

CASE REPORT

Atypical presentation of Herpes Simplex Encephalitis

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Abstract:

Encephalitis caused by herpes simplex is a common viral infection of nervous system. It typically affects temporal lobes. Involvement of sites other than temporal and frontal lobes is rare. We report a case of Herpes simplex encephalitis (HSE) effecting unusual areas of brain. A young male presented with fever, headache, confusion and hemiplegia. HSV-1 encephalitis was diagnosed by identifying viral DNA in cerebrospinal fluid (CSF) by polymerase chain reaction (PCR). MRI brain of the patient showed lesions in pons and cerebellum

Keywords: Encephalitis; Herpes encephalitis; viral encephalitis; Cerebellum; Brainstem syndrome.

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Introduction

Herpes simplex virus (HSV) is a DNA virus, which belongs to genus *simplexvirus* and has two sub types HSV-1 and HSV-2. Each subtype can affect Central nervous system (CNS), but HSV-1 more commonly so. Herpes simplex encephalitis (HSE) is the most common type of encephalitis constituting 10-20% of the total cases worldwide. Clinical features include fever, headache, and mental status changes ranging from irritability, drowsiness and confusion to coma, seizures and focal neurological deficits. It can be life threatening condition urging early diagnosis and treatment. Mortality is 5-15% despite treatment. Encephalitis is diagnosed by the combination of investigations, CSF analysis showing lymphocytic pleocytosis and brain imaging i.e. CT scan showing hypodensity

of the involved region, but MRI is more specific. Confirmation of HSE can be done by Brain biopsy or detection of HSV-1 DNA in CSF, which is more specific.

Sixteen years old boy was admitted to emergency department of Fauji foundation hospital during the month of October 2022, with complaints of fever for 3 days, altered sensorium and vomiting for 1 day. There was history of unsteady gait followed by decreased movement on right side of body. At presentation, patient had pulse of 76/min. BP 125/70mmHg, respiratory rate of 18/min and temperature was 99°F.

On examination patient was irritable with Glasgow Coma score (GCS) of 9/15, with decreased movement of the right half of the body (power 3/5). Neck was soft, Babin-ski's sign was positive on right side; pupils were bilaterally equal and reactive to light. Nystagmus was noted with fast component towards left. There was no rash. Rest of the systemic examination was normal. Initial laboratory workup on admission showed Total leukocyte count of $17.78 \times 10^3/\text{ml}$ with 86% neutrophils and 10% lymphocytes, Hb12.2 g/dl, platelets 228,000/ μl . Malarial parasite film and dengue serology were negative. Renal, liver function tests and serum electrolytes were normal. Generalized tonic-clonic fit was observed during hospital stay. Patient was admitted in Intensive care unit and empirical treatment for meningoencephalitis was started including anti-bacterial, steroids and acyclovir 30mg/kg/day as per standard protocol along with supportive treatment.

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CT scan brain was done to rule out any space occupying lesion, which showed mild brain oedema. Lumbar puncture(LP) was done, and CSF was sent for routine analysis, Gram stain, culture and viral PCR, results of which are as follows: clear watery appearance, TLC $1/\text{mm}^3$ lymphocytic, proteins: 30g/dl , glucose: 54mg/dl , Gram stain and bacterial culture was negative. Plasma glucose after LP was 118mg/dl . Viral PCR of CSF was positive for DNA of Herpes simplex virus 1. Patient's consciousness improved with treatment. There was hemiplegia with power of $1/5$ in right upper and lower limb. MRI brain was done which showed meningeal enhancement most prominent along left parietal lobe. Multiple enhancing, abnormal intensity lesions were seen in cerebellum and pons bilaterally, appearing hypo intense on T1WI and hyper intense on T2WI (figure 1) and FLAIR (figure 2). These lesions had diffusion restriction on DWI. After 14 days of treatment, patient improved but was still drowsy, with some residual behavioural changes including irritability and increased appetite; hemiplegia improved with power of $4/5$ in right upper and lower limb. In view of these findings, acyclovir treatment was further extended after discussion with neurologist. After 21 days of treatment patient recovered fully and was ambulant without any support. Patient was discharged and followed up as outpatient.

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Figure 1: MRI brain T2 WI, Saggital sections showing hyperintense lesion in pons (a) and cerebellum (b), (c) coronal section showing hyperintense lesion in left cerebellar hemisphere.

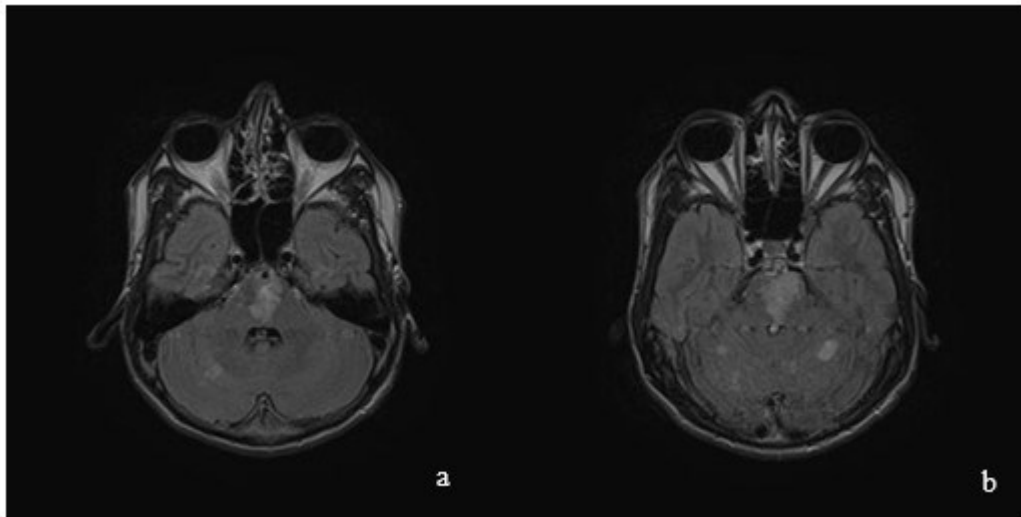
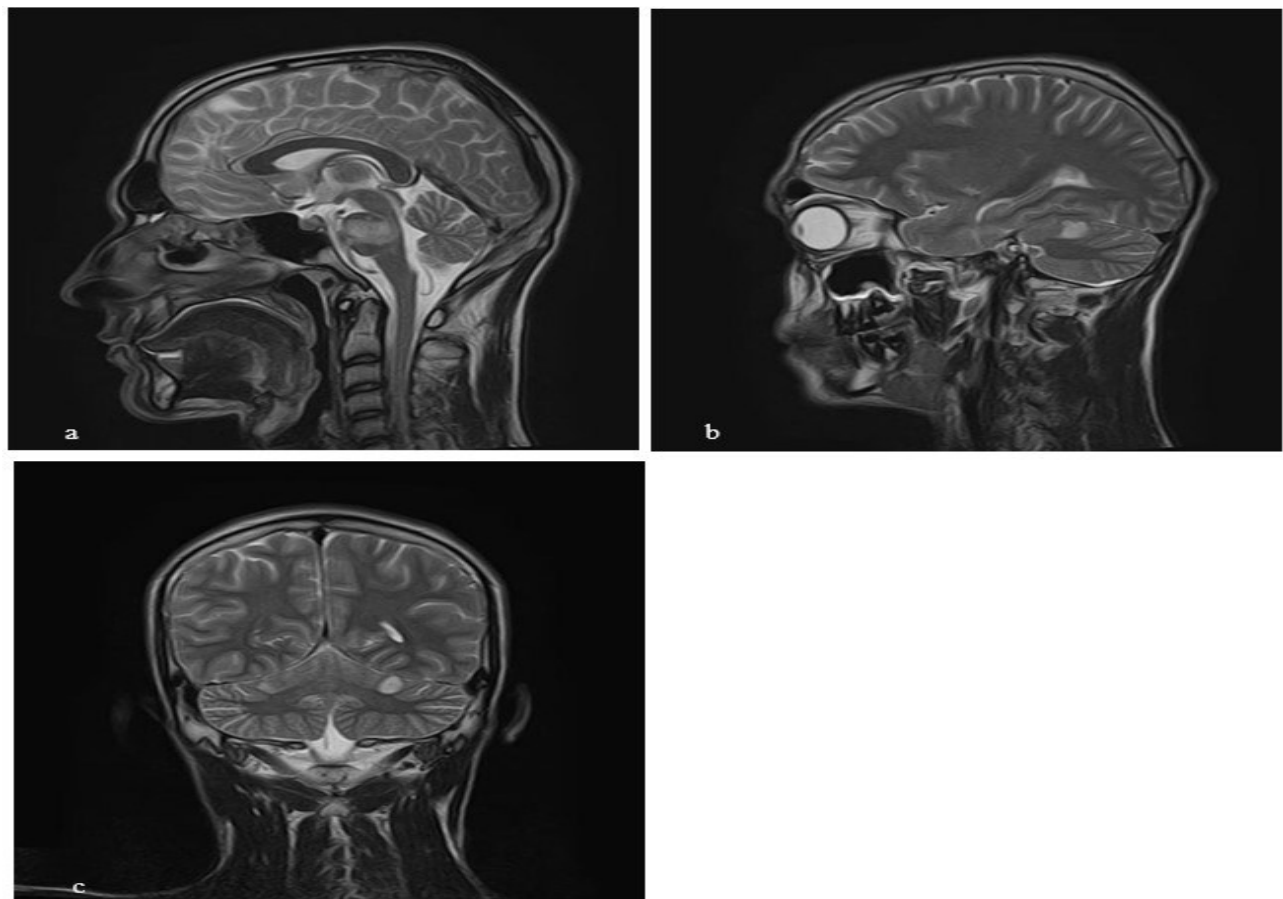


Figure 2: MRI brain FLAIR images axial sections at the level of pons and cerebellum showing bilateral hyperintense lesions.



Discussion

Herpes simplex encephalitis is most common type of viral encephalitis. The virus has predilection for temporal lobes, which are involved in about 80% cases. Most frequent extra temporal site is frontal lobe. Involvement of brain outside temporal and frontal lobes is uncommon, brainstem involvement is albeit rare. These areas are mostly affected in addition to temporal lobe(1). Isolated extra temporal involvement is uncommon(2). Livorci et al reported that out of patients having involvement of atypical brain sites, only 29% patients had isolated brainstem involvement; while brainstem involvement was a part of multifocal involvement in 71% (3). Despite being uncommon, brainstem involvement is significant, in that it increases the mortality and long term sequelae of the condition(3), emphasizing the need for quick diagnosis and timely treatment. On other hand, diagnosis of this condition is difficult due to the lack of specific signs and symptoms to differentiate from other causes of encephalitis. CSF PCR for HSV-1 is confirmatory test and MRI shows necrotic changes in affected areas, but these investigations may be normal in early phase of disease. Atypical presentations further complicate the picture. So, it requires high index of suspicion to improve disease prognosis, because atypical sites especially brainstem involvement further compromise the prognosis. In 1990 Matsumura et al reported first case of HSV-1 encephalitis affecting pons(4), another case was reported having typical and atypical MRI findings with involvement of cerebellum and pons in addition to temporal lobes(5). Combined involvement of both cerebellum and pons Without affecting temporal lobes was noted in very few cases(6). 2 cases of isolated acute Herpes simplex cerebellitis were found in literature (7). Our patient also had Herpes simplex encephalitis with cerebellar and pontine involvement in isolation of typical areas of brain. Our patient needed longer treatment as compared to that given in routine. But whether this atypical presentation affects treatment duration, is not clear and further cases are required to confirm this aspect. This presentation although rare, should be kept in mind because it has a significant impact on both short- and long-term prognosis of the patients.

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