Fatal hemophagocytic lymphohistiocytosis associated with influenza B virus in a toddler - a case report and review of the literature

Straka Kalliopi, Tsirogianni Chrysanthi, Zeki Anna, Vaggelakoudi Eleni, Michailoudi Evi,
Barbaressou Chariklia

Paediatric Intensive Care Unit, Aghia Sofia Children's Hospital, Athens, Greece

Children's Hospital
"Agia Sofia"

Background and aims

Hemophagocytic lymphohistiocytosis (HLH) is a hyperinflammatory syndrome caused by uncontrolled proliferation and activation of macrophages and lymphocytes.

Virus-associated hemophagocytic syndrome (VAHS) is a severe complication of viral infections often life-threatening.

Methods

We present the case of a 18-month old toddler with VAHS triggered by Influenza B virus with prolonged fever who was admitted to PICU due to seizures.

Results

A female patient with history of failure to thrive presented with 3 weeks of fever, diarrhea and rash with increased inflammation markers that didn't respond to multiple antibiotic courses. PCR from NPA sample was positive for Influenza B virus so oseltamivir was added to the treatment without clinical improvement. Meanwhile, a marked increase in ferritin, triglycerides, liver enzymes was found with concurrent anaemia, thrombocytopenia, leucopenia, coagulopathy, multiple effusions and the toddler was transferred to our hospital for continuation of treatment with suspected HLH following influenza B infection. Bone marrow aspiration showed aplastic bone marrow. Shortly after her admission the clinical course was complicated by seizures and reduced level of consciousness so she was admitted to our PICU.

Initial brain CT was normal but a few hours later she developed rapid deterioration of her neurological status with absent brainstem reflexes with the new brain CT showing signs of cerebral edema and herniation. ICP monitoring and treatment for intracranial hypertension were promptly started. Dexamethasone and IVIG were given as suggested in the HLH-2004 protocol. Bone marrow biopsy had characteristics similar to HLH.

Despite full intensive care support, brainstem tests were positive for brain death and the patient expired on day 6 of hospitalization.

Conclusions

- Infection from Influenza virus can be complicated by HLH.
- Due to the non-specific manifestations of HLH, clinical suspicion and early diagnosis are of crucial importance for the outcome.

REFERENCES

1. Virus-associated hemophagocytic syndrome as a major contributor to death in patients with 2009 influenza A (H1N1) infection. Gernot Beutel¹ et al
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