CASE REPORT

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Adult Subacute Sclerosing Panencephalitis Presenting as Cortical Blindness

Vaibhav Deorari, Neeren Ravela**, Prabhat Kumar***, Ashutosh Biswas*****

Abstract

Subacute sclerosing panencephalitis (SSPE) is a neurodegenerative disease of childhood and young adolescence. SSPE in adults is uncommon and can have varied presentations ranging from behavioural changes, myoclonus, seizures and cognitive decline. Cortical blindness as a presenting feature of SSPE is uncommon in adults. Herein, we present a case of fulminant SSPE in an adult who presented with cortical blindness.

Key words: Encephalitis, measles, myoclonus.

Introduction

Measles virus is a RNA virus of the paramyxoviridae family which spreads via nasopharyngeal secretions. Measles, despite being a vaccine preventable disease, is still common in developing countries like India. SSPE is a chronic progressive neurodegenerative disorder which occurs after 2 - 10 years of primary infection[1]. The manifestations of SSPE are generally seen between 5 - 15 years of age and the annual incidence of laboratory diagnosed cases of SSPE was estimated to be only 2.14 cases per million populations in India[2]. Adult onset SSPE is uncommon and cortical blindness as a presenting feature is even rarer.

Case report

A 21-year-old male presented with complaints of painless progressive loss of vision in bilateral eyes for 15 days and abnormal behaviour in the form of inappropriate talking, irritability and laughter for 7 days. The relatives also noted that he was having intermittent jerky movement of all four limbs for last 3 days. On examination, he had decerebrate posturing of all four limbs and speech was incomprehensible. Fundus examination showed bilateral disc pallor with background erythema. Bilateral plantar response was extensor and myoclonic jerks were noted in all limbs.

Blood investigations showed normal haemogram, renal function tests, liver function tests and serum electrolytes. MRI brain showed FLAIR and T2 hyperintensities and T1 iso-intensity in right occipital lobe and right precentral gyrus with no diffusion restriction, no blooming and no post-contrast enhancement (Fig. 1). MR spectroscopy of right occipital lesion showed increased choline and lactate and decreased N-acetyl aspartate (NAA) peak (Fig. 2). Electroencephalogram was abnormal with background slowing and generalised periodic discharges. Cerebrospinal fluid (CSF) analysis was acellular with sugar of 123 mg/dl and protein of 209 mg/dl. CSF sample sent for gram stain, cryptococcal antigen and GeneXpert MTB was negative. CSF PCR done for HSV1 and 2, adenovirus, enterovirus, cytomegalovirus and Japanese encephalitis viruses were negative. CSF was also negative for various autoimmune encephalitis antibodies.

He was electively intubated for airway protection and empirically started on broad spectrum antibiotics, anti-tuberculous drugs and anti-epileptics. A brain biopsy taken from right occipital region showed meningeal infiltration by mature lymphocytes, subpial gliosis, loss of neurons in cortex and reactive gliosis along with periventricular collection of lymphocytes and plasma cells. There were inclusions within the neurons and oligodendroglial cells. Brain biopsy was suggestive of subacute sclerosing meningoencephalitis (SSPE). Subsequently, CSF and serum sent for anti-measles IgG antibodies too came positive, although there was no evidence of measles in the past.

A final diagnosis of SSPE was made and he was started on oral Isoprinosine (100 mg/kg/day) and intrathecal interferon therapy (1,00,000 units/m^2 of body surface area), given five days in a week. However, there was worsening of myoclonic jerks for which clobazam was added. He also developed autonomic dysfunction during hospital stay. He was discharged after 45 days of hospital stay. His jerks, vision and sensorium had no improvement after two months of follow-up.

Senior Resident, **Junior Resident, ***Assistant Professor, ****Professor, Department of Medicine, AIIMS, All India Institute of Medical Sciences, Ansari Nagar, New Delhi - 110 029.

Corresponding Author: Dr Prabhat Kumar, Assistant Professor, Department of Medicine, AIIMS, All India Institute of Medical Sciences, Ansari Nagar, New Delhi - 110 029. Phone: 9968123167, E-mail: drkumar.prabhat@gmail.com.

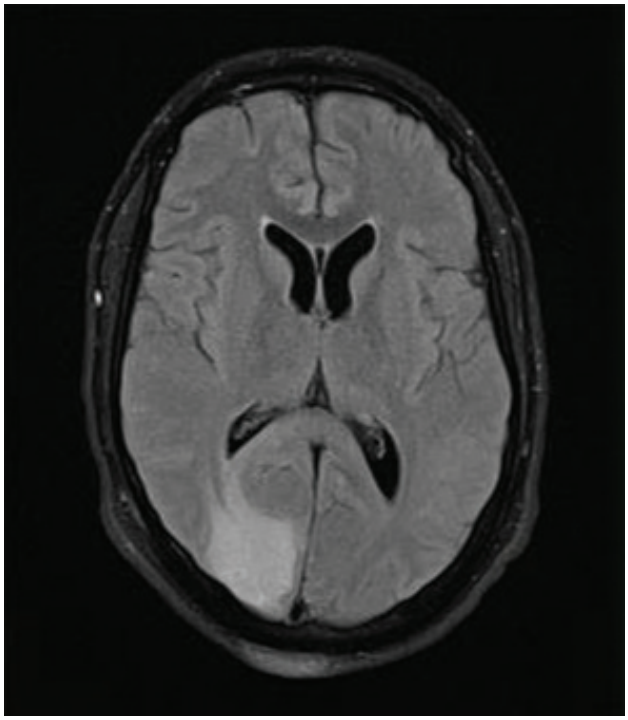

Fig. 1: *T2 FLAIR magnetic resonance image showing hyperintensity in right occipital region.*

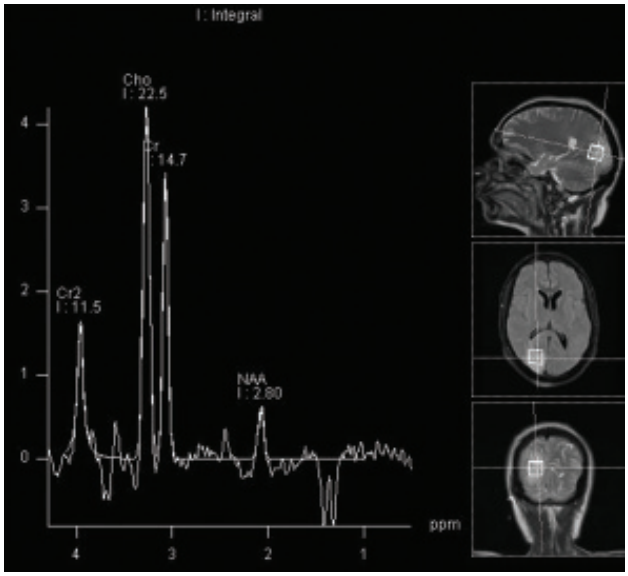

Fig. 2: *MR spectroscopy of right occipital lesion showing increased choline and lactate peak and decreased N-acetyl aspartate (NAA) peak.*

Discussion

Measles is a highly contagious disease, which presents with upper respiratory tract infections and, less commonly, can have neurological manifestations. Neurological manifestations include post-measles encephalitis, inclusion body encephalitis, subacute sclerosing panencephalitis (SSPE) and transverse encephalitis. SSPE is frequently reported in childhood and has low prevalence in adult patients. As reported by a study, age of presentation ranged from 3 to 35 years with mean age of 12 years. They reported latent period of 2.5 years to 34 years with mean latent period of 9.5 years from measles infection to SSPE diagnosis[3]. The most common presenting manifestation in adult SSPE includes myoclonus followed by behavioural changes[3]. Cortical blindness at presentation is common in children, but is rare in adult individuals. The study done by Prashanth *et al*, and Jagtap *et al*, showed that only 3 and 2 adult patients had cortical blindness, repectively[4,5].

The diagnosis of SSPE can be made by Dyken's criteria, which include two major and four minor criteria. Major criteria include 1). Elevated cerebrospinal fluid (CSF) anti-measles antibody titers in greater than or equal to 1:4 or ratio greater than or equal to 1:256 in serum, and 2) typical or atypical clinical history (typical includes acute/rapidly progressive, subacute progressive, chronic progressive, and chronic relapsing-remitting, while atypical includes seizures, prolonged stage I, and unusual age of presentation that is either in infancy or adulthood). Minor criteria include the following: 1) Typical EEG findings that include periodic, generalised, bilaterally synchronous and symmetrical high-amplitude slow waves that recur at regular intervals of 5 - 15 seconds called periodic complexes. 2) Increased CSF globulin levels greater than 20% of the total CSF protein. 3) Characteristic histopathological findings on brain biopsy include neuronal degeneration, gliosis with lymphocytic infiltration and presence of inclusion bodies in neuronal and glial cells. 4) Specialised molecular diagnostic test to identify wild-type measles virus mutated genome. For SSPE diagnosis two major and one minor criterion is usually sufficient[6].

There is no definite treatment for SSPE and death invariably occurs within 1 - 3 years of disease onset[7]. However, certain anti-virals and immunomodulators are believed to prolong duration of life in these patients. Isoprinosine, an anti-viral drug is given at a dose of 100 mg/kg alone or with intrathecal interferon-α. The dose of interferon-α is 1,00,000 units/m^2 of body surface area, given five days in a week for six weeks. The combination of these two is believed to be the most effective treatment available for SSPE.

To conclude, the diagnosis of SSPE often gets delayed as it has a long latent period and can have myriad presentations. It is important for clinicians to be aware of the various typical and atypical manifestations of SSPE. Early diagnosis and appropriate treatment might result in favourable outcome. However, the only way to prevent this life-threatening disease is by achieving universal immunisation in childhood.

References

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