We have found HLH in cases like enteric fever (3), other blood culture proved sepsis(2), pneumonia(1), primary immunodeficiency related(1), pyrexia of unknown origin(1), sJIA(2), acute pancreatitis(1), Infectious mononucleosis(1), congenital cytomegalovirus (CMV) infection associated(1).

Fever and hepatosplenomegaly were present in all the cases & other findings like rash (5), respiratory distress (4), lymphadenopathy (3), jaundice (1), bleeding (2) & seizures (2) were also present.

**Laboratory features:**

Anaemia (four needed blood transfusion) & thrombocytopenia and hyperferritenaemia were present in all cases. Neutropenia present in 4 cases but others also showed gradual fall in total counts from leukocytosis towards normal range with clinical deterioration.

Other lab features like low fibrinogen level (10), high triglyceride (9), raised liver enzymes (12), hyponatremia (7) were also present. Fall in ESR present in 6 cases including the sJIA cases.

Bone marrow showed evidence of hemophagocytosis in seven cases.

**Treatment:**

All patients received supportive treatment including the treatment of cause.

Total 10 cases were infection related of which 4 were treated with IVIG and 3 were treated with steroid and rest 3 responded with treatment of the cause only.

One case was primary immunodeficiency (PID) related [leukocyte adhesion defect] which was treated with IVIG.

All of the sJIA (2) cases received steroid for macrophage activation syndrome and one was treated with cyclosporine also

**Outcome:**

Most of the cases (8) recovered with specific treatment (gamma/steroid) but three cases recovered with supportive treatment only. Unfortunately two children died - the case of immunodeficiency died of liver failure & the case of CMV infection died of profuse bleeding due to altered coagulation profile not responding to treatment with fresh frozen plasma and both of them died within one week of diagnosis.

**Conclusions**

- HLH not that uncommon in hospitalized children.
- A mind of suspicion for HLH -In a case of non remitting fever despite the appropriate treatment, Deterioration of clinical condition despite the appropriate treatment.
- Timely diagnosis & intervention can save lives.

**Young People’s Health Special Interest Group**

**CHALLENGES TO THE TRANSITION OF YOUNG PEOPLE WITH COMPLEX HEALTH CONDITIONS TO ADULT SERVICES**

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