CORRESPONDENCE

Comment: Clinical Pathology Correlation: Clinical First, then Pathology, and Lastly Immunochemistry

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I read with interest the case reported by Wakusawa et al. (1); however, I consider this case to be misdiagnosed, due to the common pitfall of lack of clinical-pathological correlation. Neither the clinical picture nor the pathological image is typical of necrobiosis lipoidica. The pigmented, indurated lesion, previous history of early-onset insulin-dependent diabetes, and microscopic evidence of histiocyte infiltration are more consistent with a diagnosis of pigmented hypertrichotic dermatosis with insulin-dependent diabetes (PHID) syndrome (OMIM 602782; http://www.omim.org/entry/602782). Alternative titles include POEMS syndrome in childhood, H syndrome, Faisalabad histiocytosis, and familial Rosai-Dorfman disease, now all described under the same name of "histiocytosis-lymphadenopathy plus syndrome" or "SLC29A3 spectrum disorder" because they all share the same SLC29A3 gene mutation.

Our department has encountered 3 such cases in the last 10 years.

Response to the Comment by Kan Wu

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Dr Wu suggests that our case of necrobiosis lipoidica could be histiocytosis-lymphadenopathy plus syndrome, such as pigmented hypertrichosis with insulin-dependent diabetes mellitus syndrome (PHID) or H syndrome. First of all, PHID is characterized by predominantly antibody-negative insulin-dependent diabetes mellitus associated with pigmented hypertrichosis and variable occurrence of other features of H syndrome. H syndrome is described as a disease that involves multi-systemic organs, including the skin (2). Concerning skin presentation, hyperpigmentation and hypertrichosis is described. Our presented case, though hyperpigmented areas might resemble H syndrome, did not present hypertrichosis. Concerning other organs, the patient did not have hepatosplenomegaly, healing loss, heart anomalies, hypogonadism (she had a child), short stature (her height, 170.4 cm, is a little taller than average for Japanese women), and hallux vulgus/flexion contracture. Only insulin-dependent diabetes mellitus (IDDM) (antibody-positive) is compatible for H syndrome.

Moreover, Dr Wu claims that our case is not typical necrobiosis lipoidica. In fact, our case presented palisading granuloma, composed of histiocytes and degeneration of collagen. Although the description of histological findings of H syndrome, "lymphoplasmocytic infiltrate in the dermis, atrophy of skin appendages and coarse collagen fibres in the subcutaneous fat", can be similar to the histological findings of necrobiosis lipoidica, in our presented case, there is no subcutaneous lesion. In addition, the granuloma-composing cells in our case contain not only CD68+ cells, but $CD163^+$ skin-resident macrophages. This phenotype of macrophages can be found in the typical necrobiosis lipoidica, but not in sarcoidosis. Although, the clinical cutaneous findings in our case may be similar to PHID or H syndrome, the patients' medical background is quite different from those with H syndrome.

REFERENCES (for both papers)

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