Migraine with aura: visual disturbances and interrelationship with the pain phase. Vågå study of headache epidemiology

Introduction

Migraine with aura [1] (classic migraine [2]) is probably the first recognised headache. The prominent position of the visual phenomena has also probably led to the term: “ophthalmic migraine” [3].

Abstract

In the Vågå study of headache epidemiology, 1838 or 88.6% of the available 18–65-year-old inhabitants of the commune were included. Everyone was questioned and examined personally by the principal investigator (OS). There were 178 cases of various types of visual disturbances during the migraine attack, which corresponds to 9.7% of the study group. The prevalence among females was 11.9% and among males 7.4%; female/male ratio was 1.70, as against 1.05 in the total Vågå study population. By far the most frequently occurring visual disturbance pattern was (A) 1. Visual disturbances → 2. pain-free interlude → 3. pain phase (in 78% of the cases). Other frequent patterns were: (B). Visual disturbances, but no pain phase (24%); and: (C) 1. Pain phase → 2. visual disturbances (23%). Evidently, in the solitary case, there might be more than one visual disturbance pattern. The most frequently occurring solitary visual disturbances were: scintillating scotoma (62%) and obscuration (33%); but also more rare ones were identified, like anopsia, autokinesis (movement of stationary objects), tunnel vision and micropsia. Among the non-visual aura disturbances, paraesthesias and speech disturbances were the most frequent ones. The prevalence of migraine with aura seemed to be considerably higher than in similar studies. This also includes studies that have been carried out with a face-to-face interview technique.

Key words

Migraine with aura • Migraine • Scintillating scotoma • Obscuration • Anopsia

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phase comes first and then the visual phenomena [7–9].
Generally, however, it is asserted that although this sequence has been observed, the visual-phenomena-first type is the more frequent pattern [1, 3, 7].

There is, however, at least one prominent exception to these overall concepts. Klee [4], in his studies concerning "severe migraine", maintains that the visual disturbances appear at the pain maximum in the majority of cases. Klee studied 150 migraineurs: 50 extremely carefully ("primary series") and 100 in a somewhat more cursory fashion ("secondary series"). "At least one type of visual symptom" during attacks was found in 76% and 47% of the cases, respectively (Table 6, IV [4]). It is remarkable that in 62% and 31% in the "primary" and "secondary" series, respectively, visual disturbances occurred during the pain phase. Also, Selby and Lance [10] frequently found pain-phase visual phenomena, i.e., in 22 of 59 cases (37%).

Klee’s study concerned "severe migraine". Detailed information is available concerning the aura phase in migraine, in general. However, little is known about the visual disturbance pattern at the grass-roots level. Is the pain maximum/visual disturbance combination less frequent than in "severe migraine"? Klee also described macropsia and micropsia in his "severe" cases, as well as movements of stationary objects in the visual fields (so-called autokinesis) [11]. How frequent would such phenomena be in an unselected, grass-roots population?

These items, as well as the prevalence of migraine with aura per se at the grass-roots level, are the main topics of this communication.

Material and methods

During a two-year period, 1995–1997, every 18–65-year-old inhabitant in the Vågå commune in the mountainous area of southern Norway was invited to participate in this study of headache prevalence. The study comprised 1838 parishioners (F: 942; M: 896); this represents 88.6% of the available individuals [12]. Each parishioner was subjected to a face-to-face interview and a physical/neurological examination, totalling 60–75 min. In problem cases, a new appointment would be allotted. The structured interview was based upon a questionnaire, filled in by the principal investigator, OS. The study was approved by the regional ethics committee and State Data Inspectorate, and all participants signed a participation approval.

The categorisation of the visual phenomena as being migrainous was done at the examination, and – accordingly – it was based upon the IHS criteria in current use at the time [1].

Which specified visual phenomena did the patients experience? A propagating "crescent" of the homonymous type (scintillating scotoma); obscuration only; anopsia (dimness of vision, without "positive" visual phenomena); "unformed flashes of light"/star-shaped figures (or photopsias); changing patterns. Rare visual phenomena were also a topic, such as: macropsia; micropsia; "tunnel vision"; deficiency as regards distance estimation; autokinesis and metamorphopsia. Not all of these questions could invariably be satisfactorily answered in this setting.

The sequence of the events was detailed: which came first, the visual phenomena (and similar phenomena) or the pain? How frequent were visual phenomena without any accompanying pain? The duration of the pain-free interval after the visual phenomena – if any – was roughly estimated. The other aura symptoms were likewise searched for: localised sensibility disturbances, motor weakness and speech disturbances.

We have adopted Klee’s [4] definition of obscuration: during obscuration, the affected individual can just barely see – and identify – some visual field structures. With anopsia, no structures whatsoever can be seen.

Headache intensity was graded according to a 0–6+ system, described elsewhere [13]: 1+="minimal unpleasantness"; 2+="heaviness"/"discomfort"; 3+="mild"; 4+="moderate"; 5+="intense" ("severe") headache and 6+="excruciatingly severe headache". Stages 3–5+ correspond to the IHS stages [1]. Pain per se was not a main item in the present communication.

The figures given herein represent lifetime prevalence. The estimates given reflect what the migraineurs felt was the usual pattern(s). Cases of what we considered as definite migraine with aura were included.

So-called jabs were also searched for, and "CF" or "features indicative of cervical abnormality" was calculated [14].

The exact percentage of absence/presence of visual disturbance during attacks, intraindividually, was not considered an appropriate item in this retrospective study.

Results

Prevalence of the visual disturbances

There were 178 parishioners with visual disturbances, most of them combined with a migrainous pain. Based on the actual figures, the prevalence was 9.7% (178 out of 1838) (Table 1). There were 112 females and 66 males, the gender ratio being 1.70 as against 1.05 in the total Vågå study material. The prevalence among females was 11.9% (112 out of 942) and among males 7.4% (66 out of 896). The latter figures differ statistically (p=0.0012; Fisher’s exact test). The mean age at examination, both sexes, was 43.2 years. Those with scintillating scotoma only constituted 6.1% of the series.

The panorama of major visual phenomena is set forth in Table 1. If the rare and partly bizarre visual disturbances are added (Table 2), there would be a total of 233 visual manifestations. Thus, several parishioners had more than one type of visual disturbance.

Scintillating scotoma experienced in one side of the visual field was almost invariably interpreted by the actual
In 178 migraineurs, a total of 204 visual disturbance patterns were likely to have existed. Other, rare visual disturbances are mentioned in Table 2.

### Table 1 The most commonly occurring visual disturbances

| Type of visual disturbance       | N   | Per cent | F   | M   | F/M ratio |
|----------------------------------|-----|----------|-----|-----|-----------|
| Scintillating scotoma            | 112 | 62       | 70  | 42  | 1.64      |
| Obscurations                      | 58  | 33       | 33  | 25  | 1.32      |
| Photopsia                        | 30  | 16       | 19  | 11  | 1.73      |
| Anopsia                          | 4   | 2        | 2   | 2   | 1         |
| Total                            | 204 |          |     |     |           |

F/M ratio is calculated as follows:

F/M ratio = M / F

### Table 2 Rare and partly bizarre visual disturbances in migraine with aura

| Type of visual disturbance       | N   | Male | Female | Ratio F/M |
|----------------------------------|-----|------|--------|-----------|
| Autokinesia                      | 8   | 5    | 3      | 0.6       |
| Depth vision failure             | 5   | 4    | 1      | 0.25      |
| Tunnel vision                    | 4   | 2    | 2      | 1         |
| “Half” persons/half visual field | 4   | 0    | 4      | –         |
| Micropsia                        | 4   | 2    | 2      | 1         |
| Macropsia                        | 1   | 0    | 1      | –         |
| Metamorphopsia                   | 2   | 2    | 0      | –         |
| Dioplopia                        | 1   | 1    | 0      | –         |
| Total                            | 29  |      |        |           |

F/M ratio is calculated as follows:

F/M ratio = M / F

### Scintillating scotoma, obscurations and photopsias

Scintillating scotoma and obscurations are the two most frequently occurring phenomena. In several cases, the visual disturbance seemed to alternate between scintillating scotomas and obscurations, from one attack to another. Photopsias were present in 30 cases. Deleting photopsias entirely would not alter the situation much, because in every case except eight, photopsias coexisted with scintillating scotomas and/or obscurations, at least occasionally. In two out of the eight individuals that lacked other visual symptoms, there were sensory phenomena during the initial phase of attacks.

Moreover, in those with solitary photopsia, it appeared prior to the pain phase and not at other times.

### Anopsia

Anopsia had been noted by four parishioners. Three of them had at some time experienced scintillating scotoma or obscurations, lasting ≥20 min. In only one of the four cases anopsia was the only visual disturbance, the anopsia lasting 10–15 min; there was no subsequent pain. In this case, the link to migraine as such is probably relatively weak. This case could accordingly have been rejected as a migraine+aura. Anopsia nevertheless coexisted with dysphasia in the latter case.

The anopsia episode appeared at the peak of pain in one of them; generally, there seemed to have been one solitary episode (with one exception). There may be a transition, back and forth, between obscurion and anopsia during the solitary attack. Whether one or the other of these two manifestations is expressed may be a question of degree and not necessarily of nature.
The combination obscuration-anopsia is the reason why anopsia is included in Table 1.

Rare visual disturbances

A number of rare, visual disturbances were present, among them well-known ones, such as micropsia, macropsia and tunnel vision (Table 2). Most of the rare visual phenomena appeared prior to pain, but in one of the cases of tunnel vision and one of the micropsia cases, respectively, these phenomena generally appeared during the pain phase (see later). In all cases, except two cases of micropsia, there were other, concurrent visual disturbances. Autokinesis and failing depth vision were the two “most frequently occurring ones of the rare phenomena” (Table 2). For the sake of completeness, a case of diplopia is also mentioned. This 52-year-old male was fairly certain that the diplopia was additional to the obscuration, and there were multiple episodes within 30 min. At other times, he had fairly typical migraine without aura attacks.

Sensory changes, dysphasia, motor disturbances and other non-visual phenomena

As such phenomena were not infrequently temporally connected with the visual disturbances, they are also detailed. A minor delay between the various components could, of course, not be timed exactly [15].

Paraesthesias – maybe occasionally also with slight sensory deficits – were the most frequent, non-visual phenomena (Table 3). A face/upper extremity location seemed to be the most frequent one (Table 4). The sum of the solitary sensory phenomena exceeded the number of cases by far; thus, in the solitary case, there might be more than one location of the phenomena (Table 4).

The speech disturbance encountered most frequently seemed to be difficulty with finding words and wrong usage of words: usage of words related to, but different from, the desired one, phonetically and/or semantically, i.e., “paraphasias” [16–18]. There may possibly have been dysarthria in one case. Of course, we may have erred in the distinction between dysphasia and dysarthria, but probably not to a large extent.

The rarely noted motor complaints (n=2) generally seemed to appear more or less simultaneously with the visual complaints, typical scintillating scotomas, sensory symptoms and dysphasia. Motor phenomena could apparently also appear repetitively during a given attack (n=1). The motor complaints were mild – face and arm (but no hemiplegia) – and outlasted the visual complaints clearly in the one case, by 30–90 minutes. The last-mentioned, long-lasting episodes occurred only during pregnancy (see Table 3). For this reason, it was not rubricated under hemiplegic migraine. Nor were there any familial cases.

Duration, visual phenomena

The duration of the visual phenomena varied considerably, i.e., from <5 min up to one hour or more. Most frequently, the visual phenomena seemed to last from >15 to ≤35 min (67% of the cases; Fig. 1). In a few cases, the scintillating scotoma could last <5 min. An interesting pattern is no. II (Fig. 2), where multiple, about 2-min-long visual phenomena – “mini-teichopsias” finally might spark a seemingly regular, but particularly severe, migraine attack. When summed up, the total duration of the visual disturbance in these patients might equal the duration of a “regular” tei-
chopsia. In contrast, visual phenomena could last 60–120 min, or longer. In most of the long-lasting ones, there were scintillating scotoma \((n=5)\) or obscuration \((n=3)\). In one of the latter ones, there might also be “half-figures”/“half persons” and paraesthesias (one half of the tongue).

Visual disturbances without a pain phase

This subgroup (Fig. 2, pattern I) is the second largest group of all patterns. The various types of visual disturbances encountered in this subgroup are set forth in Table 5. Apparently, in many of the cases, the picture had not been uniform through the years. In childhood/adolescence, there might have been the more traditional picture, with subsequent headache (which conceivably might have been of a relatively low degree). With age, pain tended to disappear. No exact information exists as regards the frequency of this development.

Scrutiny also showed that among those who initially contended that they had no pain, a minor fraction admitted that they occasionally could have “minimal unpleasantness” (stage I, scale 0–6+, see Methods). They even could have “heaviness/discomfort”, i.e., stage II, the first stage of head pain, and with procrastination. Usually, however, there was “no pain”. It has been a notion that this pattern is most common among men. In the present series, however, there was a female/male ratio of 1.8. If the visual phase only consists of a hard-to-evaluate phenomenon, e.g., micropsia, with no subsequent headache phase at that, then the link to migraine may be at stake.

Visual disturbances at the peak of pain

Visual disturbance at the peak of the pain phase (Table 6) was the third largest group \((n=26, \text{i.e., } 15\%); \text{pattern VIII, Fig. 2.} \) If all those are included that had visual disturbances during the pain phase, but not necessarily at the peak of the pain curve, the number of cases would be 41, i.e., 23\% (Fig. 2). The most frequently encountered visual disturbances were scintillating scotoma and obscuration (Table 6). The duration of visual disturbances varied largely – from “a few minutes” to 0.5–1 h. In three solitary cases, the disturbances could last “for hours”, “to the end of the attack”, “the whole day” (all obscurations).

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**Table 4** Localisation of sensory changes during attack

| Paraesthesias in | No. |
|------------------|-----|
| Face             | 17  |
| Arm              | 14  |
| Finger           | 10  |
| Lip              | 8   |
| Tongue (1/2)     | 6   |
| Total            | 55  |

The sum of solitary phenomena exceeded the no. of cases of paraesthesias \((n=34; \text{Table 3})\), indicating that there could be more than one localisation of paraesthesias in each migraineur.
The array of visual disturbances, both when the pain phase was lacking (Table 5) and when the visual disturbances appeared during the pain phase as such (Table 6), bore considerable resemblance to the panorama of visual disturbances during regular migraine with aura attacks (Table 1).

### Table 5 Types of visual disturbance in those with no ensuing headache

| Type of visual disturbance | N |
|----------------------------|---|
| Scintillating scotoma      | 30|
| Obscuration                | 12|
| Photopsia                  | 3 |
| Anopsia                    | 1 |
| Micropsia                  | 1 |
| Total                      | 47 |

*Pattern I, Figure 2

*There were 42 individuals (24%) in this subgroup

### Table 6 Types of visual disturbances at the peak of pain

| Type of visual disturbance | N |
|----------------------------|---|
| Scintillating scotoma      | 14|
| Obscuration                | 10|
| Photopsia                  | 5 |
| Anopsia                    | 1 |
| Micropsia                  | 1 |
| Tunnel vision              | 1 |
| Total                      | 32 |

*Pattern VIII, Figure 2

*There were 26 individuals in this subgroup

The array of visual disturbances, both when the pain phase was lacking (Table 5) and when the visual disturbances appeared during the pain phase as such (Table 6), bore considerable resemblance to the panorama of visual disturbances during regular migraine with aura attacks (Table 1).

### Interrelationship between duration of visual disturbance phase and the subsequent pain-free interval

As already detailed, there was considerable variation as regards duration of visual phenomena, i.e., from ≤5 min to >1 h (Fig. 1). The symptom-free interval prior to the pain phase also seemed to vary considerably in length. Could the aura and pain-free interval mutually influence each other as regards duration?

Long-lasting visual disturbance has in this context been defined as a disturbance, lasting ≥20 min. “Short interlude” has been defined as no pause or even overlap between visual disturbances and pain. There might be a tendency to an inverse correlation between the duration of the visual phase and pain-free interval (short visual aura phase being linked to a long-lasting pain-free interval and vice versa). It should, nevertheless, be emphasised that although there seemed to be a tendency, there were exceptions to this coarse rule. Also, the interlude may be less important in the patients’ estimation than the impressive visual disturbances and the hard-to-cope-with pain phase. The duration of the interlude may have been more superficially treated by the patients than the two other phases. Moreover, the fact that the attacks, in part, were far back in time, adds further uncertainty. The viewpoint forwarded by the patients on whether or not there really was a pain-free interlude between the visual and pain phases comes into quite another category of likelihood.

### Validation

The migraine with aura part of the questionnaire was validated with two different tests [12]. For both, sufficient time had been allowed to elapse to hinder any memory from the first examination on the part of the principal investigator (OS). Recheck of 100 consecutive records (investigation A), was carried out in a blinded fashion (blinded as to name, sex, family history and diagnoses). Five participants were diagnosed as migraine+aura on examination I and on examination II, rendering a high kappa value.
In the second test (investigation B), 41 parishioners were rechecked in an absolutely blinded way with regard to migraine with aura, on average 14.8 months after examination I. Five participants were diagnosed as migraine+aura cases on examination I. All these five were re-diagnosed as migraine with aura on examination II. However, we were astonished to learn that two other citizens also proved to have had visual disturbances at the time of investigation I – and before that. They did, however, not respond positively to our inquiry about visual disturbances on examination I, but did so on investigation II. The questions asked were the same on both occasions.

Two out of seven patients accordingly failed to notify us: 29%. The kappa value would accordingly be much lower than for the observation on records (A). The second investigation pinpoints the weaknesses of investigation A. As only the records were checked with method A, there may also among these have been individuals who “forgot” to tell about their visual abnormalities. Only what is already in the record can be checked by method B.

One more individual forgot to tell about his visual disturbances during examination A. In this case, we were told about it right after examination A. This made such an impression that there would actually be no test of pattern recognition on investigation B. This patient was accordingly excluded in this context.

Other findings

Jabs were present in 44.9% of the cases. The mean CF value (features indicative of cervical abnormality) was 0.91+; mean Vågå series 0.79+ on a scale 0–5.0+, see Methods). One 18-year-old male noticed red and hot ears bilaterally during attacks (spontaneous information).

Discussion

Prevalence of visual disturbances in migraine

In the present study, there was a high prevalence of migraine with aura. The prevalence of visual disturbances in migraine is, to some extent, influenced by two independent factors:

1. The eagerness/experience of the investigator in uncovering such symptoms.
2. The assessment whether some visual phenomena really are migrainous in nature.

As regards (1), in the validation section of the present study, failure to detect visual disturbances has been described (n=2, out of 41 cases). This certainly can derive from some type of negligence on the part of the investigator.

As regards (2), are, for instance, micropsias as such indicative?/pathognomonic? of migraine? Even dark spots in the visual field [4, 6] (“mouches volantes” or myodesopsia [19]) have in some contexts been considered significant in migraine. Dark spots have been totally disregarded in the present context, even though some migraineurs claim that they only occurred before the attack pain.

Heyck [3] puts it this way: “However, sometimes metamorphopsia and primitive visual hallucinations are considered to be migraine aura”. His statement is open to interpretations. Photopsias belong to another category. The IHS [1, 20] does not use this term, but uses: “star shaped figures”. Both Lance [8] and Klee [4] use photopsia (“flashes of light”/“stars”) as a term for migrainous visual disturbances. In the present work, photopsia has been accepted as being migrainous in nature (see Results). Alvarez [6] (“light spots”) and Peatfield and Rose [21] (“flashes as “circles””) have produced evidence that such phenomena appear in anoptic migraineurs. The IHS [1] also mentions “scotoma without positive phenomena”, in other words pure obscurations. Also, Hachinski et al. [5] found that among 100 migraine children with visual disturbances, 77% had: “transient blindness, blurring of vision, and varied scotomas.”

In the present context, inclusion or not of e.g., photopsias as the sole visual manifestations would not influence the prevalence figures appreciably, due to the small number of cases with photopsia as the solitary visual phenomenon.

In spite of the reservations already made, the figures obtained herein should be regarded as being minimum figures. Three individuals in the control series (B) did not inform us about their visual symptoms. These are hardly exceptional cases; such cases would probably be present in the rest of the series as well.

Lifetime prevalence of visual disturbances in migraine would seem to be around 9.7%. This is a figure considerably higher than in two comparable studies, based on a face-to-face interview technique, i.e., 1.6% [22] and 6% [23], respectively. One of these values is 3.8 times higher than the other, although both were based on IHS criteria, in itself an astonishing fact. In the literature, there is a highly varying prevalence. Such studies have mostly been done by questionnaire technique or telephone interviews, and not by personal contact. They have also partly been performed prior to the advent of the IHS migraine criteria. Moreover, the Vågå population is largely untreated/not regularly treated for the migraine, and this not only concerns specific medication, but also – to a certain extent – non-specific painkillers.

There was a clear female preponderance in the present material: female/male ratio 1.7. This corresponds largely
with Rasmussen’s results from Copenhagen [24], which showed a female/male ratio of 1.67.

Migraine aura has been specifically studied in a meticulous way in several investigations in recent years [e.g., 25, 26]. Many of the facets studied in the present context have been illuminated in such studies. Space does not allow a detailed treatise of all the observations and a comparison with the present observations. There is a striking difference regarding the frequency of migraine with aura, respectively 4.1% [26] and 9.7% (present study). The vast differences in design may, at least partly, explain these differences as regards observations. For further detail, the reader is referred to these communications [25, 26].

Groups of interrelationship between visual disturbances and headache

Visual disturbances during the pain phase were the third most frequent pattern (Fig. 2). These results concur with those of Klee [4] and Selby and Lance [10]. But the frequency was rather less than that reported by Klee [4] (see Introduction). There may be various, alternative explanations for this: Klee’s study [4] was more profound and meticulous in this respect than the present one. Moreover, Klee selected the most “severe” cases [4]. Our series – and other series – were not selected in that way. In other words, he had no cases of aura+/pain−. A more correct approach would, therefore, probably be to compare his cases with our cases of pain+/aura+, i.e., 178 – 43 = 135 cases. Then, a figure for visual disturbances during pain phase would be 30%, a result closer to that of Klee.

Mean intensity of attacks, as assessed in the present series, according to our grading system, 0–6+ [13], was approximately 4.0+, in other words a “moderate” intensity. This figure also comprised those with visual disturbances, but without pain (a total of 24%), so that the average pain in those with a pain phase would be appreciably higher. Klee [4] used a particular terminology to describe the visual phenomena, thus “corona phenomenon”, “flickering”, “unclear vision” and metamorphopsia (distorted contours). These terms may largely correspond to scintillating scotoma, but he does not use the latter term [4]. Differing nomenclature might influence the visual disturbances subgrouping, but not necessarily the grouping of visual phenomena into the “prior to headache”/“during headache” groups. We have no adequate explanation for the difference between the Klee data and our own, as regards the frequency of visual disturbances during the pain phase.

Jabs in migraine with aura

Raskin and Schwartz [27] found a high prevalence of jabs in migraine: 42%. Interestingly, a similar prevalence of jabs was found among migraine plus aura patients in Vågå: 45%. The prevalence of jabs in the entire series was 35.2% [12]. Thus, the prevalence of jabs in migraine plus aura may seem to be somewhat higher than in controls, also in Vågå.

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