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Title of the presentation: Haemophagocytic lymphohistiocytosis in a case of T cell lymphoma with gastrointestinal bleeding

Authors and Institute: Dr. Pareekshith R Rai, Dr. Shoban H, Dr. Santhosh Babu, Dr. Shyamkumar NK
Christian Medical College, Vellore.

Case report- Presentation

A 13-year-old boy who was a known case of extra-nodal NK/T- cell lymphoma of the orbit on Vincristine-Asparaginase and Prednisolone chemotherapy was admitted with febrile neutropenia. He suddenly developed multiple episodes of haematochezia with compensated shock (4 g% Hb drop). He was resuscitated, transfused blood and started on inotropes. Upper GI endoscopy showed an esophageal diverticulum. Colonoscopy showed altered blood with active ooze from the IC junction. CT abdominal angiography showed active contrast extravasation in the terminal ileum.

Case report : diagnosis

He was taken up for embolization under general anesthesia. SMA angiogram showed active extravasation from a branch of the ileocolic artery which was selectively cannulated and embolized with 10% glue. He was doing better post procedure and was being monitored in the ICU. Three days post procedure he again developed hematochezia and was noted to also have severe thrombocytopenia which required multiple transfusions. Ferritin level was found to be 2584 ng/ml. He was diagnosed to have Hemophagocytic Lymphohistiocytosis, likely secondary to NK/ T cell lymphoma

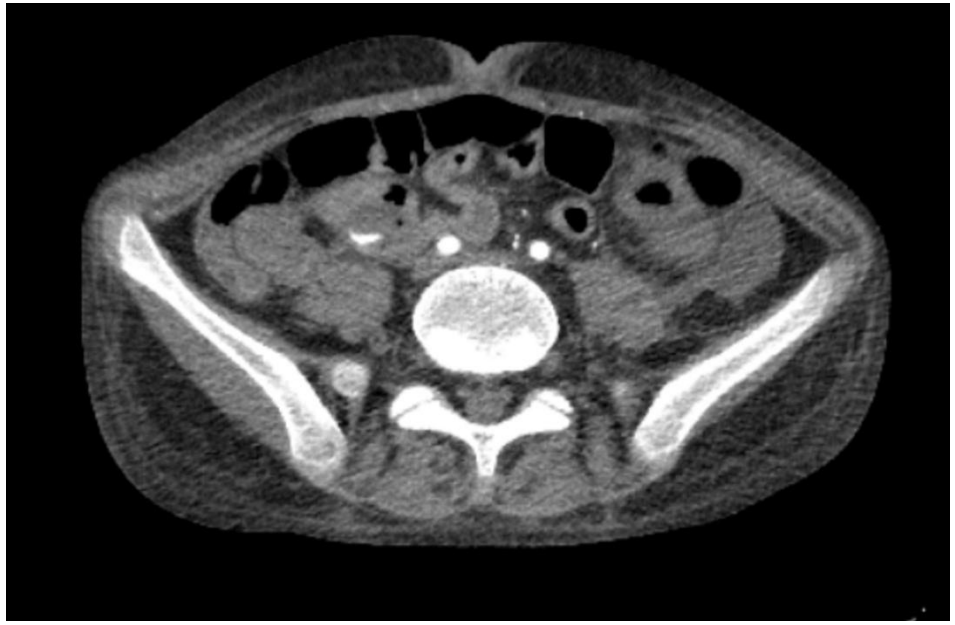
Case report- outcome

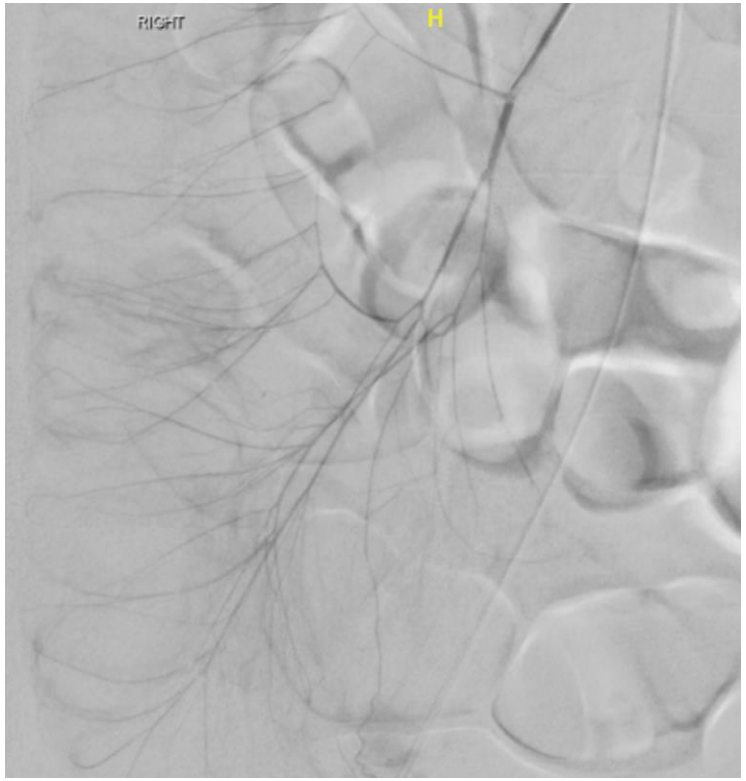
He was taken up for surgery this time but during intubation he developed haematemesis and scopy showed perforation of the esophageal diverticulum for which primary repair was done. Enteroscopy did not show any ooze and the terminal ileum was empirically resected. Post-surgery he had persistent haematochezia but was clinically stable. CT angiography did not show any abnormality. He was managed conservatively but had multiple episodes of hematochezia and recalcitrant thrombocytopenia. In view of the poor prognosis, his parents decided to pursue comfort care and take him home against medical advice. Unfortunately, he passed away on his way home.

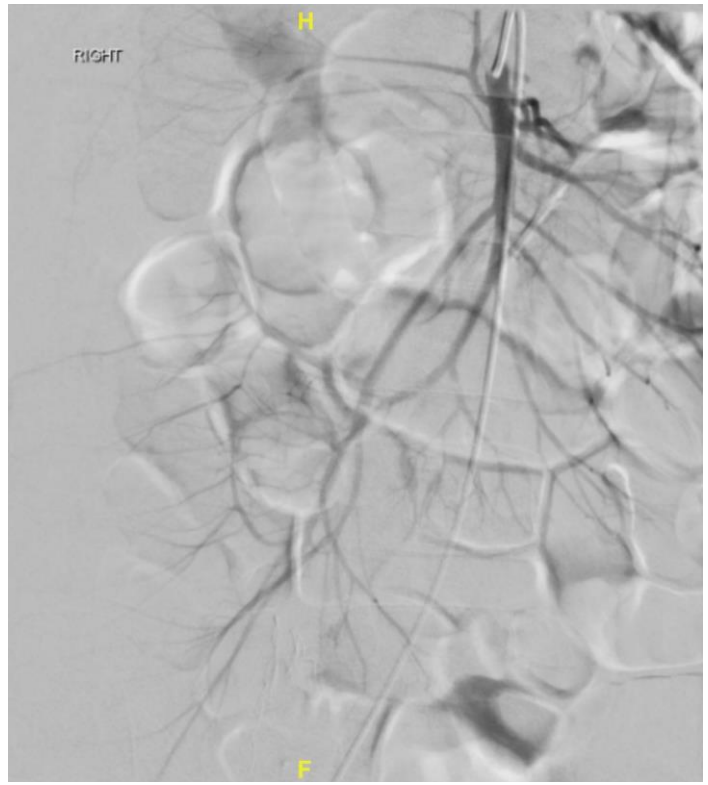
Review of Literature:

- HLH is a hyperinflammatory state characterised by cytokine deregulation, impaired function of cytotoxic T-cells (CTL), and natural killer (NK) cells in addition to hemophagocytosis. Characteristic features include unremitting fever, cytopenias, hepatosplenomegaly, and elevation of typical HLH biomarkers.
- Gastrointestinal bleeding can occur in approximately 10% of patients with HLH. When it does occur, it is known to cause bleeding from anywhere along the gastrointestinal tract due to transmural infiltration by lymphohistiocytes. Mortality rates in patients with gastrointestinal bleeding can be as high as 66%.

Representative images:







Conclusion:

- Hemophagocytic lymphohistiocytosis (HLH) is a hyper-inflammatory syndrome characterized by pathologic activation of cytotoxic T lymphocytes and macrophages. GI bleeding can rarely occur with HLH due to transmural infiltration by lymphohistiocytes with localized ulcers and diffuse mucosal infiltration.
- Angioembolisation and surgery have been only partially successful in treating it due to the high recurrence rate.
- This case report and other studies highlight the need for us to better understand this condition and develop effective treatments for it.

References:

1. Patel MM, Abrol RP, Gandhi R, Pavurala R, Aguirre J. S2272 A Rare Presentation of GI Bleeding Associated With Hemophagocytic Lymphohistiocytosis. Official journal of the American College of Gastroenterology| ACG. 2021 Oct 1;116:S969.
2. Knauft J, Schenk T, Ernst T, Schnetzke U, Hochhaus A, La Rosée P, Birndt S. Lymphoma-associated hemophagocytic lymphohistiocytosis (LA-HLH): a scoping review unveils clinical and diagnostic patterns of a lymphoma subgroup with poor prognosis. Leukemia. 2024 Feb;38(2):235-49.