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T-cell acute lymphoblastic leukemia with involvement of extramedullary sites, including pericardium

Short title: Pericardium involvement by T-ALL

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T-cell acute lymphoblastic leukemia (T-ALL) is a rare, aggressive hematological malignancy. The outcome of relapsing patients is poor and only a minority (<10%) achieve long-term survival [1–3]. Relapse patterns differ; however, extramedullary relapses are common, with the central nervous system being the most frequent site [2]. Here we present the case of a patient with extramedullary presentation of T-ALL both at diagnosis and at relapse, including within the pericardium.

A 36-year-old male was diagnosed with T-ALL with primary extramedullary involvement of the pleural cavities and possibly the pericardial sac. He received treatment according to PALG-ALL7 protocol, which resulted in complete remission, with negative minimal residual disease. Subsequently, the patient underwent allogeneic hematopoietic cell transplantation from a fully matched family donor after TBI/Eto/Cy conditioning. Six months later, the patient was readmitted for nonspecific left eye pain, left eyelid infiltration (**Figure 1A**) and cervical lymphadenopathy. His performance status was good, he denied other ailments.

Diagnostic procedures were performed. The bone marrow examination did not show infiltration with T-ALL. Cerebrospinal fluid examination led to exclusion of neuroinfection and T-ALL recurrence within the central nervous system. Positron emission tomography/computed tomography revealed metabolically active infiltrations within the eyelid, the lacrimal gland (**Figure 1B**) and cervical lymph nodes located on the left-side, as well as mediastinal lymph nodes, and the pericardium (**Figure 1C–D**). The pericardial infiltrate was associated with pericardial fluid up to 30 mm, surrounding the ascending aorta (up to the height of the arch) and pulmonary trunk. Echocardiography showed no signs of tamponade (**Figure 1E–F**). Histopathological sample of the salivary gland tumor was positive for T-ALL. Even before the start of causative treatment, the patient developed atrial fibrillation, and eventually cardiac tamponade. Symptomatic treatment was administered (propafenone, prednisone, colchicine) along with chemotherapy according to the FLAM protocol (fludarabine, cytarabine, mitoxantrone), which resulted in partial remission. Subsequently, the patient received nelarabine-based therapy as a “sandwich” strategy (first a combination with cyclophosphamide, and etoposide, afterward monotherapy due to prolonged and severe pancytopenia, and finally a combination) and eventually achieved complete remission, which was confirmed by positron emission tomography/computed tomography scan. The patient was qualified for salvage haploidentical hematopoietic cell transplantation from his father, after Flu/Treo/Thiotepa conditioning with post-transplant cyclophosphamide along with mycophenolate mofetil and tacrolimus. The early post-transplant period was complicated by cytokine release syndrome grade 3 and very severe sinusoidal obstruction syndrome/veno-occlusive disease. The patient required defibrotide for sinusoidal obstruction syndrome/veno-occlusive disease and renal replacement therapy for acute kidney injury. Additionally, signs of epidermolysis became apparent on physical examination. The patient died on +9 day after transplantation due to multiorgan failure.

To summarize, although T-ALL can involve extramedullary sites, localization in the pericardium is very rare, with only few cases reported so far, e.g. [4]. It should be noted, as

shown in the reported case, that T-ALL may involve atypical sites. Performance of a full differential diagnosis, accounting for post-transplant lymphoproliferative disease, infectious or bleeding complications, second primary malignancies [5], as well as relapse of the underlying disease being an indication for allogeneic hematopoietic cell transplantation is mandatory in such cases.

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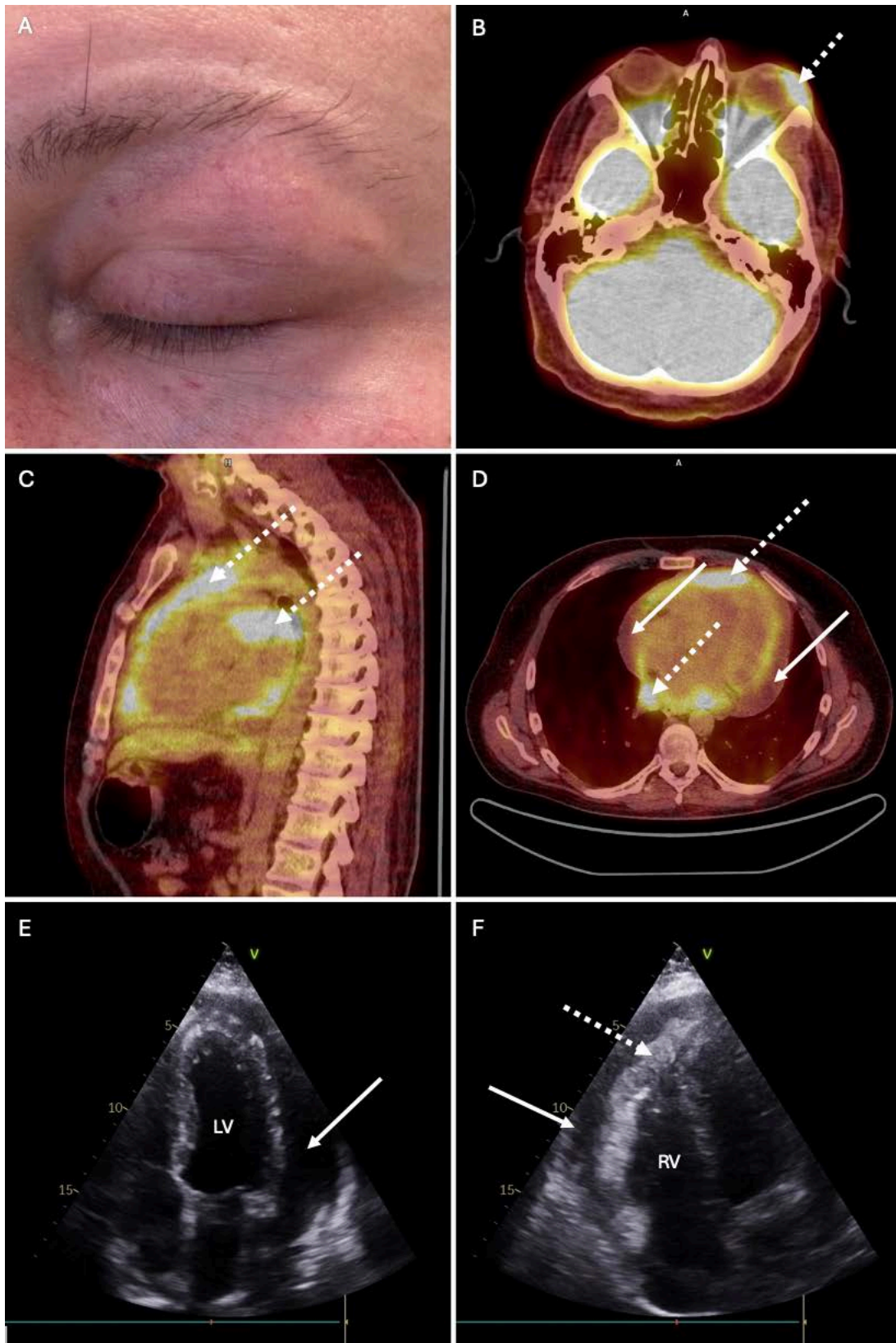


Figure 1. A. Left eyelid infiltration by T-ALL. B. PET/CT: active infiltrations within the eyelid and left lacrimal gland. C. D. PET/CT: active infiltrations within the pericardium associated with pericardial fluid. E. TTE: apical 4 chamber view. F. TTE: modified apical chamber view. Solid arrows point at pericardial fluid, the dotted arrows point at possible leukemic infiltrate

Abbreviations: CT, computed tomography; LV, left ventricle; PET, positron emission tomography; RV, right ventricle; T-ALL, T-cell acute lymphoblastic leukemia; TTE, transthoracic echocardiography