PAEDIATRIC DERMATOLOGY

LANGERHANSIAN HISTIOCYTOSIS: ABOUT 5 CASES

Ait El Hadj Hasna [1] - *Farid Nafissa* [1] - *Akhdari Nadia* [1] - *Amal Said* [1] - *Hocar Ouafa* [1]

Arrazi Hospital, Mohamed Vi University Hospital, Dermatology, Marrakech, Morocco [1]

Background: Langerhansian histiocytosis (LH) is a rare condition characterized by infiltration of one or more organs by Langerhans-type dendritic cells. LH has a polymorphic clinical presentation. We report five cases of LH revealed by cutaneous lesions.

Observation: We report two female patients and three male patients aged between 5 months and 48 years. Clinical history of patients noted chronic diarrhea in one case, death in siblings at 8 months for rash in one case, diabetes insipidus and stato-kinetic cerebellar syndrome in another case, a polydrug-polydypsic syndrome in one patient, a patient followed for ulcerative colitis. The reason for consultation was a respiratory gene with bilateral tonsillar swelling in one case, a haemorrhagic and tumoral syndrome in one case, an objectified cutaneous involvement in all patients, central nervous system involvement in two patients, bone waiting in patient, pulmonary involvement in one patient and lymph node involvement in all patients. The histological diagnosis was obtained most often on a cutaneous biopsy. In 2 cases cerebral magnetic resonance imaging revealed post-pituitary involvement. The treatment of all our patients was based on chemotherapy with vinblastine and corticosteroid. The clinical outcome was favorable in 3 patients, two deaths occurred after the first perfusion of chemotherapy, due to deterioration of the general state and disease's progression.

Key message: LH is a rare disease. It is important that the care is done in a homogeneous and multicentric way.