*Case Report*

*Iran J Pediatr*
*Dec 2007; Vol 17 ( No 4), Pp:375-378*

# The Case Report of a Survival from Ekiri Syndrome

**Seyed Mehdi Monajemzadeh [*1], MD; Sheedeh Assar [2], MD; Ali Akbar Momen [2], MD**

1. Pediatrician, Department of Pediatrics, Ahwaz Jondishapur University of Medical Sciences, Iran
2. Pediatric Neurologist, Department of Pediatrics, Ahwaz Jondishapur University of Medical Sciences, Iran

Received: 16/04/07; Revised:06/08/07; Accepted: 10/09/07

## Abstract

***Background:*** Lethal toxic encephalopathy of shigellosis (Ekiri syndrome) is a rare complication of the shigella infection presented with fever, severe toxicity, seizure and diffuse brain edema, coma and death in the absence of dehydration and is nearly always lethal. This report is about a child who has survived the Ekiri syndrome.

***Case presentation:*** A three-year old child was admitted to the emergency ward because of fever, one attack of generalized tonic-clonic seizure, drowsiness and diarrhea with no signs of dehydration. The differential diagnosis was meningitis, shigellosis and atypical febrile convulsion. All test results for blood, urine, stool and cerebro-spinal fluid samples were normal except for serum sodium (119 mmol/l) and stool exam (many RBCs). He was treated with anticonvulsants because of the recurrent seizures and ceftriaxone with probability of shigellosis. The emergency brain CT scan showed diffuse brain edema. After 48 hours the consciousness was improved. Stool culture showed the growth of Shigella flexneri. Second brain CT scan revealed a dramatic decrease of the brain edema.

***Conclusion:*** Because neurological symptoms are among the extra intestinal complications of the shigellosis, it is recommended to put this disease in the list of differential diagnosis of unexplained neurological signs in endemic areas and consider the specific medications in the treatment programs.

***Key Words:*** Ekiri syndrome, Encephalopathy, Shigella, Shigellosis, Seizure

## Introduction

The first reports of the Ekiri syndrome in year 1949 have described it as bacterial dysentery in a child, which is complicated by low serum calcium and tetanus, and so calcium supplements have been recommended to be added to the nutritional regimens of the Japanese children.[1]

According to the high incidence rate (45%) of the extra intestinal complications of the shigellosis[2], this syndrome was considered in upcoming reports to be one of the extra intestinal complications of shigella infection and was also known as the cause of death of the shigellosis patients.[3,4] Because of the high incidence rate of

*** Correspondence author;**
**Address:** Golestan Hospital, Ahwaz Jundishapur University of Medical Sciences, Department of pediatrics, Golestan St, Ahwaz, IR Iran
**E-mail:** monajemzadhmd@yahoo.com

the gastroenteritis in Khoozestan province (south west of Iran) and the relative high incidence rate of shigellosis (14% of gastroenteritis cases)[5], several patients die due to the Ekiri syndrome each year. This report is about a male child who has survived the Ekiri syndrome.

## *Case Presentation*

A three-year old male child was admitted to the emergency ward because of fever and one attack of generalized tonic-clonic seizure with post ictal drowsiness. Diarrhea was another complaint at the time of admission. After admission, the patient had two more attacks of tonic-clonic seizure and the drowsiness became more severe.
The patient had no history of pre and postnatal problems and although he had growth retardation (weight =11 Kg), the developmental milestones were normal. The patient was admitted with the probable differential diagnosis of meningitis, shigellosis and atypical febrile convulsion and so the blood, urine, stool and cerebro-spinal fluid (CSF) samples were taken for routine laboratory tests and culture. All test results were normal except that serum sodium was 119 mmol/l and stool exam revealed many red blood cells (RBCs) per high power field. Blood and CSF cultures showed no growth. Blood sugar was within normal range and complete blood count showed white blood cells (WBC) count of 6000/ml, hemoglobin 13mg/dl and normal platelet count. CSF also showed normal sugar and proteins and no cells.

The patient was transferred to the intensive care unit because of his general condition and was treated with Phenobarbital initially and Dilantin and rectal acetaminophen later because of the recurrent seizures. With the probability of shigellosis he was administered 70 mg/kg of ceftriaxone. Fluids were administered as two thirds of the maintenance dose because the patient had no signs of dehydration. An emergency brain CT scan showed a diffuse brain edema at this stage (Fig 1) whereupon the patient received dexamethazone and cimetidine.

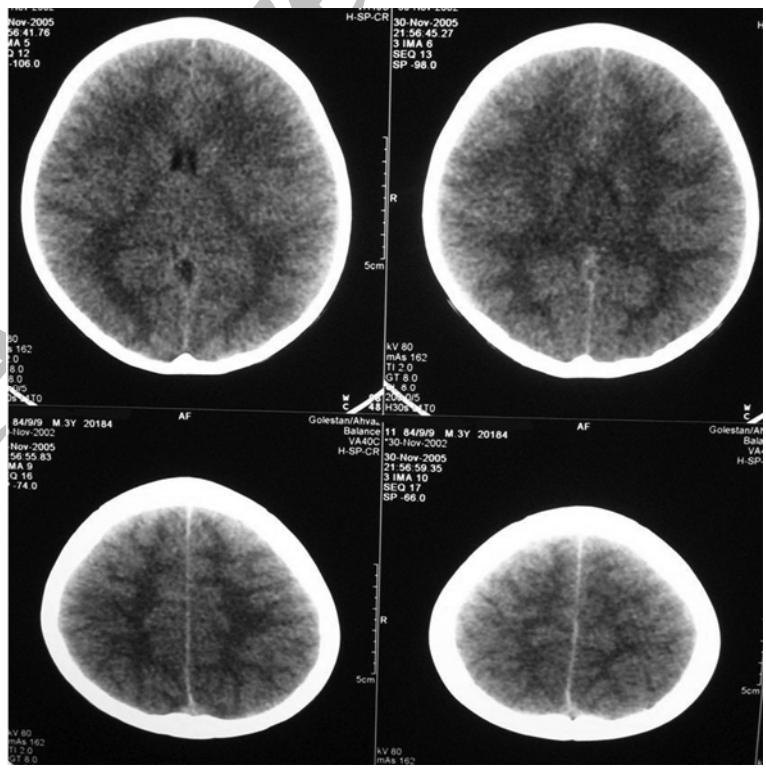


**Fig 1-** CT scan showed a diffuse brain edema

He was also intubated because of respiratory disorders and was monitored by pulse oximetry, EKG monitoring and serial checking of the vital signs. Liver function tests were done and fresh frozen plasma (FFP) was administered because of the impaired prothrombin time.

The patient was extubated after about 48 hours following the improvement of consciousness and the beginning of spontaneous respiratory movements. The result of the stool culture indicated the growth of Shigella flexneri. Ekiri syndrome was diagnosed in this patient according to the general course of the disease, brain edema, hyponatremia, lacking dehydration and the positive stool culture for Shigella flexneri.

The second brain CT scan revealed a dramatic decrease of the brain edema (fig. 2). The patient was transferred to the pediatrics ward and anticonvulsive drugs were switched to oral anticonvulsives and the ceftriaxone, dexamethasone and cimetidine disconnected. After 48 hours the patient was discharged on Phenobarbital tablets and was ordered to return for follow up. After 1 year follow up, the patient is doing well and no sequelae are identified.

## Discussion

Shigellosis is a hygienic problem in the whole world. It is estimated that 165 million people a year are infected by its responsible organism, 99 percent of which are from developing countries and 69 percent are the children under the age of five.[6] 1.1 million people die each year because of shigellosis, 60 percent of them being children under five years of age.[6,7] The general manifestations of shigellosis are often high fever, toxicity, watery diarrhea or diarrhea with blood and mucus, abdominal cramps and tenesmus.[6,8] In a number of patients shigella infection is presented with the symptoms of CNS involvement including decreased consciousness level and seizures[8,9] and sometimes the lethal toxic encephalopathy of the shigellosis can be the only manifestation of the disease in the absence of intestinal and metabolic symptoms. Headache is a dominant, alarming and malignant symptom in these cases. [2,10,11] In our own experience vomiting is also one of the initial symptoms in this group of shigellosis patients.

Shigellosis has been diagnosed in this patient according to the presenting symptoms and has

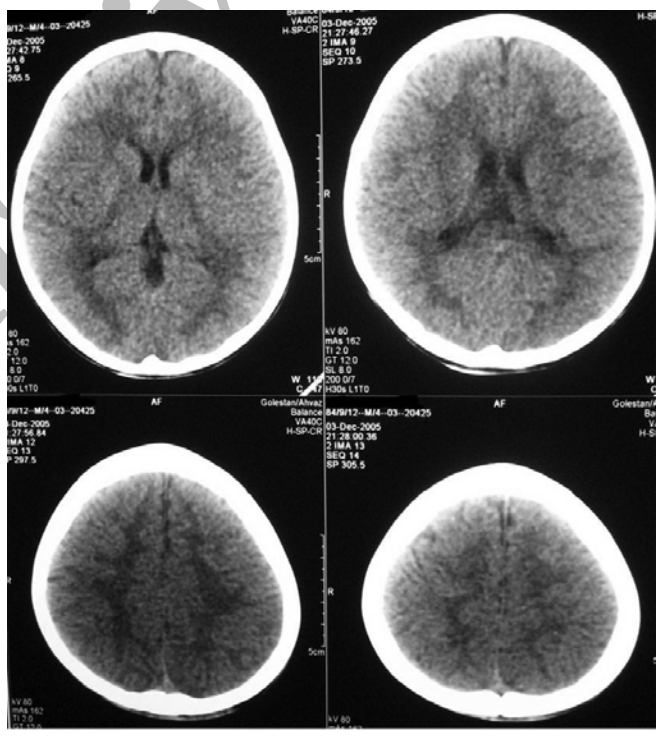

**Fig 2-** The second brain CT scan revealed a dramatic decrease of the brain edema

been treated with ceftriaxone early in the treatment course. The diagnosis of the brain edema has been based upon the emergency brain CT scan and dexametazone was administered as soon as possible. It seems that the early administration of these two drugs, controlling seizures and correction of hyponatremia have rescued the patient. So, it is recommended to prevent brain edema to help patients survive the disease. [11]

There are similar reports of some shigellosis patients who initially presented with encephalopathy and brain edema (according to brain CT results) without GI symptoms and have been treated empirically with ceftiaxone, dexamethasone and acyclovir (with the probability of herpes) before stool culture results were ready, and the patients survived the condition.[2] Our patient had also brain edema, which worsens the prognosis but fortunately remained alive because of early treatment. Whether any other factor caused the patient to survive is not probable but it seems that awareness of this entity and considering it in differential diagnosis led to correct management with prompt administration of dexamethazon to reduce brain edema and antibiotics to treat infection resulted in survival of our case.

## Conclusion

Because neurological symptoms with different degrees of severity are among the extra-intestinal complications of shigellosis[2,8], it is recommended to put this disease in the list of differential diagnosis of unexplained neurological signs in endemic areas and place specific medications to reduce brain edema and to control infection in the treatment programs.

## References


1. Dodd K, Buddingh G, Rapoport S. The Etiology of Ekiri: a highly fatal disease of Japanese children. Pediatr. 1949;3(1):9-19.
2. Somech R, Leitner Y, Spirer Z. Acute Encephalopathy preceding shigella infection. Isr Med Assoc J. 2001;3(5):384-5.
3. Bennish M. Potentially Lethal Complication of shigelloses. Rev Infect Dis. 1991;13(suppl 4):S319-24.
4. Groen A, Freier S, Passsell JH. Lethal toxic Encephalopathy due to childhood shigellosis in a developed country. Pediatr. 1992;89(6 pt 2):1189-93.
5. Tajedini S. Isolation and determination of antibiotic resistance of E.P.E.C. and Shigella associated with childhood diarrhea in Ahwaz. PhD Thesis, AhwazUniversty of Medical Sciences. 1994; p:71.
6. Niyogi SK. Shigellosis. J Microbiol. 2005; 43(2):133-43.
7. Dipika Sur T, Ramamurthy J, Deen J, et al. Shigellosis. Indian J Med Res. 2004;120(5): 454-62.
8. Hall CB, Kimberlin DW. Shigella infections. Red Book 2006. 27th ed. Elk Grove Village, IL; American Academy of Pediatrics. 2006; Pp:589-90.
9. Wasif A, Dhar U, Mohammed A, et al. Central nervous system manifestations of childhood shigellosis. Pediatrics 1999; 103(2):e18.
10. Dieu-Osika S, Tazarourte-Pinturier MF, Dessemme P, et al. Fulminant encephalopathy due to Shigella flexneri. Arch Ped. 1996;3(10):993-6,
11. Platz F, Barets HG, Fleer A, Gemke RJ. Lethal encephalopathy complicating childhood shigellosis, Eur J Ped. 1999; 158(7):550-2.