HEADACHE AS A SOLE MANIFESTATION OF NON-CONVULSIVE STATUS EPILEPTICUS

Abstract:
Non-convulsive status epilepticus (NCSE) may present with several manifestations, many of which may not be obvious. The most important tool for the diagnosis of NCSE is the electroencephalograph (EEG) pattern. This is a case report of a boy 9 years of age presenting with severe and continuous headache. He had received chemotherapy for histiocytosis, diagnosed at the age of 3.5 years. No evidence of central nervous system (CNS) involvement was found. He was diagnosed as having NCSE and following anticonvulsant therapy the headache and EEG abnormality disappeared completely within 24 hours. Headache and seizure disorder may coexist in different situations; to our knowledge this maybe the first report of NCSE with headache as a sole manifestation.

Keywords: Non-convulsive status epilepticus, headache

Introduction
Prolonged epileptic states without convulsions were initially recognized in the nineteenth century. The first clinical and electrographic description of NCSE was given by Lennox (1). Following this initial report, a number of terms have been used to describe similar phenomena; these included petit mal status, absence status, spike and wave stupor, non-convulsive status epilepticus, etc (2,3). Manifestations of NCSE may vary, very often being subtle; they may show one or more of the following features:
1) A variation in level of consciousness, from drowsiness to confusion or even stupor. 2) A change or fluctuation in behaviors for no apparent reason. 3) Semi-purposeful movements and actions; these actions importantly are clearly not the child's usual actions. 4) A change in speech or even a loss of speech. 5) Motor automatisms, involuntary and uncontrolled jerky or twitchy movements of the face, mouth or limbs.

In a recent article, Husain et al argue that severely impaired mental state, ocular movements abnormalities and the patient's history could suggest a diagnosis of NCSE (4). D. Audenino et al studied 50 adult patients with NCSE and found that there are no exceptional clinical features, characteristic of this condition (5). The diagnosis of NCSE needs EEG evidence of bilaterally synchronous continuous spike-and-wave discharges during the ictal event (1). Another diagnostic point to consider in regard to NCSE is that the EEG at the time of ictal event should
represent a definitive change from the pre-ictal state (1). Any suspicion of NCSE is the most important clinical indication for performing an emergency EEG (6). Because of the differing EEG patterns and the pleomorphic clinical features, a diagnosis of NCSE is only possible with an expert integration of EEG findings and clinical data, as emphasized by Niedermeyer and Ribeiro (7). The present report describes a child with NCSE, who complained of headache as the sole symptom. Our case was of interest with respect to his unique manifestation. A review of corresponding literature is included.

Case report:
This 9-year old boy is the first child of healthy related parents; pregnancy and delivery were uneventful. At 3 years 6 months of age, he was evaluated for proptosis and was diagnosed as having histiocytosis and after which chemotherapy was begun. Brain computed tomographic scan (CT-Scan) with and without contrast, bone survey, abdominal sonography and bone marrow aspiration revealed normal results. Orbital CT-Scan revealed a mass in the right small wing of the phenoid and temporal bone junction and the posteriolateral angle of the right orbit. After 2 years, he had two generalized tonic-clonic seizures, and because of cerebrospinal fluid (CSF) pleocytosis, intrathecal treatment was started. 9 months later, he was admitted to continue chemotherapy protocol. On the second day of hospitalization, the patient complained of, severe and continuous bilateral headache, although he had no significant prior history of headache; his physical examination including fundoscopy was unremarkable, but he was very irritable. Body temperature, heart rate, blood pressure and respiratory rate were normal. Lumbar puncture demonstrated a CSF opening pressure of 130 mmH2o, leukocytes of 0 cell/mm3, glucose 51 mg/dl and protein 12 mg/dl. The complete blood count and electrolytes were normal. A cranial magnetic resonance imaging (MRI) revealed brain atrophy. An awake EEG performed during headache revealed bilateral synchronous continuous spike and slow waves discharges (Fig.1). The patient was diagnosed with NCSE and started on intravenous diazepam 0.2 mg/kg every 4 hour for 2 days, while continuing his anticonvulsant medication, Phenobarbital 3mg/kg/day. Twenty-four hours after initiation of diazepam his headache disappeared and EEG had normalized completely (Fig.2). The patient was discharged on oral Phenobarbital; During 11 months of follow-up visits, he reported no headaches or seizures.

Discussion
The association between headaches and seizures is complex and may be multifactorial (8). Many clinical interrelationships exist between headache and epilepsy, as given below:
1) In epilepsy, headache can be seen as a pre-ictal, ictal or post-ictal phenomenon. Migraine may trigger epilepsy (migralepsy) or epilepsy may initiate headache. Karaale-Savrum F. et al studied 109 patients with partial onset and 26 patients with generalized onset seizures. They found that interictal headaches were present in 50 (40.7%) of 125 patients. Seventy-nine (58.5%) patients had peri-ictal headache, of which eleven patients had pre-ictal headache; three had ictal headache and 56 of these had post-ictal headache (9); this association has been reported in other studies (10,11,12).
2) Seizure and headache seem to be associated in certain syndromes such as childhood epilepsy with occipital spikes (13). In benign rolandic epilepsy, some studies have reported seizures and headache (14).
3) Both headache and seizures may occur in the same individual with a common underlying cause, such as head trauma or an arteriovenous malformation (8). Approximately 1% of epileptic patients report headache as a seizure manifestation (seizure headache). In some children, although, this is very uncommon, headache is the only feature of their seizure disorder. In patients with paroxysmal headache attacks, EEG recording during headache shows continuous epileptiform activity (15). In the case presented here, the patient developed continuous, severe headache without any evidence of CNS involvement. The diagnosis of NCSE was based on the EEG pattern during headache and complete response to anticonvulsant drug (both clinically and electro physiologically).
To our knowledge, this is the first report of NCSE, who presented with only headache. This case emphasizes the need to keep NCSE in mind in the differential diagnosis of acute headache in children who are recognized as being at risk for developing this condition and the need for an urgent EEG is emphasized.
References: