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Case Report

Macrophagic Activation Syndrome Related to Measles in a Child

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Abstract

Measles is a highly contagious infection whose frequency has been clearly reduced thanks to vaccination. It can rarely lead to complications, which are represented mostly by pneumonia, diarrhea or otitis, less commonly, encephalopathy and hepatitis. Macrophagic activation syndrome is a severe systemic manifestation which can rarely been associated with measles. We present a case of haemophagocytic syndrome caused by measles in a 14-month boy.

Keywords: Haemophagocytic Syndrome; Measles; Child

Introduction

Haemophagocytic lympho histiocytosis (HLH) is a potentially fatal syndrome with a systemic inflammatory condition caused by immune dysregulation. It is a rare disease with an incidence of 1.2 cases per million individuals per year [1]. It can either be primary, which occurs in patients with underlying genetic causes, or secondary, which is associated with infections, autoimmune disorders or malignancy [2]. Viral infections are the most common infectious etiologies associated with HLH such as *Epstein Barr virus*, *Herpes Simplex Virus and Cytomegalovirus* [3] but few cases of HLH caused by measles were reported [4].

Measles, a highly contagious infection, should always be suspected in front of fever and maculopapular rash. It can rarely lead to complications, which are represented mostly by pneumonia, diarrhea or otitis. Less commonly, encephalopathy, hepatitis or HLH [5]. Here, we report a rare case of HLH caused by measles.

Case Presentation

A 14-month boy was hospitalized for a 4-day-history of fever, cough and dyspnea. On admission, his body temperature was 39°C, his pulse rate was 145 beats per minute and his respiratory rate was 70 breaths per minute. He had signs of respiratory distress with inspiratory crackles on pulmonary auscultation. Abdominal palpation showed hepatomegaly. Laboratory findings on admission revealed an elevated C-reactive protein (308 mg/L), anemia (9.6 g/L) and hyponatremia (129 mEq/L). Aspartate aminotransferase concentration was 110 IU/L, while alanine aminotransferase and renal function were normal. Chest X-ray showed an opacity in the right upper lobe evoking a pneumonia. The patient was treated with intravenous 3rd generation cephalosporin and symptomatic treatment. The evolution was marked by the appearance of a generalized maculopapular skin rash. The diagnosis of measles was suspected and the baby was transferred to measles unity. The patient remained highly febrile for the next 2 days and developed splenomegaly. He had an altered states of consciousness which required mechanic ventilation. Brain Computed Tomography and lumbar puncture results were normal. Further laboratory investigations showed an elevated ferritin level (630 ng/L), hypertriglyceridemia (3 mmol/L) and a high lactic dehydrogenase at 460 U/L. Bone marrow aspiration showed macrophages with haemophagocytic activity. Measles serology was performed which revealed positive IgM

antibodies. The diagnosis of HLH and an acute respiratory failure caused by measles was confirmed. The patient was treated with intravenous immunoglobulin at a dose of 2 µg/kg. The evolution was marked by the improvement of the clinical condition and the occurrence of apyrexia 48 hours after initiating the treatment. C-reactive protein decreased to 145 then 58 mg/L, ferritin level was 389 ng/L and liver function were normal, 5 days apart. The patient was discharged from the hospital after completely recovered from its acute respiratory distress.

Discussion

We reported a case of HLH and acute respiratory distress caused by measles.

Measles, a highly contagious infection, should always be suspected in front of fever and maculopapular rash especially in case of epidemic. Actually, measles, a vaccine preventable disease, continue to increase through these years. Measles cases rose by 300% in the first 3 months of 2019, compared to the same period in 2018 [6]. The diagnosis should always be suspected, due to the current epidemic measles state, in front of any febrile maculopapular rash, even among vaccinated children. Measles can rarely lead to complications which occur in 10 to 40% of the cases and are more severe among the very young children. Complications are represented mostly by pneumonia, diarrhoea or otitis; less commonly, encephalopathy, hepatitis or HLH [6]. Pneumonia, which can either be caused by the measles virus itself or by secondary viral or bacterial pathogen, accounts for 60% of measles associated death [7]. Our patient presented an acute respiratory distress, elevated inflammatory markers and an abnormal chest X-ray which evoked pneumonia. But he had also an altered states of consciousness, which might be explained by measles neurological complications. In fact, measles encephalitis was noted in approximately one in 1000 patients, typically on day 5 of the rash and was fatal in approximately 10% of the cases [7]. But the presence of hepatosplenomegaly, a very important biological inflammatory syndrome with minor signs such as hyponatraemia and hepatic cytolysis in a child whose clinical condition worsened made us think of HLH. In HLH, cerebral symptoms can vary from neck stiffness to coma. Pleocytosis and an increased protein concentration in cerebrospinal fluid are the most common laboratory findings [8].

Clinical findings represented by fever and hepatosplenomegaly, laboratory parameters such as hypertriglyceridemia and increased ferritin level associated with haemophagocytosis in bone marrow aspiration confirmed the diagnosis of HLH based on the HLH-2004 criteria [9]. In the acquired HLH, a driving factor, which is measles virus in our case, cause the uncontrolled macrophage activation [2]. Previous studies showed that interferon-γ, increased during measles, activates macrophages and causes hemophagocytosis, which explain the HLH caused by measles in the present case [5].

In acquired HLH, treatment of the underlying etiology should be started without delay as it influences the prognosis of the disease. The administration of vitamin A to all children with measles is recommended once daily for 2 days at a dose of 50000, 100000 and 200000 IU for children aged less than 6 months, between 6 and 11 months and ≥ 12 months, respectively [10]. Our patient didn't receive vitamin A due to its lacking in our country. As for HLH treatment, the basic principle is to combine immunosuppressive and cytotoxic therapy to target the hyper inflammatory state, which include includes dexamethasone, etoposide and cyclosporine [2].

The prognosis of HLH depends basically on the underlying disease, the prompt diagnosis and treatment. However, the mortality rate of children with acquired HLH still ranges between 8 and 22%.

Conclusion

The case we presented satisfied the HLH-2004 criteria for diagnosis of HLH. Clinical diagnosis of measles was confirmed by serology. HLH caused by measles is a very rare syndrome. The diagnosis should be considered in all cases of persistent fever with elevated ferritin level. Its myriad presentation and various laboratory finding might be associated with those of the underlying disease. Therefore, the diagnosis of HLH should always be bearded in mind.

Conflict of Interest

We have no conflict of interest.

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