Babinski’s Lost Legacy: A Remarkable Case of a Not-So-New Clinical Construct

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Dear Editor,

Psychiatric disorders during migraine attacks are not uncommon. In fact, as long ago as 1890, Babinski described *De la migraine ophtalmique hystérique* based on four migraine cases with visual aura and hysteria. However, this nosological entity is often overlooked due to the arduous historical task of defining conversion disorder (CD). Nonetheless, migraine associated with CD, or Babinski’s migraine (BM), is a well-documented subset of migraine with aura (MWA), which requires an interdisciplinary approach involving the neurology, psychiatry, and psychology fields. Here we report a noteworthy BM case formed by migraine with brainstem aura (MBA) and CD.

A 45-year-old woman presented in another hospital with a past medical history of pharmacoresistant seizures in her adolescence, after trying two tolerated schedules with carbamazepine and phenytoin. Notwithstanding, she reached a seizure-free status in adulthood despite being off antiepileptic medication. Besides, she also had a family history of MWA (in her mother and sister). The findings of magnetic resonance imaging (MRI) of the brain and neck, blood and cerebrospinal fluid (CSF) studies, and video-electroencephalography (video-EEG) were normal, as were those of cardiac investigations.

She consulted our department for a second medical opinion, complaining of a 1-year clinical presentation of daily, self-limited episodes that were elicited by emotional stress, which had developed gradually and were characterized by recurrent stereotyped, generalized paresthesia, binocular diplopia, and slurred speech. These spells occurred sequentially, leading to altered consciousness about 1 hour before an intense throbbing occipital headache began, with aggravation of the pain by head movement. They occurred in conjunction with changing symptoms such as obtundation, mutism, eyes closed with opening resistance, flexo-extension movements, asynchronous pelvic contortion and kicking, axial and appendicular pseudodystonias, and opisthotonus. Each attack typically lasted 90 minutes, between which she was completely well.

A neurological examination after suggestion (Supplementary Video 1 in the online-only Data Supplement) confirmed the previous observations. The findings of CSF studies, prothrombotic screening, electrocardiography (ECG), Holter ECG, brain MRI and MR angiography, EEG, and video-EEG were normal. A psychiatric evaluation resulted in a diagnosis of MBA (Table 1) added to persistent CD (Table 1) with nonepileptic psychogenic seizures (PNES); that is, BM. She was started on dexketoprofen at 25 mg thrice daily, flunarizine at 5 mg daily (increased to 10 mg per day after 6 months), lorazepam at 1 mg during an acute crisis, fluoxetine at 20 mg daily as maintenance therapy, and psychotherapy. During one of the psychotherapy sessions, the patient reported that she had been sexually abused as a child.

After 5 years of follow-up appointments she achieved a significant reduction of headaches and the frequency, length, and intensity of PNES. Finally, she remained free of attacks. Currently, this situation persists stable.

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Table 1. I: diagnostic criteria for migraine with brainstem aura (code 1.2.2, ICHD-3 beta version, 2013). II: diagnostic criteria for conversion disorder (functional neurological symptom disorder) (code 300.11, DSM-5)*

<table>
<thead>
<tr>
<th>I. MBA</th>
<th>II. CD</th>
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<tr>
<td>Previously used terms</td>
<td>Conversion disorder.</td>
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<tr>
<td>Basilar artery migraine, basilar migraine, basilar-type migraine.</td>
<td>Coding note</td>
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<tr>
<td>Description</td>
<td>The ICD-9-CM code for conversion disorder is 300.11, which is assigned regardless of the symptom type. The ICD-10-CM code depends on the symptom type (see below).</td>
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<tr>
<td>Migraine with aura symptoms clearly originating from the brainstem, but no motor weakness.</td>
<td>Diagnostic criteria</td>
</tr>
<tr>
<td>A. At least two attacks fulfilling criteria B–D.</td>
<td>A. One or more symptoms of altered voluntary motor or sensory function.</td>
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<td>B. Aura consisting of visual, sensory and/or speech/language symptoms, each fully reversible, but no motor or retinal symptoms.</td>
<td>B. Clinical findings provide evidence of incompatibility between the symptom and recognized neurological or medical conditions.</td>
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<tr>
<td>C. At least two of the following brainstem symptoms</td>
<td>C. The symptom or deficit is not better explained by another medical or mental disorder.</td>
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<tr>
<td>1. Dysarthria.</td>
<td>D. The symptom or deficit causes clinically significant distress or impairment in social, occupational, or other important areas of functioning or warrants medical evaluation.</td>
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<td>2. Vertigo.</td>
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<td>3. Tinnitus.</td>
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<td>4. Hypacusis.</td>
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<td>5. Diplopia.</td>
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<td>6. Ataxia.</td>
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<td>7. Decreased level of consciousness.</td>
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<td>D. At least two of the following four characteristics</td>
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<td>1. At least one aura symptom spreads gradually over ≥5 minutes, and/or two or more symptoms occur in succession.</td>
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<td>2. Each individual aura symptom lasts 5–60 minutes.</td>
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<td>3. At least one aura symptom is unilateral.</td>
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<td>4. The aura is accompanied, or followed within 60 minutes, by headache.</td>
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<tr>
<td>E. Not better accounted for by another ICHD-3 beta diagnosis, and transient ischaemic attack has been excluded.</td>
<td></td>
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Notes

1. When motor symptoms are present, code as 1.2.3 Hemiplegic migraine. (F44.4) With weakness or paralysis. (F44.4) With abnormal movement (e.g., tremor, dystonic movement, myoclonus, gait disorder). (F44.4) With swallowing symptoms. (F44.4) With speech symptom (e.g., dysphonia, slurred speech). (F44.5) With attacks or seizures. (F44.6) With anesthesia or sensory loss. (F44.6) With special sensory symptom (e.g., visual, olfactory, or hearing disturbance). (F44.7) With mixed symptoms.

2. When for example three symptoms occur during an aura, the acceptable maximal duration is 3×60 minutes.

3. Aphasia is always regarded as a unilateral symptom; dysarthria may or may not be.

Comments

Specify symptom type

Specify if

Originally the terms basilar artery migraine or basilar migraine were used but, as involvement of the basilar artery is unlikely, the term migraine with brainstem aura is preferred. Acute episode: symptoms present for less than 6 months. Persistent: symptoms occurring for 6 months or more. Without psychological stressor.

There are typical aura symptoms in addition to the brainstem symptoms during most attacks. Many patients who have attacks with brainstem aura also report other attacks with typical aura and should be coded for both 1.2.1 Migraine with typical aura and 1.2.2 Migraine with brainstem aura. With psychological stressor (specify stressor).

Many of the symptoms listed under criterion C may occur with anxiety and hyperventilation, and therefore are subject to misinterpretation.

MBA is a rare and underdiagnosed subtype of MWA. These patients suffer from a constellation of negative and/or positive signs and symptoms (excluding motor weakness) that are referable to the brainstem or posterior circulation territory. This clinical scenario can be challenging, and raise differential diagnosis with a host of conditions including complex partial seizures, multiple sclerosis, vertebrobasilar transient ischemic attacks, posterior fossa tumors, and encephalitis.3,5,6 Furthermore, CD constitutes a scarcely known pathology that is shared between psychiatry and neurology. PNES can be found as one of the symptoms accompanying CD. First described by Charcot in 1892 under the diagnosis of hysteroepilepsy, still nowadays PNES are a diagnostic, psychopathological, and psychotherapeutic problem.7 Moreover, a PNES diagnosis is eventually obtained in 25–30% of patients diagnosed with intractable epilepsy.8 These cases warrant a thorough evaluation including MRI and video-EEG. Some common PNES findings are long episodes, predominance among women, young age at onset (usually between 10 and 35 years), and preservation of ictal consciousness. The motor activity is not consistent with a recognizable epileptic pattern, and consciousness is recovered immediately once the episode has finished.8,9 A key aspect of the paradigm underlying BM is that the conversive response is structured in a dramatic constellation of corporeality symptoms and produced by frustration, with BM appearing when the consciousness is impaired. This phenomenon may be caused by involvement of the brainstem reticular formation and/or deficient brain integration of emotion processing, executive control, and motor networks. In addition, pain in the form of headache as well as hypnosis would act as elicitation factors that decrease consciousness to reach the hysterical threshold.1-9

In conclusion, we suggest including BM as a diagnostic subcategory within MBA. Rescuing this Babinski’s legacy that has been omitted for decades will help in designing better diagnostic approaches and applying optimal therapies that can improve the quality of life of these patients.

**Supplementary Video Legend**

Diagnosing postsuggestion PNES. The suggestion process using hypnosis consisted of the following steps:
1. The patient signed a consent form.
2. Her sister was present so that we could confirm whether the attack triggered by suggestion was similar to any of the consultation episodes.
3. The patient was cautioned about the likelihood of suffering a new spell during the test.
4. In a relaxed atmosphere the patient was requested to maximally concentrate while her glabella was gently beaten with a reflex hammer. This intervention induced an episode within 5–10 minutes.
5. The episode was terminated by combining suggestion with pressing the thumb over the glabella area.

**Supplementary Materials**
The online-only Data Supplement is available with this article at http://dx.doi.org/10.3988/jcn.2016

**Conflicts of Interest**
The authors have no financial conflicts of interest.

**REFERENCES**