Case report

Congenital toxoplasmosis associated with severe intracerebral calcification and West syndrome

Kundu GK[1], Nandi ER[2], Saha A[3], Habib A[4], Rahman MM[5]

Introduction

Most children with congenital toxoplasmosis is an embryo foetopathy. It has been described all over congenital toxoplasmosis are developmentally normal[1] but up to four percent die or have evidence of permanent neurological damage or bilateral visual impairment during the first years of age[2,3] It is in this context that West syndrome can develop, and may be defined as a triad of menifestations infantile spasm and developmental delay and hypsrrythmic patern of EEG.[4] Here this treatable and academic case, congenital toxoplasmosis & West syndrome was reported.

Key words: Toxoplasmosis, calcification, West syndrome

Case report

Momo, a 9 months old immunized female infant of middle class family, only issue of nonconsanguineous parents, admitted in Paediatric Neurology department of BSMMU with the complaints of repeated convulsion for 2 months which occurred in clusters usually 5-6 episodes per day. Each episode consisted of 10 to15 spasm and characterized by sudden contraction of neck and extremities on to the trunk, more on awakening from sleep, not associated with deterioration of consciousness and bowel bladder incontinence. Mother also told that her child was unable to sit yet. There was no history of perinatal asphyxia but mother gave the history of death of child in perinalal period. On examination, she was less interested to surroundings. Vital signs were normal. There was no skin change. Weight & supine length were within normal limit but OFC was 40 cm which was below 3th centile. Neurological examination revealed that bulk of the muscle was normal on both side, tone was increased, jerks were increased and planters were extensor on both side. Developmental examination showed that social smile was absent and neck control was not achieved. A diagnosis of West syndrome was made clinically which was confirmed by an EEG showing hypsarythmia. (Figure-1) The patient was treated by prednisolone 2 mg/kg daily and Vigabatrin for a several weeks and convulsion was subsided to about fifty percent.

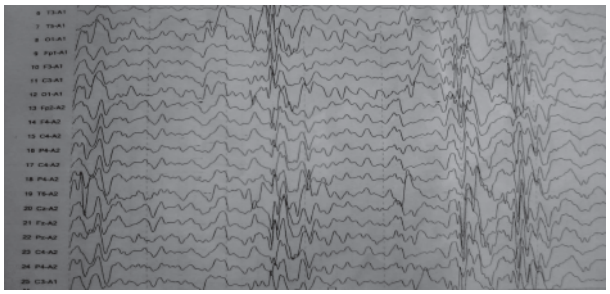

Figure-1: *EEG with hypsarythmic tracing*

A cerebral computed tomography (CT) reported severe parenchymal calcifications compatible with cerebral toxoplasmosis (Figure-2). TORCH screening revealed positive toxoplasma lgM. Anti-CMV IgG was ractive, CMV DNA was negative. Ophthalmological examination

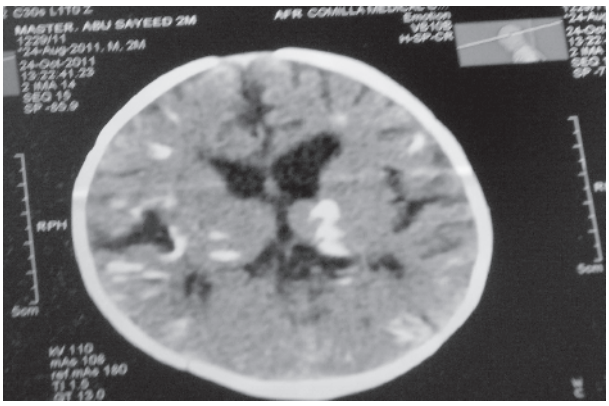

Figure-2: *CT scan of brain showing intracerebral calcification, cerebral atropy and ex-vacu dilatation*

1. * Dr Gopen Kumar Kundu
Assistant Professor, Paediatric Neurology
Bangabandhu Sheikh Mujib Medical University
Dhaka. Email:gopen.kundu@gmail.com
2. Dr Eva Rani Nandi
Registrar, Obstetrics & Gynaecology
Shaheed Shurawardi Medical College , Dhaka
3. Dr Anamika Saha
Paediatrician
4. Dr Ahsan Habib
Assistant Professor, Department of Neurology
Bangabandhu Sheikh Mujib Medical University
Dhaka
5. Professor Dr Md Mizanur Rahman
Professor & Head, Paediatric neurology
Bangabandhu Sheikh Mujib Medical University
Dhaka

** For correspondence*

revealed chorioretinal change in both eyes. An ENT examination was also performed; no evidence of hearing loss was reported. Considering these clinical, serological and imaging report, the diagnosis of a congenital toxoplasmosis complicated by West syndrome was made. The treatment of this patient was started with sulfadiazine, 100 mg/kg/daily, pyrimethamine 1 mg/kg/daily, and folinic acid 25 mg weekly but patient condition was not improved and gradually deteriorated after several weeks and unfortunately died on third week of treatment.

Discussion

Toxoplasmosis is a zoonosis, the definitive host is the cat and all other hosts are incidental. Toxoplasma gondii is an obligate intracellular protozoan that can be acquired orally, transplacentally, parenterally by transfusion or from a transplanted organ. In normal children, infection may be asymptomatic. Congenital transmission occurs when immunologically susceptible mother acquires infection during gestation. Transmission may occur transplacentally or during labor. Of untreated maternal infections acquired in the first trimester, approximately 17% of fetus is infected, usually with severe disease. Of untreated maternal infection acquired during 3rd trimester, approximately 65% of fetuses are infected usually with mild disease at birth.[1] Transmission is less frequent when infection is acquired before 10th week of gestation.

Congenital infection during neonatal period may present as IUGR, prematurity, jaundice, thrombocytopenia, peripheral retinal scars, hepatosplenomegaly, microcephaly, convulsions, mental retardation, strabismus and cerebrospinal fluid pleocytosis along with the classical triad of chorioretinitis, hydrocephalus and cerebral calcifications. Infection may result in hydrops fetalis and even perinatal death[1]. Sequelae are most often ocular. Ocular lesions may recur during childhood and adolescence.

Lesions suggestive of chorioretinitis, the high concentration of IgG in the mother and the child, and the presence of cerebral calcifications are compatible with a positive diagnosis of toxoplasmosis, which is very likely to be congenital considering neurological and ophthalmological signs during the first months of life. Obviously the presence of congenital toxoplasmosis infection is defined by the persistence of specific IgG antibodies beyond 11 months of age; and the absence of congenital infection is defined by undetectable IgG after 2 months of age in the absence of anti-toxoplasma treatment.[5,6] This is our clinical case, which is 25 months of age and presented with very high concentration of IgG characteristic of toxoplasma gondii.

All infected newborns should be treated whether or not they have clinical manifestations of the infection. Infants should be treated for 1 year with oral pyrimethamine (2 mg/kg/day for 2 days, then 1 mg/kg/day for 2-6 months, then 1 mg/kg/alternate day), Sulfadiazine (100 mg/kg/day in divided doses) and leucovorin (5-10 mg/kg/day on alternate day). Prednisone (1mg/kg/day) can be used when active chorioretinitis involves the macula or otherwise threatens vision or CSF protein is > 1gm/dl.

West syndrome may be idiopathic or symtomatic. In symptomatic case, there are identifiable brain lesions are present as like as our case. This form is more disabling than an idiopathic or isolated West syndrome.[7,8] The diagnosis of West syndrome is made clinically by the existence of flexor spasm , sudden onset between the age of 3 to 7 months, and by EEG presenting generally disorganized and irregular traces, while the CT scan of brain detect the cerebral lesion, which is the cause of the disease. [9-11]

Congenital Cytomegalovirus infection is a differential diagnosis of congenital toxoplasmosis in which there is significant intracerebral calcification in periventicular region[9,12] Primary prevention is very important and can be done through awareness raising and education of women of child-bearing age. The removal of cats, proper cooking of meat, of hands hygiene and drinking water hygiene should be encouraged in our developing countries.[13,14]

In the presence of West syndrome in a patient one should search for congenital toxoplasmosis as an etiology. A multidisciplinary follow-up of this child is required, Primary prevention should be encouraged in pregnant women for congenital toxoplasmosis in developing countries.

References

1. Behrman RE, Kliegman RM, Jenson HB, editors. Nelson Textbook of Pediatrics, 17th ed. Philadelphia : WB Saunders; 2004. pp. 1144-53
2. Remington JS, Klein JO, editors. Infectious Disease of the Fetus & Newborn Infant. Philadelphia, 4th ed. Philadelphia: WB Saunders; 1995. pp. 140-267
3. Salt A, Freeman K, Prusa A. Determinants of response to a parent questionnaire about development and behaviour in 3 year olds: European multicentre study of congenital toxoplasmosis. BMC Pediatr. 2005; 5:21

4. Guerina NG, Hsu HW, Meissner HC, et al. Neonatal serologic screening and early treatment for congenital Toxoplasma gondii infection. The New England Regional Toxoplasma Working Group. N Engl J Med. 1994; 330:1858–1863
5. Roger J, Bureau M, Dravet CH, Genton P, Tassinari CA, Wolf P. Les syndromes épileptiques de l'enfant. 3eme édition. Herts UK: John Libbey and Co Ltd publishers. 2002. pp. 47–64
6. Lebech M, Joynson DH, Seitz HM, et al. Classification system and case definitions of Toxoplasma gondii infection in immunocompetent pregnant women and their congenitally infected offspring. European Research Network on Congenital Toxoplasmosis. Eur J Clin Microbiol Infect Dis. 1996; 15:799–805
7. Thoumsin H, Senterre J, Lambotte R. Twenty-two years of screening for toxoplasmosis in pregnancy: Liege-Belgium. Scand J Infect Dis. 1992; 84:84–85
8. Wallon M, Peyron F, Lebech M, Petersen E, Gilbert R, Dunn D. Prenatal treatment and the risk of congenital toxoplasmosis: preliminary findings from two cohort studies. Pediatr Res. 1997; 42:400
9. Wirden M, Botterel F, Romand S, Ithier G, Bourée P. Intérêt du dépistage en post-partum de la toxoplasmose congénitale après primo-infection maternelle en fin de grossesse. J de Gynécologie Obstétrique et Biologie de la Reproduction octobre. 1999; 62:566-9
10. Dollfus H, Dureau P. Congenital toxoplasma chorioretinitis transmitted by preconceptionally immune women. Br J Ophthalmol. 1998; 82:1444–144517. 11. Ronday MJ, Stilma JS, Barbe RF, et al. Aetiology of uveitis in Sierra Leone, West Africa. Br J Ophthalmol. 1996; 80:956–961
11. Ronday MJ, Stilma JS, Barbe RF. Aetiology of uveitis in Sierra Leone, West Africa. Br J Ophthalmol. 1996;80 :956–961.
12. Aubert François, Guittard Philippe. Essentiel Médical de poche. 2nd édition. Universités Francophones. Paris, France: Ellipses Aupelf/Uref Publisher; 1995; 45: 448–451.
13. Kanski Jack J. Clinical ophthalmology, a systematic approach. 6th ed. Oxford, UK: Butterworth Heinemann Elsevier Publisher; 2007
14. Bowie WR, King AS, Werker DH. Outbreak of toxoplasmosis associated with municipal drinking water. Lancet . 1997; 350 :173–177