PAEDIATRIC DERMATOLOGY

HEPATOSPLENIC T-CELL LYMPHOMA PRESENTING WITH SYMMETRICAL AXILLARY CAPILLARITIS

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Background: A 10 year old boy presented to the haematology outpatients with thrombocytopenia and fluctuating neutropenia associated with fevers and worsening of the rash. Weight loss and night sweats were not initially reported. He presented with a fluctuating eruption was noted over both his axillae and upper torso. There was no history of trauma or friction. On examination, he had symmetrical, macular petechiae with no associated pigmentary changes, atrophy or induration. He also had palpable splenomegaly.
A punch biopsy performed from the affected area was consistent with capillaritis demonstrated by superficial dermal red blood cell extravasation with mild perivascular lymphohistiocytic inflammatory infiltrate. There was no evidence of haemosiderin pigment deposition (Perls stain negative). There was no evidence of an atypical lymphoid infiltrate in this biopsy.
Bone marrow aspiration revealed abnormal mature T cell population consistent of both CD4 and CD8 with gamma delta positivity. Cytogenetics demonstrated abnormal clone detected with isochromosome 7q, consistent with the diagnosis of Hepatosplenic T-Cell Lymphoma (HSTL).

Observation: Hepatosplenic T-Cell Lymphoma (HTSL) is a rare, but aggressive peripheral T-cell lymphoma composing of neoplastic mature T cells characterised by CD2, CD3, CD7 and CD16 expression. HSTL often express gamma/delta T cell receptor and clonal rearrangement can often be demonstrated.
Only few case reports have described asymptomatic purpura to be a presenting feature. In one case, an atypical lymphocytic infiltrate was demonstrated in skin biopsies with positivity for gamma delta TCR marker. Although thrombocytopenic purpura affecting the face and neck has been described, to the extent of our knowledge, this is the first case of HSTL presenting with axillary capillaritis.

Key Message: Asymptomatic non-palpable purpura can be a rare presenting feature of hepatosplenic T-cell lymphoma. Dermatologists can play a pivotal role in the early recognition of this rare, but aggressive, haematological malignancy.