Abstract:
The most serious consequence of infection by Herpes simplex virus (HSV) is that of encephalitis. Herpes virus causes protean manifestations in the central and peripheral nervous system. We report a case of Herpes simplex encephalitis followed by two rare complications, acute retinal necrosis (ARN) and bilateral sudden sensorineural hearing loss (SSNHL). Acute retinal necrosis also known as Kirisawa's uveitis is a progressive necrotizing retinopathy caused by herpes simplex virus (HSV) or varicella zoster virus (VZV). Postulated causes for SSNHL include viral cochleitis, microvascular events, and autoimmune disorders.

Keyword: herpes encephalitis, acute retinal necrosis, acute sensorineural hearing loss

INTRODUCTION
The most serious consequence of infection by Herpes simplex virus (HSV) is that of encephalitis. It is considered that HSV is the most frequent cause of sporadic necrotizing encephalitis. The clinical picture of herpes simplex virus encephalitis (HSVE) is the spontaneous onset of fever, headache, confusion, and seizures in a child or adult. The pathogenesis of HSVE is not clear. In some cases the virus from the periphery and the brain were identical, but in others were not identical. The distribution of HSV antigen concentrated in the olfactory and limbic system, suggests a possible exogenous olfactory origin. Latent HSV genomes in the central nervous system (CNS) in normal individuals at autopsy were largely confined to olfactory bulbs, pons, and medulla. Occasionally, retinal necrosis has been reported coincident with or after episodes of HSVE (1). Acute retinal necrosis also known as Kirisawa's uveitis is a progressive necrotizing retinopathy caused by herpes simplex virus (HSV) or varicella zoster virus (VZV) (2). Varicella-zoster virus or herpes simplex virus type 1 cause acute retinal necrosis syndrome in patients older than 25 years, whereas herpes simplex virus type 2 causes acute retinal necrosis in patients younger than 25 years. A history of central nervous system infection in a patient with acute retinal necrosis syndrome suggests that herpes simplex virus is likely to be the viral cause (3). The etiology of most cases of sudden sensorineural hearing loss is unknown.
hearing loss (SSNHL) is uncertain (4). Postulated causes for SSNHL include viral cochleitis, microvascular events, and autoimmune disorders. Herpes simplex virus may be a significant etiologic factor in SSNHL, analogous to its possible role in Bell's palsy.

**CASE REPORT:**

A 33 years old lady without any co morbid illness developed fever, headache and vomiting. She was hospitalised on the third day of her illness for two days and was discharged with partial resolution of her symptoms. Three days later she again developed high grade fever with numbness of left side of her body, swaying to left side and twitching of left hand and left side of face and was readmitted. Patient was referred to us two days later as her symptoms were worsening and she developed altered sensorium. On admission patient was drowsy with left hemiparesis involving the face, hemi anaesthesia and left hemi ataxia. Her fundus showed papilledema and she had neck stiffness. Her complete blood count, renal function tests, liver function tests and chest x ray were normal. Her MRI brain showed T2 / FLAIR non suppressible hyperintense lesions in medial part of temporal lobe bilaterally, medulla, pons and cerebellum.

**MRI-FLAIR showing hyperintense lesions in B/L temporal lobe, pons, medulla & cerebellum.**

Her serology for HIV and VDRL were negative. Diagnostic work up for inflammatory diseases including ANA, ANCA and serum angiotensin converting enzyme (ACE) levels were negative. Her CSF protein was mildly elevated (74 mg/dL) with 15 lymphocytes. Her CSF was positive for IGM anti – HSV antibodies. She was treated with acyclovir for 3 weeks. She was ambulant with mild left hemiparesis on discharge. Three weeks later patient was readmitted with sudden painless loss of vision in her left eye and hard of hearing. Her visual acuity was reduced to perception of light in her left eye and her optic fundus showed features suggestive of acute retinal necrosis. Her audiogram showed bilateral sensorineural hearing loss. Repeat CSF analysis was acellular with normal protein and sugar and was negative for IGM anti – HSV antibodies. Repeat MRI brain showed partial resolution of the previous lesions. She was treated with Acyclovir and steroids in consultation with ophthalmologist. Intra tympanic glucocorticoids were administered in view of poor response to systemic steroids. Her vision in the left eye improved to counting of fingers and her deafness remained static on discharge.

**DISCUSSION:**

The diagnosis of ARN is usually based in clinical features. Patient presents with red eye peri orbital pain, hazy vision or decreased vision, appearance of floaters and decreased colour vision. Examination findings include anterior uveitis (iritis), panuveitis, episcleritis or scleritis, keratic precipitates - fine or granulomatous, occlusive retinal vasculitis involving arteries and veins, one or more focus of retinitis, resulting in necrosis with discrete borders located in the retinal periphery with
circumferential spread, vitreitis and optic neuropathy. The use of polymerase chain reaction (PCR) in aqueous humor samples is useful to identify the etiology of the disease. The mainstay of its treatment is antiviral therapy against such as intravenous acyclovir or oral valacyclovir. Systemic and topical corticosteroids together with antiviral therapy are used as an anti-inflammatory treatment to minimize damages to the optic nerve and retinal blood vessels. Because the majority of severe cases of the disease show occlusive retinal vasculitis, a low dosage of aspirin is used as anti-thrombotic treatment. Vitreo-retinal surgery is useful to repair rhegmatogenous retinal detachment, one of the main late-stage complications. Moreover, recent articles have reported some encouraging results of prophylactic vitrectomy before rhegmatogenous retinal detachment occurs. The efficacy of laser photocoagulation to prevent the development or extension of rhegmatogenous retinal detachment is controversial. Despite these treatments, the visual prognosis of acute retinal necrosis is still poor. To prevent fellow eye involvement, intravenous acyclovir is followed by oral acyclovir for 14 weeks. Alternatives to acyclovir include ganciclovir, foscartern, famciclovir, brivudine, and valganciclovir (5). Complications of acute retinal necrosis may include retinal detachment (50%), anterior ischemic neuropathy, central retinal artery occlusion, cataract (from inflammation or steroids) and glaucoma - from inflammation or steroids. Visual prognosis is guarded if retinal detachment, anterior ischemic optic neuropathy, or central retinal artery occlusion occur (6). Strong evidence for the efficacy of any treatment option for patients with sudden sensorineural hearing loss (SSNHL) is not available. Glucocorticoids are considered first-line therapy for SSNHL and may be administered systemically (generally orally) or locally via intratympanic installation. Intratympanic glucocorticoids are often reserved for use when hearing does not improve after a trial of systemic therapy. Intratympanic glucocorticoids can also be used as initial therapy in patients in whom high-dose systemic glucocorticoids should be avoided, such as in patients with diabetes. In addition to glucocorticoids, treatment with an antiviral agent is suggested for treatment of possible HSV-1 infection. The prognosis is poor in patients with profound hearing loss across all frequencies; approximately three-quarters of such patients have no recovery of hearing (7). Auditory rehabilitation may be required for patients with permanent hearing loss. Acute retinal necrosis and bilateral Sensorineural hearing loss could cause a devastating functional impairment to the patient. Though acute retinal necrosis has been reported in patients following HSV encephalitis, simultaneous occurrence of ARN and bilateral SSHL has not been reported so far in the published literature.

**Bibliography:**


