Acute cholestatic liver failure in haemophagocytic lymphohistocytosis secondary to lymphoma in an elderly patient: A case report

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Introduction:

Haemophagocytic lymphohistocytosis (HLH) is a rare and serious haematological condition characterized by severe inflammation and tissue destruction secondary to abnormal immune system activation. It can be primary (hereditary) in infants/young children or secondary; often associated with infections, malignancy and auto immune disorders.

Case summary:

An 85-year-old patient was admitted to our hospital following a fall, generalized weakness for two weeks and persistent fever. Past history was only notable for hypertension. He had no past or current features of autoimmune disease. Examination revealed no organomegaly but fevers up to 39.9 deg C. Investigations showed a raised CRP, cholestatic LFTs, anaemia (Hb 118g/L) and normal procalcitonin. Further tests are shown in Table 1. CT chest, abdomen and pelvis showed small volume retrocrural lymphadenopathy. MRI liver and MRCP showed no significant abnormalities. Hepatitis A, B, C and E screens were negative. EBV and CMV IgG titers were positive, IgM negative.

Despite negative cultures he was given multiple courses of broad-spectrum antibiotics with no noticeable response. H score was 200 (80-88% HLH probability), so a trial of steroids was started. Bone marrow biopsy showed appearances consistent with marrow involvement by classic Hodgkin lymphoma (EBV-positive) and associated haemophagocytosis. Unfortunately, the patient developed COVID, his condition deteriorated and a decision to palliate was made.

Table 1

	On admission	Maximum
CRP (mg/L)	86	196
Ferritin (U/L)	4655	>33000
ALP	415	1398
Bilirubin (umol/L)	28	339 (conjugated 89)
AST (U/L)	58	63
Platelets (x 10^9/L)	70	24
Triglyceride (mmol/L)	2.2	3.6
LDH (U/L)	283	320

Conclusion:

- Our case demonstrates an unusual presentation of HLH with a predominant cholestatic picture of liver function derangement and no hepatobiliary cause identified on imaging. The diagnosis was reached by exclusion of infection and clinical suspicion from a rising ferritin and falling platelet count, although no single parameter is diagnostic of HLH.
- Another challenge met during diagnosis was the absence of organomegaly (splenomegaly). The initial ferritin was also only moderately raised, emphasizing the need to repeat relevant investigations.
- When diagnosis of HLH is suspected, especially in adults, search for underlying conditions, including infection, haematologic malignancies and rheumatologic conditions should be included in the work up to allow early treatment. However, the prognosis is poor in those with an underlying malignancy.
- HLH appears less common in the elderly. A 2019 review identified 71 cases of HLH in the literature in patients over the age of 50. There was a male predominance and infection appeared the most common precipitating cause. Leukemia and lymphoma were the precipitating cause in 24%. Overall mortality was 62%.
- This case demonstrates the need to have a high index of suspicion for HLH, particularly in the elderly, when comorbidities may cloud the clinical picture.

References

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