Isolated vision loss and disappearing lesions as the manifestation of subacute sclerosing panencephalitis

Sir,

Subacute sclerosing panencephalitis (SSPE) is a devastating disease of central nervous system (CNS) caused by persistent mutant measles virus infection. Although it is a rare disease in developed nations, it is of great concern in developing countries. Patients present with progressive cognitive decline and myoclonus.[1] Visual manifestations are present in 10-50% of patients but they usually occur concurrently with above neurological findings. Rarely visual symptoms may precede neurological manifestations.[2] Here we report a case of SSPE that had presentation in the form of acute vision loss and remained undiagnosed for 2 years until he developed typical manifestations of SSPE.

This 14-year-old boy presented to us with 6 months history of myoclonic jerks involving all four limbs. There was a history of gradually deteriorating cognitive functions, decreased speech output, stiffness of all four limbs and urinary and stool incontinence since last 1 month.

Relatives gave history of acute onset diminution of vision in left eye around 2 and half years back. Review of medical records at that time revealed visual acuity of 6/60 and disc edema in left eye. Gadolinium enhanced magnetic resonance imaging (MRI) of brain and cerebrospinal fluid examination (CSF) was normal. A diagnosis of left eye papillitis was kept. Patient was given a short course of oral steroids. Over next 2 months, patient's vision in left eye deteriorated further to finger counting till two feet. Fundoscopy in left eye then showed disc pallor. Optical Coherence Tomography (OCT) of left eye at that time showed thinning of retina including macula [Figure 1a].

Six months back when patient started developing the myoclonic jerks, he underwent MRI of brain. It showed multifocal confluent subcortical white matter hyperintensities in T2 weighted and fluid inversion recovery (FLAIR) sequences without contrast enhancement involving centrum semiovale bilaterally and left occipital region [Figures 1b1 and 1b2]. A diagnosis of acute disseminated encephalomyelitis was kept. Patient was given a short course of intravenous methylprednisolone with antiepileptic drugs.

Now, when we examined the patient, he had myoclonic jerks involving limbs, face, and trunk occurring at frequency of 12-15 per minute. Patient was akinetic and mute. Fundus examination showed temporal pallor of optic disc on right side, optic atrophy, pigment-splinters, and contracture of internal limiting membrane on left side [Figures 1c1 and 1c2]. Tone was increased and deep tendon reflexes were exaggerated in all four limbs. Planters were bilaterally flexor.

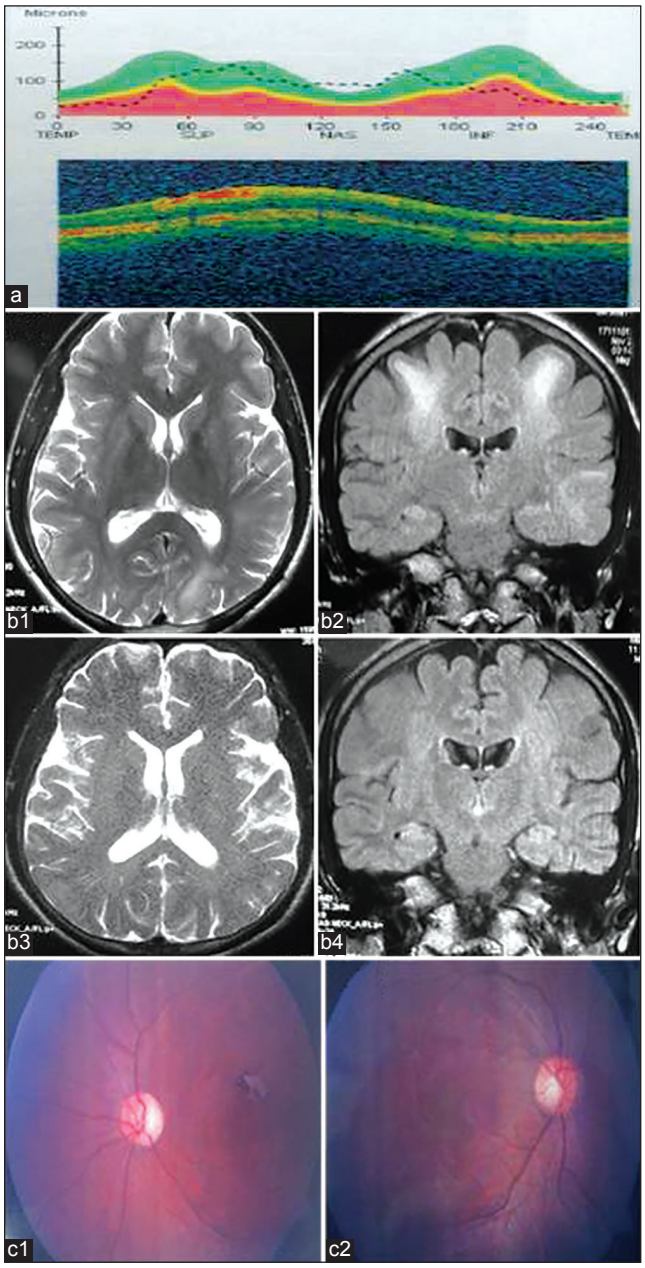

Figure 1: (a) Optical Coherence Tomography of left eye showed retinal thinning; (b1) MRI of brain axial T2 weighted image showing left occipital subcortical white matter hyperintensity; (b2) Coronal (FLAIR) imaging showing bilateral parietal subcortical white matter hyperintensities; (b3 and b4) Repeat MRI done 6 months later shows complete resolution of the lesions; (c1) Left eye fundus photograph showing pigment-splinters and contracture of internal limiting membrane associated with pallor of optic disc on temporal side; (c2) Right eye fundus photograph showing pallor of optic disc on temporal side

Electroencephalogram (EEG) showed bilaterally symmetrical, synchronous, periodic, high voltage bursts of polyphasic, stereotyped delta waves. A provisional diagnosis of SSPE was kept. CSF examination was positive for anti-measles IgG antibodies. MRI of brain was completely normal implying complete resolution of previous white matter lesions [Figures 1b3 and 1b4]. Patient was started on weekly intrathecal interferons and oral clonazepam and valproate. Relatives were explained about passive physiotherapy and other supportive care for the patient.

Visual manifestations are present in 10-50% of patients and they usually occur concurrently with above neurological findings but rarely may precede it; however it is very rare to have long latency between ophthalmological and neurological manifestations.[2,3] The causes include disc edema (papillitis or papilledema), optic atrophy, visual field defects, nystagmus, supranuclear gaze palsy, retinitis, chorioretinitis, macular pigment epithelial abnormalities, macular edema, macular haemorrhage, gliotic retinal scar, and internal limiting membrane contracture.[4]

This case report not only emphasise that isolated visual symptoms could be the presenting symptoms of SSPE but also the fact that there could be long latency (2 years in our case) between visual and typical neurological features of SSPE. We wish to emphasize that SSPE should always be kept as a differential diagnosis in patients presenting with optic neuritis or chorio-retinitis in appropriate clinical settings.

Spontaneous resolution of white matter lesions while the patient was deteriorating clinically (clinic-radiological dissociation), a very rare fact, is worth reporting because it may mislead radiologist or neuroradiologist.[5] It emphasize the fact that SSPE should be kept as a differential diagnosis of spontaneously disappearing white matter lesions especially in appropriate clinical circumstances.

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DOI: 10.4103/0972-2327.144045