SKIN CANCER (OTHER THAN MELANOMA)

PRIMARY CUTANEOUS GAMMA-DELTA T-CELL LYMPHOMA WITH LONGLASTING REMISSION

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Observation: A 60-year-old woman, who had suffered from a primary cutaneous gamma-delta T-cell lymphoma (GD-TCL) in complete remission (CR) from 2010 after two lines of systemic treatment and allogeneic stem cell transplantation from unrelated donor (complicated by mild, self-limiting GVHD), complained the occurrence of new, rapidly growing erythematous patches and plaques on her lower limbs.
A skin biopsy was performed, with the aim of typing a possible relapse of GD-TCL vs subcutaneous GVHD. The diagnosis of GD-TCL was made. Due to the CD30 antigen expression by a significant % of tumour cells, an off-label treatment with brentuximab vedotin (BD, anti-CD30 immunotoxin) was started while considering a new allotransplantion procedure.
BD was administered (1.8 mg/kg i.v. q21) for 16 cycles. A clinical CR was obtained already after 6 cycles, with non-remarkable side effects.

Background: GD-TCL is a rare and aggressive entity (median survival 15 months), clinically featured in the skin by erythematous plaques and nodules, frequently undergoing ulceration. Histologically, it is characterized by a dermo-epidermal infiltrate of CD4+ (less frequently CD8+), gamma-delta TCR + T cells, with variably prominent angiocentricity and possible subcutaneous involvement. In this latter case, the differential diagnosis with subcutaneous panniculitis-like TCL is made on the basis of TCR gene rearrangement and immunohistochemistry with TCR probes.
The preferred therapeutically approach is polichemotherapy, possibly followed by allogeneic stem cell transplantation.

Key message: In most cases, GD-TCL is recalcitrant to (and rapidly relapsing after) radio- and chemotherapy.
The interest of the present case is related to the longlasting complete remission after allotransplantation and the very good and rapid response to treatment with brentuximab vedotin.