PAEDIATRIC DERMATOLOGY

CASE REPORT: LANGERHANS CELL HISTIOCYTOSIS IN A 4-MONTH OLD MALE

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Background: Langerhans cell histiocytosis (LCH) is a rare histiocytic disorder which can be localized in the skin or affect various organs. It is commonly seen in children aged 1-14 years old with an incidence of 3-5 per million cases. Skin involvement is the second most commonly involved organ system and is found in 40% of patients.

Observation: We report a 4-month-old male who initially presented with erythematous plaques with yellowish-brown crusts found on the face and scalp which eventually became generalized. Residual hypopigmentation was noticed upon resolution of some lesions. Biopsy findings of diffuse dermal infiltrates of Langerhans cells is consistent with LCH. Immunohistochemical staining with CD1a, a specific marker for Langerhans cells, showed a positive result confirming the diagnosis of LCH. Whole abdominal ultrasound, bone marrow aspirate smear, creatinine, AST, bilirubin levels and albumin showed normal findings. This is a case of a single-system Langerhans histiocytosis. However, specialist was unsure of whether to attribute the finding of anemia as physiologic or as part of the LCH. Due to this, Vinblastine, first-line treatment, in combination with Prednisone was subsequently started and 50% improvement was seen after 2 weeks of treatment with subsequent complete clearance. Disease recurrence is monitored every 6 months after clearance of lesions.

Key message: Skin-limited Langerhans cell histiocytosis is rare. It accounts for 5% of cases of LCH and most commonly seen in newborns or infants. Specific markers such as CD1a or CD207 are needed to confirm the diagnosis. Topical steroids have no proven efficacy. Therefore, for widespread involvement, systemic steroid in combination with Vinblastine or low-dose Methotrexate can be given. Occurrence of Langerhans histiocytosis in children below 1 year of age signifies poor prognosis.

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