



DERMATOPATHOLOGY

MYELOID LEUKEMIA AND CUTANEOUS HISTIOCYTOSIS: FORTUITOUS ENCOUNTER?

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Background: Histiocytosis encompass highly heterogeneous proliferative disorders of the mononuclear phagocyte system. Recently they have been reclassified by the Histiocyte Society (2016) and reviewed in WHO classifications of hematopoietic (2017) and skin tumours (2018). These entities range from single-system and often self-limiting to multisystem and life-threatening forms. Recently the frequent association of Non-Langerhans Cell Histiocytosis with myeloid neoplasms has been highlighted.

Observation: We hereby describe three cases of histiocytosis who subsequently developed acute myeloid leukemia. All of them were male, with a mean age of 53 years old. Patient (pt.) 1 was diagnosed with Indeterminate cell histiocytosis, pt. 2 with diffuse Reticulohistiocytosis and pt. 3 with a mixed histiocytosis. Pt.1 and pt.2 present with generalized papulo-nodular skin eruption. Instead, pt.3 initially presents oral mucous membrane lesions and one year later displayed bone and skin involvement. Leukemic evolution occurred in pt.1 and pt.3 following chemotherapy while pt.2 developed acute myeloid leukemia after two years of observational follow up. In pt.1 and pt.2, both the conventional and molecular cytogenetics approaches detect clinically significant chromosome abnormalities. Both pt.1 and pt.3 died few months after the diagnosis of leukemia, whereas pt.2 is currently alive, in complete remission and good general conditions after chemotherapy and allogenic stem cell transplantation.

Key message: Cutaneous histiocytosis are bone-marrow derived disorders which may be considered to some extent as skin manifestation of dysmyelopoiesis (i.e. result of an altered genetic background). Therefore, in some cases, histiocytosis patients are at increased risk of developing myeloid neoplasms such as myeloid leukemia and myeloproliferative/myelodysplastic neoplasms. For such patients (especially adults with generalized skin eruption) is highly recommended a tight follow up and constant





surveillance regarding clinical manifestation of leukemia (e.g. recurrent infection, anemia, pancytopenia).

