PERMANENT SCOTOMA IN MIGRAINE*

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In 1949 Graveson[1] described several cases of what he called 'retinal arterial occlusion' in migraine, a condition which had been described previously but which occurs so seldom that its existence is remembered by very few ophthalmologists or physicians who are concerned with such cases. It is known that retinal arterial spasm occurs in the early phases of migraine, but the opportunity for the investigator to examine the fundi at the time seldom presents itself. When the spasm is severe, an occlusion of the affected vessel may occur, leading to a permanent defect in the fields of vision. I wish to add another instance of this condition which was referred to me with this diagnosis already made by Dr. L. Stein.

CASE REPORT

The patient was a well-nourished, placid, intelligent and apparently healthy young woman aged 23 years. She gave a history of having had migraine since about the age of 10 years. At that time each migrainous attack consistently manifested itself with headaches and nausea, preceded by an aura which she calls 'squiggles', consisting of either hemianopic manifestations where, for example, only half the hand would be seen, or zigzag formations of many colours. These would occur in both eyes simultaneously, would last only a short time and would be followed by the headaches which lasted approximately a day. She used to obtain relief by lying down. For 8 years then, each attack ran a similar course. For the last 5 years she has been more or less free of the headaches and nausea, but her 'squiggles' have persisted. These have recurred at irregular intervals.

Two weeks before examination she had a further attack of 'squiggles', no different in any way from any of the previous attacks, except for the fact that when she awoke next morning, she discovered a strip in the visual field where she could not see, in the right eye only. This scotoma has persisted and was the reason for consulting her physician in the first place. There is no family history of migraine, nor is she or her family subject to allergic conditions.

Clinical Examination of the Eyes

The visual acuity in the affected eye was 6/5 but she was only able to read the letters on the right-hand side of the chart, the letters on the left-hand side being missing. She was emmetropic and orthophoric. The pupils were equal and active to light, both direct and consensual and to accommodation-convergence, and the reaction was well sustained. The tension of the eyes was digitally normal. Close examination of her fundi, by both ophthalmoscopy and slit-lamp biomicroscopy, failed to show any vascular or fundal defect to account for the scotoma plotted. The scotoma consisted of an area roughly circular in shape infero-nasally and close to the fixation point. The fields of red, green and blue were similar, but only the white is illustrated (Figs. 1 and 2). The opposite (left) eye was entirely normal.

Dr. Stein reported that the blood pressure was 140/90 mm.Hg. Urinalysis was normal, haemoglobin was 12 G/100 ml., ESR was 6 mm./hour, fluoroscopy of heart and chest was normal, and general investigation indicated no physical disease, no evidence of cranial arteritis or any special reason for carrying out a carotid angiogram. An occlusion of a branch of the central retinal artery was suspected by him.

To sum up, this patient has had recurrent scotomata of similar nature during her migraine attacks for years. After each attack, the scotoma would disappear, but this particular scotoma

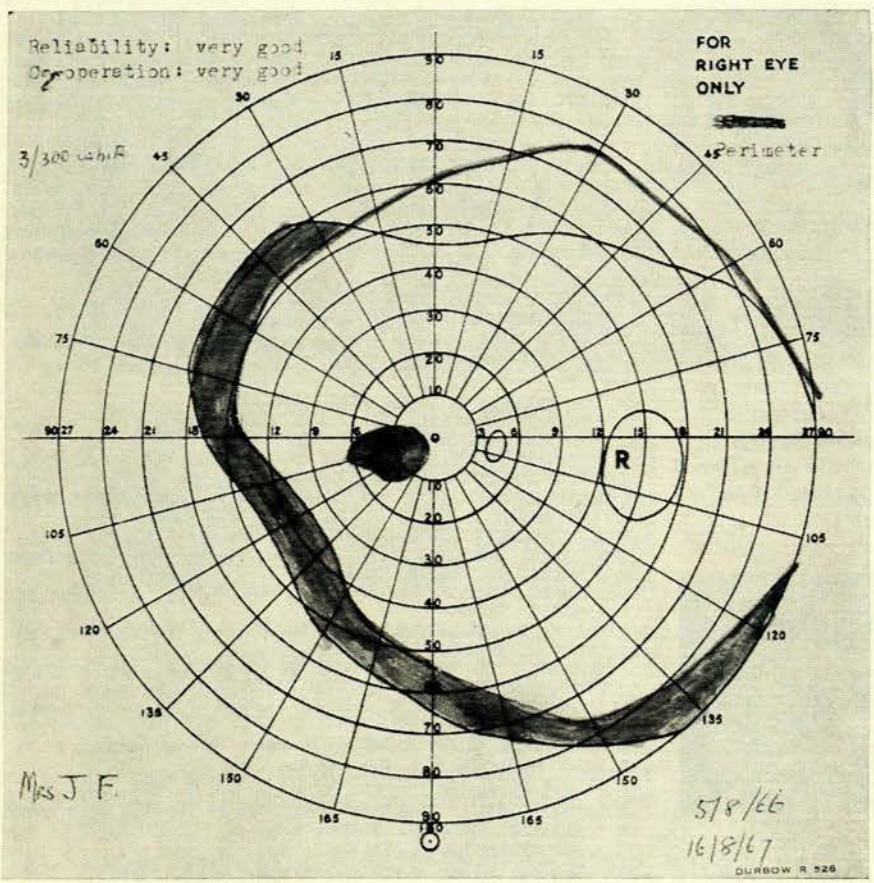

Fig. 1. Peripheral visual field, right eye, tested with a 3/300 white test object showing scotoma nasal to blind spot.

*Paper presented at the 46th South African Medical Congress (M.A.S.A.), Durban, July 1967.

identical with the episodic scotomata has persisted, and has not changed in appearance over a period of 12 months. The 'squiggles' still occur perhaps 2 - 3 times a month, with intervals of 2 - 3 months between attacks.

DISCUSSION

Clinical and experimental observation suggests that the symptoms of migraine result from an alteration in the calibre of the blood-vessels in the head and that both intracranial and extracranial vessels take part in this process. In a fully developed attack 2 stages are recognized:

1. The stage of pre-headache symptoms, e.g. visual disturbances, paraesthesia, etc., which are thought to be due to functional disturbances of cortical cells, secondary to the anoxaemia produced by intracranial angiospasm.
2. The stage of headache produced by an excessive dilatation particularly of extracranial arteries which stimulate the pain nerve-endings accompanying these vessels.

In 1870 Raymond first propounded the vascular theory of migraine. But it is only in the 1940s that adequate proof of the existence of vasospasm has been obtained.[2] While the mechanism of headache production is no longer in doubt, evidence that the prodromal symptoms are the result of intracranial vasospasm has largely been indirect.

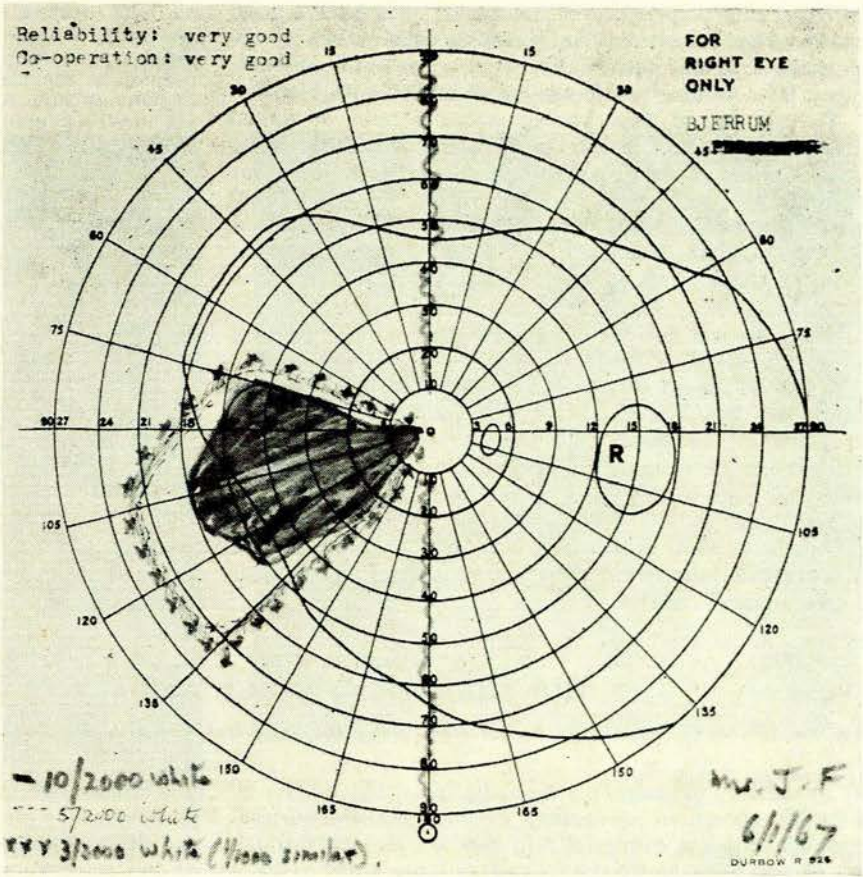

Fig. 2. Central visual field, right eye, tested with 3, 5 and 10 mm. white test objects at 2·0 metre distance. An absolute scotoma extends nasally from the blind spot.

Alvares[3] has shown that the frequency of scotoma during a migraine attack is estimated by different investigators as varying between approximately 33% and 60%. A few cases are on record where the scotoma during an attack could even be mapped out on a perimeter. The scotoma is temporary in nature, e.g. patients have noticed a hemianopia when driving and have had to stop the car and wait until the vision returned in order to avoid an accident. In the passage of time, some migrainous persons, especially the elderly, find that headache and nausea disappear eventually and only scotomata are left. Alvares considers that a number of scotomata are due perhaps to cerebral arterial sclerosis with little strokes.

On the other hand, there are numerous records of patients who have been left with a permanent homonymous hemianopic visual field defect after a migraine attack. Adie,[4] for instance, described 8 cases of this nature. Rich[5] described a homonymous quadrantanopia and mentioned other cases in the literature. He assumed that in such patients arterial spasm has been so intense as to lead to thrombosis and subsequent cerebral softening.

Though proof of the occurrence of vasospasm in the early stages of the migraine attack is largely circumstantial,[6-8] the occasional development of retinal arterial occlusion offers more direct and visible evidence, for it seems altogether probable that such thrombosis is secondary to spasm of the affected vessels. Higgins[9] found the retinal arteries reduced to mere threads with distension of veins in one case. He could tell when vision was returning by watching the enlargement of the arteries.

Very few cases have been published where a unilateral permanent scotoma has resulted from a migrainous attack, following a local vascular occlusion. Galezowski (1882)[10] was probably the first to record such an occurrence. He described 4 patients with uni-ocular loss of vision occurring in relation to attacks of headache. These cases showed conditions such as thrombosis of the central artery, neuroretinitis and consecutive optic atrophy. Lohlein[11] described one case, and Hunt[12] described a case with a central scotoma and oedema of the nerve head. Gronvall[13] has recorded a case of a girl aged 18 years who had loss of the upper half of her vision, and her retina showed an area of retinal oedema between the disc and macula and an occlusion of the inferior retinal artery. The oedema subsided after a month and the field defect showed a slight improvement, but the arterial occlusion remained permanent. Walsh[14] described several patients who suffered from transient attacks of unilateral (and bilateral) blindness which was sometimes followed by migrainous headaches. Finally, Grimsdale[15] 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. The vessel, as a result of treatment, later became visible again and refilled with blood.

To sum up, of these known recorded cases of retinal arterial obstruction in migraine, 4 have occurred in the central artery of the retina giving rise to severe uni-ocular blindness, 4 occurred in peripheral branches producing sector field defects, and one (Hunt's) case developed central scotoma, possibly the result of an occlusion of an intraneural branch of the central retinal artery. Graveson's 4 cases showed no signs of local retinal, or generalized arterial, disease nor was there evidence of any possible source of emboli.

Migraine should therefore be considered as a possible cause of those cases of retinal occlusion occasionally encountered in practice in which no cerebrovascular disease can be found. The condition is probably not as uncommon as its recorded occurrence may suggest. The present case is just one more instance of the sudden development in a migrainous individual of a permanent unilateral scotoma presumably due to arterial occlusion of an intraneural vessel.

SUMMARY

A 23-year-old woman, otherwise perfectly fit and well, developed a permanent uni-ocular paracentral scotoma in the same position as she has had evanescent scotomata as part of her migrainous aura for the past 13 years. Unilateral scotomata are rare in migraine but can occur, and are presumably due to the occlusion of a retinal vessel during the period of retinal arterial spasm.

I wish to thank Dr. L. Stein for referring this case, Dr. D. Sevel for helpful comments, and Mrs. P. Webster for her assistance.

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