

## EBV-induced fulminant hepatic failure treated with liver transplantation

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### ABSTRACT

Viral hepatitis is the leading cause of fulminant hepatitis. Infectious mononucleosis caused by primary infection of EBV is a self-limiting lymphoproliferative disease, and shows concomitant clinical features such as pyrexia, cervical lymphadenopathy, liver dysfunction and hepatosplenomegaly. Even though approximately more than 90 percent of all humans are infected with EBV it rarely causes hepatitis and even if it does it is usually benign and it rarely causes hepatic failure in which the outcome has a high mortality rate. We report a case of fulminant hepatic failure in an immunocompetent 3.5 years old girl caused by primary EBV infection that was treated by orthotropic liver transplantation. This observation emphasizes that EBV must be known as a possible cause of fulminant hepatitis and that liver transplantation is probably the unique therapeutic option to avoid a usually fatal course.

**KEY WORDS:** Transplantation, EBV, Hepatitis.

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### INTRODUCTION

Viral hepatitis is the leading cause of fulminant hepatitis. EBV is a member of the herpes virus family.<sup>1</sup> Infectious mononucleosis caused by primary infection of EBV is a self-limiting lymphoproliferative disease, and shows concomitant clinical features such as pyrexia, cervical lymphadenopathy, liver dysfunction and hepatosplenomegaly.<sup>1</sup> In rare cases, EBV establishes a latent infection in B lymphocytes and

runs a chronic course and shows infectious mononucleosis like symptoms repeatedly.<sup>2,3</sup> This syndrome, named chronic active Epstein-Barr virus infection (CAEBV), carries a very poor prognosis, and most CAEBV patients die from haematological or non-haematological disorders within a few years.<sup>4,5</sup> EBV may cause Infectious mononucleosis, nasopharynx carcinoma, Burkitt lymphoma, Hodgkin disease, lymphoproliferative disease after transplantation and "oral hairy leukoplakia" disease in "human immunodeficiency virus (HIV)" serology positive patients.<sup>3,4</sup> Even though approximately more than 90 percent of all humans are infected with EBV it rarely causes hepatitis and even if it does it is usually benign and it rarely causes hepatic failure in which the outcome has a high mortality rate. We report a case of fulminant hepatic failure in an immunocompetent 3.5 years old girl caused by primary EBV infection that was treated by orthotropic liver transplantation.

### CASE REPORT

A 3.5 year old girl was admitted to our emergency room with vomiting, abdominal pain and jaundice.

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She was the second child of nonrelated parents and didn't have any history of known disease at the presentation. She had been evaluated for hepatitis in another clinic about 15 days before admission. The serology for hepatitis A was negative although SGPT, SGOT and Total bilirubin values were elevated. The patient was for evaluation. She had in her eyes and her skin color was yellowish.

On physical examination hepatomegaly (11 cm), splenomegaly (4 cm) was found. The other systems were found to be normal. On differential diagnosis direct-indirect coombs, brucella, Wright and gruber widal agglutination was performed and all was found negative. Wilson disease was thought on the differential diagnosis and eye examination suspected the beginning of Kaiser Flescher ring. Plasma copper and ceruloplasmin levels studied and found equivocal. ANA, ASMA, LKM, IgG, ESH and CRP were evaluated for autoimmune hepatitis and found negative. Only EBV-VCA IgM was found positive in the serology for TORCH.

On MRI T1 sequence hyperintensity was seen basal ganglia and found compatible with chronic liver disease after consultation with paediatric neurology. On physical examination hepatomegaly was 6 cm on 4th day, 3 cm on 6th day and nonpalpabl on 7th day. On the morning of the 8th day she developed an inclination to sleep, unresponsiveness to verbal stimulation but on the other hand she was responding to painful stimulation, babinsky (-).

Cefotaxime+Amikasin IV therapy was initiated after taking bloodculture and gentamycine was given for the purpose of oral decontamination. It's seen that AFP: 289,41 ng/ml, PT:75, INR:5,78 and Fresh Frozen Plasma was administered. Hepatic coma was thought and patient was transferred with a doctor and an ambulance to a intensive care unit. The patient underwent a living donor liver transplantation (LDLT) from her mother because of fulminant hepatitis. After the 4th day of transplantation jaundice disappeared. The levels of AST, ALT, total bilirubin, PT and INR decreased to normal levels.

## DISCUSSION

EBV infection rarely results in hepatic failure.<sup>4</sup> Hepatic failure secondary to EBV infection is generally seen in childhood and life expectancy is four

months after the disease distinctively observed. Hepatic failure is the most important determinant factor for mortality. Lymphocytosis and Downey cells are seen in laboratory findings. Decrease in plasma levels of AST, ALT and Total Bilirubin, presence of dehydration are signs for hepatic involvement.<sup>3</sup> Autoimmune diseases such as systemic lupus erythematosus and Sjogren syndrome following EBV infection was also reported, and EBV infection appears to be a key factor in the initiation of autoimmune disorders at least in some cases.<sup>1</sup>

Mortality prevalence is extremely high in hepatic failure.<sup>5</sup> Acute hepatic failure caused by primary Epstein-Barr virus (EBV) infection has been reported in the literature in 16 cases, with an overall mortality of 87%.<sup>6</sup> It's reported that treatment by orthotropic liver transplantation in fulminant hepatic failure caused by primary EBV infection is really successful. The most important factor which is affecting the success of the therapy is the timing of liver transplantation.<sup>7</sup> This observation emphasizes that EBV must be known as a possible cause of fulminant hepatitis and that liver transplantation is probably the unique therapeutic option to avoid the usual fatal course. High index at suspicion should be held for EBV infection in case of fulminant hepatitis and if there are no contraindications liver transplantation should be taken as soon as possible to prevent the expected fatal course.

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