Short reports

Liver failure and Epstein-Barr virus infection

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SUMMARY A 5 year old boy developed liver failure secondary to infection with Epstein-Barr virus. He was subsequently shown to have a partial C4 complement deficiency. The importance of considering Epstein-Barr virus as a cause of fulminant hepatic failure and the need to assess immune state in such an event is emphasised.

Case report

A 5 year old boy was referred because of increasing jaundice and confusion. He had been unwell for four days, initially he was just listless but he subsequently lost his appetite and started vomiting. On the morning of referral he was vomiting profusely and complained of abdominal pain, by the afternoon he was delirious and jaundiced. There was nothing of note in his medical or family history. He had had no contact with hepatitis but had been on a school trip to a farm one week earlier. He had received no blood transfusions nor had he taken any drugs recently. On arrival at hospital he was drowsy and responded only to painful stimuli (grade IV coma). He was restless and had bite marks on his arms (self-inflicted). His pupils were dilated but reacted to light; his fundi were normal. He had generalised hyper-reflexia and extensor plantar responses. He was deeply jaundiced with hepatic fetor, he was also dehydrated and had a blood pressure of 125/75 mm Hg and a temperature of 36.9°C. Neither his liver nor his spleen were palpable. He had no lymphadenopathy and his tonsils were normal.

Table Results of tests for serum antibodies to Epstein-Barr virus

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<thead>
<tr>
<th>At presentation</th>
<th>Time after presentation</th>
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<tbody>
<tr>
<td></td>
<td>2 Months</td>
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<tr>
<td>IgM to EB viral capsid antigen</td>
<td>+</td>
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<tr>
<td>IgG to EB viral capsid antigen</td>
<td>+</td>
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<tr>
<td>IgM to EB virus nuclear antigen</td>
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It has been suggested previously that failure to develop atypical mononuclear cells during Epstein-Barr virus infection may represent a defect in cell mediated immunity. There have been several reports of cases of fatal infectious mononucleosis where an immune defect has been identified, the most well known being those individuals with the X linked recessive lymphoproliferative syndrome (Duncan’s disease). A severe illness consequent to Epstein-Barr virus infection in an individual with a complement deficiency, however, has not previously been reported. Individuals who are deficient in C4 may have an impaired immune response to viral antigen that could account for the severity of the illness experienced by this child.

It is of concern that individuals with C4 deficiency have been reported to develop systemic lupus-like illness in later life but at present the child reported here is well with no apparent sequelae from the liver failure or Epstein-Barr virus infection.

It seems remarkable that he survived considering the severity and rapid progression of his illness. It is felt that only 15–25% of children will survive grade IV hepatic coma.

We report this case in order to indicate the importance of considering Epstein-Barr virus as a cause of hepatic failure and the need to assess immune state if this is the case.

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References


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