Extensive Cardiac Infiltration in Acute T-Cell Lymphoblastic Leukemia: Occult Extra-Medullary Relapse and Remission after Salvage Chemotherapy.

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Short Title Extensive cardiac infiltration in acute T-cell lymphoblastic leukemia

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A 38 year-old man was admitted with vasculitic rash and horizontal diplopia nine months post allogenic stem cell transplantation for T-cell acute lymphoblastic leukemia (ALL). Magnetic resonance (MR) of the orbits showed enlarged extra-ocular muscles and an echocardiogram, to exclude cardiac embolic sources, showed bi-ventricular hypertrophy and speckled myocardium. A 12-lead ECG showed diffuse T-wave inversion (Panel 1A). A cardiovascular MR (CMR) showed preserved bi-ventricular systolic function, septal left ventricular (LV) hypertrophy (max 21 mm, normal <12mm) (Panel 1B) with increased LV mass (88 g/m², normal 48-77 g/m²). Multiple areas of myocardial and pericardial infiltration (n=9, largest 18x50mm) were noted on advanced tissue characterisation, showing markedly increased T1 values on native T1 mapping, and non-enhancing after contrast administration (Panel 1C and D, white arrows). The CMR findings raised the suspicion of acute cardiac involvement of ALL. A MR-guided biopsy of the swollen ocular left rectus muscle confirmed the diagnosis of relapsed acute T-cell ALL (Panel 2A and B), further supported by evidence of 0.09% T-lymphoblasts on bone marrow flow cytometry and low level central nervous system (CNS) disease on lumbar puncture. As all the investigations confirmed acute multi-organ ALL relapse (heart, extra-ocular muscles, bone marrow and CNS), systemic chemotherapy with nelarabine and intra-thecal cytosine arabinoside were started. Diplopia rapidly improved, and a repeat orbit MR 14 days after commencement of chemotherapy showed complete resolution of extra-ocular muscle enlargement. A 1-month repeat CMR to assess cardiac response demonstrated complete resolution of all nodular lesions with normalization of LV wall thickness (10 mm) and mass (60 g/m²) (Panel 3 B, C and D), and of the widespread T-wave inversion on ECG (Panel 3A).

Declaration of Interest
None of the authors has disclosure to declare.

Role of the Funding Source
No funding source was involved.