

Retinitis as the presenting feature of subacute sclerosing panencephalitis in an Indian male: A case report

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Measles virus is a rare but important cause for acute retinitis as it can eventually lead to the fulminant complication of SSPE. We report a case of a young Indian male with acute viral retinitis who subsequently developed SSPE. It is of paramount importance to consider measles virus and SSPE as a cause in an immunocompetent young adult with necrotizing viral retinitis

Key words: Measles, subacute sclerosing panencephalitis, viral retinitis

Subacute sclerosing panencephalitis (SSPE) is a progressive inflammatory disease of the central nervous system caused by a persistent, aberrant measles virus infection. The annual incidence rate varies from one to four per million population in developed countries.^[1] In India the annual incidence rate is reported to be as high as 21 per million population.^[2] We report a case of fulminant retinitis, which evolved to present as SSPE after 18 months.

Case Report

A 24-year-old Asian Indian male presented to our uvea clinic with a complaint of difficulty in vision since 1 month. Systemic history was unremarkable. His best corrected visual acuity in right eye was 6/24 for distance and N8 for near and in the left eye was counting fingers at 1 m with near vision less than N36. Anterior segment was unremarkable in both eyes. He had bilateral vitreous haze and fundus examination revealed necrotizing hemorrhagic retinitis in the right eye and acute retinitis in the left eye [Fig. 1]. Fluorescein angiogram showed early blockage of background fluorescence, followed by late staining of the retinitis lesions [Fig. 2]. SD-OCT showed thinning of inner retinal layers with hyporeflective spaces in outer retinal layers with increased reflectivity of retinal

layers, more in right eye than left eye [Fig. 3]. Aqueous tap analysis was negative for cytomegalovirus, varicella zoster virus, herpes simplex virus, and chikungunya virus. With a working diagnosis of necrotizing herpetic retinopathy, he was empirically started on oral valacyclovir (1 gm thrice/day) and oral prednisolone (60 mg/day) in a weekly tapering dose. He was regularly followed up and at 1 year of presentation his vision improved with best corrected vision of 6/18, N6 in right eye and 3/60, N36 in left eye, with retina showing healed lesions, however he had developed disc pallor in the left eye. Eighteen months after initial presentation, the patient returned with sudden deterioration of vision in the left eye. On evaluation, right eye vision was same but left eye vision had dropped to hand motion close to face. On ocular examination, right eye was quiet with healed retinal lesions as before. Left eye had developed a subtotal retinal detachment with nasal thinned out retina with breaks [Fig. 4]. Patient underwent pars plana vitrectomy with silicon oil injection. Surgery, and postoperative period were uneventful. At 6 weeks postop, his vision in left eye had improved to 2/60, N36, right eye was stable. At this visit the patient complained of involuntary jerky movements on the left side of his body. He was referred for a neurologic evaluation. On evaluation by neurologist, he was diagnosed to have myoclonus typical of SSPE and was advised electroencephalogram, neuroimaging, and serum and cerebrospinal fluid evaluation. EEG report showed generalized epileptiform activity with myoclonic jerks suggestive of SSPE. CSF titers for HSV, cryptococcus, and CMV were negative; however, CSF titer for measles IgG was 1:512 (normal <1:4) and IgM was 1:32 (normal <1:4). Serum panel for measles IgG was 144.83 U/ml (normal <8 U/ml) and IgM was 1.48 U/ml (normal <8 U/ml). The patient was diagnosed to have SSPE and was put on tablet valproate for his myoclonus.

On follow up, he maintained 6/24, N8 vision in the right eye and 2/60, N36 in the left eye. Anterior segments of both eyes were quiet. Right eye had healed atrophic retinal lesions close to fovea. Left eye was oil filled with disc pallor and healed atrophic retinal lesions [Fig. 4]. Neurologically, he had persistent unilateral myoclonic jerks.

Discussion

Viral retinitis is commonly caused by herpes group^[3] of viruses, namely, cytomegalovirus, herpes simplex, varicella zoster, and occasionally by Epstein Barr virus^[4] and West Nile virus.^[5] Diagnosis is based on clinical presentation and molecular biologic study of ocular fluids. In immunocompetent patients who develop neuropsychiatric symptoms associated with ocular symptoms it is imperative to consider measles virus as a probable cause.

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SSPE is a rare complication of measles virus infection. Diagnosis is based on a typical clinical picture: personality and behavioural changes followed by myoclonic seizures, paresis, dyspraxias, memory impairment, language difficulties, blindness, and eventually obtundation, stupor and coma along with characteristic EEG changes, raised titers of measles antibodies in blood and CSF and typical histopathological findings on brain biopsy or autopsy.^[6] Raised antimeasles antibody titers of 1:256 or greater in serum, and 1:4 or greater in cerebrospinal fluid are considered diagnostic.^[7] In adults, the disease has a more aggressive course and is rapidly fatal in majority of the patients.^[8] Ocular involvement is seen in about 50% of cases.^[9]

Ocular manifestations of SSPE include optic neuritis,^[10] viral retinitis,^[9,11,12] chorioretinitis,^[13-15] and cortical blindness.^[6,16] The most characteristic lesion is a hemorrhagic necrotizing retinitis.^[14,17,18]

The disease process affects the retina first with secondary involvement of the retinal pigment epithelium and choroid. Retinitis appears as a ground-glass whitening of the retina with ill-defined margins and a mottling of the underlying RPE. This process slowly involves most of the posterior pole and peripheral retina. There may be associated retinal changes such as edema, hemorrhage, detachments, venous dilatation, vascular occlusions, and retinal pigment epithelial detachments. There is little or no vitreous inflammation or involvement of retinal vessels. Optic nerve involvement in the form of mild disc pallor to total secondary optic atrophy is usually seen. These

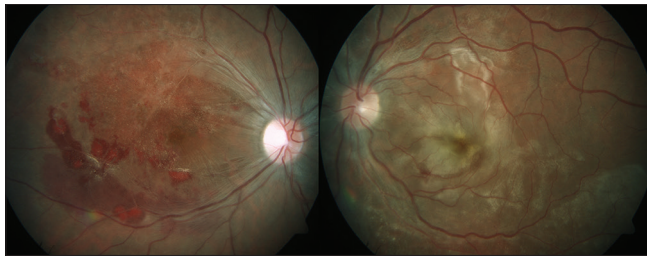


Figure 1: At first visit, right eye had hemorrhagic necrotizing retinitis and left eye had acute retinitis involving the posterior pole

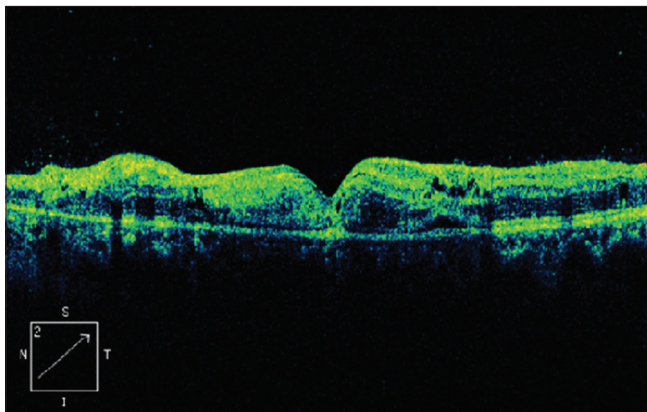


Figure 3: SD-OCT of the right eye (top) shows tissue loss in the inner retinal layers with hyporeflective spaces. OCT of the left eye (bottom) shows retinal edema, hyperreflectivity of inner retinal layers with hyporeflective spaces

features help in distinguishing measles retinitis from other inflammatory retinitis.^[9]

SD-OCT findings of viral retinitis have been described as hyperreflectivity of retinal layers, retinal layer disruption, interruption of ellipsoid zone, epiretinal membranes among other inflammatory changes.^[9] OCT findings in measles retinitis have not been described in literature.

In our patient, initially we saw a picture of hemorrhagic necrotizing retinitis in the right eye and an early retinitis with ground glass appearance in the left eye [Fig. 1]. OCT findings show more disruption of retinal layers and tissue loss in right eye whereas left eye shows hyperreflectivity of retinal layers with schitic spaces and early RPE loss [Fig. 3].

In a multicentric study for treatment of SSPE, about 30–35% cases were found to benefit with a combination of weekly

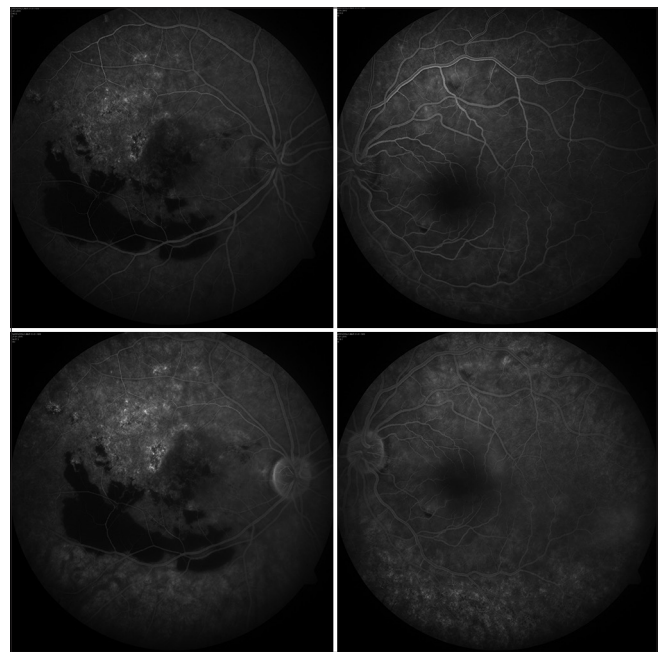


Figure 2: Early phases of angiogram show blockage of background fluorescence and late phases show staining of retinitis lesions. Changes are suggestive of a more acute stage of retinitis in the left eye with diffuse hyperfluorescence and a more necrotic stage in the right eye with staining

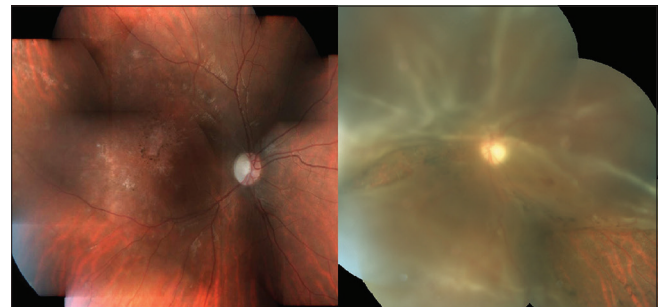


Figure 4: On followup, right eye shows healed retinal lesions with pigment changes. Left eye developed a rhegmatogenous retinal detachment, note the thinned out retina with large break nasally. Left eye had also developed disc pallor

intrathecal INF- α and daily oral isoprinosine. Benefit was defined as either slower progression, prolonged survival, or, less likely, clinical improvement. Early diagnosis and initiation of treatment was considered as a factor aiding in improved clinical outcomes.^[6]

Conclusion

As seen in previous case reports, ocular findings of retinitis very often precede the neurological symptoms, usually within 1–8 months. As the patient was asymptomatic for 18 months, the development SSPE could be unrelated to retinitis. However, an ophthalmologist plays a very important role in such cases as the suspicion of measles as the cause of viral retinitis and an early referral to the neurologist may help in an early diagnosis of SSPE, targeted treatment and aid further research into this grim ailment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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