

Photo Essay

Unilateral macular chorioretinitis in subacute sclerosing panencephalitis studied by spectral domain optical coherence tomography



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Subacute Sclerosing Panencephalitis (SSPE) was described by Dawson in 1933–34. It is a subacute inflammatory and neurodegenerative encephalitis related to the measles (rubeola) virus and usually affects children and young adults. The ocular manifestations of SSPE were described and classified by Robb and Watters in 1970.¹ These include papillitis, optic atrophy, chorioretinitis and cortical blindness. Various cases of SSPE with chorioretinitis have been reported.^{2,3} However, only a few cases have been described with a unilateral macular chorioretinitis in SSPE.⁴ We describe a case of SSPE with a unilateral isolated macular chorioretinitis, studied by Spectral domain Optical coherence tomography.

An 11-year-old male child presented with gradual decrease in scholastic performance for 3 months, spontaneous falls without loss of consciousness for 2 months and progressively increasing jerky movements of the body along with slippage of objects from hands for 2 months. The patient also complained of decrease in vision in right eye for 3 months. There was no history of trauma, fever, seizures, limb weakness or bladder/bowel dysfunction. There was a past history of febrile illness with rash 3 years back. The rash started from the face and progressed to the trunk in 1–2 days; and involved the trunk more than the extremities. The patient had never received any vaccination or immunization.

On examination, the patient was conscious and oriented to time, place and person. His general physical examination was within normal limits. The neurological examination showed frequent myoclonic jerks of all the limbs, trunk and neck, aggravated with activity. There was no limb weakness, cranial nerve dysfunction or sensory loss.

On ocular examination, he had a best corrected visual acuity (BCVA) of 10/200 in the right eye and 20/20 in the left eye. The anterior segment was unremarkable, and there were no cells or flare in the anterior chamber in both the eyes. The lens was clear and there was no vitritis in either eye. The pupils were normal in size and reaction in both the eyes. The fundus of the right eye showed hyperpigmentation at the macula, while the disc, blood vessels and peripheral retina were unremarkable (Fig. 1). The fundus of the left eye was normal. Optical coherence tomography (OCT) of the right eye performed at the fovea (Spectral domain OCT), with six radial scans revealed severe thinning of all layers of retina with photoreceptor damage (Fig. 2). The OCT scan of the left eye was unremarkable.

The patient was referred to our neurology department, where a complete neurological examination, haematological examination and a cerebrospinal fluid (CSF) tap was done. The blood picture showed a normal total leucocyte count (TLC) with lymphocytosis (50% lymphocytes). Other blood chemistry studies like liver function tests (LFT), kidney function tests (KFT), serum calcium, magnesium, sodium, potassium, copper and ceruloplasmin levels were normal. The CSF tap showed clear fluid with 2–3 cells/ μ l, all lymphocytes, and a protein concentration of 47 mg/100 ml. The CSF electrophoresis showed the γ -globulin fraction to be 43% and the IgG titres against measles to be 1:512. The patient underwent a magnetic resonance imaging (MRI) of the brain, that revealed a normal study (Fig. 3).

On the basis of the clinical and ocular features, the elevated γ -globulin levels in CSF, elevated anti-measles IgG

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Fig. 1. Fundus photograph of the right eye showing hyperpigmentation at the macula, suggestive of healed macular chorioretinitis.

titres in CSF and a normal MRI, the diagnosis of Subacute sclerosing panencephalitis was made. The patient was started on oral valproate and clonazepam and kept on a close follow up.

Comment

Subacute sclerosing panencephalitis (SSPE) is a progressive inflammatory disease of the central nervous system caused by a persistent aberrant measles virus infection. Its annual incidence is higher in developing countries as a larger proportion of the total population is younger than 2 years of age. In India, the annual incidence rate is reported

to be as high as 21 per million. The clinical features consist of personality and behavioral changes and worsening school performance, followed by myoclonic seizures, dyspraxias, paresis, memory impairment, language difficulties, blindness, and eventually coma.⁵ Most patients give a history of having acquired measles before 2 years of age. The latent period between measles infection and SSPE is 6–8 years in most cases but may range from 3 months to 18 years. Once SSPE is identified, clinical progression is variable. Death usually occurs within 1–3 years after the onset of symptoms.

Ocular findings have been described in about 50% of patients with SSPE, out of which 88% are bilateral. These include papillitis, optic atrophy, chorioretinitis and cortical blindness.¹ The most characteristic lesion however, is a necrotizing retinitis. Occasionally described as a chorioretinitis, it appears to preferentially affect the retina with secondary involvement of the retinal pigment epithelium and choroid. This process may progress to involve most of the posterior pole and peripheral retina. There is little or no vitreous inflammation or involvement of retinal vessels. These features help in distinguishing SSPE from other inflammatory retinal disorders such as toxoplasmosis and Behçet disease. Visual loss can also occur due to cortical lesions. Basal ganglia involvement is seen in 20–35% cases on MRI.⁵ In our case, however, the MRI study was normal at presentation.

Most cases of chorioretinitis reported in SSPE are bilateral.^{2,3} However, few cases, like our case, have been described that have an isolated unilateral chorioretinitis.⁴ Hence, SSPE should be kept in mind in young children presenting with unilateral chorioretinitis as well, especially in developing countries.

Conflict of interest

The authors declared that there is no conflict of interest.

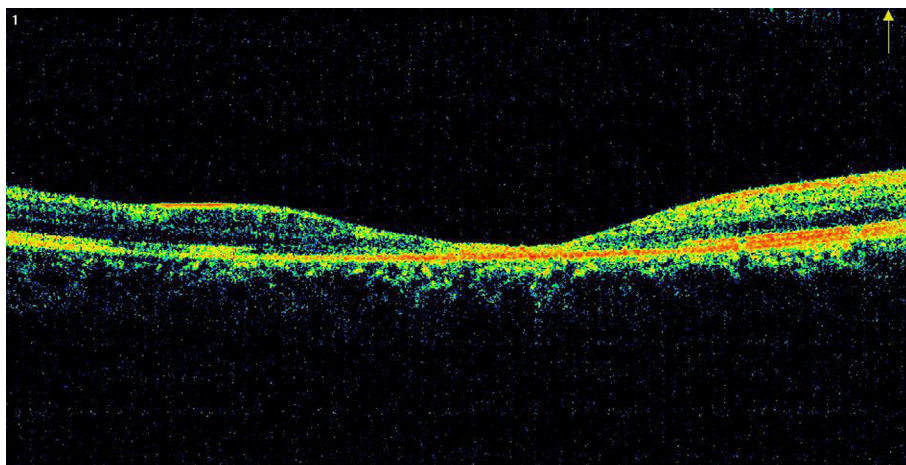


Fig. 2. Spectral domain Optical coherence tomography of right macula showing severe thinning of all retinal layers at the fovea.

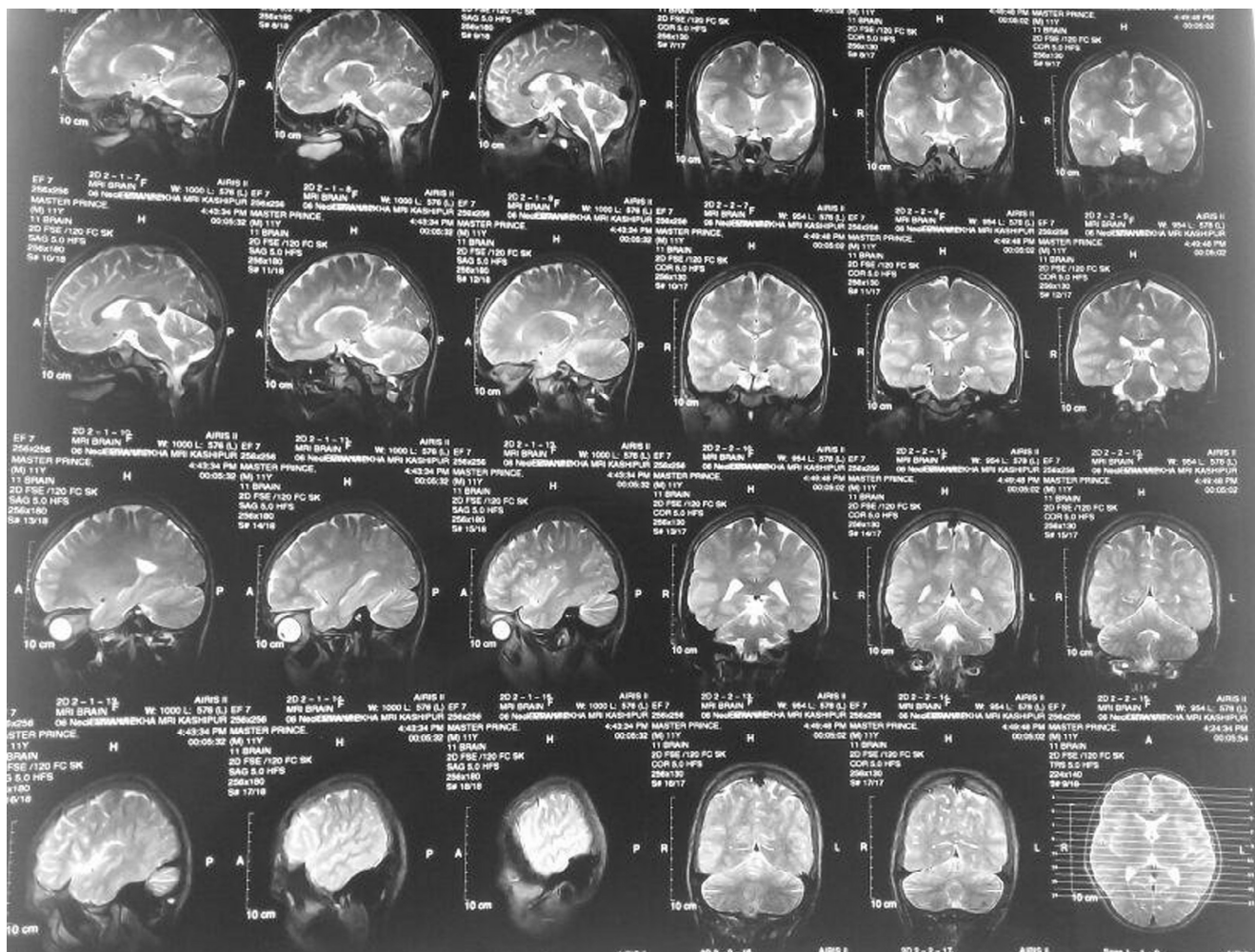


Fig. 3. Magnetic Resonance imaging (MRI) of the brain showing a normal study.

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