DOI: 10.14260/jemds/2015/580

CASE REPORT

SUBACUTE SCLEROSING PANENCEPHALITIS: A RARE NEUROLOGICAL DISORDER IN PREGNANCY

Minakeshi Rana[1], Varinder Mohan Rana[2], Abha Singh[3], Seema Singhal[4]

HOW TO CITE THIS ARTICLE:
Minakeshi Rana, Varinder Mohan Rana, Abha Singh, Seema Singhal, "Subacute Sclerosing Panencephalitis: A Rare Neurological Disorder in Pregnancy". Journal of Evolution of Medical and Dental Sciences 2015; Vol. 4, Issue 23, March 19; Page: 4033-4036, DOI: 10.14260/jemds/2015/580

ABSTRACT: Subacute Sclerosing Panencephalitis (SSPE) is a rare, chronic progressive demyelinating disease. Incidence of subacute sclerosing panencephalitis has declined after vaccination but annual incidence is quite high and in India its Incidence is 21 per million population. We had a case of term pregnancy with a history of forgetfulness, abnormal behaviour, abnormal movement of body and altered sensorium. Detailed history, neurologic examination, serology (CSF & serum) and MRI brain clinched the diagnosis. Patient underwent LSCS for fetal distress. She was put on trial of interferon-alpha 2b but she developed superadded infection and died.

KEYWORDS: Subacute sclerosing panencephalitis (SSPE), Measles, Pregnancy, Myoclonic, MRI, Interferon-alpha 2b.

INTRODUCTION: Subacute Sclerosing Panencephalitis (SSPE) is a rare, chronic progressive demyelinating disease of the central nervous system associated with a chronic non-permissive infection of brain tissue with measles virus. Fewer than 10 cases per year are reported in the United States.[1] the incidence has declined substantially after introduction of measles vaccine. The annual incidence is still quite high but variable among developing countries. Saha et al reported an annual incidence of 21 per million population in India.[2] in comparison with 2.4 per million population in Middle East.[3,4] It is a slow virus infection caused by defective measles virus. The term subacute sclerosing panencephalitis has been used since Greenfield suggested it in 1960 to designate a condition due to a persistent virus involving both grey matter and white matter.[5] Most patients give a history of primary measles infection at an early age which is followed after a latent period of 6-8 years by the development of a progressive neurologic disorder. 85% of patients are 5-15 years of age at time of diagnosis.

The initial symptoms are usually subtle and include intellectual deterioration and behavioural changes without any apparent neurological signs and findings. As disease advances non – specific manifestations evolve into disturbances in motor dysfunction and development of periodic stereotyped myoclonic jerks. Myoclonic jerks initially involve head and subsequently trunk and limbs. Patients frequently develop pyramidal and extra-pyramidal signs. Few patients may develop ataxia, dystonia and dyskinesia. Generalised tonic – clonic seizures and partial seizures may also occur. Ocular and visual manifestations are reported in 10% - 50% of patients, which include cortical blindness, chorioretinitis and optic atrophy. Once myoclonus is evident the clinical diagnosis is seldom a problem. However, subtle behavioural changes at an early stage of disease are frequently missed by relatives. Many such patients are often treated by a psychiatrist at this stage. We report a case which was referred from psychiatric department with diagnosis of Primigravida with 9 months amenohorea and Catatonic Schizophrenia.

DOI: 10.14260/jemds/2015/580

CASE REPORT

CASE REPORT: A 20 - year old female referred from the Psychiatric department of LHMC & SKKH, New Delhi with amenorrhea nine months and in early labour. Patient was unable to give the history. Patient's mother revealed that there was a history of forgetfulness, abnormal behavior, and abnormal movements of body and altered sensorium for the last two months. There was difficulty in walking, swallowing, speaking and also incontinence of urine. Past history revealed history of measles at age of 8 years.

On general physical examination patient was drowsy, mute, disoriented with time, place and person. Her pulse rate was 80 bts/ min, Blood pressure 120/80 and she had mild pallor. She was unable to perceive even hand movements or even a beam of light. Examination of optic discs revealed no abnormality. Pupils were of normal size and both direct and indirect light reflexes were normal. Her gait was mildly ataxic, she had generalized hypertonia, all deep tendon reflexes were exaggerated, and both plantars were up going. On careful observation the patient had periodic stereotyped left sided hemimyoclonic jerks involving her left shoulder, arm and leg. She had also subtle hemifacial jerks with simultaneous closure of both eyes.

On obstetrics examination, per abdomen uterus was of term size, fetal heart rate was 140bts/min, P/V examination: Os 2cm, cervix early effacement, vertex -3 station and pelvis was normal in shape and size.

Laboratory work up did not reveal any abnormality in blood and urine. While the patient was being investigated, she went into labour and caesarean section (LSCS) was done for fetal distress after neurological and anaesthetic consultation.

Electroencephalography (EEG) showed a characteristic periodic pattern with bursts every 3 to 8 seconds of high voltage, sharp slow waves followed by periods of attenuated flat background. MRI brain revealed subcortical hyperintensities in T2 -weighted images involving right frontal and bilateral parieto-occipital lobes (Fig. 1). Both serum and cerebrospinal fluid were strongly positive for anti-measles Ig G antibodies. Serum copper was 270mg/dl (70-140mg/dl.) and serum ceruloplasmin was 0.843.

Figure 1: MRI brain showing subcortical hyperintesities in T2 – images involving right frontal and bilateral parieto-occipital lobes.

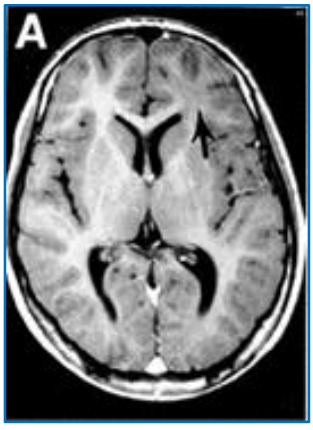

Fig.1

DOI: 10.14260/jemds/2015/580

On the basis of her investigations and clinical co-relation the diagnosis of subacute sclerosing panencephalitis (SSPE) was made. She was put on trial of interferon-alpha 2b 3miu twice a week for 8 weeks. Patient's attendant was told about the doubtful benefits of Interferon alpha 2b. Total five doses of interferon alpha were given and patient developed superadded infection. Interferon was withheld. Her autonomic instability worsened, she developed severe pulmonary infection, and died.

DISCUSSION: Subacute sclerosing panencephalitis (SSPE) is a slow virus disease caused by persistent infection by mutant measles virus. It affects children, it is uncommon after 18 years of age and the disease has a more aggressive course in adults. It rapidly progresses during pregnancy. It has been suggested that relative older age of presentation and unusually rapid neurological deterioration are partially due to immunological and hormonal alternations of pregnancy. Most of the patients with SSPE survive for 1-3 years after diagnosis, with a mean survival of about 18 months. In acute fulminant SSPE, the disease rapidly evolves leading to death within three months of diagnosis. Approximately 10% of patients have such fulminant course.[6] In rapidly evolving SSPE various stages of disease cannot be recognized. The exact mechanism producing an acute fulminant course is not known. Several factors such as exposure to measles at an early age, viral virulence, impaired host defence mechanisms and concurrent infections with other viruses have been suggested as factors responsible for producing a rapid course of the disease.[7,8,9]

The disease is still common in developing and underdeveloped countries. One of the most important limitations in treatment of SSPE is difficulty in recognizing early manifestations of the disease, when the inflammatory changes are still reversible. Diagnosis is difficult in pregnancy. The diagnosis is based upon typical CSF changes and a characteristic EEG pattern. The diagnosis of SSPE can be made if patients fulfil three of the five criteria given by Dyken PR.[10]

1. Clinical progressive, subacute mental deterioration with typical signs like myoclonus.
2. EEG, periodic, stereotyped high voltage discharges.
3. Cerebrospinal fluid raised gammaglobulin or oligoclonal bands.
4. Measles antibodies raised titre in serum(>1:256) and CSF(>1:4)
5. Brain biopsy suggestive of panencephalitis.

Treatments available are still costly and only available at few centres in the world. Moreover these treatments are not curative and only help in buying time for these patients. The families of patients with SSPE have a lot of physical, psychological and economical stresses to endure. A great deal of external support is required for these suffering families to cope with these stresses. At present effective measles vaccine seems to be the only solution to the problem of this deadly neurological disorder.

REFERENCES:

1. Swoveland PT, Johnson KP. Subacute sclerosing panencephalitis and other paramyxovirus infections. In: Mckendall RR, ed. Handbook of clinical neurology.Vol 12 (56).Virus diseases. Amsterdam: North Holland Publishing Company, 1989: 417-37.
2. Saha V, John TJ, Mukundan p,etal. High incidence of subacute sclerosing panencephalitis in South India.Epidemiol infect 1990; 104: 151-6.

DOI: 10.14260/jemds/2015/580

3. Radhakrishnan K, Thacker AK, Maloo JC, etal. Descriptive epidemiology of some rare neurological diseases in Benghazi, Libya. Neuroepidemology 1988; 7: 159-64.
4. Yakub BA. Subacute sclerosing panencephalitis: early diagnosis, prognostic factors and natural history. J Neurol SCI 1996; 139; 22-34.
5. Greenfield JG. Enecephalitis and encephalomyelitis in England and Wales during last decade.Brain1950; 73: 141-66.
6. Risk WS, Haddad FS. The variable natura history of SSPE: a study of 118 cases from Middle East. Arch Neurol; 1979: 56: 610-14.
7. Pe Benito R, Naqvi SH, Arca MM, etal. Fulminating subacute sclerosing panencephalitis: case report and literature review. Clin Pediatr (Phila) 1997; 36: 149 -54.
8. Alexander M, Singh s, Gnanamuthu c,etal SUBACUTESCLEROSING PANENCEPHALITIS:CT andMR imaging in a rapidly progressive case. Neurology India 1999; 47: 304-7.
9. Gokcil Z, Odobasi Z, Aksu A, etal. Acute fulminant SSPE. Clinical and EEG features. Clin Electroencephalogra 1998; 9: 43-8.
10. Dyken PR: Subacute Sclerosing Panencephalities. Current status Neurol Clin. 1985; 3: 179-196 [Pubmed].

AUTHORS:

1. Minakeshi Rana
2. Varinder Mohan Rana
3. Abha Singh
4. Seema Singhal

PARTICULARS OF CONTRIBUTORS:

1. Senior Resident, Department of Obstetrics & Gynaecology, LHMC, New Delhi.
2. Junior Resident, Department of Medicine, LHMC, New Delhi.
3. Professor, Department of Obstetrics & Gynaecology, LHMC, New Delhi.
4. Associate Professor, Department of Obstetrics & Gynaecology, LHMC, New Delhi.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Minakeshi Rana,
H. No. 24, Lane 51,
Greater Kailash,
Jammu.
E-mail: minakeshirana76@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None

Date of Submission: 20/02/2015.
Date of Peer Review: 21/02/2015.
Date of Acceptance: 07/03/2015.
Date of Publishing: 18/03/2015.