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Hemophagocyticlymphohistiocytosis (HLH): A systematic review on approach to diagnosis and management of HLH as an emerging non-malignant disease with high morbidity and mortality in children and adults.

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Hemophagocyticlymphohistiocytosis (HLH) is a rapidly progressive, life-threatening syndrome of excessive immune activation. Prompt initiation of treatment for HLH is essential for the survival of affected patients. Although haematologist oncologists will treat these patients, but general paediatricians, immunologists, gastroenterologists, infectious men and even internists and adult oncologists should be familiar with diagnosis of this disease which has high morbidity and mortality; and its treatment before progression of neurologic complications is helpful. Presentation of HLH will be mentioned to be considered in the differential diagnosis of critically ill patients with prolonged fever, splenomegaly, cytopenia, jaundice, coagulopathy, etc. Method: English language large data bases including PubMed Central (Medline), EMBASE, Science Direct, ISI, ISC and Google Scholar were reviewed using these keywords: HemophagocyticLymphohistiocytosis, Diagnosis and/or treatment of HLH, genetic predisposition to HLH, Children or Adults with HLH. Fifty seven review articles were enrolled. Results: OVERVIEW AND INDICATIONS FOR TREATMENT: Hemophagocytic lymphohistiocytosis (HLH) is a dynamic disorder of unchecked insusceptible initiation and tissue harm. In the event that left untreated, patients with HLH make due for just a couple of months, because of dynamic multi-organ disappointment. Regularly, the best boundary to treatment and a fruitful result for people with HLH is a deferral in finding. A few parts of the clinical presentation of HLH add to this postponement, including the uncommonness of the disorder, the variable clinical presentation, and the absence of specificity of the clinical and research facility discoveries. Diagnostic criteria for HLH include molecular testing consistent with HLH or 5 of 8 of the following criteria: fever, splenomegaly, cytopenias affecting ≥2 lineages, hyperferritinemia, hypertriglyceridemia and/or hypofibrinogenemia, hemophagocytosis (in bone marrow, spleen, or lymph node), impaired NK cell function, and elevated soluble CD25 (sCD25) (ie, sIL2R). Additional discoveries that are regular are transaminases, coagulopathy, hypernatremia, edema, rash, hypoalbuminemia, hoisted lactate dehydrogenase (LDH), C-responsive protein, and d-dimer, expanded low-thickness lipoprotein, diminished high-thickness lipoprotein, lifted cerebrospinal liquid protein and cells, and neurologic indications running from central shortages to adjusted mental status. Any patient with suspected HLH ought to be seen by a haematologist, and the individuals who are intensely sick ought to be exchanged eminently to an office where they can get HLH treatment.

Biography

I was born in 30/12/1959 in North of Iran. I studied there until Diploma, and then in 1978 I was entered to Shiraz University of Medical Sciences. After 9 years training of Medicine in 1988 I was accepted as Paediatric Resident, then I had 2 years practice as Paediatrician. From April 1992 till July 1994 I was trained as PediatricHematologist –Oncologist and then I became a Scientific Member of Shiraz University of Medical Sciences. I have more than 30 publications in the field of Haemostasis, anemia and Paediatric Oncology. Now I'm Member of Board Certification of PediatricHematology - Oncology of Iran.

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