Atypical Migraine in Clinical Practice: Are We Missing It?

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Abstract

In countries like India, many migraine patients presenting to primary care clinics fail to fulfill standard (ICHD 3) migraine diagnostic criteria. Since they do not present with typical ICHD 3 migraine diagnostic symptoms, it is necessary to define the criteria for atypical migraine. This would ensure that the patients receive the right treatment approach, both non-pharmacological and pharmacological. Looking for triggers, family history, activity affected and absolute normality in between attacks, past episodes of episodic syndromes, prodromal and oculonasal autonomic symptoms will help in identifying the migraine origin of these headaches.

Keywords: Atypical migraine, ICHD 3, migraine variants, triggers

Introduction

Headache is largely a subjective disorder and hence the diagnostic classification (ICHD 3) designed for it is more helpful in clinical research than in clinical practice. Many patients fail to fit in the existing rigid criteria. The 3rd edition of the International Classification of Headache Disorders (ICHD-3) has classified migraine under Group 1 with sub classification for each migraine subtypes.[1] But in primary care clinical practice, typical attacks either of migraine with aura or without aura are considered as the standard migraine presentation and any variations in presentation may be missed or confusing. Probable migraine diagnosis too can be confusing with typical episodic tension type headaches. As per ICHD 3 recommendation, if a particular patient fulfills two different sets of diagnostic criteria, all other available information should be used to decide which of the alternatives is the correct or more likely diagnosis. This could include, the longitudinal headache history, (how and when did the headache start ?), the family history, the effect of drugs, menstrual relationship, age, gender and a range of other features. Unfortunately, ICHD 3 experts have not mentioned anything about these range of other features.

Variations in the Standard Migraine Presentation

Variations in the standard migraine presentation include but not limited to all the following types of disorders:[1]

- Typical aura without headache
- Migraine with brainstem aura
- Hemiplegic migraine
- Retinal migraine
- Complications of migraine (status migrainosus, persistent aura without infarction, migrainous infarction, migraine aura-triggered seizure)
- Probable migraine
- Episodic syndromes that may be associated with migraine (recurrent gastrointestinal disturbance, cyclical vomiting syndrome, abdominal migraine, benign paroxysmal vertigo and benign paroxysmal torticollis)
- Vestibular migraine (in the Appendix).

Prevalence of Probable/Atypical Migraines From Recent Studies

To estimate the prevalence of migraine in Germany, a cross-sectional telephone-based survey was conducted among adults (N = 5,009) between October 2019 and March 2020. The headache/symptom characteristics were measured using the diagnostic criteria defined in the ICHD. It showed that 14.8% of women and 6.0% of men meet all the diagnostic criteria for migraine. In addition, 13.7% of women and 12.0% of men have probable migraine.[2] This indicates that migraine is not always typical.

Atypical Migraines - Indian Experience

In countries like India, more than 90% of headaches are managed by General practitioners, Consultant physicians, Otorhinolaryngologists, Ophthalmologists, Pediatricians, and Alternative medicine practitioners and many get drugs from the pharmacies without any doctor consultation. These practitioners may not be updated on the ICHD 3 and a large number of them...
are unaware of the existence of this headache classification and diagnostic criteria. Each primary care physician manages 100–200 patients daily out of which 10 to 15% will have headache as one of the symptoms. Overlapping and confusing statements in the diagnostic classification of the most common primary headache syndromes, migraine without aura and Episodic tension-type headaches make it difficult to diagnose headache disorders in such busy practice set up. I would like to share my findings from an observational study (n = 1000) of atypical migraine patients recorded over a period of last 18 years in my clinic at Cherthala, Alleppey, Kerala, India.

NEED FOR DEFINING ATYPICAL MIGRAINE

A migraine diagnosis is made when all the diagnostic features given in ICHD3 are fulfilled. Migraine-like attacks missing one of the features required to fulfill all criteria for a type or sub-type of migraine coded in ICHD, and not fulfilling criteria for another headache disorder are classified as Probable migraine.[1] When the attacks are missing two diagnostic features listed in ICHD-3 or do not include all the typical phases of migraine, then Atypical migraine may be diagnosed. Lesser duration or lesser number of episodes with no typical migraine symptoms (nausea or vomiting or phonophobia and photophobia) may be seen. Some of them get only phonophobia not photophobia. The ICHD3 has not provided any guidance for diagnosis of attacks missing two diagnostic features or do not include all the typical phases of migraine. Even typical migraines are missed in primary care practice and atypical are missed more often. Currently the atypical migraine gets classified under Group 14 (Headaches unspecified or not classified elsewhere).[1] This is not justified as we have many clinical indicators to diagnose migraine in patients with atypical presentations.

LIMITATIONS OF ICHD3

Confusing and overlapping statements- Group 1 and 2 (migraine without aura and Episodic tension-type headache 1.1 and 2.1 and 2.2) have more than one overlapping statements. Similar number of episodes, duration, moderate intensity, phonophobia, photophobia, and mild nausea are also listed in criteria for chronic tension-type headaches. Migraine is classically unilateral and when autonomic symptoms are present, gets confused with Trigeminal Autonomic Cephalalgias (TACs -Group 3).[1,3] As brief migraine episodes are common in the southern part of India, differentiation from TACs becomes very difficult in busy clinical practice.[4]

Well-known Migraine Variants have been omitted in ICHD 3. Alice in Wonderland syndrome (AIWS) is a rare disorder caused by migraine presenting with distorted perceptions of time and space, vision (micropsia/macropsia), hearing, and somesthetic sensations (body feeling bigger).[5] Confusional migraine and Migraine equivalents, though well documented, are not mentioned in ICHD 3. So also, Migraine transformation to a mild-to-moderate tolerable headache in the elderly.

Some Migraine subtypes are listed in the Appendix section of ICHD-3 rather than Group 1 –Migraine Aura status, Visual snow, and Vestibular migraine belong to this category. In Vestibular migraine (sub-type very common in clinical practice), triggers cause vestibular symptoms instead of Headache.[6] Hence can be easily missed.

PROPOSED CRITERIA FOR ATYPICAL MIGRAINE

Migraine-like attacks missing two of the features required to fulfill all criteria for a type of migraine coded in ICHD 3 or migraine attacks without typical phases of migraine with presence of any one of the following-

1. Presence of prodromal symptoms- stiff neck or generalized edema, restlessness, euphoria, aggression, yawning and dysphasia, photophobia, blurring, or difficulty concentrating
2. Presence of autonomic symptoms
3. Recurrent activity affected headache with absolute normality after each episode
4. Common or well-known migraine triggers precipitating headache attacks
5. Family history of migraine, probable migraine or vestibular migraine
6. Sleep aborting recurrent attacks
7. History of episodic syndromes in the past.
8. Like in ICHD3 (not attributed to another disorder), red flags to be ruled out and if present investigated.

CONCLUSION

Diagnosing Atypical migraine might be challenging in a busy practice. These presentations are common in primary care practice in countries like India. Many are treated symptomatically by practitioners without a migraine diagnosis, thus missing an opportunity to approach migraine treatment with non-pharma measures like trigger avoidance and lifestyle modifications. The currently available abortives and preventives are also not utilized to get a better outcome. Going out of the box of ICHD3 by considering, common and well-known migraine triggers precipitating headaches, family history of migraine or vestibular migraine, recurrent activity affected headaches with absolute normality in between attacks, prodromal, and autonomic symptoms, family history of migraine, probable migraine or vestibular migraine, history of episodic syndromes in the past, will help in identifying the migraine origin of these atypical headaches.

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Conflicts of interest
There are no conflicts of interest.

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