PAEDIATRIC DERMATOLOGY

HALO-LIKE PHENOMENON OF MONGOLIAN SPOT AROUND THE CAFÈ AU LAIT SPOTS IN NEUROFIBROMATOSIS TYPE 1

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Background: Neurofibromatosis type 1 (NF1) is an autosomal dominant genetic disorder, probably one of the most common neurocutaneous disease known by dermatologists. It is characterized by cutaneous manifestations that include café-au-lait spots, freckling on flexural areas, and cutaneous neurofibromas. The halo phenomenon in Mongolian spot surrounding the cafè au lait spot is a particular event, rarely described previously in literature. Histological finding evidenced dendrites of dermal melanocytes not clearly seen in the whitish halo-like zone and in the dermis of the cafè au lait spot, assuming that dendrites seemed to disappear at
these sites. Herein we present two children affected by NF1, confirmed by genetic examination, presenting Mongolian spot since birth.

Observation: Two children presented with a Mongolian spot localized on the back and chest area. The children gradually revealed the appearance of cafè au lait spots that particularly in the gluteal area were characterized by a halo-like phenomenon. The halo area presented the same color of the normal skin. What is the reason?

Key message: The etiology of this phenomenon is unknown. Two are the major hypothesis about the origin of this event: (i) inflammatory or (ii) autoimmune. The interesting fact is that the halo zone around the cafè au lait spots is not depigmented skin, but has the same color as the normal skin. The reason of this event is unknown, and further investigation is needed to explain it.