SKIN MANIFESTATIONS OF INTERNAL DISEASE

PRIMARY CUTANEOUS ANAPLASTIC LARGE CELL LYMPHOMA:A RARE ENTITY

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Background: CD30+ cutaneous lymphoproliferative disorders (CLPDs) include a spectrum of diseases: lymphomatoid papulosis (LyP), PC-ALCL and borderline conditions. Primary cutaneous anaplastic large cell lymphoma (PC-ALCL) is a non-Hodgkin T cell lymphoma that present in the skin, without systemic involvement at the time of diagnosis and in the next six months. Over a 30-year timespan, only 157 cases of primary localized CD30+ cutaneous lymphoproliferative disorders were documented in an analysis of the Surveillance, Epidemiology, and End Results (SEER) database. Patients with PC-ALCL classically present with solitary, grouped or multifocal nodules that persist for at least 3 to 4 weeks. Lymphomatoid papulosis is a chronic recurrent papulonodular skin eruption characterized by spontaneous resolution unlike PC-ALCL. Expression of anaplastic lymphoma kinase (ALK) is highly suggestive of systemic ALCL rather than PC-ALCL. The treatment varies according to the staging. Surgical excision or local radiotherapy are the most common therapies for solitary lesions. In multifocal disease, low-dose methotrexate represents an option.

Observation: A 65-year-old gentleman presented with a 2-month history of an asymptomatic proliferating ulcer on his left thigh that started as a small papule resembling an insect bite. Physical examination showed a well-demarcated, clean-based ulcer with rolled edges, measuring 5×7. Incisional biopsy showed a proliferation of large tumor cells infiltrating the deep dermis. An Immunohistochemistry study showed a positive reaction to CD30 and was negative for ALK. CT scan of the chest, abdomen and pelvis did not show any systemic localization. Large excision with a margin of 2 cm confirmed the initial diagnosis of PC-ALCL. Complete remission was achieved with no recurrence after 16 months of follow up.

Key message: Primary cutaneous anaplastic large cell lymphoma is a rare entity. The disease should be detected at early stage for better outcome treatments.