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Hepatitis and Herpetic Encephalitis Complicated by Macrophage Activation Syndrome in an Immunocompetent Patient

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1. Abstract

Herpetic hepatitis and encephalitis are rare but severe forms of liver and brain infections caused by the herpes simplex virus (HSV). Often underdiagnosed due to their nonspecific symptoms and rarity, these infections can be life-threatening and lead to multiple complications. Although these conditions primarily affect immunocompromised individuals, rare cases have been reported in immunocompetent patients. Early recognition and treatment with antivirals like Acyclovir are crucial to improving the prognosis and preventing complications in patients with these conditions. We present a rare case of herpetic hepatitis and encephalitis in an 81-year-old immunocompetent patient experiencing a generalized primary infection with herpes simplex virus type I, complicated by macrophage activation syndrome. The patient's condition improved with Acyclovir treatment. The generalized herpes infection and its complications in this patient underscore the importance of increased clinical vigilance and early diagnosis, using techniques such as PCR to detect herpes viruses in cerebrospinal fluid and serum. Prompt administration of appropriate antiviral treatments like Acyclovir has proven notably effective, although prognosis largely depends on the timeliness of treatment initiation.

2. Introduction

Herpetic hepatitis is a rare but serious form of liver infection caused by the herpes simplex virus (HSV). Often underdiagnosed due to nonspecific symptoms and its rarity, this condition can cause hepatic cytolysis and be life-threatening. Although herpetic hepatitis mainly affects immunocompromised individuals or pregnant women in late gestation, rare cases have been reported in immunocompetent patients. Clinical signs include fever, abdominal pain, nausea, and jaundice, making diagnosis challenging without strong clinical suspicion and appropriate testing.

Herpetic encephalitis, on the other hand, is a rare but severe brain infection caused by HSV, most commonly associated with HSV-1. It presents with acute neurological symptoms such as headaches, seizures, confusion, and may rapidly progress to coma [1-7]. Neurological complications may include permanent cognitive deficits, memory impairment, and even death if treatment is not promptly initiated. Herpetic encephalitis should be strongly suspected in any patient presenting with headache, confusion, or febrile coma. Early recognition and treatment with antivirals like Acyclovir are essential to improve prognosis and prevent complications in patients with these conditions. Continuous monitoring and post-treatment follow-up are also necessary to manage potential long-term complications and assess the patient's neurological and hepatic recovery.

3. Case Report

An 81-year-old man was admitted to the emergency department with intense frontal headaches and morning vomiting for four days, along with gait and speech disturbances, right abdominal pain, and bloody diarrhea. His medical history included only childhood ap-

pendectomy, and he had no chronic treatments, substance abuse, or known allergies. A retired construction worker living alone, he was fully autonomous. Upon arrival, the patient was hemodynamically stable, normocardic, normotensive, and afebrile. Neurologically, he had a Glasgow score of 15, no sensory or motor deficits, a widened base of support, and a cautious gait. No meningeal stiffness was observed. Abdominal examination revealed a hypogastric mass without urinary symptoms. Laboratory tests showed acute renal failure with hyperkalemia, and urinalysis was unremarkable. Urinary ultrasound showed dilation of both renal pelvicalyceal systems, suggesting urinary retention. Brain CT showed no intracranial masses.

The combination of acute renal failure and urinary retention led to a diagnosis of acute urinary retention with associated headaches, nausea, vomiting, and diarrhea. To optimize care, the patient underwent bladder catheterization, rehydration, and close monitoring of urine output. After improvement in renal function and urine output, he was transferred to internal medicine for further care. On day 2 of hospitalization, a fever spike to 40°C prompted empiric treatment with Ceftriaxone and Amikacin. Urine culture and blood cultures revealed extended-spectrum beta-lactamase-producing Escherichia coli, leading to a switch to Piperacillin-Tazobactam. By day 10, liver enzyme abnormalities emerged. On day 12, a sudden worsening of liver function raised suspicion of immune-allergic hepatitis from Piperacillin-Tazobactam, prompting a switch to Aztreonam. A viral workup for hepatitis was ordered, and the patient was transferred to infectious disease care for specialized management. Within 24 hours of transfer, liver function deteriorated further, with transaminases reaching 100 times the normal level. Viral testing confirmed ambiguous HSV1 IgG with a positive HSV1 viremia at 20 CT, indicating possible primary HSV1 infection. Neurologically, meningeal stiffness was detected, and the patient showed obnubilation without focal signs, leading to lumbar puncture, which revealed a non-inflammatory cerebrospinal fluid with slight lymphocytosis and positive HSV1 detection at 38 CT. Given the widespread HSV1 infection, intravenous Acyclovir was initiated. Hematologically, pancytopenia appeared, accompanied by hyperferritinemia and hypertriglyceridemia, typical of macrophage activation syndrome. A bone marrow aspirate confirmed a few activated macrophages with rare hemophagocytosis. The final diagnosis was herpetic hepatitis and encephalitis complicated by macrophage activation syndrome in the context of generalized primary HSV1 infection.

4. Discussion

Herpes simplex virus 1 (HSV-1) is part of the Herpesviridae family, transmitted through direct skin or mucous membrane contact, with humans as its primary reservoir. HSV-1 has neurocutaneous tropism, with latency in sensory ganglia neurons, explaining periodic reactivations. Quantitative HSV PCR testing in blood is rapid, non-invasive, sensitive, and specific. Primary infection occurs in about 80% of children, with 90% of adults being seropositive. Adult primary infection in an immunocompetent patient is extremely rare, as in this case. Primary infection in adults is typically asymptomatic or mild, but in rare cases can manifest with more severe symptoms like hepatitis or herpetic encephalitis. Common manifestations include labial, oral, or genital herpes, herpetic keratitis, and neonatal herpes. Only 14 cases of severe HSV hepatitis have been reported in immunocompetent patients, with over 137 cases of HSV hepatitis identified overall. Clinical characteristics include fever (98%), coagulation disorders (84%), and encephalopathy (80%).

Antiviral treatment involves intravenous Acyclovir at a dose of 10 mg/kg every eight hours for at least ten days, while HSV viral load remains positive. Empiric Acyclovir administration is advised as soon as the diagnosis is suspected, with discontinuation if the diagnosis is ruled out [7]. Mortality and liver transplant need were 88% in untreated patients, significantly higher than the 51% rate in treated patients ($p < 0.001$). Our patient showed clinical improvement under treatment; HSV viremia and liver enzymes decreased rapidly with therapy such as shown in Figure 1. Unfortunately, most patients experience delayed diagnosis, increasing mortality, with 74% of cases resulting in death, including 51% of treated patients versus 88% of untreated cases. Most cases (58%) were diagnosed at autopsy. testing. In this case, lumbar puncture was performed late after discovery of herpetic hepatitis, likely due to initial encephalitis, followed by generalized infection and hepatitis. Without early treatment, herpetic encephalitis can lead to an 80% mortality rate. Macrophage activation syndrome (MAS) complications in this patient are typical of HSV infection, as previously described in the literature. This condition is related to inappropriate stimulation of macrophage cells in the bone marrow and lymphoid system due to the inflammatory response caused by the virus. The diagnosis is based on cytological or histological examination revealing hemophagocytosis.

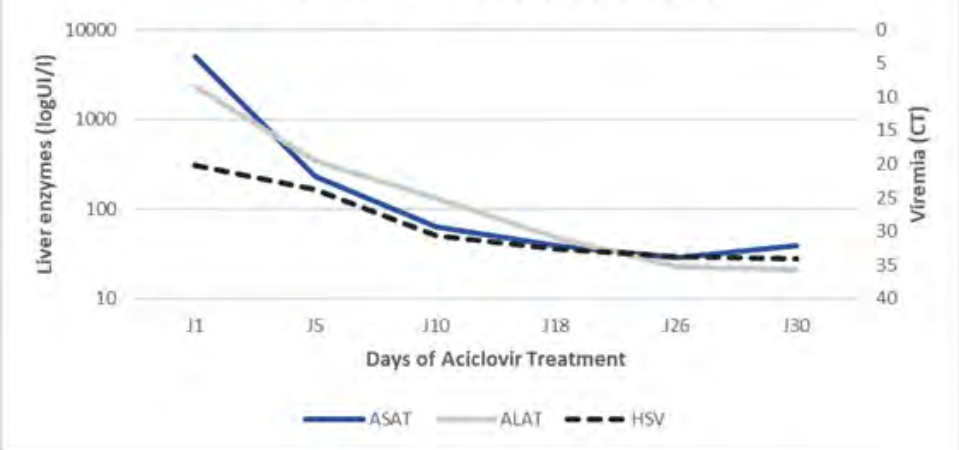

Figure 1: Evolution of the liver enzymes and viremia with the treatment

5. Conclusion

Herpes simplex virus 1 (HSV-1) infection presents a clinical challenge due to nonspecific symptoms, which can delay diagnosis and treatment, increasing morbidity and mortality. Severe manifestations such as herpetic encephalitis or hepatitis require heightened vigilance and prompt intervention with empiric antiviral therapy. The importance of early management, as shown in this case, highlights the need to test for this virus through sensitive methods like PCR in serum or cerebrospinal fluid at the first signs, including fever, coagulation abnormalities, or headache, to prevent severe complications and fatal outcomes associated with this infection.

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