DERMATOPATHOLOGY

UNCOMMON VARIANTS OF NON-LANGERHANS CELL HISTIOCYTOSIS

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Background: Histiocytosis are rare disorders of the mononuclear phagocyte system, characterized by a derangement in differentiation, proliferation or function of monocytes and dendritic cells. They have been recently reclassified in five different groups based on a combination of clinical, histopathological and radiographic findings. Specifically, Non-Langerhans Cell Histiocytosis are among the rarest and most challenging histiocytoses to be diagnosed and managed as several entities display large clinical-pathological overlap. Moreover, this remarkable issue is still poorly investigated.

Observations: We describe four extraordinary cases of Cutaneous Non-Langerhans Cell Histiocytosis whose clinical-pathological findings did not fulfill the diagnostic criteria for any specific disorder. The first patient was a 32-year-old male, who initially presented with a face-limited papular eruption, akin to pediatric Benign Cephalic Histiocytosis and later manifesting a generalized skin involvement, as in Generalized Eruptive Histiocytosis. Moreover, histopathology could not discriminate between Xanthogranuloma and Reticulohistiocytosis. The second case was a 41-year-old woman with multiple comorbidities, who displayed a papular eruption limited to head and limbs associated with arthralgia of distal joints. Although the clinical picture was suggestive for Multicentric Reticulohistiocytosis, skin biopsy showed features intermediate between Rosai-Dorfman Disease and Adult Xanthogranuloma. The third subject was a 14-year-old woman who developed a sudden xanthogranulomatous papulo-nodular skin eruption 10 days after HPV-vaccination. Molecular analysis of skin biopsy tested positive for HPV-6 type. The fourth patient was a 47-year-old woman who presented disfiguring nodular facial lesions. Complessive clinical findings could not discriminate between Progressive Nodular Histiocytosis and Xanthoma Disseminatum.

Key message: The high heterogeneity of Non-Langerhans Cell Histiocytosis shows how

their differential diagnosis, prognostic stratification and therapeutic approach may be incredibly troublesome. Therefore, overlapping cases may benefit of a revision of nomenclature and classification, perhaps considering looser criteria (as already proposed for Langerhans-Cell Histiocytosis) to avoid misdiagnosis and allowing the development of standardized management.