

# A Case Report of Sudden Loss of Vision in Viral Encephalitis - Review of Literature

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**Abstract:** *We present a case of a 36-year-old woman with sudden loss of vision that was initially diagnosed as cortical blindness. After extensive workup herpes simplex virus type 1 (HSV-1) was detected in the patient's cerebrospinal fluid (CSF) by polymerase chain reaction (PCR). Our patient's clinical features, response to treatment as evidenced by improvement in vision, cognition and CRP, as well as the CSF PCR result, are convincing evidence of atypical HSV-1 encephalitis masquerading as an episode of sudden loss of vision.*

**Keywords:** sudden loss of vision, atypical HSV-1 encephalitis, polymerase chain reaction, cortical blindness, generalized tonic clonic seizures, normocytic hypochromic anemia

## 1. Introduction

Herpes simplex encephalitis (HSE) is an acute or subacute illness that causes both general and focal signs of cerebral dysfunction. Brain infection is thought to occur by means of direct neuronal transmission of the virus from a peripheral site to the brain via the trigeminal or olfactory nerve. The exact pathogenesis is unclear, and in fact patients with HSE may have a prodrome of malaise, fever, headache, and nausea, followed by acute or subacute onset of an encephalopathy whose symptoms include lethargy, confusion, and delirium. However, no pathognomonic clinical findings reliably distinguish HSE from other neurologic disorders with similar presentations.

## 2. Case Report

A 36-year-old female presented with a 6-day H/O fever associated with headache and neck pain, followed by an episode of generalized tonic clonic seizures. She stated that she woke up with blurred vision that developed into loss of vision in both eyes. She stated that she had no significant medical issues or any history of chronic medical conditions or surgeries. She had no recent travel, animal exposure, corticosteroid intake, chemotherapy, or radiation therapy. Physical examination showed a blood pressure of 150/70 mm hg, pulse rate of 75/min, respiratory rate of 20/min, temperature of 99.5°F. Neurological examination was normal. On ocular examination - direct and consensual pupillary light reflexes and fundus were normal. Her vision was CF 112 m in both eyes at presentation. There were no external ophthalmic lesions. Computed tomography (CT) showed hypodense area in frontal lobes bilaterally with perilesional edema. MRI brain has evidence of bilateral frontal white matter edema with mass effect on adjacent cortical sulci. MRV suggests cerebral venogram normal. EEG normal. Complete blood picture revealed normocytic hypochromic anemia. Lumbar tap showed a normal CSF. Intravenous acyclovir (1000 mg every 8 hours)

and intravenous ceftriaxone (2 g daily) were initiated empirically and antiepileptic eptoin 100 mg TID was started. Result of CSF herpes simplex virus (HSV) DNA by PCR was positive. Levels of vitamin B12, folate, and thyroid-stimulating hormone were all within normal limits

## 3. Discussion

HSE is the most common non-epidemic encephalitis and accounts for 5-10% of all cases of encephalitis(1). HSE is most common and severe in children and elderly people. One third of patients are aged under 20 years and half are over 50 years at presentation(2). HSV-1 encephalitis is more common in adults and HSV-2 infection is more common in neonates. Herpes simplex encephalitis (HSE) is an acute or subacute illness that causes both general and focal signs of cerebral dysfunction. Brain infection is thought to occur by means of direct neuronal transmission of the virus from a peripheral site to the brain via the trigeminal or olfactory nerve(3). The exact pathogenesis is unclear, and in fact patients with HSE may have a prodrome of malaise, fever, headache, and nausea, followed by acute or subacute onset of an encephalopathy whose symptoms include lethargy, confusion, and delirium. However, no pathognomonic clinical findings reliably distinguish HSE from other neurologic disorders with similar presentations(4)

The following are typically the most common symptoms of HSE:<sup>(5)</sup>

- Fever, Headache, Psychiatric symptoms, Seizures, Vomiting, Focal weakness, Memory loss.

Neurons are quickly overwhelmed by a lytic and hemorrhagic process distributed in an asymmetric fashion throughout the medial temporal and inferior frontal lobes. HSV-1 can affect immunocompetent individuals also unlike HSV-2 which affects immunocompromised individuals. The exact mechanism of cellular damage is unclear, but it may involve both direct virus-mediated and

indirect immune-mediated processes. The ability of HSV-1 to induce apoptosis (programmed cell death, or “cellular suicide”) in neuronal cells, a property not shared by HSV-2, might explain why the former causes virtually all cases of herpes simplex encephalitis in immunocompetent older children and adults.<sup>[6,1]</sup>

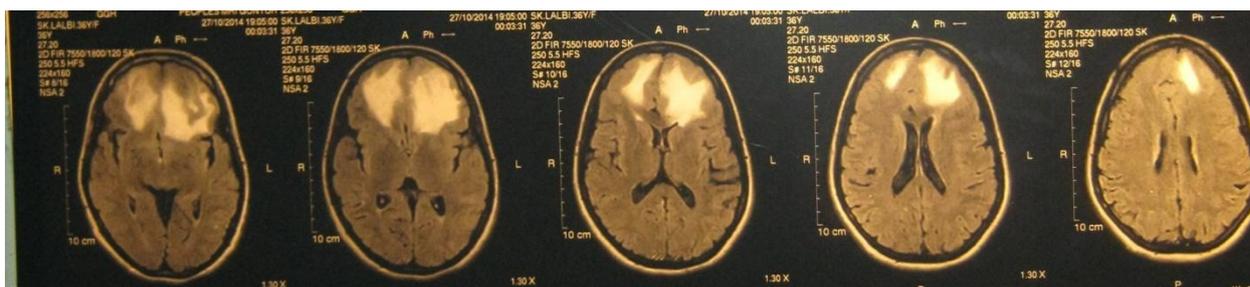
Routine laboratory tests are generally not helpful in the diagnosis of HSE but may show evidence of infection or a renal disease. The diagnosis can be confirmed only by means of PCR or brain biopsy<sup>[7]</sup>. Investigations that may be helpful in patients with suspected HSE include the following<sup>[8]</sup>

- Serologic analysis of blood or CSF: Retrospective diagnosis only; not for acute diagnosis and management
- Tzanck preparations of vesicular lesions: For confirmation of HSV in neonates with HSE
- Quantification of intrathecal antibodies: For evidence of CNS antibody response

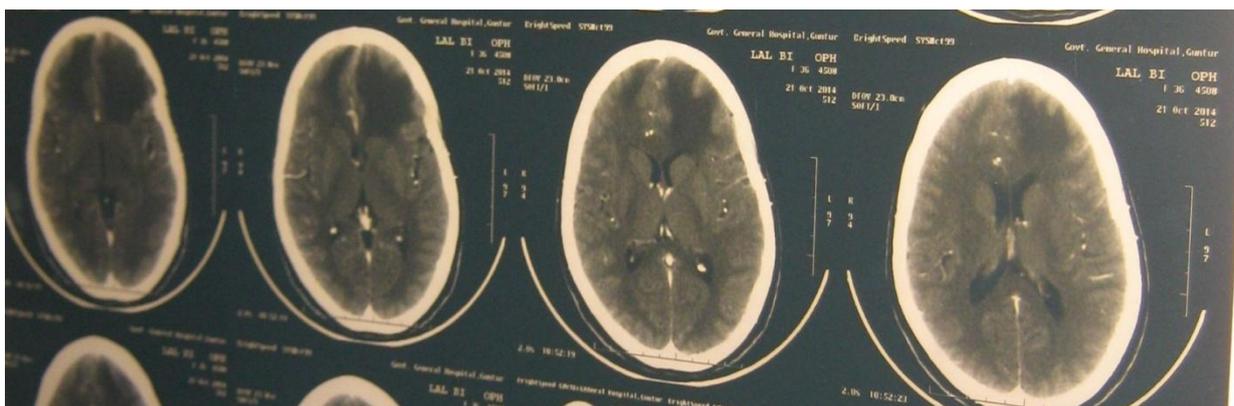
#### 4. Imaging Tests

The following are imaging studies used in the evaluation of suspected HSE:

- MRI of the brain: The preferred imaging study



**Figure 1:** MRI brain suggesting of bilateral frontal white matter oedema with maass effect on adjaacent cortical sulci



**Figure 2** CT scan suggesting bilateraal frontal lobe oedema with peri lesional oedema with mass effect

- CT scanning of the brain: Less sensitive than MRI
- EEG: Low specificity (32%) but 84% sensitivity to abnormal patterns in HSE

#### 5. Procedures

- Lumbar puncture for CSF analysis
- PCR assay of CSF for HSV-1 and HSV-2: Essentially replaced brain biopsy as the criterion standard for diagnosis<sup>[8]</sup>
- Brain biopsy: Diminishing role; rarely used in current practice for either confirming diagnosis of HSE or establishing alternative diagnosis.

Without treatment, HSE results in rapid death in approximately 70% of cases; survivors suffer severe neurological damage.<sup>[7]</sup> When treated, HSE is still fatal in one-third of cases, and causes serious long-term neurological damage in over half of survivors. Twenty percent of treated patients recover with minor damage. Only a small population of survivors (2.5%) regain completely normal brain function.<sup>[8]</sup> Earlier treatment (within 48 hours of symptom onset) improves the chances of a good recovery. Rarely, treated individuals can have relapse of infection weeks to months later.

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