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CD4+/CD56+ Hematodermic Neoplasm Presenting in the Skin: A Tunisian Case Report and Current Review of the Literature

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Author(s)

Yosra Ben Youssef, Nessrine Ben Sayed, Kmira Zahra, Abir Gmidène, Naouel Ben Salah, Atef Ben Abdelkader, Nejia Brahem, Hlima Sennana, Colanda Belajouza, Abderrahim Khelif

ABSTRACT

The CD4+/CD56+ hematodermic neoplasm is a rare aggressive systemic neoplasm for which effective therapies have not yet been established, it is clinically characterized by cutaneous involvement with spread to bone marrow, blood and poor prognosis with current chemotherapy regimens. Our objective is to report diagnosis and treatment difficulties of CD4+/CD56+ hematodermic neoplasm. We describe here a Tunisian man who presented with subcutaneous ulcerated lesion localized in the right leg and multiples generalized nodules. Skin biopsy showed an atypical lymphoid cell infiltration with an angiocentric pattern and extensive necrosis by immuno-histochemical analysis, these cells were positive for CD4, CD56, granzyme B and negative for CD8, CD123, CD20 and CD30. T-cell rearrangement and Epstein-Barr-virus (EBV) in situ hybridation studies were negative. The patient underwent 5 cycles chemotherapy SMILE regimen monthly sandwiched with radiotherapy on the residual lesions of the right leg with great tolerance but he relapsed within 8months with skin, blood, bone marrow, lung, and cerebrospinal involvement. Based on these findings, the patient was diagnosed with CD4+/CD56+ hematodermic neoplasm (blastic NK-like T-cell lymphoma) treated with one course of hyper-CVAD regimen, he died within 20 days with a septic chok. Despite the use of L-Asparaginase and radiotherapy the prognosis is very poor; we suggest the exploration for highly active drugs, hematopoietic stem cell transplantation (HSCT) is crucial to improve survival.

KEYWORDS

Blastic NK-Like T-cell Lymphoma; Hyper-CVAD; SMILE; Prognosis; Treatment

Cite this paper

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