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GRBT050-62 Olesen- 2057G GRBT050-Olesen-v6.cls August 3, 2005 19:44

Chapter 62

Basilar Migraines and Retinal Migraines

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Although the second edition of the International Classification of Headache Disorders (1), preserved the basic classification of migraine without aura, the classification of migraine with aura and it subtypes, as well as rarer forms of migraine, were considerably restructured. Given the range of disorders that present with features of migraine, a systematic approach to the less common disorders that are part of the migraine spectrum is essential to good clinical management and to research (2,3). In this chapter we review two of these headaches, basilar-type migraine, a migraine with aura subtype, and retinal migraine, a rarer form of migraine.

BASILAR-TYPE MIGRAINE

IHS Code and Diagnosis

1.2.6 Basilar-type migraine

WHO Code and Diagnosis

G43.103

Short Description

Migraine with aura symptoms clearly originating from the brainstem and/or from both hemispheres simultaneously affected, but no motor weakness.

Previously Used Terms

Basilar migraine
Basilar artery migraine
Bickerstaff migraine

Basilar-type migraine (1.2.6) is a new term, replacing *basilar migraine* (4). The change is intended to remove the implication that the basilar artery (or its territory) is involved (5). The distinguishing feature of basilar-type migraine is a symptom profile that suggests posterior fossa involvement.

Basilar-type migraine was first brought to attention by Bickerstaff (6), after seeing a teenager whose aura symptoms were long lasting, bilateral, and very prominent. Shortly after, the same clinician saw an elderly man, with aura symptoms that rapidly progressed into coma and death. Autopsy showed thrombosis in the basilar artery. After collecting data on 34 patients, Bickerstaff postulated that "some prodromal symptoms . . . are not in the territory of the internal carotid, but of the basilar artery. The premonitory symptoms are different and reflect brainstem dysfunction. Symptoms consist of bilateral loss or disturbance of vision, ataxia, dysarthria, vertigo, tinnitus, and bilateral peripheral dysesthesias, followed by occipital headache. The syndrome is commonest in adolescent girls" (6,7).

Studies on the pathophysiology of basilar migraine are scarce. For years this headache was thought to originate from a transient disturbance in the vertebrobasilar circulation (6–8). Almost two decades ago, neurophysiology studies using electroencephalogram suggested the neurophysiologic basis of basilar-type migraine, by detecting a typical photoconvulsive response (9). Other studies focused on the vascular mechanism of basilar-type migraine. La Spina et al., in 1996 (10), documented a basilar-type migraine attack using transcranial Doppler (TCD), electroencephalography (EEG), and single-photon emission computed tomography (SPECT). In the aura phase, the patient had bilateral blindness and ataxia. Doppler ultrasound revealed reduced mean flow velocity of the posterior cerebral arteries, EEG showed posterior slow activity, and SPECT documented hypoperfusion in the right parietal and occipital regions. During the headache phase with normal neurologic examination, TCD showed an increase in the mean flow velocity of both posterior cerebral arteries and the EEG revealed increased in occipital slow activity. When headache subsided, EEG slow activity resolved and the TCD findings were normal.

P1: KWW/KKL P2: KWW/HCN QC: KWW/FLX T1: KWW
GRBT050-62 Olesen- 2057G GRBT050-Olesen-v6.cls August 3, 2005 19:44

Current migraine theories consider migraine as a state of neuronal hyperexcitability, and that neuronal events mediate both the aura and headache phases of migraine (11–13), although studies specifically investigating basilar-type migraine are still lacking. These findings support transient focal reduction of cerebral blood flow during the aura phase. It remains to be determined if the vascular changes are the cause or, as in other forms of migraine with aura, the consequence of neuronal dysfunction. To date, mutations in *CACNA1A,* encoding a neuronal calcium channel subunit, and *ATP1A2,* encoding a catalytic subunit of a sodium-potassium-ATPase, have not been found in subjects with basilar-type migraine (14).

Classification

In classifying basilar-type migraine, the following criteria are required (1):

A. At least two attacks fulfilling criteria B through E.
B. Fully reversible visual and/or sensory and/or speech aura but no motor weakness.
C. Two or more fully reversible aura symptoms of the following types:
  1. Dysarthria
  2. Vertigo
  3. Tinnitus
  4. Decreased hearing
  5. Double vision
  6. Ataxia
  7. Decreased level of consciousness
  8. Simultaneous bilateral visual symptoms in both the temporal and nasal field of both eyes
  9. Simultaneous bilateral paresthesias
D. Headache that meets criteria B through D for migraine without aura (1.1) begins during the aura or follows aura within 60 minutes
E. Not attributed to another disorder

It is important to note that if distinct motor weakness is present, the diagnosis should be hemiplegic migraine even when "basilar symptoms" are present (hemiplegic migraine sufferers have basilar-type symptoms in 60% of cases) (15).

Clinical Features

Basilar-type migraine affects all age groups with the usual female predominance, but is most common among teenage girls (16,17). A distinguishing feature of basilar-type migraine is the bilateral nature and posterior fossa origin of many of the associated symptoms (16), differentiating it from typical migraine. Visual aura, if present, is usually followed by one of the distinguishing symptoms of this disorder. A hemianopic field disturbance can rapidly expand to involve both visual fields, leading at times to temporary blindness that may precede other symptoms (17). Alterations in consciousness may be present, especially in younger subjects (18). Basilar-type migraine with prominent alteration of consciousness is often called *confusional migraine,* a term not adopted by the second edition of the International Classification of Headache Disorders (1).

Occipital headache appears more frequent in basilar-type migraine than in typical migraine with aura (19). Headache is most frequently throbbing. In one case series, nausea was present in most patients (83%). Two studies showed that alteration of consciousness (45% and 75%), vertigo (41%, 63%), bilateral visual disturbances (48%, 86%), bilateral sensory changes (14%, 61%), and ataxia (17%, 63%) are common symptoms in basilar-type migraine (16,19).

Differential Diagnosis

The diagnosis of basilar-type migraine should be considered in patients with paroxysmal brainstem disturbances. Adolescents with family history seldom require further investigation, but in this age range basilar-type migraine may be difficult to differentiate from complex partial seizures or from postepileptic state. Aura symptoms are often followed by severe, throbbing occipital headache and vomiting. Usually infrequent, they can last for 1 to 3 days. Rarely, attacks may be complicated by cardiac arrhythmias and brainstem stroke (20,21).

The differential diagnosis, besides occipital lobe epilepsy, includes posterior fossa tumor or malformation, urea cycle defects, and mitochondrial disorders (21). When vertigo is prominent, occurring at times alone, other causes of vertigo should be considered, including Ménière disease. If weakness is present, familial or sporadic hemiplegic migraine should be considered.

In basilar-type migraine first presenting in persons over the age of 50, vascular pathologies should be excluded. In such cases, magnetic resonance imaging and angiography should be performed (22).

Treatment

No acceptable "standard of treatment" for migraine prophylaxis currently exists for patients who have basilar-type artery migraine (23). Largely anecdotal, some reports suggest this disorder may be aggravated by treatment with β-blockers to the point of precipitating ischemic strokes (24). Many specialists recommend a calcium-channel blocker such as verapamil; flunarizine (10 mg per day), which is not available in the United States, may be a useful alternative. Antiepileptic drugs (divalproex sodium, topiramate) have also been recommended.

P1: KWW/KKL P2: KWW/HCN QC: KWW/FLX T1: KWW
GRBT050-62 Olesen- 2057G GRBT050-Olesen-v6.cls August 3, 2005 19:44

Acute management must be approached with caution; nonsteroidal anti-inflammatory drugs or combination analgesics should first be tried. Restricting use of triptans in basilar-type and hemiplegic migraine is based on the concept that neurologic symptoms associated with basilar-type migraine may be caused by vasoconstriction, consequently increasing the risk of brain infarction. However, Klapper et al. treated 13 patients with basilar-type migraine or migraine with prominent aura with triptans; all responded positively with no significant adverse effects (25). Even so, triptans cannot be recommended in patients with basilar-type migraine.

RETINAL MIGRAINE

IHS Code and Diagnosis

1.4. Retinal migraine

WHO Code and Diagnosis

G43.81

Short Description

Repeated attacks of monocular visual disturbance, including scintillations, scotomata or blindness, associated with migraine headache.

Previously Used Terms

Ocular migraine
Ophthalmic migraine
Anterior visual pathway migraine

In 1882, the first documented case of a retinal defect associated with migraine was called *ophthalmic megrim* (26); the term *retinal migraine* was first used in 1970 (26). In 1988 the first edition of the International Headache Society established diagnostic criteria for retinal migraine, which required transient unilateral visual loss of less than 60 minutes associated with headache (4).

Retinal migraine is rare; strictly monocular visual phenomena occur in only 1 in 200 migraine sufferers (27). Epidemiologic studies of retinal migraine are unavailable, and extreme caution is required when exclusively diagnosing retinal migraine. Not infrequently, subjects with migraine with aura experience episodes of fully reversible visual symptoms including positive features (e.g., flickering lights, spots, or lines) or negative features (i.e., loss of vision) not associated with pain (28). However, the visual symptoms are homonymous in typical aura without headache (IHS 1.2.3). In retinal migraine, they are strictly unilateral.

A family history of migraine may be present in approximately 25% of probands with retinal migraine (a similar rate is found in other forms of migraine) (27,29). The pathogenesis of this disorder remains to be determined. Neuronal spreading depression has been documented in the retina of chicken and other animals (30,31). Given the monocular nature of retinal migraine, a prechiasmatic origin may be inferred (32). Some authors explain retinal migraine by the involvement of the posterior ciliary vasculature, therefore producing true monocular, rather than homonymous, visual disturbance (32,33). One hypothesis assumes that an imbalance of endothelium-derived relaxing and contracting factors (including peptide endothelin-1 and cyclooxygenase products, such as thromboxane A_2 and prostaglandin H_2) could be important for the development of retinal migraine and retinal infarction (34).

Several reports of retinal vasoconstriction associated with migraine with aura have been published (35–37). A recent study described a patient with retinal migraine as the first symptom of CADASIL (38). Solomon et al. found that a significant proportion of subjects with retinal migraine have subclinical retinal infarction (39). This being the case, instability of retinal neurons might trigger spreading depression. Therefore, the distinction between retinal migraine as a primary disorder or retinal infarction remains to be established.

Classification and Clinical Features

A. The classification of retinal migraine requires at least two attacks fulfilling criteria B and C (1).
B. Fully reversible, monocular, positive and/or negative visual phenomena (e.g., scintillations, scotomata, or blindness) confirmed by examination during an attack or (after proper instruction) by the patient's drawing of a monocular field defect during an attack
C. Headache fulfilling criteria B through D for 1.1 Migraine without aura begins during the visual symptoms or follows them within 60 minutes
D. Normal ophthalmologic examination between attacks
E. Not attributed to another disorder

A new diagnosis of retinal migraine is rarely made on clinical grounds alone. Appropriate investigations are necessary to exclude other causes of transient monocular blindness (see Differential Diagnosis).

Retinal migraine is characterized by repeated attacks of blindness or scotoma lasting less than 1 hour, associated with headache. Patients may experience monocular blackouts, with or without flashes. Patients are usually less than 45 years old and often have a history of other manifestations of migraine headache (27,40). It is the monocular nature of the transient visual deficit that characterizes retinal migraine. The prognosis seems to be the same as migraine

P1: KWW/KKL P2: KWW/HCN QC: KWW/FLX T1: KWW
GRBT050-62 Olesen- 2057G GRBT050-Olesen-v6.cls August 3, 2005 19:44

with aura (41). Recurrent attacks are expected unless the treatment is successful.

Differential Diagnosis

Most importantly, embolism from the carotid artery (transient ischemic attack [TIA]) or other causes of transient monocular blindness must be ruled out by appropriate investigation (32). Both TIA and retinal migraine are characterized by focal deficits that usually last less than 1 hour. Even though there are a number of distinguishing clinical features between typical migraine aura and visual deficits caused by TIAs, these distinctions are blurred in the assessment of retinal migraine. All patients should have a full ophthalmologic, neurological and vascular workup (39,41,42).

The differential diagnosis between retinal migraine and TIA may be more difficult than initially considered. A recent study assessed subjects with a previous diagnosis of retinal migraine. The majority had subclinical retinal infarction or rarely optic nerve ischemia (39). According to the International Headache Society criteria, these patients should be coded as migrainous infarction (1.5.4), a complication of migraine.

Treatment

Almost all patients with retinal migraine should be placed on migraine prevention. Even though large series on the literature are not available, reports on the efficacy of calcium-channel blockers and anticonvulsant agents on the prevention of retinal migraine do exist. Magnesium (400 mg twice a day) and riboflavin (400 mg daily) may be tried (43). Isoproterenol has been reported to alleviate and abbreviate the duration of retinal migraine attacks (44). Although not universally accepted, some specialists recommend that even when no vascular cause is found, prophylaxis with aspirin is indicated because it is effective in both disorders.

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P1: KWW/KKL P2: KWW/HCN QC: KWW/FLX T1: KWW
GRBT050-62 Olesen- 2057G GRBT050-Olesen-v6.cls August 3, 2005 19:44

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