

THE MIGRAINOUS SYMPTOMS OF CEREBRAL ANGIOMATA

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That cerebral angiomata may simulate migraine is well known but the frequency of such a mode of presentation is not clearly established (Mackenzie, 1954). In the last 10 years many patients with migraine have been investigated by carotid angiography in the Royal Infirmary, Manchester, but very few have been shown to harbour an angioma. This has led to the impression that the relationship between migraine and angioma has been overstated. The first part of this study concerns 70 patients with proven intracranial angiomata.

CASE MATERIAL

The patients were investigated in the Departments of Neurology and Neurosurgery at the Royal Infirmary, Manchester, between the years 1949 and 1960. The angiomata were localized by carotid and vertebral angiography. Contrast radiography with air or Myodil and by craniotomy gave additional information in some cases.

DEFINITIONS

For clinical purposes, the lesions can be described as more or less circumscribed, arteriovenous communications consisting of a collection of tortuous dilated blood vessels.

In order to simplify the analysis of symptoms, migraine has been divided into the following four grades:—

GRADE 1 Alternating hemicrania, accompanied or preceded by such visual symptoms as teichopsia, flashes of light or hemianopia, and often by paraesthesia, pareses and dysphasia, followed by nausea or vomiting.

GRADE 2 The same as grade 1, except that the headache, visual, sensory, or other focal symptoms, always affected the same side of the head or limbs. The term 'fixed migraine' may be used for this grade.

GRADE 3 Recurrent headache, often hemicrania.

GRADE 4 Headaches without special characteristics.

The diagnosis of migraine would be inevitable in grade 1, probable in grade 2, possible in grade 3, but a mere label in grade 4.

INITIAL SYMPTOMS

It is clear that the diagnosis of cerebral angiomata is most likely to be difficult at the time of the first symptoms. A diagnosis of idiopathic migraine at this stage might allow the underlying lesion to remain undetected for months or years.

The first symptoms in the 70 patients are shown in Table I. Of 70 patients, only 11 had recurrent head-

TABLE I
ANALYSIS OF SERIES BY FIRST SYMPTOM

<i>First Symptom</i>	<i>Number of Cases</i>
Subarachnoid haemorrhage	35
Cerebral haemorrhage (strokes)	7
Epilepsy (focal or generalized)	9
Progressive paresis (hemiparesis)	5
Visual defect (scotoma)	1
Aphasia	1
Face pain	1
Headache	11
Total	70

ache, unaccompanied by complications as a first symptom. When this group of 11 patients was studied further it was clear that some of them were unlikely to be diagnosed as migraine. Details of the 11 patients are shown in Table II. Amongst them were three patients who could be placed in the category of grade 2 migraine but none in that of grade 1 migraine.

HEADACHE The following are brief illustrative case reports of patients whose initial symptom was headache.

Case 1 At the age of 12, this woman began to have attacks of flashes and black spots in the vision, with paraesthesiae in the arms and tongue, dysphasia and

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TABLE II

ELEVEN PATIENTS WITH HEADACHE AS FIRST SYMPTOM								
Grade of Migraine	Localization of Headaches	Duration of Headaches	Visual Effects	Paraesthesiae	Dysphasia	Vomiting	Signs	Complications
2	Left side only	50 yr.	++	+	+	+	Slight	—
2	Right side only	13 yr.	++	+	—	+	+	Tinnitus
2	Left side only	2 yr.	—	+	+	—	—	—
3	Right side only	15 yr.	—	—	—	—	+	Subarachnoid haemorrhage
3	Generalized	1 yr.	—	—	—	—	+	Subarachnoid haemorrhage
3	Generalized	1 yr.	—	—	—	+	+	Subarachnoid haemorrhage
3	Bifrontal	6 yr.	—	—	—	+	+	Subarachnoid haemorrhage
4	Occipital	5 mth.	—	—	—	—	+	Subarachnoid haemorrhage
4	Generalized	6 wk.	—	—	—	—	+	Subarachnoid haemorrhage
4	Generalized	8 wk.	—	—	—	—	+	Subarachnoid haemorrhage
4	Bifrontal	7 wk.	—	—	—	—	+	Subarachnoid haemorrhage

diplopia, followed by left-sided headache which was aggravated by stooping. At the age of 62, when her symptoms were worse, a large left parietal angioma was demonstrated. The only abnormal physical signs were nystagmus and depression of the left corneal reflex. This patient had 'fixed migraine'. The headaches were always unilateral on the left side of the head, but the paraesthesiae and visual symptoms were bilateral or generalized.

Case 2 In childhood this woman had periodic vomiting. At the age of 13 episodic headaches began; they were accompanied by flashing lights in the left visual field and followed by right-sided headache. At the age of 25 she had tinnitus and a number of attacks of focal paraesthesiae in the right hand and left leg during the headaches. On first examination at the age of 25 there were abnormal neurological signs, the left ankle jerk being increased and the left plantar response extensor.

A right carotid angiogram was negative at that time but one year later further angiography showed a large right occipital angioma. Although there were many features of migraine the headache and visual symptoms were fixed.

Case 3 This woman began to suffer from periodic episodes of left temporal headache at the age of 30. Each attack was accompanied by paraesthesiae in the right arm, dysphasia, and vomiting. She experienced numbness in the gums on both sides during these attacks which occurred once every eight weeks for two years.

Carotid angiography revealed an angioma in the left parietal region of the brain. In this patient headaches were 'fixed' and the left sensory symptoms and dysphasia suggested a left parietal lesion.

Case 4 This man began to have periodic right hemicrania each month at the age of 24. There were no other symptoms. At the age of 39, after a few months of tinnitus, he sustained an acute subarachnoid bleed.

Angiography and craniotomy showed an angioma in the right temporal lobe of the brain.

Case 5 This woman began to have severe headaches when she was 33 years of age. They were most evident in the mornings and were exacerbated at menstruation. There were no other migrainous features. One year later she had a severe subarachnoid haemorrhage with left

hemiparesis. Two more haemorrhages followed in the next six weeks.

Vertebral angiography showed an angioma in the left lobe of the cerebellum.

Case 6 At the age of 52 this woman began to suffer from attacks of vomiting, accompanied by generalized headache, which lasted one or two days. One year later she had an episode of unconsciousness due to a subarachnoid haemorrhage.

A large left parietal angioma was shown by carotid angiography.

Case 7 At the age of 9 years this man began to have periodic headaches and vomiting. The headaches were in the frontal or bitemporal regions. A severe cerebral episode, which was probably a subarachnoid haemorrhage, was diagnosed as 'cerebrospinal fever' at the age of 15. Three years later a partial right homonymous hemianopia suddenly developed. When he was 27 he had a sudden subarachnoid haemorrhage. Examination revealed a complete right homonymous hemianopia with macular sparing. He had also nominal dysphasia, nystagmus, extensor plantar response in the right foot, and a bruit to the left side of the skull.

There was a large angioma in the left parietal, temporal, and occipital lobes.

Case 8 At the age of 27 this man began to have headaches each morning. Exacerbations occurred every three to four weeks for five months. He then sustained a subarachnoid haemorrhage with loss of consciousness and this was followed by paresis of the right leg and later by epilepsy without focal features. There was a skull bruit.

Carotid angiography showed a large left frontal angioma.

Case 9 This man of 53 had periodic attacks of sweating which were followed by frontal headaches. The attacks continued for six weeks and were followed by five major epileptic seizures.

Angiography revealed a large left frontal angioma.

Case 10 For two years this man of 46 had periodic generalized throbbing headaches which lasted five hours each morning. He then fell unconscious after blowing his

nose and remained comatose for 11 days. One month later, a central scotoma suddenly developed in the right eye; this was caused by a vitreous haemorrhage.

Carotid angiography and craniotomy revealed an arteriovenous angioma which communicated with the right ophthalmic artery and the superior longitudinal sinus.

Case 11 This man of 47 had bilateral frontal headaches for only seven weeks before a subarachnoid haemorrhage occurred.

From this description it may be observed that only the first three or four patients had headaches which were likely to be called 'migraine'. Case 1 had symptoms for 50 years before the angioma was diagnosed. All the usual migraine symptoms had been experienced, yet the headache was constantly on one side of the head. Case 2 also had symptoms for many years and although suggestive of migraine two features were suspicious, namely, that the headache was always on one side of the head and that she had an extensor plantar response on one side. The diagnosis of migraine might have been considered in Case 3 during the first two years of symptoms. There were no abnormal signs but again the headache was constantly on one side of the head. Case 4 also had 'fixed' headache but 15 years elapsed before a complete diagnosis was made.

A diagnosis of migraine in the other six patients would probably not have been made because of the short period of headache, lack of other migrainous symptoms, or the presence of abnormal neurological signs.

The site of the angiomatous malformation in each of the 11 patients whose first symptom was headache is shown in Table III. When the headache was constantly on one side of the head the position of the pain was a good localizing sign of the angioma. Generalized or bilateral headaches were caused by angiomas in many positions.

Focal symptoms were usually a good guide to the localization of the cerebral lesion. In Case 1, who had multiple migrainous symptoms, there was a large left temporal angioma, but it was very deep, in fact situated centrally in the Circle of Willis. Its site may account for the widespread effects, which included flashes of light in the vision, paraesthesiae in both hands, arms, and both sides of the tongue, and of vomiting. The headaches were made worse by stooping or exercise. Case 2, who had left-sided visual flashes of light, like those of migraine, had an angioma which was partially situated in the right occipital lobe. Case 3, who had an occipital angioma, also experienced left homonymous hemianopia.

SUBARACHNOID HAEMORRHAGE Only one patient had a syndrome resembling migraine in those 47 patients whose first symptom was a subarachnoid haemorrhage.

Case 12 This man had a subarachnoid haemorrhage at the age of 16 and was unconscious for one week. For the next eight years he was free of symptoms and then began to have frequent early morning headache. The pain was near the right eye and was accompanied by epiphora, nausea, and vomiting. Some headaches occurred over the left frontal region. There were no visual or paraesthetic symptoms. These headaches continued for many years. For eight years before the age of 56 there was no nausea or vomiting with the attacks.

At the age of 56, a left carotid angiogram showed a large posterior parietal and a large occipital angioma, which extended into the choroid plexus of the left lateral ventricle. Although this man had no visual or sensory symptoms of migraine, he did have periodic headache, nausea and vomiting, which could have been diagnosed as migraine. The fact that he had a subarachnoid haemorrhage early in life pointed to the underlying lesion.

OTHER NEUROLOGICAL SYMPTOMS In the seven

TABLE III

SITE OF ANGIOMATOUS MALFORMATION IN 11 PATIENTS WITH HEADACHE AS FIRST SYMPTOM				
Patient	Grade of Migraine	Site of Headache	Site of Angioma	Site of Focal Symptoms
1	2	Left	Left 'central Circle of Willis' and temporal	—
2	2	Right	Right parieto-occipital	Left visual field Right tinnitus Right hand Left leg Right hand Dysphasia
3	2	Left	Left parietal	—
4	3	Right	Right temporal	—
5	3	Generalized	Left cerebellar	—
6	3	Generalized	Left parietal	—
7	3	Bilateral	Left { Parietal Occipital Temporal	Right visual field
8	4	Occipital	Left frontal	Right leg
9	4	Frontal	Left frontal	—
10	4	Generalized	Right ophthalmic and frontal	Right eye (central scotoma)
11	4	Bifrontal	Right side close to the origin of the straight sinus	—

patients whose first symptom was cerebral haemorrhage, none had a syndrome resembling migraine afterwards.

In the five patients whose first symptom was a progressive paresis or other permanent neurological sign, only one had symptoms which resembled migraine.

Case 13 At the age of 21, this man had a sudden right homonymous hemianopia which was permanent. A few weeks later he began to have right-sided temporo-occipital headaches which were rather persistent, though at times periodic and accompanied by nausea. In the next three years he had two attacks of numbness in the right arm and right side of the face; in one of these there was dysphasia. On examination he had a right homonymous hemianopia, extensor plantar response in the right foot, and a systolic bruit in the left temporal region of the skull.

Carotid angiography revealed an extensive left-sided parieto-occipital angioma.

Of the nine patients who had epilepsy as the first symptom, none developed migrainous symptoms later. No migraine syndrome occurred in the patients who had central scotoma, aphasia, or facial pain as the first symptom.

The second part of this study concerns all patients with migraine or intracranial angiomas who were seen in the period 1955-60 in the Department of Neurology at St. Bartholomew's Hospital, London.

The 300 patients with migraine were consecutive and unselected. All had a complete physical examination including auscultation of the neck and skull and radiological examination of the skull. Of these patients, 23 were considered to require arterial angiography.

It would serve no purpose to reproduce the clinical details of all these 23 patients. The symptoms and signs for which angiography was performed were as follows: Fixed headache (5), focal sensory symptoms (10), focal visual effects (2), focal motor symptoms (3) or hemiparesis (2), dysphasia (2), diplopia (1), ataxia and vertigo (1), Horner's syndrome (1), attacks of unconsciousness or epilepsy (7). One gave a history of subarachnoid haemorrhage, two had extensor plantar responses, and eight had focal electroencephalographic changes.

In spite of these features none showed an angiomatous malformation; indeed the arteriograms, often bilateral and sometimes including the vertebral as well as carotid systems, were all essentially normal.

There were four patients who had intracranial angiomas, the only four in five years. Only one had symptoms resembling migraine.

Case 14 For 26 years this man aged 51 had attacks

of left frontal headache, followed by right homonymous teichopsia and right-sided paraesthesiae. On two occasions he lost consciousness during attacks. Examination revealed a bruit over the left side of the skull in addition to sensory and reflex changes in the right arm and leg. There was a left parietal angioma which was partially calcified. The early symptoms, although migrainous, were always 'fixed' and focal.

The other three patients with angiomas had no headaches and no symptoms resembling migraine. One, a man of 49, had epilepsy. A bruit was heard over a fronto-parietal angioma. Another, a man of 52, had brain-stem and cerebellar symptoms caused by angiomas. He had telangiectasia of the mouth, lips, and nose. The third, a woman of 38, had two focal epileptic attacks without headache or visual symptoms. Migraine was not even considered. There was a parietal angioma.

Thus, in the final analysis of 300 cases of migraine and four cases of intracranial angioma seen in five years, only one patient with angioma presented in a way resembling migraine (Case 14), and of 23 patients investigated as angioma 'suspects' none showed such a lesion.

It must be noted that these four patients with angiomas and the 300 with migraine concern only neurological patients and not neurosurgical.

DISCUSSION

Cushing and Bailey (1928) did not stress migraine as an important symptom of vascular tumours of the brain although eight of 12 patients they described had headache at some time. Their Case 9 faintly resembles migraine in that there were attacks of blurred vision in the right homonymous field. This patient had pain in the left temporal region and numbness of the right hand. The symptoms were 'fixed' and focal. Involuntary movements of the right arm and speech disturbances were also experienced. Their Case 11, who had a left occipital angioma, developed a right homonymous hemianopia and left optic atrophy after seven years of left-sided headache. None of the remaining patients exhibited the migraine syndrome.

Dandy (1928) described eight patients who had cerebral angiomas; none had headache as a first symptom and seven had focal epilepsy. He also analysed all the cases of angiomas which had been described before 1928 and found only three which had recorded headache as a first symptom. They had other features, including fits and hemiplegia.

Olivecrona and Riives (1948) considered that little change in symptomatology had been observed since their original paper in 1936. The cardinal signs of angioma being fits, subarachnoid haemorrhage, and

hemiplegia, they considered that although headache was a frequent complaint it was not characteristic. They noted that Baruk (1931), Deutsch and Friedman (1938), and Hyland and Douglas (1938) had recorded single examples of migraine in patients with cerebral angioma. In all three the angioma was situated in the occipital lobe. Olivecrona and Riives found only one patient who had migraine; he also had focal epilepsy.

Northfield (1940), in a study of 14 cases of cerebral angioma, found that headache was outstanding in eight; one had episodes of headache for 37 years and another for 22 years; visual hallucinations occurred in only one patient. The headache was not necessarily a presenting feature and, of the 14 cases, seven had epilepsy and seven intracranial haemorrhages.

Ray (1941) commented that headache was not an outstanding symptom in the records of cases that had been reported by others and that often it was not thought important enough to ascribe to any particular mechanism.

Mackenzie (1953) found that in 24% of 50 patients headache was an early symptom. Follow-up of the 12 patients showed that they had suffered from it for as long as they could remember and in seven there were migrainous features from the onset. The headache and aura were often persistently one-sided but aura was prolonged and might continue after the headache. Headache in these 12 patients was a presenting feature, not the first symptom. In the four who had had it as long as they could remember, one had subarachnoid haemorrhage at the age of 10 and another at the age of 15 years; both had epilepsy. Another had haemorrhage at the age of 16 and the last of the four had hemianopia for 23 years. In the seven who had a history suggestive of migraine all were focal, five had epilepsy, and one had subarachnoid haemorrhage. Although the presenting symptom was headache few of these patients would be diagnosed as having migraine because of these features or complications. Seven of the 12 had hemiparesis and one had proptosis.

Mackenzie (1954) stated that the relative frequency of each mode of presentation was not yet established. In a series of nine posterior fossa angiomas described by Logue and Monckton (1954), none had migraine.

Potter (1955), in a study of 58 patients, found that 18% had headache as the first symptom. These patients had a relatively long survival period. All 10 survived over 10 years, eight over 20 years, five over 30 years, and one for 54 years, after the onset of symptoms. He considered that the headache was often 'migrainous'.

Paterson and McKissock (1956) observed that periodic headaches resembled migraine and occurred in 15% of 110 patients with cerebral angiomata. However, six of these had subarachnoid haemorrhages

and nine had epilepsy before the first attendance. Only four of them had migraine as an isolated symptom before diagnosis and all of these were atypical because one had a cerebral haemorrhage before the headache began, two had epilepsy, and two had permanent homonymous hemianopia. They also found that in one third of those with a history of migraine there was a family history of the same complaint. Nine per cent of the whole series had such a family history, which suggested that migraine might be something which occurred independently of the angioma.

Sutton (1957) found that 10 out of 100 patients with cerebral angioma had periodic migraine. Six of the 10 had homonymous field defects, though all had constantly homonymous visual aura. The angiomata were situated in or near the occipital lobe.

In the series of Dimsdale (1957), headaches occurred in 26%. One patient is mentioned to have had migraine only. Many of the patients had epilepsy or intracranial haemorrhages. The frequency of headache or migraine as a first symptom was not recorded.

Sabra (1959) found that 40% of his patients had recurrent headaches, in half of them preceded by visual phenomena simulating migraine.

Most writers agree that cerebral angiomata may simulate migraine. Visual phenomena, similar to those of migraine, are prone to occur if the lesion is in or near the occipital lobe. It is not possible to discover the frequency of periodic headaches as a first symptom in some of the series of cases which have been published, though it is probably not high. As a presenting feature headache is not uncommon though by this stage many of the patients have had complications such as fits or subarachnoid haemorrhage and on examination many have abnormal neurological signs. Even in those cases which closely simulate migraine there are in most the features of persistently one-sided headache or sensory symptoms. In the present series of 70 patients not a single case completely simulated idiopathic migraine. Although 11 (16%) patients complained of headache as a presenting symptom, only three of them had 'migraine' and in these the headaches were always on one side of the head; two of the three had abnormal physical signs. The other eight patients had none of the sensory phenomena of migraine and all were atypical in that the symptoms tended to be 'fixed' or there were early complications or abnormal neurological signs.

The group of 59 patients whose presenting symptom was not headache contained only two cases which resembled migraine; one of them had a longstanding right hemianopia and the other gave a history of subarachnoid haemorrhage. Even in

these two patients the headache and other symptoms were persistently one-sided.

The second part of the investigation has indicated the infrequency of intracranial angiomas in a series of cases of migraine. It is of course impossible to be quite certain that a patient does not harbour an angioma without resorting to multiple angiograms. This is neither necessary nor practicable. Even in 23 patients, investigated because of alarming or focal symptoms and in whom one could fairly suspect an underlying lesion, no angiomas were found. Only one patient seen in the five-year period had an angioma with symptoms resembling migraine, yet these symptoms were focal and there were abnormal signs.

CONCLUSION

Although the symptoms of cerebral angioma may simulate migraine, particularly when the lesion is in or near the occipital lobe of the cerebral hemisphere, it is unusual for the complete syndrome of 'idiopathic migraine' to be produced either as a first symptom or a later one. Investigations of a patient with headache are unlikely to reveal an angioma of the brain unless the headache and aura are per-

sistently one-sided or unless examination reveals abnormal neurological signs or a cranial bruit.

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