Case Report

Catastrophic Childhood Temporal Lobe Epilepsy

Giancarlo Di GENNARO¹, Antonio SPARANO¹, Addolorata MASCIA¹, Fabio SEBASTIANO¹, Vincenzo ESPOSITO², Pier Paolo QUARATO¹

¹Epilepsy Surgery Unit, IRCCS Neuromed, Pozzilli (IS), Italy
²Department of Neurosurgery, University of Rome “La Sapienza”, Rome, Italy

Abstract

Focal epilepsy, more frequently of extratemporal type, in children can present with a rapidly progressing course of intractable, severe epilepsy, associated to cognitive regression or stagnation. We present a typical example of such a patient with focal seizures due to a temporal lobe cortical lesion of developmental origin. Brain MRI revealed abnormalities in the right temporal lobe and Video-EEG monitoring revealed episodes clinically characterized by epileptic spasms followed by automotor seizures, with EEG findings suggestive of a right temporal lobe focus. Surgical resection resulted in excellent outcome.

Keywords: presurgical evaluation, epilepsy surgery

Introduction

The term catastrophic epilepsy is used to describe the clinical picture of severe, intractable epilepsy, usually with daily seizures and developmental stagnation or regression, typically in infancy or in the early childhood.

Usually this clinical picture may be found in generalized epileptic encephalopathies such as West syndrome and Lennox-Gastaut syndrome. However, catastrophic epilepsy may be found in extra-temporal lobe focal epilepsies often due to cortical development malformations¹. In these cases epilepsy surgery should be considered, often after invasive investigations. In the best scenario, the chance for freedom from seizures after surgery for catastrophic focal epilepsy due to malformation of cortical development appears to be in the range of 50% . Occasionally, in children with catastrophic...
epilepsy a diagnosis of temporal lobe epilepsy (TLE) can be made with the possibility, like in adulthood, of resective surgery with good seizure outcome.

We are reporting a case of a child with catastrophic TLE mimicking a severe symptomatic generalized epilepsy who underwent temporal lobectomy with excellent seizure outcome.

**Case Presentation**

The patient is a 7 year old boy who was referred to us for medically resistant epilepsy with daily events, occurring since age 2 months. He was the product of an uncomplicated twin pregnancy and delivery. Birth weight was 2.6 Kg and head circumference was within normal limits. There was no history of other medical diseases including CNS infections, febrile convulsions or head injuries. The family history was negative for seizure disorders or other neurological diseases.

The mother reports that the child had no problems until approximately 2 months of age when she began to notice brief paroxysmal episodes characterized by behavioral arrest associated with face blushing and eyelid fluttering. The baby was promptly evaluated by a pediatrician who made a diagnosis of epilepsy and started an antiepileptic therapy with phenobarbital. Eventually these episodes became more frequent and within 6 months after the onset she reported a frequency of 20-30 events per day, characterized by sudden abduction and extension of both arms and legs flexion followed, in some instances, by behavioral arrest, unresponsiveness, eyelid fluttering, face blushing, eye deviation to the back and slightly to the left. These episodes lasted no more than 10 seconds occurring mainly upon awakening and in cluster. Therefore, the patient was referred to a local neurologist who diagnosed idiopathic infantile spasm and treated with valproic acid in combination with phenobarbital with no major improvement. A course of ACTH was also instituted without noticeable benefit. In the course of several years the patient developed medically refractory epilepsy with daily seizures resulting in frequent falls. Furthermore, a severe diffuse cognitive dysfunction with behavioral disturbance became evident.

At age 7 the patient was referred to us in order to evaluate a possible surgical therapy. A presurgical non invasive diagnostic protocol included: review of medical history, neurological examination, brain magnetic resonance imaging (MRI) and Video-EEG monitoring. At admission the neurological examination revealed a global cerebral dysfunction with severe mental retardation and hyperactivity; no focal signs were noted. Neuropsychological testing was not possible because of his mental status.

A brain MRI (1.5 Tesla magnet) showed a dilation of the right lateral ventricle including the temporal horn, with thickening and poor gray-white matter differentiation of the anterior right temporal neocortex, suggesting a cortical dysplasia (Fig 1 A).

Approximately 20 hours of video-EEG monitoring (Telefactor Corp, Conshohoken, PA) were obtained. Interictal EEG recording revealed diffuse background slowing with frequent generalized slow spike- and polyspike-and-wave complexes and rare epileptiform activity over the right temporal region (Fig 1 B,C,D). A total of 9 seizures were recorded, clinically characterized by an epileptic spasms (ES) followed by, behavioral arrest, unresponsiveness, eyelid fluttering, face blushing, eye deviation to the back and slightly to the left. These episodes lasted no more than 10 seconds occurring mainly upon awakening and in cluster. Therefore, the patient was referred to a local neurologist who diagnosed idiopathic infantile spasm and treated with valproic acid in combination with phenobarbital with no major improvement. A course of ACTH was also instituted without noticeable benefit. In the course of several years the patient developed medically refractory epilepsy with daily seizures resulting in frequent falls. Furthermore, a severe diffuse cognitive dysfunction with behavioral disturbance became evident.

At age 7 the patient was referred to us in order to evaluate a possible surgical therapy. A presurgical non invasive diagnostic protocol included: review of medical history, neurological examination, brain magnetic resonance imaging (MRI) and Video-EEG monitoring. At admission the neurological examination revealed a global cerebral dysfunction with severe mental retardation and hyperactivity; no focal signs were noted. Neuropsychological testing was not possible because of his mental status.

A brain MRI (1.5 Tesla magnet) showed a dilation of the right lateral ventricle including the temporal horn, with thickening and poor gray-white matter differentiation of the anterior right temporal neocortex, suggesting a cortical dysplasia (Fig 1 A).

Approximately 20 hours of video-EEG monitoring (Telefactor Corp, Conshohoken, PA) were obtained. Interictal EEG recording revealed diffuse background slowing with frequent generalized slow spike- and polyspike-and-wave complexes and rare epileptiform activity over the right temporal region (Fig 1 B,C,D). A total of 9 seizures were recorded, clinically characterized by an epileptic spasm (ES) followed by, behavioral arrest, unresponsiveness and subtle motor automatisms, lasting up to 1 minute. During the episodes, EEG showed a generalized polyspike-and-wave complex coinciding with the ES, followed by rhythmic 1hz spike-and-wave discharge in the right fronto-temporal region, evolving
in a sharp contoured theta activity in the same region. Postictal slowing was more pronounced in the right hemisphere (Fig 1E). Anatomo-electro-clinical correlations pointed to a right temporal epileptogenic zone and therefore a right standard temporal lobectomy was performed. Pathology showed a cortical dysplasia of cytoarchitectural type.

After 18 months follow-up (at the time of this report) the child is seizure free; he remains severely mentally retarded although his hyperactivity is improved.

**Discussion**

We reported a child with catastrophic epilepsy in whom non-invasive presurgical investigations allowed us to disclose a temporal lobe focus that was successfully resected with freedom from seizures and substantial improvement of the hyperactivity and attention disturbances. Despite a good seizure outcome, our patient remained severely mentally retarded. This can be explained, other than with the prolonged uncontrolled seizure disorder also with the possibility of a more diffuse cortical developmental disorder not detectable by MRI.

Although several studies reported that seizure semiology in childhood TLE is similar to that seen in adulthood\(^2\), nevertheless various age-dependent ictal motor phenomena, including diffuse tonic components or ES, have been rarely observed\(^3\). In this condition, childhood TLE may present with daily seizures often resulting in falls, associated with cognitive stagnation or regression, and behavioral disorders, leading to a clinical picture of catastrophic epilepsy.

Despite a well localized temporal epileptogenic zone as demonstrated by the excellent seizure outcome obtained after a temporal lobectomy, the reported child exhibited generalized and symmetrical ES, often inaugurating seizures with automotor phenomenology. ES have been described in children with epilepsy due to focal lesions and it was reported that approximately two third of children with West syndrome may develop focal seizures, mostly originating in temporal lobe.

We think that in childhood TLE the ES or other generalized motor phenomena may be an expression of age-dependent seizure pattern, probably due to limbic system immaturity. In animal epilepsy models the application of kainic acid and pilocarpin on the cortical surface of the limbic structures...
is able to induce age-dependent generalized ictal motor phenomena\textsuperscript{4} comparable to those seen in childhood TLE, probably as a consequence of a rapid and extensive subcortical extratemporal activation. Furthermore, Moshe (1981)\textsuperscript{5}, analyzing the afterdischarge thresholds in hippocampal-kindled rat pups, have demonstrated that the limbic system is more resistant to synchronization at early age: this could explain why temporal lobe seizures in childhood tend to have less signs of limbic system involvement than in adulthood.

In conclusion childhood TLE may rarely present as a catastrophic epilepsy with some generalized clinical and electrographic features resembling, according to the age, West or Lennox-Gastaut syndrome\textsuperscript{6} that may discourage a possible resective epilepsy surgery favoring, instead, palliative surgical treatments such as callosotomy, often resulting in poor seizure control with potential cognitive and behavioral side effects.

In spite of the relative resistance of immature neurons to epilepsy-induced brain damage, in fact, seizures in the immature brain do produce significant and often irreversible alterations of the developing brain\textsuperscript{7}. Seizures may perturb a wide range of developmental phenomena that are activity dependent, including cell division, migration, sequential expression of receptors, formation, and probably stabilization of synapses. Indeed seizures can modify a wide range of unique processes that take place during development and are essential for the correct formation and wiring of the circuitry. The migration of neurons, the arborization of the neurites, the formation of synapses, or the removal of redundant processes are all essential processes that are activity dependent and may be disturbed by seizures. These alterations in normal neuronal connectivity can result in long-term consequences in, seizure susceptibility, learning and memory, and risk for subsequent seizure-induced injury\textsuperscript{8}. Therefore we think that in all catastrophic childhood epilepsies, an early presurgical evaluation should be performed in order to identify a possible focal epileptogenic zone that may be amenable to resective surgery. Further studies, however, are needed to establish if early surgical intervention may prevent permanent cognitive dysfunction in children with catastrophic epilepsy with focal onset.

References


Correspondence
Giancarlo Di Gennaro

Epilepsy Surgery Unit, IRCCS Neuromed, Pozzilli (IS), Italy

e-mail: gdiGennaro@neuromed.it

Received by: Apr 20.2005
Revised by: July 28.2005
Accepted: Sept 07.2005

The Online Journal of Neurological Sciences (Turkish) 1984-2005
This e-journal is run by Ege University Faculty of Medicine, Dept. of Neurological Surgery, Bornova, Izmir-35100TR as part of the Ege Neurological Surgery World Wide Web service.
Comments and feedback:
E-mail: norolbil@med.ege.edu.tr
URL: http://www.jns.dergisi.org
Journal of Neurological Sciences (Turkish)
ISSNe 1302-1664