change in the clinical signs after operation, the left visual acuity remaining good.

CASE IV.—*Meningioma en plaque, presenting with monocular blindness, with negative investigations, including ventriculography and angiography.*

G. V., female, aged 51. In May 1948, thirteen months before examination, the patient noticed progressive deterioration in the vision of the right eye, and within ten months the eye was blind. There were no further symptoms, and the vision of the left eye had remained unaltered. She had had occasional headaches for years, not worse recently.

Clinical examination revealed a blind right eye, with a pupil fixed to direct light but reacting well consensually. The right optic disc was pale, with blurred edges, and the upper temporal vessels were sheathed. The left fundus was normal, the visual field full to 3/2000, and the visual acuity 6/9 (J2 with glasses). Sense of smell was normal, and there was no exophthalmos. The rest of the examination of the nervous system was normal.

The cerebrospinal fluid was under a pressure of 140 mm., contained 60 mg.% of protein, and no cells. The Wassermann reaction was negative in both cerebrospinal fluid and blood. Radiographic examination of the skull was normal, but the right optic foramen was slightly larger than that on the left side. There was no evidence of erosion of the walls of the right optic foramen, and the radiologist considered it was within the limits of normality. Air pictures showed good filling of the ventricular system, including the pontine and chiasmatic basal cisterns, and there was no evidence of any filling defect in

this region, nor of any abnormality of the ventricular system. A right percutaneous carotid arteriogram showed good filling of the internal carotid, anterior and middle cerebral arteries, with no abnormality. The electro-encephalogram was normal.

In view of these negative findings, it was decided to explore the region of the right optic nerve, and this was undertaken by Mr. Wylie McKissock on August 4, 1949. There was a plaque of pinkish-red vascular meningioma arising from the bone immediately above and in front of the sella, and from the anterior wall. A small nodule of tumour extending back to the sella, lying between the optic nerves. The tumour extended two-thirds round the right optic nerve, from within downwards, and the whole removal of the tumour could not be achieved. Microscopy of the tumour proved it to be a meningeal endothelioma.

CASE V.—*Meningioma arising from anterior clinoid process presenting as monocular blindness, with negative investigations, including air studies. Post-operative recovery of vision to* 6/9.

R. T., female, aged 36. In July 1946 the patient noticed gradually increasing loss of vision of the right eye, and within three months the eye was almost blind. The vision in the left eye remained unaltered. Neither pain nor headache had been prominent symptoms. At the onset there had been occasional aching pain behind the right eye and in the right cheek, and she had also had occasional right temporo-occipital headaches since the vision began to fail. There had been no vomiting, and no other symptoms referable to the central nervous system except that her sense of smell had never been good. Her general health had remained good.

Examination in March 1947 showed slight pallor of the right optic disc, and vision in the right eye was reduced to hand movements in the upper field (fig. 7). The right visual field

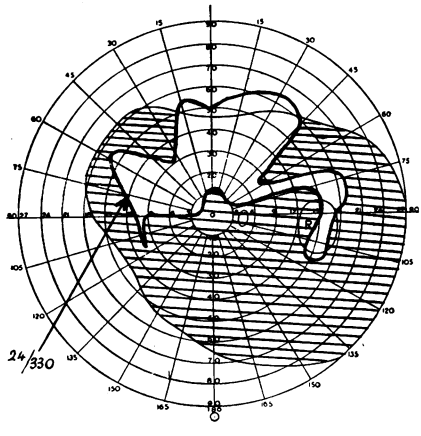

FIG. 7.—Visual field of Case V, before operation.

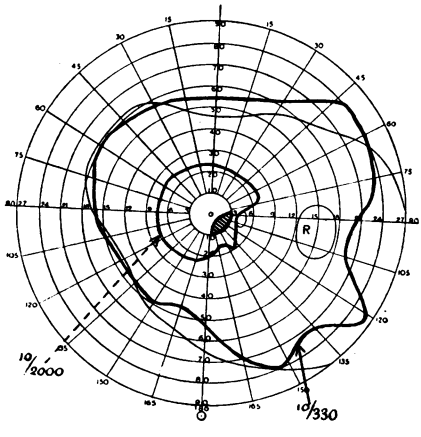

FIG. 8.—Visual field of Case V, three weeks after removal of meningioma.

showed severe loss centrally and inferiorly, the only intact vision being in the periphery of the upper nasal field. The right pupil reacted poorly to direct light but well consensually. On the left side, the optic fundus was normal, the visual field full, and visual acuity 6/5. Ocular movements were full, and there was no exophthalmos. Sense of smell was diminished on the right side, but normal on the left side. There were no further abnormal signs in the central nervous system or elsewhere.

Further investigation showed normal skiagrams of the skull and optic foramina, normal cerebrospinal fluid (pressure 150 mm.), and negative blood Wassermann reaction. Air encephalography showed good filling of a normal cerebral ventricular system.

On May 30, 1947, operation was undertaken, in spite of the negative results of investigations (Mr. Harvey Jackson). A right frontal bone flap was turned downwards, the dura incised and the right frontal lobe elevated. A small meningioma, measuring 2 cm. long by 1 cm. wide, was found arising from the upper surface of the right anterior clinoid process

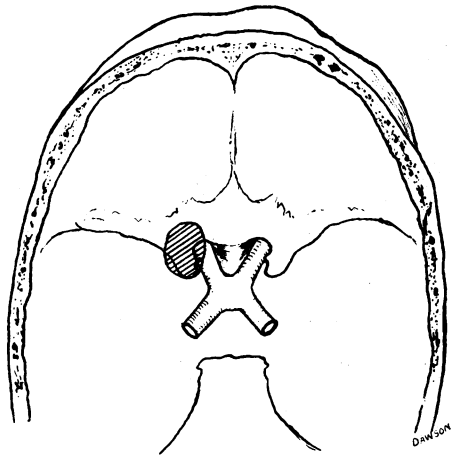

FIG. 9.—Diagram showing site of meningioma in Case V.

(fig. 9). The medial aspect of the tumour was extending over and pressing on the right optic nerve. The optic nerve was freed and the tumour completely removed. The area of attachment to the anterior clinoid process was diathermized. Bleeding was minimal and the patient stood the operation well. Microscopy of the tumour proved it to be a meningioma.

Return of vision ensued with surprising rapidity, so that within a few days the patient could read with the right eye. On June 19, 1947, nearly three weeks after operation, visual acuity was 6/9 in the right eye, and the right visual field showed a defect in the lower temporal field (fig. 8).

CASE VI.—*Meningioma of the olfactory groove with unilateral optic atrophy and central scotoma.*

Mrs. E. M., aged 46. For eighteen months before examination the patient had had severe vertical headaches, worse on coughing or stooping. For about five months she had had frequent attacks in which she was seized with a feeling of terror, followed by a horrible smell or taste lasting a few seconds. The smell was always the same, and she likened it to a bonfire of burning garden hedges. Following this smell she lost the power of her legs and sank to the ground, but did not lose consciousness. Shortly before she was examined she noticed progressive visual failure in the right eye, the left eye being unaffected.

Examination in May 1948 revealed a cheerful, euphoric patient. The vision was reduced to 2/60 on the right side, and, to confrontation tests, there was a central scotoma in the right visual field. The right optic disc and fundus were normal. The left visual acuity was 6/9 and the left optic fundus and visual field were normal. The sense of smell was diminished on the right side. The rest of the nervous system was normal on examination.

A diagnosis was made of intracranial tumour, with direct pressure on the optic nerve, probably an olfactory groove meningioma.

Unfortunately the patient was not seen again until December 1948, when she had right-sided optic atrophy with no perception of light in the right eye. The right pupil was fixed to light, but reacted well consensually. The left eye was normal, and the field full.

Radiography of the skull showed increased bony density along the central part of the floor of the anterior fossa, more marked on the right side, consistent with the presence of an olfactory groove meningioma.

A right percutaneous arteriogram showed good filling of the internal carotid, anterior and middle cerebral arteries. There was a spherical "blush" above the floor of the right anterior fossa, about 6 cm. in diameter (fig. 10). There was a marked shift to the left of the right anterior cerebral artery in the A.P. view. The appearances suggested a large right olfactory groove meningioma.

Ventriculography showed that the septum was displaced about 2 cm. to the left of the mid-line. The anterior horns were much displaced posteriorly, and the third ventricle was not filled. The appearances suggested a large right subfrontal lesion.

Operation was performed on December 23, 1948, by Mr. McKissock. A right frontal bone flap was elevated, and the right frontal lobe amputated. A large ovoid meningioma occupied most of the anterior fossa on the right side, except for the anterior 2 cm. The tumour had a wide attachment, about 2 cm. in length, to the floor of the anterior fossa in the region of the olfactory groove. The tumour was completely removed, and weighed 59 grammes ($5{\cdot}5 \times 4{\cdot}5 \times 4$ cm.). On histological examination it proved to be a meningioma.

The post-operative course was uneventful, and there was some return of vision (perception of light only) in the lower nasal field of the right eye.

On April 13, 1949, nearly four months after operation, the patient felt very well indeed. There was some return of vision in the periphery of the right nasal field, where she could count fingers. The right optic disc was pale and there was right-sided anosmia.

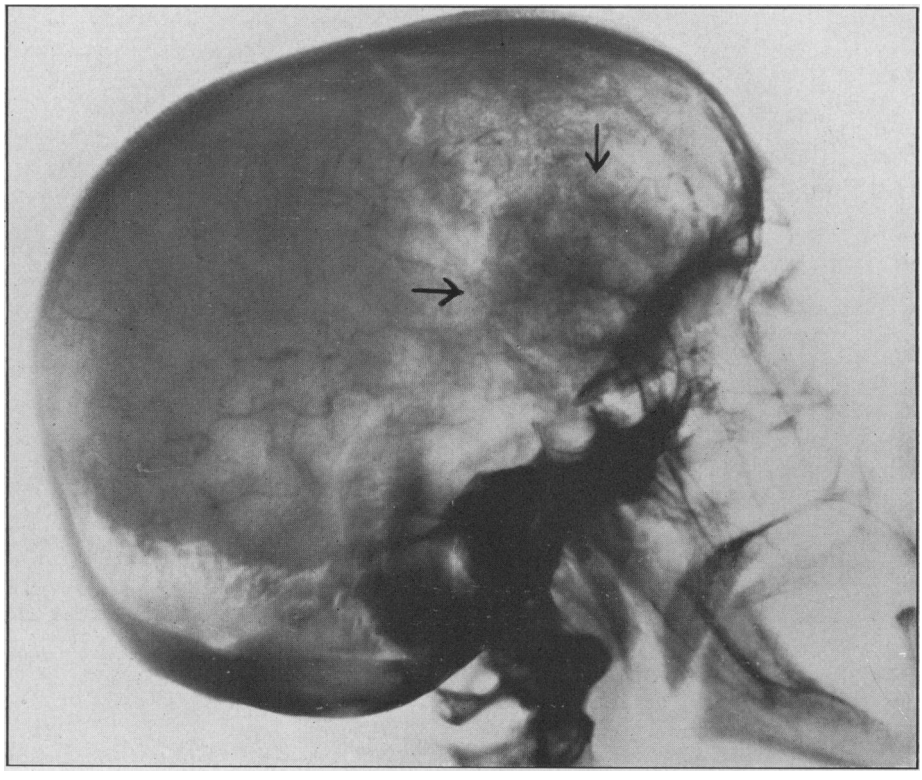

FIG. 10.—Arteriogram in Case VI, showing "blush" outlining the position of the meningioma.

CASE VII.—*Meningioma of the sphenoid with unilateral optic atrophy and slight exophthalmos.*

Mrs. M. M., aged 70. This patient complained of gradual failure of vision in the left eye starting in April 1948, five months previously. She had noticed that the visual loss was chiefly in the lower part of the left visual field. During this period, too, she had had intermittent left frontal headaches, though not severe in character. Her general health had remained good, and there was nothing of note in the past history.

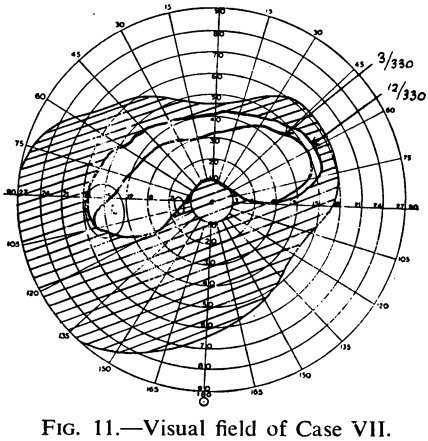

FIG. 11.—Visual field of Case VII.

Examination in September 1948 revealed a visual acuity of 6/6 partly in the right eye, but with ability only to count fingers in the remaining field of the left eye. There was a defect in the lower part of the left visual field, including fixation, the remaining vision being confined to a crescentic area in the upper field (fig. 11). The left optic disc was slightly pale, with slight blurring of the disc edges. The left pupil reacted poorly to direct light, but well consensually. There was slight left-sided exophthalmos. Sense of smell was normal, and further examination of the nervous system revealed no abnormality. The blood-pressure was 210 mm. Hg systolic, and 110 diastolic. Radiography of the skull showed sclerosis of the lesser and greater wings of the sphenoid on the left side, with involvement of the optic foramen, almost certainly due to diffuse meningiomatous involvement of the bone (fig. 12). No operation was undertaken.

Although the diagnosis was not proved by microscopy, there is little doubt that the visual loss, exophthalmos, and radiographic changes in the sphenoid are due to a meningioma, as several similar cases have been seen, some of them verified by operation.

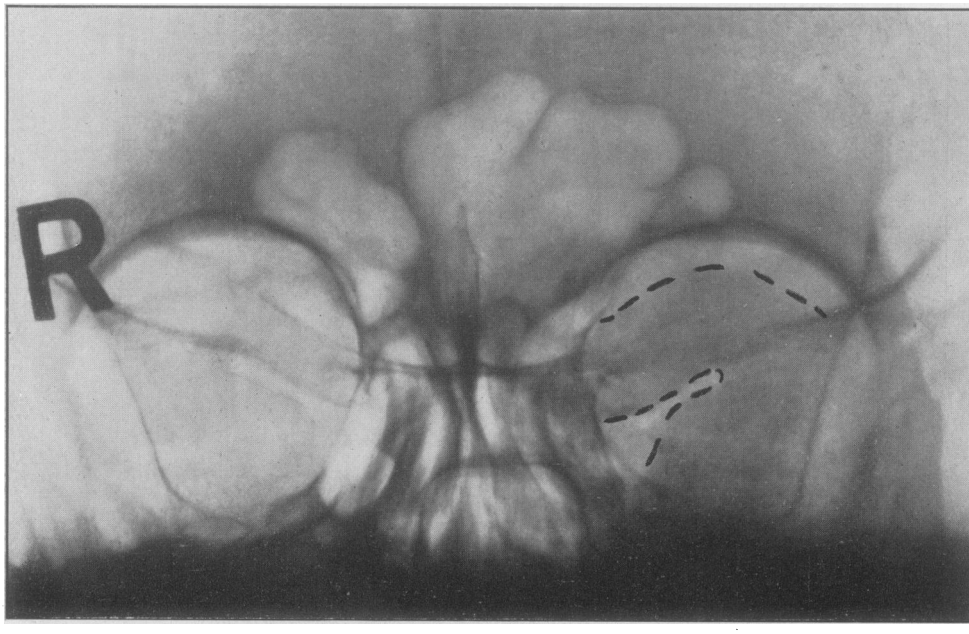

FIG. 12.—Radiograph of Case VII, showing hyperostosis of sphenoid bone on the left side due to meningioma.

CASE VIII.—*Intracranial aneurysm with pressure on left optic nerve, with spontaneous improvement in vision.*

Mrs. H. A., aged 36. Early in 1946, about two years before she was first examined, the patient suddenly developed severe left frontal headache and pain in the left eye, associated with an "influenzal cold" and fever. The fever subsided after a few days, but the headache persisted for about two or three weeks. Soon after this, the patient noticed a gradual dimming of the vision of the left eye, which progressed for some months. She noticed loss of central vision of the left eye, but she could see clearly round the edges. This loss of vision remained almost stationary for about a year, but five months before she was examined the vision in the left eye rapidly deteriorated, so that she could discern only the general shape of large objects. Her general health remained good and there were no other symptoms of note.

In May 1946 it was noted that the vision of the left eye was 6/9, and that there was a sector-shaped scotoma running from the fixation point upwards and to the right in the left nasal field. In September 1947 the vision had fallen to 2/60 in the left eye, the left optic disc was pale, and there was a central scotomatous defect in the left field spreading to the periphery in the upper part of the field.

Examination in January 1948 showed a central scotoma in the left visual field and a slight defect in the upper right temporal field (fig. 13). The visual acuity was 6/6 (corrected) on the right side, but reduced to "counts' fingers" at one foot on the left side. Both optic fundi showed evidence of myopia, with myopic crescents, and pallor of both optic discs more marked on the left side. The right optic disc was considered normal for the degree of myopia. The left pupil was slightly larger than that on the right side, and reacted poorly to direct light but well consensually. There was slight proptosis on the left side. The sense of smell was normal, and the rest of the central nervous system was normal on examination. There was no cephalic bruit, and the cardiovascular system was normal.

The cerebrospinal fluid was under a pressure of 120 mm., and of normal content, and the Wassermann reaction was negative in the blood and cerebrospinal fluid. Skiagrams of the skull and optic foramina showed no abnormality.

A myodil ventriculogram showed obliteration of the optic recess of the third ventricle, and deviation to the right of the bottom of the third ventricle, with slight elevation of the floor of the left anterior horn. These findings suggested an expanding lesion in the region of the left anterior clinoid process.

On January 12, 1948, operation was performed by Mr. Wylie McKissock, and a small left frontal bone flap was turned down. On elevation of the left frontal lobe, the left optic nerve was seen to be grossly widened from side to side, and displaced upwards and laterally by a large aneurysm, arising from either the left anterior cerebral, or anterior communicating artery. In view of the very soft wall of the aneurysm, no further operative procedure was undertaken, and the aneurysm was not needled.

On January 21, 1948, nine days after the intracranial exploration, left percutaneous arteriography was performed, which showed that the left anterior cerebral artery was elevated and pushed backwards, but no aneurysm was visualized. Five days later a right

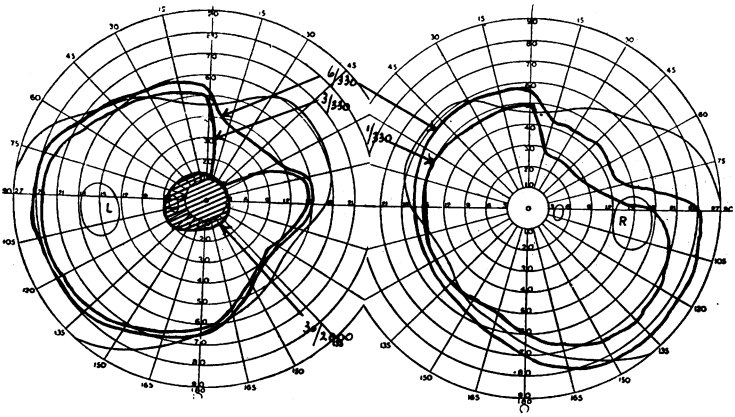

FIG. 13.—Visual fields of Case VIII (aneurysm).

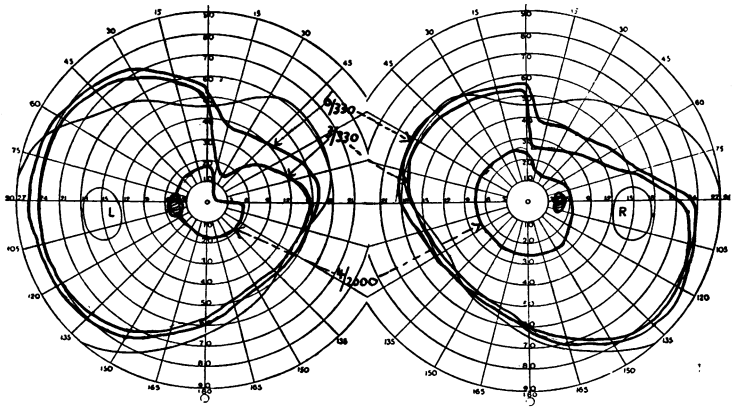

FIG. 14.—Visual fields of Case VIII, some months later, showing disappearance of central scotoma, but persistence of upper quadrantic homonymous defect.

percutaneous arteriography showed no definite abnormality, except that the anterior cerebral arteries did not fill.

The patient made an uninterrupted recovery from the operation, and in spite of the fact that no radical operative measures were taken, the vision in the left eye slowly improved. When seen on December 15, 1948, eleven months after the operation, the left visual acuity was 6/9 partly, and there was a partial right upper quadrantic homonymous defect, but no evidence of a central scotoma on the left side (fig. 14).

The improvement in the vision would appear to add further proof to the diagnosis of aneurysm.

CASE IX.—*Unilateral visual failure and central scotoma due to ? aneurysm.*

E. C., male, aged 45. Eight months previous to admission to hospital, the patient noticed gradual failure of vision of the left eye, with loss of central vision, the peripheral field being relatively intact. He had had occasional dull frontal headaches occurring about once a month for the previous four or five years, but there had been no increase in the headaches with the onset of visual failure. He had noticed no loss of sense of smell, and his general health had remained good. Past history and family history showed no relevant features.

Examination in January 1949 revealed slight pallor of both optic discs, more marked on the left side, with visual acuity of right 6/24 and left 1/60. There was a dense central scotoma on the left side, breaking through to the periphery in the lower nasal quadrant. The right field showed a slight defect in the lower temporal quadrant (fig. 15). The pupils

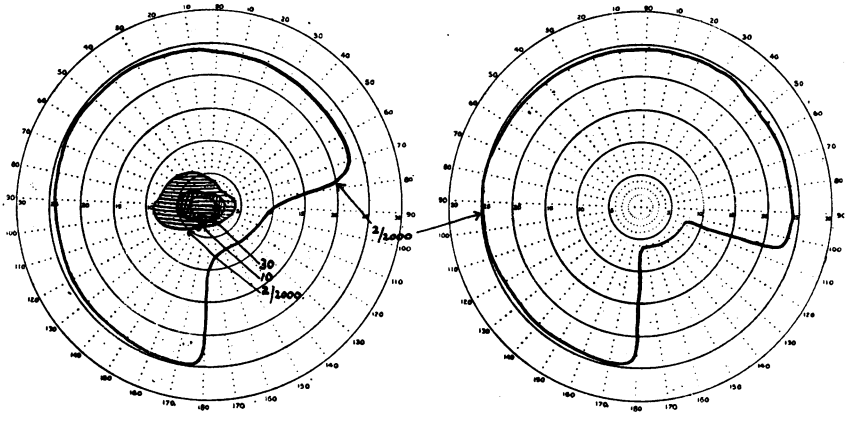

FIG. 15.—Visual fields of Case IX (? aneurysm).

reacted normally, ocular movements were full, and sense of smell was normal. The rest of the nervous system was normal on examination.

The cerebrospinal fluid was normal, the pressure being 140 mm., and the total protein 40 mg.%. Radiography of the skull showed no abnormality. Ventriculography showed a normal ventricular system, and a left percutaneous arteriography showed a normal arterial tree.

In spite of negative investigations it was considered justifiable to explore the region of the optic nerve and chiasm. On January 27, 1949, a right frontal approach (Mr. McKissock) revealed that the left optic nerve was elevated from below by an oval pinkish grey mass measuring 1 cm. in length. The tumour gave the impression of containing fluid, when it was palpated with a blunt hook. From its anterior extremity, and just medial to the optic nerve, an artery emerged and passed into the optic foramen medial to the nerve. It was considered that this tumour was probably an aneurysm. No radical treatment was undertaken.

After operation, the left-sided arteriogram was repeated, and again a normal arterial tree was demonstrated. Examination four months after the exploration showed virtually no change in the visual fields.

CASE X.—*Monocular blindness with cephalic bruit due to ? aneurysm.*

Mrs. A. J. S., aged 63. About Christmas 1948 this patient noticed a sudden onset of a beating noise in her head, which has persisted. For some weeks at this time she had right occipital headache, and for a few days there was diplopia. On March 21, 1949, nine days before she was examined, she noticed visual hallucinations in the right eye, of sudden onset, and lasting most of the day. Later the same day a grey mist appeared in the right temporal field and spread rapidly over the right eye, sparing the upper part of the field. The visual hallucinations were described as "blue lights like stars, with a red rim". Since the onset of blindness of the right eye, formed visual hallucinations occur on closing the right eye, "people waving things, most uncanny".

Examination on March 30, 1949, showed slight swelling of the right optic disc, which was also slightly pale, with a few hæmorrhages near the disc margin. Visual acuity was reduced to hand movements in the right eye, and the right visual field was reduced to a small island of intact vision in the upper temporal periphery (fig. 16). A loud systolic murmur was

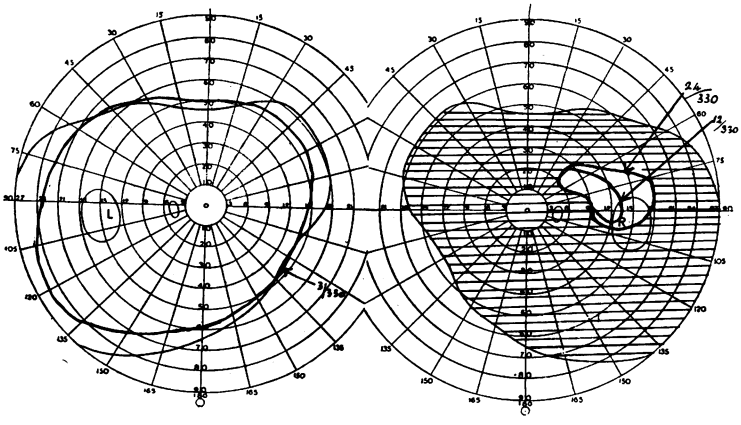

FIG. 16.—Visual fields of Case X.

audible all over the head, but was maximal on auscultation over the right eye, and was abolished by digital compression of the right common carotid artery in the neck. The right corneal reflex was reduced, but the other cranial nerves were normal. The rest of the nervous system was normal on examination, and the blood-pressure 140/90.

Radiographic examination of the skull was normal, and the cerebrospinal fluid showed no abnormality, the pressure being 120 mm. A right percutaneous carotid arteriogram showed fair filling of the common and internal carotid arteries, as well as the posterior cerebral and posterior communicating arteries, but no aneurysm was visible. There was, however, a sudden marked narrowing in the calibre of the internal carotid artery, as it passed through the cavernous sinus. This narrowing may be due to pressure of an aneurysm which does not fill with radiopaque substance.

On May 25, 1949, the right common carotid artery was ligated by Mr. Wylie McKissock, with no untoward effect. The cephalic bruit immediately ceased, but there was no improvement in the vision of the right eye.

Although the diagnosis is unproven in this case, the pathology is undoubtedly vascular, and probably aneurysmal.

CASE XI.—*Glioma of optic nerves and chiasm presenting as thrombosis of central retinal vein.*

J. B., male, aged 50. Four months before admission to hospital, the patient noticed progressive loss of vision of the right eye, so that it was blind within about two weeks. When seen at that time there was evidence of thrombosis of the central retinal vein, and the visual acuity was reduced to 6/36 in the right eye. During the next two months he developed severe pain in the eye, and three months after the onset the right eye was enucleated for secondary glaucoma. The vision in the left eye probably began to fail soon after the right eye became blind, and this failure progressed till admission to hospital.

For some weeks before admission the patient had slept a good deal, had become drowsy, and developed a huge appetite. Pain recurred behind the right orbit, and he became progressively tired and weak, and, more recently, confused.

On admission to hospital on April 6, 1948, the patient was slightly confused, drowsy, and inattentive. The right eye had been enucleated. The visual acuity in the remaining eye was 1/18, the left optic disc was slightly pale, and there was a hemianopia involving the left temporal field (fig. 17). There was slight spastic weakness of the right upper limb, and both plantar responses were extensor. Radiological examination of the skull and chest were negative.

On April 30, 1948, a right frontal lobectomy was performed by Mr. Harvey Jackson, and a tumour was seen involving both optic nerves, but no radical treatment was undertaken. The patient died two days later.

Autopsy revealed that both optic nerves were about one and a half times their normal size at the optic foramina. The left nerve gradually tapered in the orbit to normal size. The optic chiasm was greatly swollen, indefinite in outline, and incorporated in a plum-coloured thickening, which involved it and the infundibular region of the floor of the third

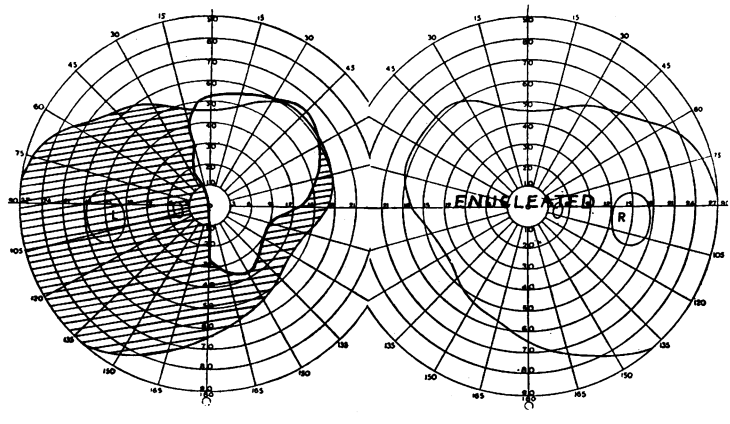

FIG. 17.—Visual fields of Case XI.

ventricle. The left optic tract was swollen for its first centimetre, and the left cerebral peduncle was one-third larger than the right. On section the chiasm and hypothalamus were occupied by a pale firm tumour, which spread to involve the cerebral peduncle and pons, and right lateral ventricle. Histological examination showed the tumour to be a glioma (glioblastoma multiforme).

I am indebted to my neurosurgical colleagues Mr. Harvey Jackson and Mr. Wylie McKissock for operative details in these cases, to Dr. Helen Dimsdale and Mr. Valentine Logue for the details and drawings of Case I, to Dr. William Blackwood for the autopsy report on Case XI, and to Dr. Hugh Davies and Dr. J. W. D. Bull for arteriographic and radiological investigations.

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Mr. Eugene Wolff said that he had long been puzzled as to why the disc became white after division or compression of the optic nerve. Also he had found no explanation in the literature. He suggested the following reason.

The optic nerve being part of the central nervous system does not, as is well known, obey the Wallerian laws of degeneration. There is in fact degeneration centrally and peripherally. The disappearance of the nerve fibres is followed by gliosis. This new-formed glial tissue then contracts and obliterates the capillaries of the nerve-head. And it is the disappearance of the capillaries which is essentially responsible for the white disc.