INFLAMMATORY SKIN DISEASES (OTHER THAN ATOPIC DERMATITIS & PSORIASIS)

TWO CASES OF HISTIOCYTOID SWEET SYNDROME: A RARE VARIANT OF SWEET SYNDROME WITH DERMAL MYELOID CELL INFILTRATION

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Background: Histiocytoid Sweet syndrome (HSS) is a rare histopathologic variant of Sweet syndrome (SS). Although clinical characteristics of HSS are almost the same as SS, the dermal infiltrate of the lesions in HSS is composed of not neutrophils but mostly of immature cells of myeloid lineage. Here we report two cases of HSS.

Observation: Histopathological findings of both cases were almost identical and typical as HSS. They showed edema in the papillary dermis, and dense granulomatous infiltration composed of lymphocytes, granulocytes, and histiocytes in the dermis without vasculitis. The histiocytoid cells in the dermis were positive for myeloperoxidase staining and CD163.
Case1: A 55-year-old previously healthy male presented with erythematous lesions developed only on the sun-exposed area with fever and conjunctival hyperemia. He had many chances of sun exposure because he was a physical education teacher. He had no episode of photosensitivity nor recent severe sunburn. His symptoms had disappeared after oral steroid therapy but relapsed two years later. Although there have been several reports of SS in which skin lesions were suggested to be photoinduced, this is the first report of sun-exposure induction of HSS lesions.
Case2: An 81-year-old male who had a five-year history of multiple myeloma was started treatment with weekly ixozomib. After four times of ixozomib treatment, he developed edematous, erythematous lesions on the face, neck, and trunk. Discontinuation of ixozomib and oral steroid therapy cleared up the skin lesions. No recurrences of HSS have been seen in the six-month follow-up period. Although bortezomib, another anti-proteasome inhibitor used for multiple myeloma, is known to induce HSS, no reports have been done with ixozomib.

Key message: As the skin-infiltrating cells in HSS belong to the myeloid lineage, this rare non-neutrophilic variant should be acknowledged. Otherwise, HSS may be misdiagnosed as a skin invasion of hematopoietic malignancies.