Single center experience of pediatric Hemophagocytic lymphohistiocytosis

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Introduction

Hemophagocytic lymphohistiocytosis (HLH) is a rare and often fatal syndrome of abnormal T-cell activation and cytokine production, which can be familial or secondary in nature

Aim

To describe the epidemiological features, outcomes and prognostic factors in diagnosis of pediatric HLH.

Methods

118 children fulfilling the inclusion criteria for HLH were identified from review of the Pediatric hospital records for period January, 2016 to December, 2024. They were revised regarding aetiology and age at time of diagnosis, viral serology, symptoms,neurological affection treament modality,morbidities and mortalities

Results

. Treatment modalities were steroid only (25%), anti-infectious agent (58%), multi-agent chemotherapy (43%) and HSCT (40%); mortality among above treatment groups were 25%, 58%, 43% and 40%, respectively. 15 patients (13%) had relapsed/refractory HLH who were treated with salvage chemotherapy and hematopoietic stem cell transplantation (HSCT). The overall mortality rate was 39%; mortality within 30 days seen in 23%. Estimated overall survival (OS) and event free survival (EFS) at 3 years were 62% and 61%, respectively.

Regarding the clinical prognosis, the severity of anaemia (p=0.001), neutropenia (p=0.007), thrombocytopenia (p=0.033), and hyperferritine mia (p

Results

Median age at diagnosis was 3 years (range13 days-15 years). Presenting features were fever (99%), hepatosplenomegaly (90%), neurological symptoms (25%), cytopenia (76%), elevated liver enzymes (67.3%), increased soluble interleukin-2 receptor) (sIL-2R) (78%) and hemophagocytosis on bone marrow (75%). Median follow-up duration was 14.5 months (3 days to 102 months). Primary HLH was identified in 27 (23%) patients. Etiology of secondary HLH was infections in 53 (45%), rheumatologic illnesses in 21 (18%),storage diseases (4%),infections (9%) and malignancies in 8 (6%) children.

Summary

Pediatric HLH is an aggressive disease with high mortality. Hyponatremia, hyperbilirubinemia, coagulopathy and increased sIL2 receptor level at diagnosis predicts

Discussion

Our study has assessed the clinical correlation of paediatric patients with haemophagocytic lymphohistiocytosis, most prevalent clinical characteristics included fever, lymphadenopathy and hepatosplenomegaly. Lab findings included cytopenias, elevated ferritin, deranged liver enzymes and raised LDH. A significantly poor long-term prognosis was identified in the sample cohort. Regarding the clinical prognosis, the severity of anaemia, neutropenia, thrombocytopenia, and hyperferritinemia was significantly associated with poor clinical progression in HLH. This is one of the preliminary studies exploring the clinical outcome of paediatric HLH