RETINAL ARTERIAL OCCLUSION IN MIGRAINE

BY

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The occurrence of retinal arterial spasm in the early phases of attacks of migraine has occasionally been described. Unfortunately, an opportunity of examining the optic fundi at such times seldom presents itself, and therefore the frequency and distribution of arterial changes are unknown.

When retinal spasm is severe an occlusion of the affected vessel may occur, leading to a permanent defect in the field of vision. This is a rare complication, but one of much greater theoretical importance than its infrequency would suggest. Such cases supply direct evidence that the pre-headache phenomena of migraine are due to vasospasm. For this reason the following cases are worth recording.

Case 1

A girl aged 17 first attended hospital in April, 1948. From the age of 7 she has had periodic "bilious attacks" without headache. Her attacks of migraine began early in 1947 and have occurred at approximately monthly intervals. Until March, 1948, they were all similar in type, beginning with a whirling sensation surrounded by coloured lights and zigzag formations which started in both eyes simultaneously in the temporal half of the fields and spread to involve central vision. This teichopsia usually lasted about 10 minutes and disappeared suddenly, to be succeeded sometimes by a throbbing headache situated in the fronto-temporal region on both sides. The headache was equally short-lived, never lasting longer than about a quarter of an hour.

In March, 1948, she suddenly developed a blurring of vision in the right eye and thought that one of her usual attacks was starting. Instead the blur persisted and no headache followed. She noticed that the visual defect was present only in the right eye, and that it was situated below and to the left of whatever she looked at. It had remained unchanged for three weeks before her first visit to hospital.

Examination.—The only abnormalities to be found were in the right eye. Ophthalmoscopy revealed an occlusion of the superior temporal retinal artery at a point about half a disk diameter from where it crossed the disk margin. From this point the vessel continued as a fine thread from which no light reflex was seen. In the macular area just above the fovea there was a small area of retinal oedema. The disk itself was normal. The field of vision showed a hemianopic defect in the lower half-field, especially on the nasal side (Fig. 1). This did not involve the fixation point, so that her visual acuity remained normal: 6/5 J1. Her cardiovascular system was normal; the blood pressure was 130/85. Blood Wassermann reaction was negative. There was no family history of migraine.

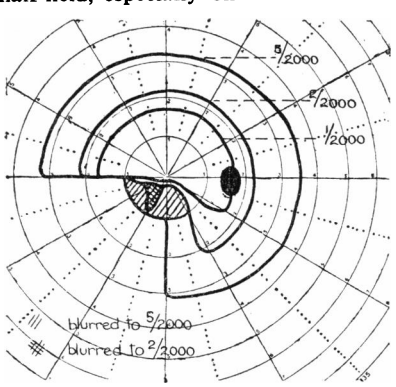

FIG. 1—Case 1. Field of vision of right eye, Aprii, 1948: showing a defect in the lower half-field, particularly on the nasal side, together with a paracentral relative scotoma in this sector.

She was seen again in December, 1948. In the intervening nine months she had had eight further attacks of migraine—all consisting of headache preceded by scintillating scotoma formation in both visual fields. The defect in the right visual field had improved a little and was now confined to the inferior nasal quadrant (Fig. 2). Fundal examination still showed an occlusion of the superior temporal retinal artery, and the superior temporal portion of the optic nerve-head was now pathologically pale. The macular oedema had disappeared. Her visual acuity remained 6/5 J1 in the right eye.

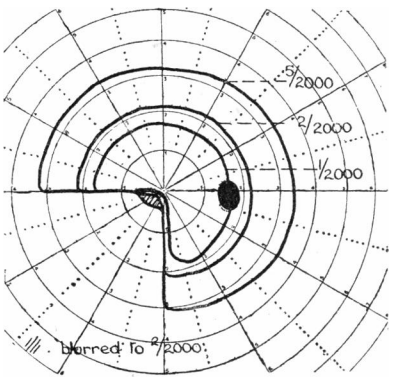

FIG. 2.—Case 1. Field of vision right eye, December, 1948: showing improvement in both the sector defect and the relative scotoma.

Case 2

A woman aged 35 was first seen in October, 1948, complaining of impaired vision in the right eye. For the previous two years she had suffered from attacks of migraine at approximately monthly intervals. These attacks would start with a scintillating scotoma in the right homonymous fields of vision, which lasted for about 10 minutes and was then followed by a bifrontal headache. This was never very severe and lasted no longer than half an hour. Only once or twice had vomiting occurred in the attacks.

In June, 1948, she had two attacks of teichopsia—flashes, spots, zigzag formations—in the right eye only. The first lasted 10 minutes, the second half an hour, but neither attack was followed by headache.

On September 21 at 11 a.m. she suddenly developed a dazzling sensation with spots and flashes of light all over the right visual field. Within a minute a patch of blackness appeared in the upper nasal quadrant of the right field. The dazzling sensation lasted until 4 p.m. and then disappeared, but the blackness remained and obscured vision in this quadrant. No headache followed the attack. She was first seen a month after this episode. In the interval there had been no subjective change in the visual defect in the right eye.

Examination.—Abnormalities were confined to the right eye. In the fundus the retinal arteries were all quite distinctly thinner than in the left eye. The inferior temporal vessel was particularly narrowed, being threadlike in appearance and showing no light reflex. The retinal veins were normal. The disk itself was normal, except for the inferior temporal sector, which was paler than the rest. The retina adjacent to the disk at this point was slightly swollen and blood-streaked. Visual acuity: 6/6 J1 right and left. The right field of vision (Fig. 3) showed a defect in the upper nasal quadrant extending a little into the upper temporal quadrant and corresponding to the occlusion of the inferior temporal artery. The cardiovascular system was normal, and the blood pressure 135/80. The blood Wassermann reaction was negative.

She was seen again two months later, in December. She had had no further attacks of migraine. The right field of vision had improved a little, though she was still blind in the upper nasal quadrant. The fundal appearances were unchanged, except that the retinal oedema had now disappeared.

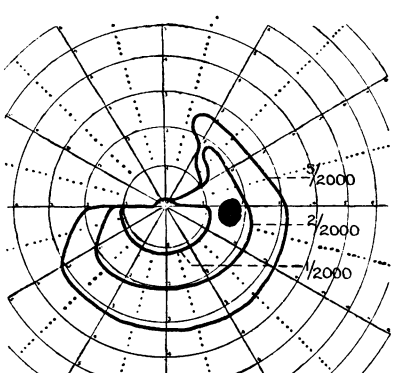

FIG. 3.—Case 2. Field of vision right eye, October, 1948: showing a defect in the upper nasal quadrant and the adjoining part of the upper temporal quadrant.

Case 3

A woman aged 31 was first seen in 1942, when she was 24 years of age. Three years previously, while washing her face one morning, she suddenly lost the sight of her right eye. There was no accompanying headache or ocular pain. For three or four hours the blindness was complete, and then vision returned in the upper half of the field, so that she was left with an inferior altitudinal hemianopia. She was then otherwise symptom-free for about six months. Thereafter, however, she began to suffer from attacks of blurring of vision in the remaining superior half-field associated with ocular pain. These attacks have occurred at intervals of about three months ever since. They begin with a sudden stab of pain over the right eye, which is followed by a sharp pain in the eyeball lasting several minutes. At the same time vision in the lower half-field becomes blurred and remains so for about three hours. There is no family history of migraine.

Examination.—In 1942 the only abnormalities were in the right eye. The field of vision showed an incomplete inferior altitudinal hemianopia with macular sparing (Fig. 4). The superior temporal and nasal retinal arteries were much smaller than the inferior vesvels in this eye. In 1948 there was no change in the field defect in the right eye or in the calibre of the affected vessels. Her right disk showed an optic atrophy limited to the superior half of the nerve-head. Visual acuity was 6/5 J1 in the right eye. Neurological examination was otherwise normal. Her blood pressure was 110/75, and the heart and peripheral arteries were normal. The blood Wassermann reaction was negative.

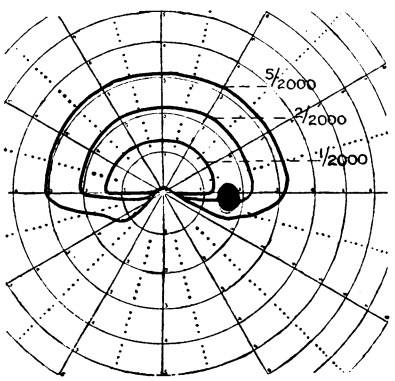

FIG. 4.—Case 3. Field of vision right eye, March, 1942: showing an inferior altitudinal hemianopia.

Case 4

A girl aged 16 was first seen in May, 1948. Between two and three years previously she had had two or three attacks of sudden transient blurring of vision in the left eye, each lasting no longer than a minute and not followed by headache. Then in June, 1946, she developed an influenza-like illness with coryza, generalized aches and pains, malaise, and fever. She went to bed, and after two days felt normal again. On the third day, while lying in bed on her right side, she suddenly saw "black clots" in the left field of vision. Within seconds she became blind in the eye except for a little blurred vision in a small central area (imperfect macular vision). No headache followed this attack of blindness, which has since persisted unchanged.

A year later she began to experience right-sided headaches. She would get pain of a throbbing type over the right brow which came on quite abruptly and lasted up to 20 minutes. At first the headaches were frequent—several times a week—and came on whenever she did a good deal of close work—e.g., reading or sewing. Latterly she had avoided these activities, with a corresponding reduction in the number of her attacks.

On two occasions, in August and September, 1947, she had attacks of scintillating scotoma in the right eye. In the first she suddenly developed a bright dazzling light covering the centre of the right field of vision. After a few minutes this broke up and expanded peripherally, leaving the centre clear, and finally disappeared from the periphery after about 10 minutes. No headache followed. The second attack was very similar but of shorter duration, and again was not followed by headache. There is no family history of migraine.

Examination.—Vision in the left eye was reduced to 6/36 J18. The optic disk was small and atrophic. The retinal arteries were all thin and did not reflect light. The macular area was normal in appearance. The changes were those of an occlusion of the central artery of the retina. The left field of vision was restricted to a small area around the fixation point (tubular vision). The left pupil did not react to direct light, whilst the right pupil failed to react consensually. The right eye was otherwise normal—visual acuity 6/5 J1. There were no other neurological abnormalities, and the cardiovascular system was normal. Her blood pressure was 135/85. The blood Wassermann reaction was negative.

Discussion

The evidence available from both clinical and experimental observation suggests that the symptoms of migraine result from alterations in the calibre of blood vessels in the head, and that both intracranial and extracranial vessels take part in this process. In a fully developed attack two stages are recognized: (1) the stage of pre-headache symptoms—e.g., visual disturbances, paraesthesiae, asphasia—which are thought to be due to functional disturbances of cortical cells secondary to the anoxaemia produced by intracranial angiospasm; and (2) the stage of headache produced by an excessive dilatation, particularly of extracranial arteries, which stimulates the pain nerve endings accompanying these vessels.

Although a vascular theory of migraine was first propounded as long ago as 1870 by du Bois Raymond, it is only in the last decade or so that adequate proof of its occurrence has been obtained. This has been largely due to the work of Wolff and his collaborators, excellently summarized in a recent publication (Wolff, 1948). Their experiments have shown quite clearly that the intensity of the migraine headache is proportional to the degree of dilatation of extracranial arteries, and that the relief from headache achieved by drugs such as ergotamine tartrate is directly related to the degree of vasoconstriction produced.

Whilst the mechanism of headache production is now no longer in doubt, evidence that the prodromal symptoms are the result of intracranial vasospasm has largely been indirect. There are numerous records of patients who have been left with a permanent homonymous visual-field defect after a migrainous attack. A recent case has been described by Rich (1948), and references to previous cases are given in his article. It has been assumed that in such patients arterial spasm has been so intense as to lead to thrombosis and subsequent cerebral softening. Where pathological proof is lacking, however, criticism may be made that such defects may be caused by an underlying vascular abnormality—e.g., an aneurysm or angioma—and that the migraine is of a "symptomatic" rather than a "primary" type. An instructive example of this is mentioned by Kinnier Wilson (1940)—that of a man of 45, a "life-long martyr to migraine," who for 20 years had had a fixed homonymous quadrantanopia, ultimately proved to be caused by a blood-vessel tumour. Adie (1930) described eight cases of permanent hemianopia following migraine attacks. Seven occurred in men—an unusually high proportion for primary migraine. One had had an attack of subarachnoid haemorrhage, which strongly suggests the existence of a structural vascular abnormality.

Evidence of the existence of vasospasm has been accumulated in other ways. James (1945) has shown that a vasodilator drug (carbachol), given regularly, is effective in reducing the frequency of attacks. Wolff in one case was able to show that the inhalation of amyl nitrite, in sufficiently low concentration to produce vasodilatation without a fall in blood pressure, would cause the disappearance of a scintillating scotoma, whereas the latter increased in size when the concentration was great enough to produce hypotension.

That these vascular changes probably produce their effects on the cerebral cortex is shown by the electroencephalographic findings of Engel, Ferris, and Romano (1945). They found that the development of hemianopic teichopsia was accompanied by alterations in the wave pattern from the posterior part of the contralateral hemisphere. In five cases examined during the initial phase of an attack slow irregular 5–7 c/s waves were recorded whilst the visual phenomena lasted and for a brief period afterwards.

Proof of the occurrence of vasospasm in the early stages of a migraine attack is therefore largely circumstantial. The occasional development of retinal arterial occlusion, however, offers more direct and visible evidence, for it seems altogether probable that such thrombosis is secondary to spasm of the affected vessel. Very few such cases have been published.

Galezowski (1882) was probably the first to record such an occurrence. He described four patients afflicted with migraine in each of whom uniocular loss of vision had occurred in relation to attacks of headache. In two, thrombosis of the central artery of the retina was found, one developed an occlusion of the superior branch, whilst the fourth showed peripheral retinal arterial thrombosis with fundal haemorrhages. Their ages ranged from 15 to 67.

Löhlein (1922) described the case of a man who had suffered from attacks of migraine since the age of 20. At the age of 37 he had a nocturnal attack of headache and vomiting; the following morning there was blindness in the left eye. Vision was reduced to the recognition of hand movements in a small peripheral portion of the field and showed no subsequent improvement. Optic atrophy developed, and the condition was considered to be due to a haemorrhage into the optic nerve. Five years later, at the age of 42, a similar attack was followed by blindness in the right eye due to extensive retinal and vitreous haemorrhage, vision later improving. At the age of 46 another attack occurred, reported by Wegner (1926), which resulted in complete blindness and total optic atrophy in the right eye. It seems probable that an occlusion of the central artery of the retina, secondary to vasospasm, was the underlying mechanism of these attacks. Retinal haemorrhage in migraine has also been recorded by Brasch and Levinsohn (1898) and by Vallery-Radot *et al.* (1937), and was seen to a slight extent in Case 2.

Hunt (1915) reported the case of a woman of 31 who had suffered from attacks of migraine since childhood. After a nocturnal attack she noticed a blurring of vision in the right eye and on examination was found to have a central scotoma and some oedema of the nerve-head. These changes regressed and after a period of two months central vision had returned to normal, although a slight pallor of the disk was present. Hunt described the condition as a retrobulbar neuritis and visualized the cause as a vascular lesion in the optic nerve.

The best-documented case is that of Grönvall (1938). His patient was a girl who had begun to suffer from attacks of scotomata and, occasionally, giddiness followed by headache, at the age of 12. At the age of 18 she developed a sudden giddiness associated with a sensation of spots before the eyes. Within a few minutes she became totally blind in the right eye. After a further ten minutes vision returned to the lower half-field of vision, but the upper half remained greyish-black. When examined the following day there was an area of retinal oedema between the disk and the macula; the inferior temporal retinal artery was occluded, and perimetry revealed a defect in the upper nasal quadrant and an adjoining portion of the upper temporal quadrant of the field of vision. The oedema subsided after a month and the field defect showed a slight improvement, but the arterial occlusion remained permanent.

Walsh (1947) records brief details of several patients who suffered from transient attacks of unilateral and bilateral blindness which were sometimes followed by migrainous headaches. One patient developed permanent occlusion of the central artery of the retina in an attack occurring four days after parturition.

Finally, Grimsdale (1940) has written an interesting description of his own personal experience of an attack of retinal arterial spasm, which resulted in an occlusion of a branch of the superior temporal vessel.

Of these nine recorded cases of retinal arterial obstruction in migraine four have occurred in the central artery of the retina, giving rise to severe uniocular blindness; four occurred in peripheral branches, producing sector field defects; and one (Hunt's case) developed a central scotoma, possibly the result of an occlusion of an intraneural branch of the central retinal artery. In the four cases recorded here, one has involved the central retinal artery and three its peripheral branches.

No age group seems particularly predisposed to these complications; there is certainly no evidence that increasing age and coincident arteriosclerotic changes render the migrainous subject liable to such sequelae, for they have affected young and old alike. In the cases described here there were no signs of local retinal or generalized arterial disease, nor was there evidence of any possible source of emboli. Cases 1 and 2 suffered from attacks of migraine before the onset of the retinal occlusion, whilst in Cases 3 and 4 the occlusion was apparently the first symptom of the disease.

Migraine should therefore be considered as a possible cause of those cases of retinal occlusion, occasionally experienced in practice, in which no cardiovascular disease can be found. The condition is probably less uncommon than its recorded occurrence might suggest. Three of the four cases here described have been seen within a period of 12 months. Their interest lies chiefly in the fact that they provide further evidence that the symptoms of migraine are the result of vasomotor disturbances in the territory of cranial blood vessels.

Summary

Four cases of retinal arterial occlusion occurring in migraine are reported. In one case the central artery of the retina was affected; in three cases occlusion occurred in a single peripheral branch.

Such cases provide evidence for the theory that the pre-headache phenomena of migraine are due to vasospasm.

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