Oral Communications

ADULT LANGERHANS CELL HISTIOCYTOSIS: A DERMATOLOGICAL DIAGNOSTIC CHALLENGE UNVEILING MULTISYSTEM DISEASE

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Introduction. Adult Langerhans cell histiocytosis (LCH) is a rare inflammatory myeloid neoplasm whose cutaneous manifestations frequently mimic common dermatoses, resulting in delayed diagnosis. We report a case in which persistent skin lesions led to the diagnosis of BRAF V600E-mutated multi system LCH associated with a clonal myeloid neoplasm.

Case report. A 67-year-old man with thrombocytopenia of undetermined etiology (2012) and scalp psoriasis (2018) refractory to topical therapy and acitretin developed confluent nodular lesions in the malar regions in June 2020. An initial biopsy was interpreted as impetiginized seborrheic keratosis. Following progressive worsening, a second biopsy (November 2020) showed acanthosis, hyperparakeratosis, and a dense dermal inflammatory infiltrate with necrosis. A presumptive diagnosis of rosacea was made, but the lesions failed to respond to antibiotic therapy. Progressive extension to the scalp and chest, together with ulcerative evolution of the facial lesions, prompted a third biopsy in June 2021, which established the diagnosis of LCH (CD1a+, S100+, BRAF V600E-mutated). Systemic prednisone (0.5 mg/kg/day), initiated while awaiting definitive histopathology, induced partial regression. Subsequent staging revealed multisystem involvement (bone, spleen, lung, ENT). Cladribine was initiated; bone marrow biopsy for persistent thrombocytopenia disclosed an associated myelodysplastic/myeloproliferative neoplasm. Cladribine induced cutaneous remission by December 2022. Cutaneous disease remained stable during follow-up; however, extracutaneous reactivation occurred, including an atypical intramuscular thigh lesion whose biopsy showed granulomatous inflammation with scattered Langerhans cells, together with FDG-PET/CT evidence of sinonasal, periorbital, and supraclavicular nodal involvement. After exclusion of an aggressive neoplasm, dabrafenib plus trametinib was planned for presumed disease recurrence.

Conclusions. This case highlights how persistent or atypical skin lesions may represent the first manifestation of multisystem disease and underscores the importance of repeated clinicopathological correlation when initial investigations are inconclusive. The main differential diagnoses are also discussed. The coexistence of a clonal myeloid neoplasm and the planned use of MAPK-targeted therapy further reflect the biological complexity of adult LCH.

References

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