DERMATOPATHOLOGY

LANGERHANS CELL HISTIOCYTOSIS AND MULTIPLE RETICULOHISTIOCYTOMAS IN A PATIENT WITH TAR SYNDROME: A NON-PREVIOUSLY DESCRIBED ASSOCIATION

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Background: Histiocytoses comprise a heterogeneous group of disorders characterized by proliferation of cells thought to be derived from monocytes, macrophages and dendritic cells in different organs. Langerhans cell histiocytosis (LCH) is a rare dendritic cell condition occurring mainly in pediatric population. LCH frequently presents as a multi-systemic disease, being the skin the second most common involved organ. LCH belongs to the L-group of histiocytosis and mutations in the Ras/Raf/MEK/ERK pathway have been proposed as the underlying pathogenic mechanism. On the other hand, reticulohistiocytoma (RH) is a benign dermal tumor and it belongs to the C-group of histiocytosis.

Observation: We present a young woman with thrombocytopenia and bilateral radius agenesis, condition known as TAR syndrome, multisystemic LCH and multiple RHs.

Key message: TAR syndrome is characterized by hypomegacariocytic thrombocytopenia, bilateral radial aplasia and could present heart, genitourinary and skeletal anomalies. Cutaneous lesions have been described. There are 5 cases of the association of TAR syndrome and histiocytosis in literature, all of them being LCH. Unfortunately the cutaneous involvement was mostly overlooked in all cases. In our case, two different histiocytosis were diagnosed in the context of TAR syndrome: a multi-systemic LCH with dermal involvement developed and multiple reticulohistiocytomas. Although overlaps between histiocytosis of the same group are well documented, the concomitant presence in the same patient of more than one type of histiocytosis from different groups is extremely rare, with approximately 31 cases reported in the English literature. To the best of our knowledge, there is no previous description of such infrequent association in the literature.