

A Case of Subacute Sclerosing Panencephalitis with Widespread Chorioretinitis

Yaygın Koryoretinitli Bir Subakut Sklerozan Panensefalit Olgusu

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ABSTRACT

The purpose of this study was to report a SSPE patient presenting with bilateral optic atrophy, maculopathy, and widespread chorioretinitis. A 14-year-old girl was admitted to our hospital with the complaint of sudden painless visual loss in both eyes with a 1-month interval. Best corrected visual acuity was finger counting level in both eyes. Bilateral optic atrophy, macular scar, and chorioretinitis affecting the whole posterior segment were observed. Encephalopathy and convulsions developed 2 months after the presentation of eye findings. She was diagnosed with SSPE according to clinical, electroencephalographic, and cerebrospinal fluid findings. SSPE should be considered in the differential diagnosis of optic atrophy and atypical widespread chorioretinitis in young patients, especially in countries where SSPE has a higher incidence rate.

Key Words: SSPE, optic atrophy, chorioretinitis.

ÖZ

Bu çalışmanın amacı, her iki gözde optik atrofi, makülopati ve yaygın koryoretinit bulgularıyla başlayan bir SSPE olgusunu sunmaktır. On dört yaşında bayan hasta, her iki gözde bir ay ara ile gelişen, ağrısız ani görme kaybı şikayeti ile hastanemize başvurdu. Düzeltilmiş uzak görme keskinliği her iki gözde el hareketlerini seviyesindeydi. Arka segment muayenesinde, her iki gözde optik atrofi, makülada skar ve tüm arka kutbu etkilemiş koryoretinit bulguları izlendi. 2 ay sonra hastada konvulsiyonlar ve ensefalopati tablosu gelişti. Klinik tablo, elektroensefalografik değerlendirme ve beyin omurilik sıvısının incelenmesi sonucunda elde edilen bulgularla hastaya SSPE tanısı kondu. Özellikle SSPE insidansının daha yüksek olduğu ülkelerde, optik atrofi ve atipik yaygın koryoretinit izlenen genç hastalarda, SSPE tanısı akılda tutulmalıdır.

Anahtar Kelimeler: SSPE, optik atrofi, koryoretinit.

INTRODUCTION

Subacute sclerosing panencephalitis (SSPE) is a rare progressive neurodegenerative disorder caused by persistent infection of the brain by an aberrant measles virus called SSPE virus.¹ SSPE is a slow virus infection and characteristic clinical manifestations include personality and behavioral changes, cognitive decline, myoclonic seizures, paresis, and eventually obtundation, stupor, and coma. Ocular involvement occurs in up to 50% of cases and optic neuritis, papilledema, chorioretinitis, homonymous visual field defects, and cortical blindness are characteristic ocular findings.¹⁻³ Herein we describe a 14-year-old girl with sudden loss of vision as an initial symptom of SSPE.

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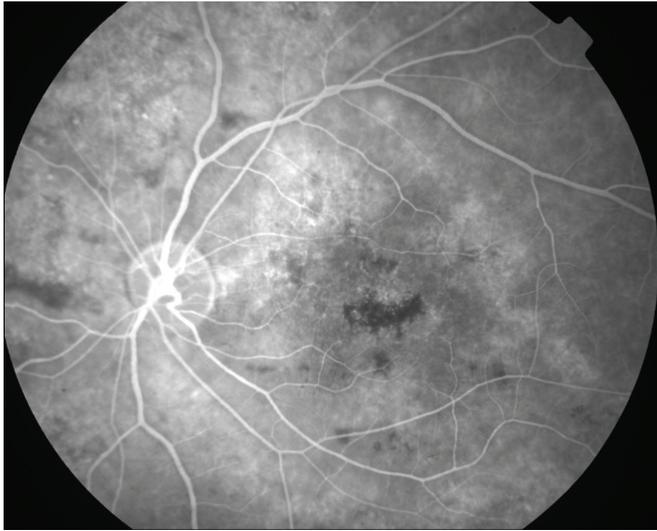


Figure 1: FA of left eye demonstrating wide spread chorioretinal scar without any signs of active inflammation and any leakage. There is intensive RPE hypertrophy in the macula.



Figure 2: The RPE hypertrophy is also visible in other quadrants and extending to the periphery in FFA of right eye.

CASE REPORT

A 14-year-old girl was admitted to our clinic with the complaint of sudden painless loss of vision in both eyes. Visual loss was encountered in her right eye first, and then after 1 month a considerable visual loss occurred in the second eye. Best corrected visual acuity was at finger counting level in both eyes. Her anterior segment examination and intraocular pressures were normal. Her fundus examination showed bilateral optic atrophy, sclerosis, and a beaten bronze appearance on the macula.

In addition, epiretinal membrane and patches of retina pigment epithelial hyperpigmentation were noted. There were no signs of active retinal vasculitis. Widespread scars as a result of necrotizing retinitis and small islands of hemorrhagic foci were also observed at the peripheral retina.

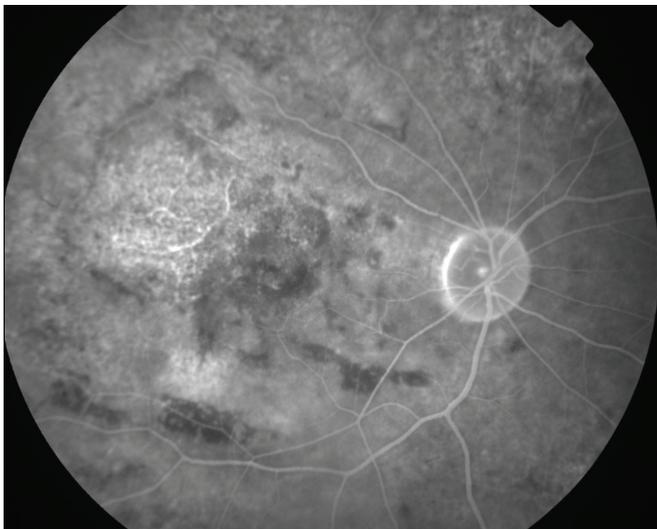


Figure 3: On the right eye staining of wide spread scar tissue on the macula.

FA revealed hypofluorescent spots at hyperpigmented areas of the macula. The OCT revealed widespread destruction of all retinal layers and thinning of the retina. ERG and EOG examinations suggested widespread rod cone degeneration. No systemic or neurological abnormalities were recorded during her physical examination. Her family history was negative for ocular or any other systemic or neurological disturbances. Her school performance was good. Laboratory investigations revealed an increased white blood cell count ($18.5/\text{mm}^3$) and ESR (26 mm/h). Liver and kidney function tests, anti-double-stranded DNA antibody, anti-nuclear antibody, blood and urine culture, tuberculin test, chest X-ray, and abdominal ultrasound were all normal. Serology for toxoplasmosis, cytomegalovirus, and Borreliosis, VDRL, and FTA was negative. Visual evoked potentials showed reduced amplitude and delayed P100 latency.

Eight weeks after presentation, she developed convulsions and encephalopathy. Lumbar puncture (LP) was performed. Cerebrospinal fluid (CSF) showed 2 cells/ mm^3 (all lymphocytes), 31 mg/mL protein, and 56 mg/dL glucose and negative results for Gram and acid-fast stains and polymerase chain reaction (PCR) for herpes simplex virus (HSV) and Epstein-Barr virus (EBV). Results of borrelia serology were negative. Antimeasles IgG titers were elevated in both serum and CSF. Immunofixation electrophoresis of CSF revealed an oligoclonal band.

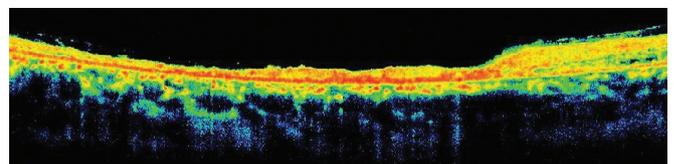


Figure 4: Massive destruction of the normal retinal structure nearly in all of the layers on the OCT images.

On T2-weighted imaging of magnetic resonance imaging (MRI) study, increased signal intensity of the white matter in the right parietooccipital region was seen. EEG showed diffusely slow background activity and periodic slow waves. These periodic generalized slow waves did not disappear after intravenous diazepam injection. The diagnosis of SSPE was confirmed by these EEG and CSF examination findings. She was hospitalized and isoprinosine (100 mg/kg) and carbamazepine (10 mg/kg) were started.

DISCUSSION

Our case is an unusual presentation of SSPE as severe visual loss preceded the typical neurological signs and symptoms. Differential diagnosis of widespread chorioretinitis, maculopathy, and optic atrophy with neurological involvement was made by excluding syphilitic retinitis and Lyme disease.

Ocular involvement of syphilis may develop at any stage of the disease. Since it can affect all the structures in the eye, it is called the 'great mimicker'. It causes a wide range of clinical manifestations such as episcleritis, panuveitis, chorioretinitis, macular edema, optic neuritis, and optic atrophy. The serum examination, which revealed negative results for both non-treponemal (VDRL) and treponemal (FTA) tests, excluded the diagnoses of syphilis.

Lyme disease is another disease that has a diverse range of ocular manifestations with CNS involvement. Therefore, it should be kept in mind in cases of chorioretinitis with neurological symptoms. Negative results of the serum and CSF examination for *Borrelia* serology also excluded the diagnosis of Lyme disease.

SSPE is a rare but feared sequela of a common viral infection.^{1,2} Various neuroophthalmic and retinal findings may be observed in SSPE patients.⁴⁻¹⁰ In 10% of cases, ophthalmic manifestations precede the neurological symptoms.³ This is explained by the presence of possible measles virus acquired neurotropism in the retina.¹¹ Necrotizing retinitis is reported as the most characteristic lesion but, in contrast to the present case, it is usually observed as focal areas of retinitis in the macula. Optic atrophy is also a rare presenting sign of SSPE and is an indicator of CNS involvement.^{12,13}

The present case is unusual, presenting with optic nerve involvement that precedes the more characteristic neurological symptoms. Although SSPE is a rare disease, its incidence is higher in our country compared with more developed countries, and so it should be considered in the differential diagnosis of atypical chorioretinitis with optic nerve involvement especially in young patients even in the absence of other neurological symptoms.

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