

# Giant hypothalamic hamartoma and dacrystic seizures

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**ABSTRACT** – Ictal crying is a rare type of epileptic seizure associated with hypothalamic hamartoma and with other lesions such as tumours, vascular malformations, hippocampal sclerosis, or cerebral infarction. We describe the case of an infant with gelastic, dacrystic and other types of seizures associated with a giant hypothalamic hamartoma, and present a video sequence of dacrystic seizures. Dacrystic episodes presented in clusters at sleep onset, initially in the form of moaning followed by face-flushing that rapidly evolved to crying, associated with a lateral and upper deviation of both eyeballs, along with clonic aspects of the eyelids. After a few seconds, the crying became less intense, she stared, and oro-alimentary automatisms became prominent along with some slow horizontal movements of the eyes and the head. Following surgery, at the age of nine months, the gelastic seizures stopped, but dacrystic seizures persisted.

[Published with video sequences]

**Key words:** hypothalamic hamartoma, dacrystic seizures, ictal crying, epilepsy

Ictal crying (dacrystic seizures) is a rare, paroxysmal phenomenon usually reported in association with hypothalamic hamartoma (HH) as well as other lesions such as tumours, vascular malformations, hippocampal sclerosis, or cerebral infarction (Wang *et al.* 1995). Crying can occur as an unconscious ictal or postictal phenomenon. Ictal crying can also occur with preserved consciousness, either as an automatic behaviour without associated affect or as part of a depressive reaction (Luciano *et al.* 1993). In the majority of previously reported cases of crying seizures, ictal activity was most prominent in the temporal lobe of the non-dominant hemisphere (Luciano *et al.* 1993). Although da-

crystic seizures have long been related to HH and gelastic epilepsy, there are few reports regarding this association (Williams *et al.* 1978, Kahane *et al.* 1994). We report the case of an infant suffering from dacrystic seizures associated with a giant HH.

## Case report

A newborn female presented, on her second day of life with a clonic seizure of the upper limbs and oro-alimentary automatisms lasting approximately one minute. She was born at term after a normal pregnancy with delivery by Cesarean section because of cranio-pelvic disproportion. Family history was unremarkable for epilepsy



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This case was presented at the II Meeting of the Spanish and Portuguese Pediatric Neurology Associations, Estoril (Portugal), 2005 and was published in an abstract form in *Rev Neurol* 2005; 41 (Separata): 23

or neurological disease. A cranial ultrasound revealed a lesion on the diencephalon. Brain MRI showed a giant, diencephalic lesion which was isointense to mildly hyperintense to grey matter, measuring four centimetres of diameter, with posterior displacement of the brainstem (*figure 1*). No gadolinium enhancement was observed. No other malformations were present. Her EEG was normal. She received treatment with phenobarbitone.

Four weeks later she presented with a history of recurrent episodes of prolonged irritability, agitation, crying, and polypnea. She also experienced episodes described as an arrest of activity, upper or lateral deviation of the eyes and generalized tonic contraction. Ictal scalp-EEG was normal except for one occasion following one of the seizures, when it showed mild slowness of the background activity due to excess of diffuse high amplitude slow waves. Sodium valproate was added to her treatment.

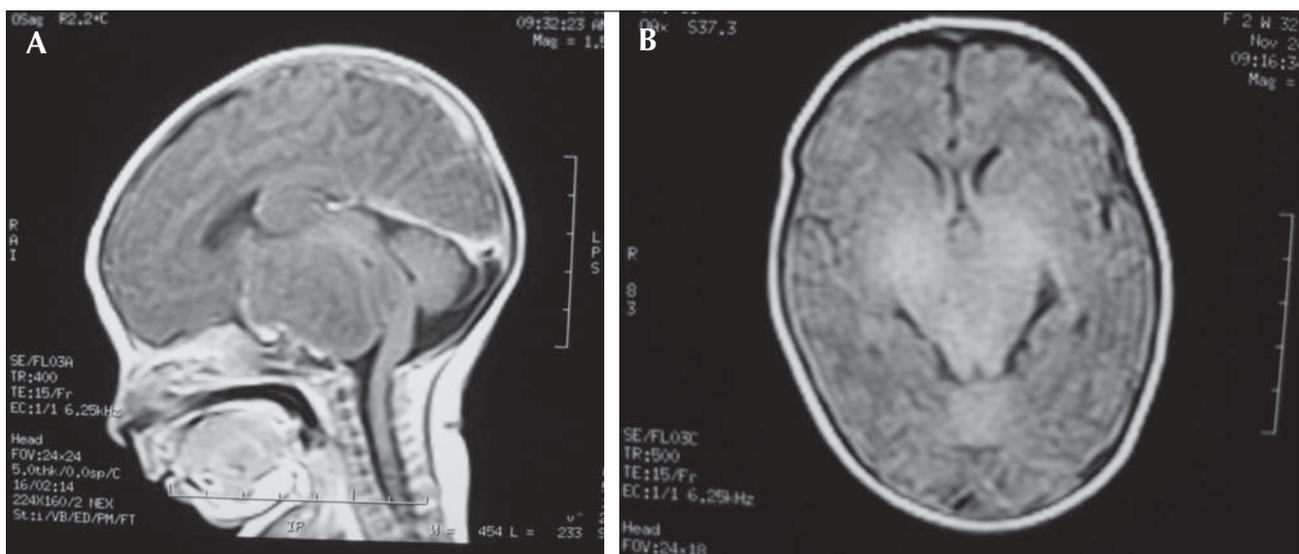
During the next few months she continued to suffer five to more than 30 daily seizures, with periods when seizures occurred as frequently as every ten minutes. Many of the seizures she presented were gelastic, with an occasional smile or mirthless laughter mixed with crying. During some of the episodes she also presented upper or lateral deviation of the eyes, unresponsiveness, staring and mild myoclonic elements of both eyelids suggesting complex partial seizures. She received treatment with topiramate, and phenobarbitone was discontinued.

From the age of five months, some degree of mental impairment became evident, she was unable to follow objects properly and she had poor interaction with her surroundings. Failure to thrive was noted and a hormone profile showed a mild elevation of LH and prolactin. She also presented several dacrytic seizures daily, all of them

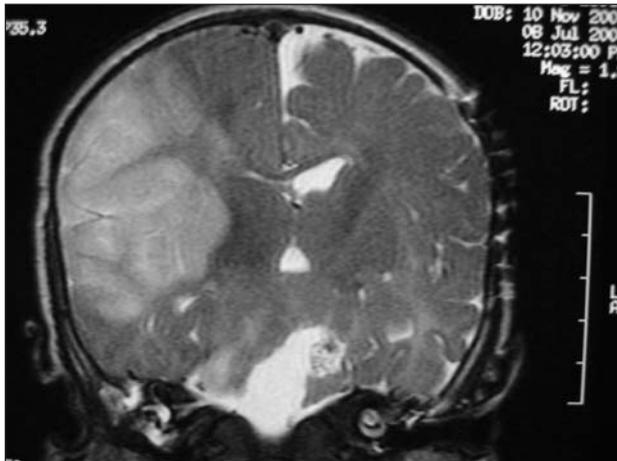
stereotyped with crying as the most noticeable element of the episode (*see video sequence*). These episodes presented as clusters at sleep onset. A typical episode while falling asleep would start with moaning followed by face-flushing. The moaning then evolved to crying with lateral and upper deviation of both eyeballs, along with clonic elements of the eyelids, which were more evident on the left side. After a few seconds the crying became less intense, she stared and oroalimentary automatisms became prominent along with some slow horizontal movements of the eyes and the head. At the end of an episode she went back to sleep. Administration of rectal diazepam did not modify the clinical presentation or the duration of the paroxysmal attack.

At the age of nine months she underwent mass resection through a subfrontal route. Surgical intervention was complicated by a subarachnoid hemorrhage secondary to the accidental section of a branch of the internal carotid artery. She developed acute left hemiparesis and a transient paresis of the third cranial nerve. A post-operative MRI scan (*figure 2*) revealed an ischemic stroke in the territory of the right middle cerebral artery. Partial resection of the tumor was achieved. Histopathological analysis of the resected tissue showed features typical of hypothalamic hamartoma.

Following surgery, the seizure frequency ranged from six episodes per hour to five seizures in 24 hours, most of them dacrytic in type. These crying seizures remained relatively stereotyped. Notwithstanding, the gelastic seizures stopped. She also developed hydrocephalus, probably as a consequence of the subarachnoid hemorrhage, that necessitated a ventriculo-peritoneal shunt. At the age



**Figure 1.** Sagittal (A) and axial (B) T1-weighted MRI sequence showing a giant hypothalamic hamartoma within the third ventricle, with a large interpeduncular component.



**Figure 2.** Coronal T2-weighted MRI sequence at the level of the third ventricle performed a few days after surgery. The hypothalamic hamartoma has been partially resected. There is a cerebral infarction in the territory of the right middle cerebral artery which affects both the cortex and underlying white matter with a mass effect of the infarcted territory.

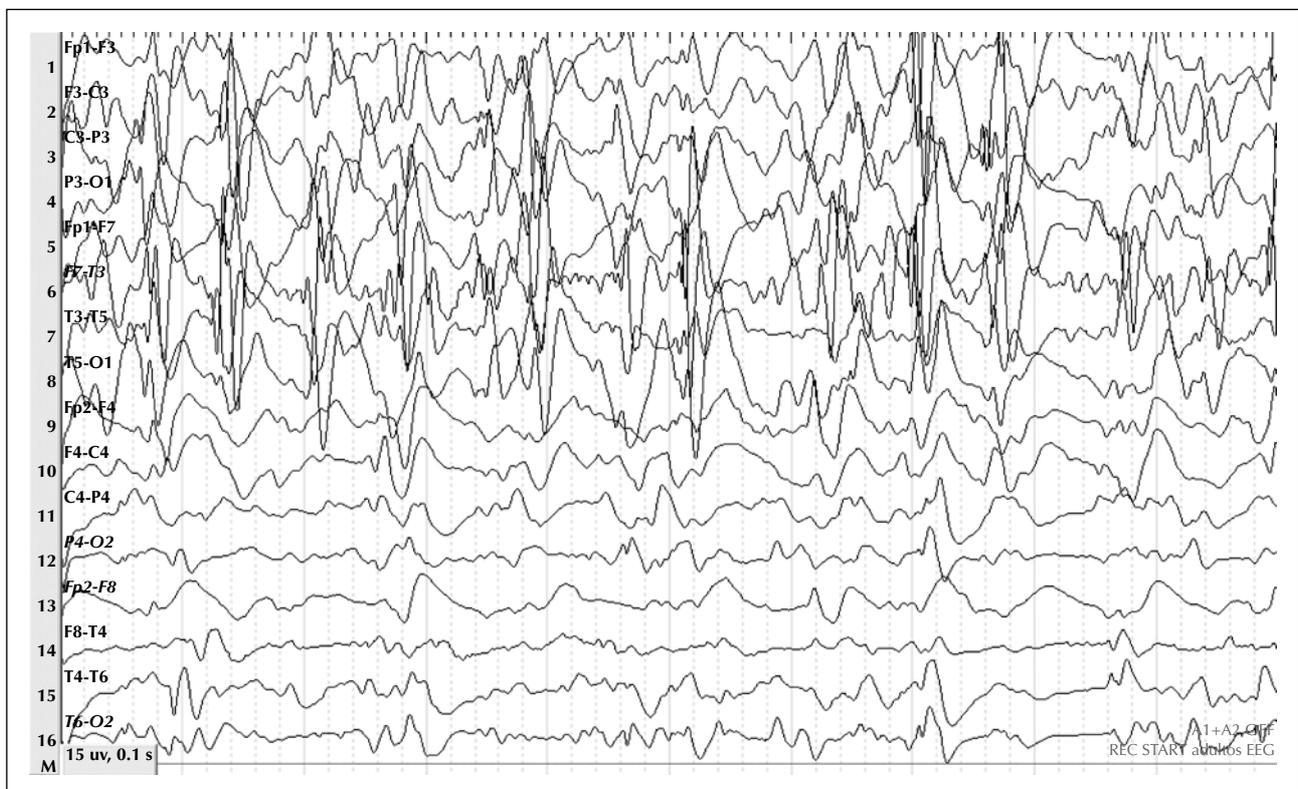
of 11 months, an EEG showed asymmetrical hypsarrhythmia.

She is now two years and nine months old, and experiences seven to eight dacrystic seizures a day. Some of these seizures are followed by a brief series of extension and abduction of both arms resembling extensor epileptic spasms. Her neurological development is poor, she is hypotonic and language has not developed.

Interictal EEG (*figure 3*) shows an epileptic encephalopathy pattern, with high amplitude slow background activity and multifocal epileptic abnormalities, predominantly in the left hemisphere, consisting of sharp waves, spikes and slow waves, all of which suggest the presence of a secondary, generalized epileptic process. She is being treated with topiramate, lamotrigine and clobazam. She also needs thyroxine supplementation. LH and prolactin serum levels normalized postoperatively.

## Discussion

Dacrystic seizures are rare, particularly in young children. Offen and coworkers (1976) described a 69-year-old patient who had paroxysmal episodes of crying which they presumed to be epileptic in origin and proposed the term “dacrystic epilepsy”. Many of the previous reports of crying seizures were of adult patients with frontal or temporal



**Figure 3.** Interictal EEG of the patient at age 2 years and 9 months showing a pattern of hypsarrhythmia (asymmetric hypsarrhythmia), with slow waves of high amplitude, spikes and sharp waves of multifocal appearance in the left hemisphere and slow background activity in the contra-lateral hemisphere.

focal epilepsy of lesional origin. The known causes of these lesional seizures were tumors, vascular malformations, hippocampal sclerosis and cerebral infarction (Luciano *et al.* 1993, Wang *et al.* 1995). Luciano *et al.* (1993) identified 11 published cases with dacrystic seizures and reported on seven adult patients with crying during video-EEG-documented, simple or complex partial seizures. Six of their seven patients had EEG evidence of a right fronto-temporal seizure focus.

Our patient was an infant with a giant hypothalamic hamartoma who presented with dacrystic and other types of seizures. HH present most often with epilepsy. In the majority of cases, epilepsy begins during the neonatal or early childhood period, usually in the form of laughter seizures for which the term "gelastic" seizures (Daily and Mulder 1957) is commonly used. Some of the patients with HH and gelastic seizures can also present rare ictal episodes of crying alone or together with laughter (Kahane *et al.* 1994, Munari *et al.* 2000). From the age of two months, he presented recurrent episodes of prolonged irritability, agitation, crying, and polypnea, initially not diagnosed as epileptic. Some months later, we identified the epileptic origin of the recurrent episodes of prolonged irritability because of their paroxysmal nature, in the absence of any precipitants and being relatively stereotyped. Arzimanoglou *et al.* (2004), described two personal cases of neonates who presented "bizarre frantic agitation, with crying and grimaces that could last for hours, later followed by more typical gelastic attacks" and related to HH. Our case illustrates the difficulty of recognizing these subtle ictal manifestations during infancy.

Ictal scalp-EEG recordings during gelastic and/or dacrystic seizures are reported as rather inconclusive (Tassinari *et al.* 1997, Kahane *et al.* 2003). During the first months of life, the several ictal and interictal scalp-EEG performed in our patient were normal. After the surgical procedure, interictal EEG consistently showed an epileptic encephalopathy pattern. We did not obtain a post-surgery ictal EEG during the dacrystic attacks. The stereotypes and recurrent nature of dacrystic, non-provoked episodes in our patient strongly suggested an epileptic nature.

Data obtained from stereotactic intracerebral EEG recordings in patients with HH have revealed that gelastic seizures (Munari *et al.* 1995) as well as dacrystic attacks (Kahane *et al.* 1994) are linked to the occurrence of an ictal discharge which remains located within the hamartoma, sparing the other investigated cerebral regions. Furthermore, the results of electrical stimulation of the mass reproduce the gelastic or dacrystic episodes (Kahane *et al.* 1997). Crying seizures in our patient recurred postoperatively, suggesting that the ictal onset zone of this type of seizures was not removed. Notwithstanding, the gelastic seizures disappeared.

More than half of the patients reported with HH also suffer from other types of seizures, mainly complex partial sei-

zures, tonic-clonic seizures, tonic seizures, falling seizures and atypical absences, that may start at the same time as gelastic and/or dacrystic seizures, although usually later (Tassinari *et al.* 1997, Arzimanoglou *et al.* 2003). These other seizure types are sometimes immediately preceded by laughing or crying attacks, as we observed in our patient, suggesting that ictal discharges within the HH could trigger other seizures which seem to originate in the cortex (Kahane *et al.* 2003). □

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