

A Case Report of Acute Necrotizing Encephalopathy of Childhood

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Article information	Abstract
<p>Article history: Received: 13 June 2011 Accepted: 24 Sep 2011 Available online: 5 Nov 2012 ZJRMS 2013; 15(4): 88-89</p> <p>Keywords: Encephalopathy Encephalitis Case report</p> <p>*Corresponding author at: Department of pediatrics, Children and Adolescent Health Research Center, Zahedan University of Medical Sciences, Zahedan, Iran. E-mail: gh_miri@yahoo.com</p>	<p>Background: Acute Necrotizing Encephalopathy of Childhood (ANEC) is a rare disease with a higher prevalence in the East Asia which is characterized with symmetrical and multifocal involvement of areas including thalamus, brainstem, cerebellum and white matter and it is associated with death as well as long-term neurological disabilities (sequel) in the individuals who survive.</p> <p>In this report, we introduce a child with acute neurological symptoms resulted by a possible infection process and explain his CT-scan and brain MRI as well as paraclinical symptoms.</p>

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Introduction

Acute necrotizing encephalopathy of childhood diagnosis is suggested in the children mostly along with acute occurrence of neurological defects. Such symptoms are often nonspecific, but the diagnosis confirms by paraclinical examinations and brain imaging.

Case Report

An Iranian 8-month-old male infant of Baluchi breed with no previous history of illness and hospitalization and fully vaccinated (as routine in Iran), has suffered from a generalized tonic clonic (GTC) seizure for 5-10 minutes as well as loss of consciousness due to having acute watery diarrhea and high fever and vomiting; he was transferred from Khash hospital to Ali Ibn Abi Taleb subspecialty hospital in Zahedan.

At beginning of admission to this hospital, the patient had BP=80/40 mm/Hg, RR=35/min, PR=150b/min and GCS=8-9. Due to loss of consciousness, he was finally intubated and hospitalized in ICU ward. In the patient’s history, there was no trauma, possible drug and food poisoning and bite as well as no history of previous admission to the hospital. The symptoms of loss of consciousness and the seizure arise and promote following to acute febrile gastroenteritis.

Serum biochemistry is indicative of normal electrolytes; however, in complementary examinations, liver tests showed an apparent increase (AST=861 U/L, ALT=1184 U/L, ALP=444 U/L). Furthermore, protein level had been increased to 394 mg/dl in CSF sample without any

evidence of pleocytosis or RBC and reduced concentration of CSF glucose. Coagulation tests and factors I and XIII levels were reported as normal. All metabolic studies including serum and urine amino acid chromatography, urine reducing substances as well as the measurement of the blood ammonia and lactate, acylcarnitine profile and fatty acids metabolism disorder were all normal. In the CT scan without contrast, symmetrical bilateral hypodensity was apparent in thalamus and caudate nuclei. Ventricles were normal and there was no evident of hemorrhage (Fig. 1).

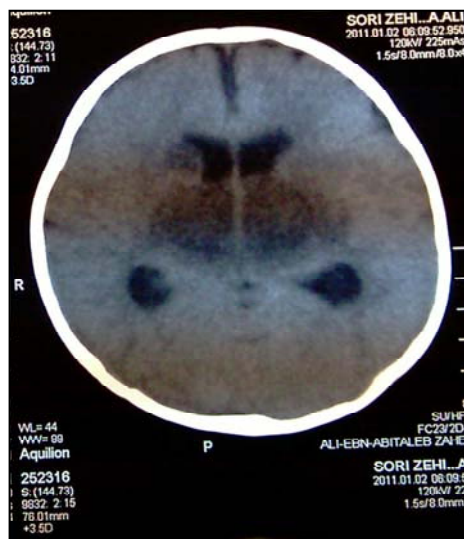


Figure 1. Symmetrical hypodensity in bilateral thalamic areas



Figure 2. Areas of increased signal are seen in the thalamus and striatum

In MRI conducted without contrast, there was an abnormal bilateral and symmetrical signal in thalamic Caudate nucleus and cerebellum which is indicative of hypoxic ischemic encephalopathy (HIE) and Acute Disseminated Encephalomyelitis (ADEM) or ANEC. No tumor or pressure effect was present.

CSF, blood and urine cultures were negative for bacterial infection. The patient was extubated as a consequence of protective measures while he had tolerated. During hospital stay, he was firstly fed by NG tube and finally oral feeding was done for him. His GCS was increased to 12-13, but there have been still problems in his limb movements, speech and communication.

Discussion

ANEC is a rare disease as observed in the children who have been already healthy and it is more prevalent in the East Asia, though its sporadic cases were found throughout the world. This disease is clinically associated with high fever, seizure, rapid decline in neurological function and high mortality. Laboratory evaluations alone are also mostly nonspecific and include increased CSF protein, increased liver tests and thrombocytopenia [1, 2]. Radiological symptoms in ANEC are specific and diagnostic including symmetrical and multifocal

involvement in thalamus, brainstem, supratentorial white matter and cerebellum [1, 3].

Thalamic signal loss is found in both T₁ and T₂ MRI that is sometimes associated with petechial hemorrhages [4]. In our patient, CT, MRI and paraclinical evidences were exactly based on the mentioned cases; however, no finding was reported in favor of petechiae.

The children suffered from ANEC show often acute neurological defects clearly and a broad range of long term cerebral sequel is seen in them. The extent of cerebral involvement, severity of petechial hemorrhages and development of cavity, cystic degeneration and cortical atrophy, all are effective on ANEC prognosis. In general, prognosis is poor in ANEC [2]. Differential diagnosis in ANEC include Rey syndrome, Leigh disease (hyperammonemia, hypoglycemia, metabolic acidosis), ADEM (CSF pleocytosis, more prevalent unilateral neurological involvement) and Japanese encephalitis (common involvement of cortex and dark matter) [5-7].

A metabolic study was conducted on the mentioned patient; lactate and ammonia serum was normal and the evaluation of fatty acid oxidation disorders, chromatography of serum and urine amino acids and urine reducing substances were resulted as normal too. ANEC etiology is still really unknown, but infectious, metabolic, immune related causes and etc were suggested.

In our patient, no special etiology was found except a possible gastrointestinal infection. Due to the viral tests inaccessibility, these causes are still unknown to the author. The patient left finally the hospital with stable vital signs, full tolerance for oral foods, GCS=13 and without seizure; though it appeared to have some defects in his movement and speech until an uncertain time with himself.

Authors' Contributions

All authors had equal role in design, work, statistical analysis and manuscript writing.

Conflict of Interest

No conflict.

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