PAEDIATRIC DERMATOLOGY

SELF-HEALING JUVENILE XANTHOGRANULOMA 'EN PLAQUE' IN AN INFANT

P Pastrana-mabanta [1] - R Francisco [2] - J Jamora [3]

Skin And Cancer Foundation, Inc., Dermatology, Metro Manila, Philippines [1] - Philippine Children's Medical Center, Dermatology, Metro Manila, Philippines [2] - Quirino Memorial Medical Center, Dermatology, Metro Manila, Philippines [3]

Background: Juvenile xanthogranuloma (JXG) is a benign, self-limited non-Langerhans cell histiocytosis, commonly occurring in infants and young children. It is characterized by solitary or multiple yellowish papules or nodules, but may have unusual morphological presentations, such as agminated, lichenoid or plaque-like.

Observation: A six month old healthy Filipino male presented during the first week of life with light brown papules and nodules on the right posterolateral trunk. Lesions progressively enlarged, coalescing into a plaque, with associated occasional mild pain on movement. The patient was initially diagnosed by skin biopsy as a Spindle cell tumor at 4 months and as Langerhan's Cell Histiocytosis 2 months later, for which he underwent a course of chemotherapy that provided no significant relief to the lesion. A third biopsy was done, which showed sheets of histiocytes with round to oval nuclei and abundant slightly eosinophilic cytoplasm with skin reactivity to CD68. It was negative for the following stains: CD1a, LCA, ALK, CD30, Desmin, and Myogenin. He was then referred to Dermatology where physical examination revealed a 7.5 cm x 2 cm plaque of clustered 2-8 mm firm pink-brown to hyperpigmented nodules on the right lower posterolateral trunk. Third biopsy was reviewed and Touton cells were noted. Clinicohistopathologic correlation was done and the diagnosis was revised to Juvenile Xanthogranuloma 'en plaque'. No intervention was advised except an Ophthalmologic examination. Periodic follow-up visits with Dermatology and Ophthalmology revealed progressive resolution of tumor and normal eye findings.

Key Message: This case highlights the atypical presentation of a relatively common disorder of childhood. A conservative approach is recommended for JXG 'en plaque' as it tends to resolve spontaneously. Dermatologists need to be aware of the variable morphologic presentations of JXG in order to avoid unnecessary invasive diagnostic procedures and aggressive therapeutic interventions.