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SKIN MANIFESTATIONS OF INTERNAL DISEASE

XANTHOMA DISSEMINATUM AND CONVULSIVE SYNDROME: CASE REPORT

Maria Camila Diaz Posada (1) - Hernando Mosquera Sánchez (1)

Foscal, Universidad Autonoma De Bucaramanga, Bucaramanga (1)

Background: Xanthoma Disseminatum is a rare disease, with 100 cases reported until 1995, slight predominance in men, up to 60% of patients develop the disease before 25 years, characterized by a symmetrical eruption of multiple papules, plaques or yellow, red or brown nodules, favoring the face and areas of flexion. The classic triad of cutaneous xanthomas, mucosal xanthomas (40-60%) and diabetes insipidus (40%) has been described. It may be associated with a monoclonal gammopathy, and the involvement of the central nervous system.

Observation: 60-year-old female patient with a history of convulsive syndrome in carbamazepine and phenytoin management, presents 20 years ago yellowish nodular lesions on the face, which involve eyelids, nasolabial fold, with subsequent dissemination to axillary, antecubital and inguinal, symmetrical, bilateral folds, not pruritic, with progressive growth that hinder the ocular opening. No weight loss or other symptomatology. Lipid profile, glycemia and electrolytes in normal ranges. Protein electrophoresis without alteration.

Biopsy of upper eyelid lesion with diffuse dermal infiltration of abundant epithelioid cells, foamy histiocytes, lymphocytes and multinucleated giant cells Touton type, has immunohistochemistry CD68 (+), S100 (-) CD1a (-), it is a histiocytosis of Non-Langerhans cells, most likely Xanthoma Disseminatum.

Brain magnetic resonance with gadolinium with multiple nodular lesions with mass effect at the level of the bridge in the temporal lobes, neurosurgery considers secondary convulsive syndrome to lesions in the brain due to underlying disease

Key message: We present a case of Xanthoma Disseminatum, a histiocytosis of Non-Langerhans cells, by clinical, histological and immunohistochemical findings, associated with compromise of the central nervous system. We recall the association with diabetes insipidus and monoclonal gammopathy, not present in the current case. The importance of cutaneous manifestations as early signs of systemic manifestations is highlighted.





