

17TH EUROPEAN DERMATOLOGY CONGRESS

March 01-03, 2018 | Paris, France

Langerhans cell histiocytosis with extensive cutaneous involvement

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Introduction: Langerhans cell histiocytosis (LCH) is a disease related to clonal proliferation of myeloid dendritic cells with morphological, immunophenotypic and ultrastructural similarities to Langerhans cells. Histiocyte Society (1987) reclassified LCH into a chronic disseminated, chronic multifocal, chronic focal and unrecognized disease.

Case report: Female, 6 years old, white, no pathological background or use of medicine, with several infiltrated papules, erythematous violaceous plaques and other residual hypochromic in trunk and limbs, with progressive increase of the disease for three years. The anatomopathological examination revealed infiltrate lymphoid of polyclonal T-cells (CD4+ and CD8+), non-granulomatous histiocytic and dendritic cells similar to LCH (S100+, CD1a+, CD68+). Systemic investigation was performed and despite the extent of cutaneous involvement, pure cutaneous LCH was confirmed, with partial response to therapy with Vinblastine and Prednisone.

Findings: LCH presents exaggerated proliferation of clonal dendritic cells with accumulation of different tissues, specially bones (80%), skin (33%) and histiocytosis (25%). It determines a variable clinical presentation according to the disease's stage, location and degree of involvement of organs and systems and can evolve from the pure cutaneous involvement to multisystemic disease (MS). The clinical evolution is unpredictable, going from spontaneous resolution to fast progression and death. From 30 to 40% of patients develop irreversible sequel from the disease or treatment, with alterations such as endocrinal, neurological, orthopedic, oral and tumoral. In general, cutaneous lesions are the first signs of LCH, manifesting as seborrheic dermatitis and refractory dermatitis: Diaper area, exudative suggestive of candidiasis or atopic.

Discussion: The final diagnosis is made by clinical manifestations with immunohistochemical and histological findings of inflammatory infiltrate composed of eosinophils, histiocytes, lymphocytes and immature myeloid dendritic cells in great numbers with similar gene expression to LCH cells (Birbeck granules CD1a+, CD207). Therapeutic options should be particularized by the extent and severity of the disease.

Biography

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