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Pure epileptic headache and related manifestations: a video-EEG report and discussion of terminology

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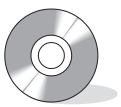
ABSTRACT – We present the first video-EEG recording of episodes of "*epileptic headache*". The case reported is that of a 9-year-old girl with brief episodes (of a few minutes) of severe frontal headache, which corresponded to the presence of concurrent spikes and slow waves, starting in the right temporal area. A dysplastic lesion of the right temporal lobe was observed by MRI and the patient received surgery, with subsequent disappearance of headaches. This case highlights ictal EEG as the main diagnostic tool for epileptic headache. We discuss the terminology regarding this type of manifestation and believe that cases without subsequent epileptic manifestations, as in the present case, should be more appropriately referred to as "*pure ictal epileptic headache*" or simply "*pure epileptic headache*". [*Published with video sequences*]

Key words: epilepsy, headache, migraine, seizure, pain, video-EEG

The terms "*epileptic headache*" (Nymgard, 1956; Grossman *et al*., 1971) and "*ictal headache*" (Laplante *et al*., 1983) were used in the first reports of the rare condition of headache occurring as the sole (or strongly prevailing) manifestation of an epileptic seizure. The term "*ictal epileptic headache*" (IEH) has recently been proposed (Parisi, 2009; Belcastro *et al*., 2011), although the use of the word "*ictal*" may be unnecessary since an epileptic manifestation is ictal *per se*.

Epilepsy and headaches, migraines in particular, coexist in many patients and occur independently. As a rule, migraine is not an epileptic phenomenon, however, headache can follow an epileptic seizure (postictal headache).

The distinguishing criteria for epileptic headache is the contemporaneous onset and cessation of headache with epileptic abnormalities on the EEG. Such headaches therefore appear to be a specific mode of expression of an epileptic seizure. During an episode of epileptic headache, head pain may be accompanied by minor symptoms and vigilance is at least partially



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preserved (the patient would not otherwise be aware of having a headache). Symptoms that indicate an epileptic manifestation, particularly motor symptoms, are absent. However, the term "*epileptic headache*" usually includes cases in which episodes of ictal head pain alone alternate with episodes in which head pain is followed by a clear epileptic manifestation, particularly a motor manifestation (Laplante *et al*., 1983; Parisi *et al*., 2007; Perucca *et al*., 2010; Dainese *et al*., 2011). Epileptic headache appears to be a rare condition, although probably under-reported in the past, since half of the cases have been reported in the last 3 years (all of which were in Italy).

Here, we present the case of a girl with epileptic headaches including the first reported video-EEG recording of attacks.

Case report

A 9-year-old girl was admitted to our clinic due to the presence of headaches with fronto-orbital pressure. The headaches had started about five months earlier with episodes of severe intensity, lasting for no more than 2-3 minutes, and without accompanying symptoms except for occasional hypersensitivity to noise. Immediately preceding hospitalisation, episodes had become much more frequent (up to 10 a day) and were accompanied by phonophobia. The girl was right-handed and neurological examination was normal. During a video-EEG recording, she complained of a sudden, rapidly increasing severe headache with frontal pain, associated with what she subsequently described as "*loud noises*" in her head, initially misinterpreted as auditory hallucinations. She was conscious and could understand the physician's questions, but she did not answer in order to avoid the increase in head pain due to her hypersensitivity to noise (*video sequence 1*). Concurrent with the headache, the EEG showed an onset of spikes and high-amplitude slow waves beginning in the right temporal area and diffusing bilaterally after a few seconds (*figure 1 and video sequence 1*). The headache lasted for less than two minutes and as the pain ceased, the EEG became normal and the girl responded immediately. No EEG anomalies were present outside the headache episode; however, in the following weeks, rare slow and sharp waves began to appear in the right temporal area. Other episodes with the same characteristics were recorded in the following days. In *video sequence 2*, we present a slightly different seizure, recorded 18 months later, characterised by reduction of mild postictal strength on the left side.

The presence of a dysplastic lesion in the right temporal cortex, anterior to the amygdala, was observed by 3-tesla MRI (*figure 2*).

The attacks subsided with carbamazepine at 800 mg/d and clobazam at 10 mg/d. However, the attacks later returned and 200 mg topiramate was added, providing relief but with the adverse effect of word finding difficulties which regressed by reducing the dose to 100 mg. Due to progressive worsening, along with drug resistance, an anterior temporal lobectomy was performed. Histological diagnosis demonstrated dysplasia type IIa. At eight months of follow-up, no further episodes were reported (post-operative outcome class 1A, according to Engel *et al*. [1993]).

Discussion

Our case was characterised by:

– brief duration (of no more than three minutes) of headache attacks;

– minimal accompanying symptoms;

– focal onset with spikes and slow waves followed by diffusion;

– initially normal interictal EEG;

– and absence of other seizure types.

Attacks responded temporarily to the antiepileptic drugs carbamazepine and clobazam. Topiramate, a drug effective against both epilepsy and migraine, was more effective, although this treatment remained insufficient.

The exclusive manifestations of headache along with normal initial interictal EEG were the cause of delay in diagnosis until the occurrence of an attack during the EEG recording. A variable delay, sometimes of years, has also been reported to occur in other cases (Laplante *et al*., 1983; Belcastro *et al*., 2011; Dainese *et al*., 2011; Fanella *et al*., 2012).

The paroxysmal EEG activity coinciding with onset and cessation of the headache indicated that the headache itself was a manifestation of an epileptic seizure, justifying the diagnosis of "*epileptic headache*".

A comparison of our case with the other cases of epileptic headache more recently reported in the literature is shown in *table 1*. The cases of Isler *et al*. (1987) and Beauvais *et al*. (2001) are not shown due to a considerable lack of data in the original reports. Similarly, other earlier cases were reported but are not included due to a lack of sufficient data (Heyck and Hess, 1955; Nymgard, 1956; Morocutti and Vizioli, 1957; Grossman *et al*., 1971). From the data shown in *table 1*, we can derive the following general characteristics of epileptic headache:

1) The headache may appear as a migraine without aura, according to the criteria of the International Classification of Headache Disorders (ICHD)-II (2004) (Parisi *et al*., 2007; Perucca *et al*., 2010; Belcastro *et al*., 2011), an unspecified headache (Laplante *et al*., 1983; Ghofrani *et al*., 2006; Dainese *et al*., 2011), or a

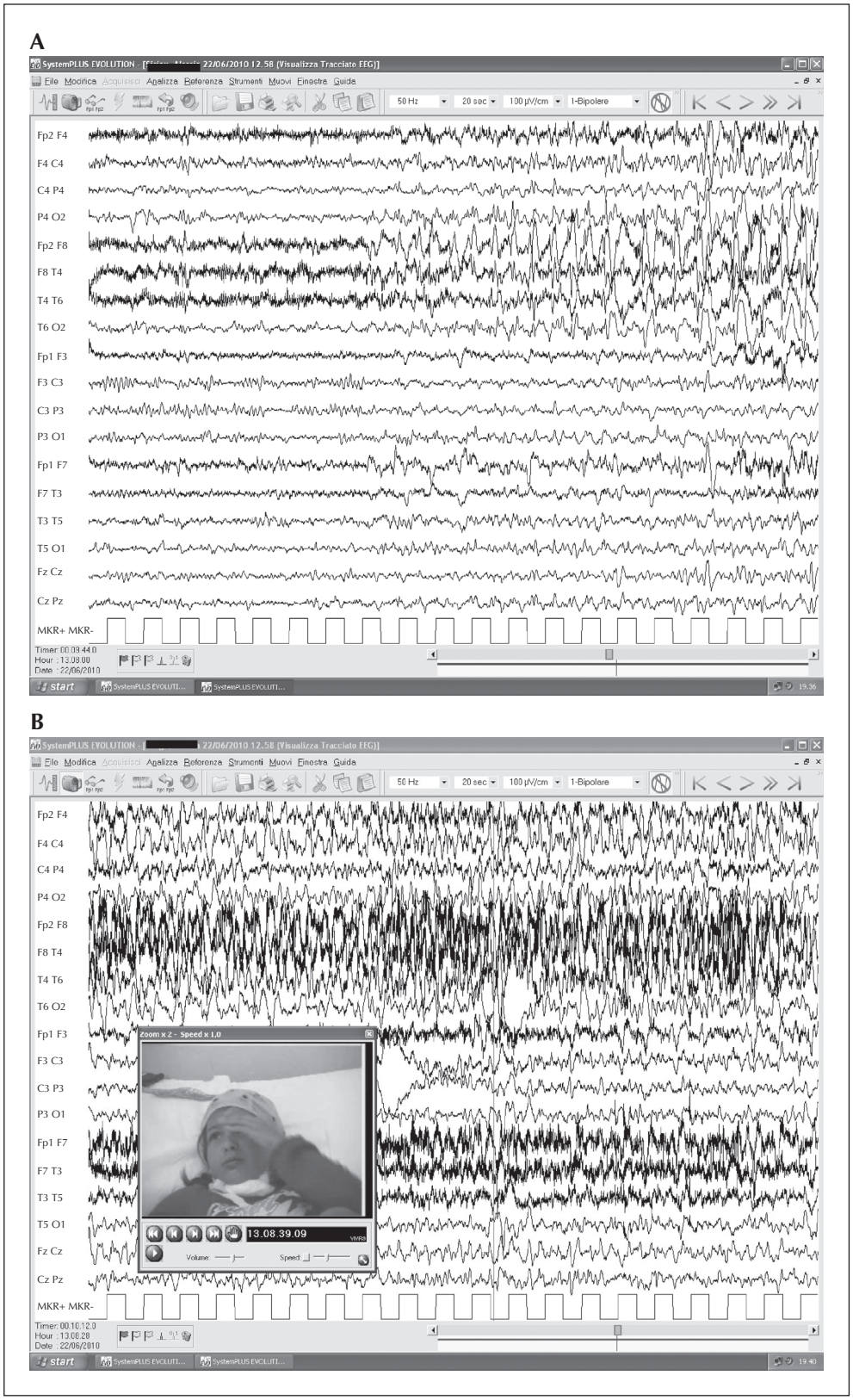

Figure 1. A) Paroxysmal activity in the right temporal area coincides with the onset of headache (time: 13.08.17). B) The patient indicates her forehead as the site of pain (time: 13.08.39). MKR: 100 μV, 1 second.

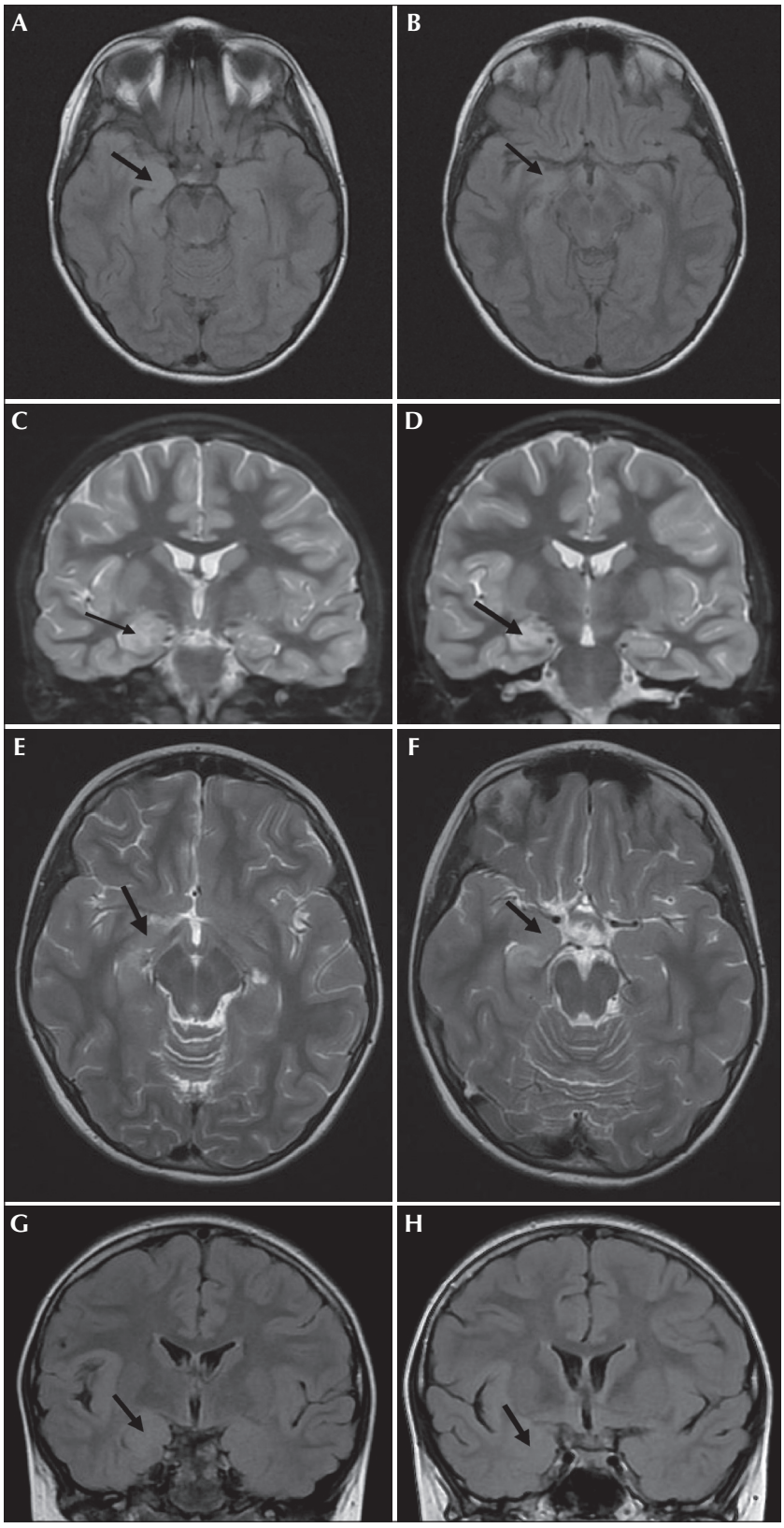

Figure 2. Focal cortical dysplasia of the right temporal cortex. Axial T2 FLAIR (A and B) and coronal T2 STIR (C and D) MRI show the dysplastic lesion (arrows) in the right temporal-mesial cortex. Axial T2 FSE (E and F) shows an increase of signal and mild tumefaction of the amygdala (arrows) and of the contiguous hippocampus. Coronal FLAIR (G and H) illustrates more clearly a blurring of the grey-white matter junction at the right temporal pole and an increase of size and signal of the amygdala (arrows).

Table 1. Main features of 10 cases of epileptic headache reported in the literature.

	Belcastro *et al.*, 2011	**Dainese *et al.*, 2011 Case 1**	**Dainese *et al.*, 2011 Case 2**	**Fanella *et al.*, 2012**	**Fusco *et al.*, 2011**	**Ghofrani *et al.*, 2006**	**Laplante *et al.*, 1983 Case 1**	**Laplante *et al.*, 1983 Case 2**	**Parisi *et al.*, 2007**	**Perucca *et al.*, 2010**	**Our case**
Age/sex	20/F	11/M	47/F	37/F	18/M	9/M	17/F	28/M	14/F	56/F	9/F
Headache characteristics	MO	left side and vertex	right frontal	tension type	MO	bilateral,	vertex	temporal	MO	1) right MA; 2) left MO	frontal
Duration	3 days	seconds	few seconds	1 hour	hours - 1-2 days	More than 24 hours	1 minute	30-60 sec.	3 days	1) <1 minute 2) hours (status)	3 minutes
Symptoms accompanying headache	phono-photophobia, nausea, vomiting	nr	nr	no	phono-photophobia; sometimes nausea, vomiting	agitation, irritability, crying, moaning	dyspnoea, headedness, sometimes could not talk	nr	phono-photophobia, nausea, vomiting	no	hyper-sensitivity to noise
Manifestations following headache	no	sometimes visual hallucinations, oculoversion, loss of contact	sometimes right arm tonic posture	no	no	no	sometimes automatisms, confusion, agitation	sometimes chewing, head-version, agitation, rarely generalised seizure	tonic left arm, head version, then GTC	1) sometimes head deviation; 2) no	no
Ictal EEG	right occipital rhythmic 11-12-Hz spikes	scalp EEG normal/ stereo-EEG spikes right occipito-parietal	left central spikes	subcontinuous GSW and GPSW	Seizure A: right fronto-central spikes; Seizure B: right occipital	continuous GSW	scalp: bitemporal/deep: right hippocampus and amygdala	scalp: normal or 3-4 Hz or rapid/deep: spikes at right hippocampus	right occipital theta and sharp waves	right temporal-occipital rhythmic 11-12-Hz spikes, then slow waves	right temporal spikes and slow waves, then diffuse

Table 1. (*Continued*)

	Belcastro *et al.*, 2011	**Dainese *et al.*, 2011 Case 1**	**Dainese *et al.*, 2011 Case 2**	**Fanella *et al.*, 2012**	**Fusco *et al.*, 2011**	**Ghofrani *et al.*, 2006**	**Laplante *et al.*, 1983 Case 1**	**Laplante *et al.*, 1983 Case 2**	**Parisi *et al.*, 2007**	**Perucca *et al.*, 2010**	**Our case**
MRI	right parieto-occipital scar / DWI: right occipital diffusion	right occipital (lingual) dysplasia	left inferior parietal tuber	normal	partial hemispherectomy; ictal SPECT Seizure B: right occipital hyperperfusion	brain atrophy	CT normal	nr	normal	bilateral occipital cortex swelling, hyperintensity	right temporal cortex dysplasia
Aetiology	post-trauma	malformation	tuberous sclerosis	idiopathic generalised epilepsy	Rasmussen's encephalitis	(histiocytosis)	temporal focal malformation?	early encephalopathy	occipital epilepsy Gastaut-type	perinatal hypoxic-ischaemic encephalopathy	dysplasia type IIa

CT: computerized tomography; DWI: diffusion weighted MRI; EEG: electroencephalogram; F: female; GPSW: generalised polyspikes and waves; GSW: generalised spikes and waves; GTC: generalised tonic-clonic; IPS: intermittent photic stimulation; M: male; MA: migraine with aura; MO: migraine without aura; MRI: magnetic resonance imaging; nr: not reported; SPECT: single-photon emission computed tomography.

tension-type headache (Fanella *et al*., 2012), not accompanied or followed by other epileptic manifestations. The head pain described appears to be distinct from somato-sensitive pain in the head area, as reported, amongst others, by Young and Blume (1983), Young *et al*. (1986), Siegel *et al*. (1999), and Charlesworth *et al*. (2009), since it is not unilateral or opposite to the epileptic focus, localised in the parietal lobe. However, Case 2 of the series of Dainese *et al*. (2011) might be included in this condition;

2) The duration of headache attacks varies considerably, lasting from seconds to days. Brief duration was described in the cases of Laplante *et al*. (1983) and Dainese *et al*. (2011), as well as in our case. A long-duration variant (more similar to migraine attack or tension-type headache) is considered as status epilepticus (Ghofrani *et al*., 2006; Parisi *et al*., 2007; Perucca *et al*., 2010; Belcastro *et al*., 2011);

3) Sometimes minor accompanying symptoms are present, some of which are the characteristic accompanying symptoms of migraine (Parisi *et al*., 2007; Belcastro *et al*., 2011; Fusco *et al*., 2011), and others which are different (Case 1 of Laplante *et al*., 1983; Ghofrani *et al*., 2006);

4) In some patients, the epileptic headache attacks are not always isolated and on some occasions are immediately followed by other epileptic manifestations, usually motor (Laplante *et al*., 1983; Parisi *et al*., 2007; Perucca *et al*., 2010; Dainese *et al*., 2011);

5) Concerning the EEG ictal pattern, different onset sites have been identified: in five cases (Parisi *et al*., 2007; Perucca *et al*., 2010; Belcastro *et al*., 2011; Case 1 of Dainese *et al*., 2011; Fusco *et al*., 2011 [second episode]), paroxysmal EEG occurred with right occipital activity; in two cases (Ghofrani *et al*., 2006; Fanella *et al*., 2012), the paroxysmal activity was generalised; in both cases of Laplante *et al*. (1983), depth electrode studies showed paroxysmal activity in the right hippocampus and amygdala (scalp EEG showed bitemporal activity in Case 1 and inconstant anomalies in Case 2); and in our case, the seizure began in the right temporal area and subsequently diffused. This variability of onset site thus appears to be characteristic of the reported population of epileptic headache and may be due to the multiple pain representation sites in the brain (Talbot *et al*., 1991; Lötsch *et al*., 2012).

6) There is considerable variability of aetiology and neuroimaging findings.

The cases reported in *table 1* presented with headache attacks not accompanied or followed by other epileptic manifestations, except occasionally in some cases, as stated in (4). In other cases, headache may be the first manifestation of an epileptic seizure, which subsequently develops with more typical manifestations (motor, sensory or autonomic) and is therefore an "*epileptic headache*". This combination may occur in occipital epilepsy (Panayiotopoulos, 1999; Panayiotopoulos *et al*., 2008), as well as in other reported cases (*e.g*. Isler *et al*., 1987; Marks and Ehrenberg, 1993; Velioglu and Ozmenoglu, 1999), in addition to those cited in (4) above. This pattern, however, does not appear frequently as is evident from the epidemiological data of the Help Study Group (2010): only 1.5% cases of "*headache… just before the onset of overt seizure or simultaneously with the other seizure manifestations*" were reported (other epidemiological studies have failed to provide more relevant data).

Thus, in order to clarify the clinical picture, for the cases reported in *table 1* and including our case, it would appear appropriate to apply the more specific definition of "*pure epileptic headache*" or "*isolated epileptic headache*".

A recent debate addressed seizure-related headaches with regards to terminology and pathophysiological mechanisms. The only international classification that defines seizure-related headaches and related manifestations is the ICHD-II (International Headache Society, 2004), since the International League Against Epilepsy classification does not include any definition. Aside from the postictal headache (7.6.2), the ICHD-II reported two denominations, "*migraine-triggered seizure*" and "*hemicrania epileptica*", described as follows:

1) "*Migraine-triggered seizure*" (1.5.5): "*A. Migraine fulfilling criteria for 1.2 Migraine with aura. B. A seizure fulfilling diagnostic criteria for one type of epileptic attack occurs during or within one hour after a migraine aura*". Furthermore, "*this phenomenon (is) sometimes referred to as migralepsy*";

2) "*Hemicrania epileptica*" (7.6.1): "*A. Headache lasting seconds to minutes, with features of migraine, fulfilling criteria C and D. B. The patient is having a partial epileptic seizure. C. Headache develops synchronously with the seizure and is ipsilateral to the ictal discharge. D. Headache resolves immediately after the seizure. Diagnosis requires the simultaneous onset of headache with EEG-demonstrated ictal discharge*".

The definition according to the ICHD-II is not consistent in any way with the kind of manifestations reported in *table 1*. However, "*hemicrania epileptica*" (ICHD-II 7.6.1) appears to be a true epileptic headache, followed by other epileptic manifestations (therefore excluding "*pure*" epileptic headache). Cases of "*hemicrania epileptica*" were only reported by Isler *et al*. (1987), who introduced this term. However, there were no further cases described in the subsequent 25 years and we therefore believe this term should be abandoned and substituted by "*epileptic headache*".

In conclusion, "*epileptic headache*" appears to be an appropriate term to clearly define the phenomenon of an epileptic (EEG-confirmed) manifestation, clinically represented by headache. All of the features discussed here may be included under the term of "*epileptic headache*"; however, if we use the term in a broader sense, it would be more appropriate to define the specific manifestations of the cases described in *table 1* as "*pure epileptic headache*" or "*isolated epileptic headache*", as they are not accompanied or followed by other seizure types.

Legends for video sequences

Video-EEG recordings of two seizures (dated 22/06/2010 and 15/12/2011). In both, the patient presses a button (causing a beep, audible on the recording) at the onset of pain. The conversation with the staff is in Italian; the translation is written below.

Video sequence 1

The patient indicates the onset of headache by pressing the button. Paroxysmal activity appears in the right temporal area, rapidly increases in amplitude, and spreads homo- and contralaterally. The patient responds to the questions.

Video sequence 2

Similar pattern recorded about one year later.

Key words for video research on www.epilepticdisorders.com
Syndrome: focal non-idiopathic temporal (TLE)
Etiology: focal cortical dysplasia (type ii)
Phenomenology: headache
Localization: temporal lobe (right)

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