Hemophagocytic Lymphohistiocytosis Presenting as Subacute Meningoencephalitis

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To the Editor: A four-year-old boy was brought with 11 d history of high grade fever, headache, vomiting and irritability. The child was diagnosed as West syndrome at 6 mo of age and successfully treated with adrenocorticotrophic hormone therapy. Subsequently, he had few infrequent seizures which were controlled on valproate. On examination, mild pallor, neck stiffness and hepatomegaly were found. Investigations revealed hemoglobin 7.7 g/dl, platelets $105,000/\text{mm}^3$, total leukocyte count $7700/\text{mm}^3$, neutrophils of 68 %, albumin 2.1 g/dl and C-reactive protein 262 mg/L. Chest radiograph and contrast enhanced magnetic resonance imaging of brain were normal. Cerebrospinal fluid analysis showed 30 cells/ mm^3 (70 % neutrophils), sugar 51 mg/dl, and proteins 69 mg/dl. He was initially treated with ceftriaxone and acyclovir. He continued to have fever and developed pancytopenia, worsening transaminitis and splenomegaly. Investigations showed serum triglycerides 294 mg/dl (normal <150 mg/dl), fibrinogen level 1.5 g/L (normal range 1.5–4 g/L), ferritin 133.4 mcg/L (normal range 7–140 mcg/dl) and 3+ hemophagocytes in the bone marrow. The child was given methylprednisolone pulse therapy followed by oral steroids and he became afebrile after 3 d. On one year follow-up child is asymptomatic.

This case highlights the fact that hemophagocytic lymphohistiocytosis should be considered as diagnostic possibility in a child with subacute meningoencephalitis.

Although various neurologic symptoms are known, meningo-encephalitic presentation at onset of hemophagocytic lymphohistiocytosis is rare. In a series by Horne et al., 37 % had neurological symptoms such as seizures, cranial nerve palsy, meningismus, altered sensorium, tone abnormalities, and upto 52 % had abnormal cerebrospinal fluid [1]. Persistence of fever, worsening pancytopenia, increasing splenomegaly, worsening triglyceridemia and hypofibrinogenemia should point towards the possibility of hemophagocytic lymphohistiocytosis [2]. This diagnosis is important because of the therapeutic potential. The index case also had West syndrome, which is a unique age dependent epileptic encephalopathy characterised by clustered infantile spasms, hypsarrhythmia on electroencephalography. Both entities, being responsive to steroids treatment and having genetic susceptibility, generate hypothesis of possible common pathophysiological link; however this needs to be investigated.

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