SKIN CANCER (OTHER THAN MELANOMA)

A RARE CASE OF SUBCUTANEOUS PANNICULITIS-LIKE-T-CELL LYMPHOMA WITH α/β T CELL PHENOTYPE WITH GREAT OUTCOME

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Background: Primary cutaneous lymphoma is rare in pediatric population, with estimated annual incidence of 0,1 in 106 habitants among children with age between 0 to 9 years. On this group, there are only few cases of subcutaneous panniculitis-like-T-cell lymphoma (SPTL) reported, with different outcomes.

Observation: A 2 year-old female patient presented with painful erythematous infiltrated plaques and nodules, some were ulcerated with necrotic crusts in the trunk, upper and lower limbs for the last 7 months. She also had bilateral periorbital oedema and erythematous infiltrated plaques on the face. Inguinal lymph nodes were enlarged and associated findings included daily fever, anemia and high level of lactate dehydrogenase. Serologies were negative for HIV, HTLV, EBV, syphilis and hepatites B and C. Skin biopsy revealed a lobular panniculitis with hystiocytes and lymphocytes, which were CD3+/CD8+/granzime+/high Ki-67 index (>90%)/CD56-/CD20-/CD4+ in rare cells/EBV-, suggesting SPTL CD8+. By flow cytometry of skin biopsy cells, we identified a CD3+/TCR α/β+/TCR γ/δ− T cell population, leading to diagnosis of SPTL with α/β T cell phenotype. She did not present hemophagocytic syndrome or extracutaneous involvement. She was treated with 4 cycles of cyclophosphamide, doxorubicin, vincristine and prednisolone with great response and is in remission for the last 9 months.

Key message: SPTL with α/β T cell phenotype is very rare with few cases reported in children. Its outcome varies from spontaneous remission to poor prognosis, specially when related to hemophagocytic syndrome. The differential diagnosis are SPTL with γ/δ T cell phenotype, histiocytic cytophagic panniculitis, hydroa vacciniform-like lymphoma and extranodal NK cell lymphoma. An accurate and early diagnosis is important to initiate treatment and improve patient's prognosis. Our patient presented great response with chemotherapy, but will need a longer follow-up to evaluate recurrence.