The use of deep skin biopsy for the identification of primary cutaneous lymphomas as a driver for secondary HLH

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HLH

Haemophagocytic lymphohistiocytosis

Cytokine storm

Primary or secondary

Unexplained sepsis-like presentation fevers >38.5oC

Rare causes of HLH include: Primary cutaneous lymphomas

Some, including intravascular large B-cell lymphoma (IVLBCL) present atypically

Tissue biopsy - skin

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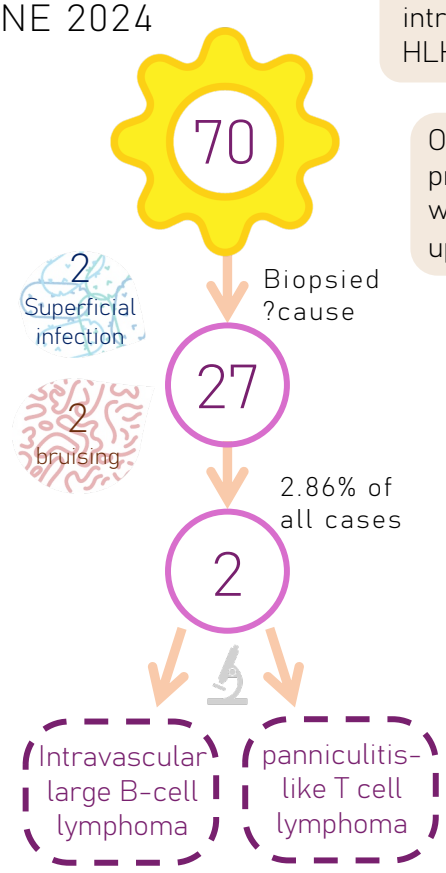
Following an index case of IVLBCL diagnosed at post-mortem, we introduced deep skin punch biopsy as routine work-up for patients with HLH of unknown driver.

Our protocol is to take a biopsy of any rash present along with three 6-8mm punch biopsies with depth to capture subcutaneous fat, from the upper arm, abdomen and thigh.



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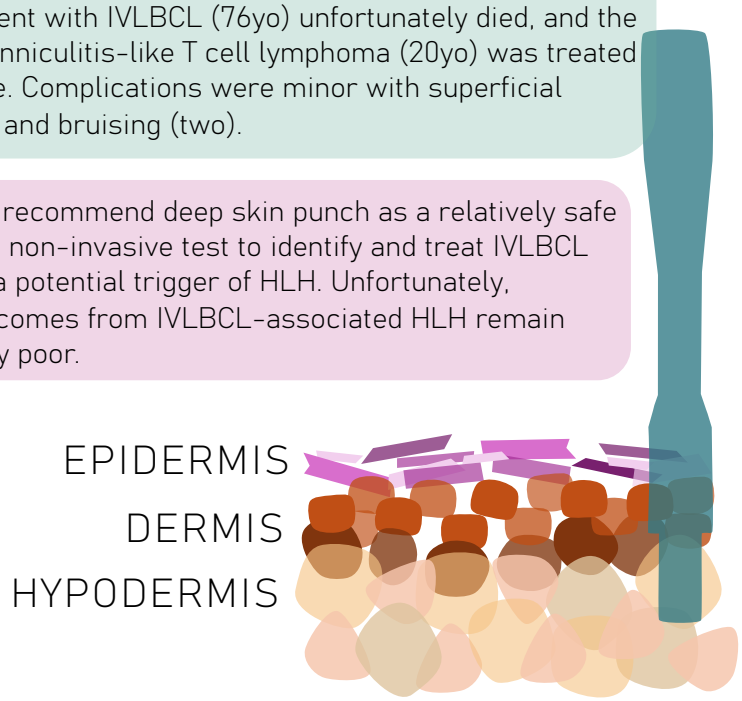
Electronic health records of all patients with confirmed HLH cared for under our team were reviewed. Data were collated on the use of deep skin punch biopsy, any associated complications, the diagnostic yield and patient outcome.

Of 70 confirmed cases of HLH in adults ≥18 years old, 27 with unknown triggers underwent deep skin punch biopsy. Two (2.86% of all) new cases of primary cutaneous lymphoma were made. The patient with IVLBCL (76yo) unfortunately died, and the patient with panniculitis-like T cell lymphoma (20yo) was treated and went home. Complications were minor with superficial infection (two) and bruising (two).

We recommend deep skin punch as a relatively safe and non-invasive test to identify and treat IVLBCL as a potential trigger of HLH. Unfortunately, outcomes from IVLBCL-associated HLH remain very poor.



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