Dermatology Practical & Conceptual

Juvenile Xanthogranuloma in a Congenital Melanocytic Nevus

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Case Presentation

A 6-year-old boy was evaluated for a 6-month history of a 2x1mm yellowish papule that developed within a congenital melanocytic nevus (CMN) located on his back. Dermoscopic examination showed a homogeneous yellowish background with localized pigmented structures. In vivo reflectance confocal microscopy revealed an area with sharp borders, characterized by thinning of the epidermis and the absence of rete ridges. At the dermo-epidermal junction, the dermal papillae did not exhibit the typical ringed pattern. Within the dermis, there was infiltration by numerous large, round to ovoid cells with foamy cytoplasm, consistent with xanthomatous histiocytes; occasional Touton giant cells were also identified. Histopathological analysis confirmed the diagnosis of juvenile xanthogranuloma (JXG) arising within a CMN (Figure 1).

Teaching Point

JXG represents a prevalent non-Langerhans cell histiocytosis, predominantly affecting infants and young children [1,2]. The precise cause of JXG remains unclear; however, it is hypothesized to arise following an unidentified physical or infectious trigger that stimulates histiocytic proliferation [1]. To the best of our knowledge, there have been no previous reports describing JXG developing within a CMN.

Dermoscopy initially suggested xanthoma as the primary differential diagnosis in our patient. Nonetheless, considering the patient's age, lack of multiple lesions and hyperlipidemia, and histopathological findings of Touton giant cells and inflammatory infiltrate, the diagnosis favored JXG. Immunohistochemical analysis revealed CD68 positivity and absence of HMB45 and S100 staining in the foamy cells, effectively ruling out balloon cell nevus. Further studies are required to elucidate the precise etiopathogenesis underlying this collision phenomenon.

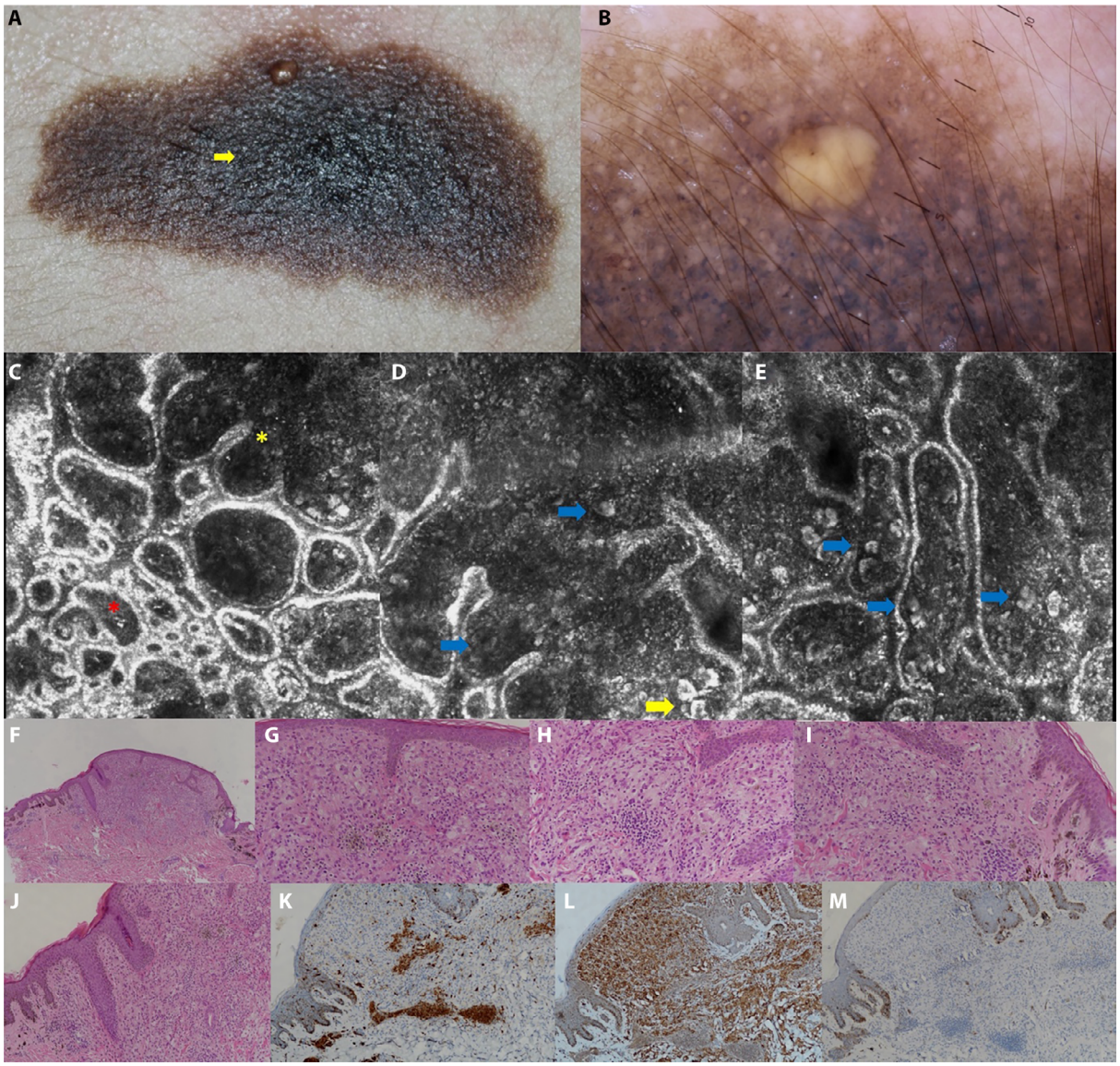

Figure 1. (A) Clinical image: yellowish papule in the congenital melanocytic nevus on the back (B) Dermoscopic image: yellowish homogenous background and focal pigmented structures (C-E) Two different components showing xanthomatous proliferation (yellow asterisk) and congenital nevus (red asterisk). Within the xanthomatous part, dermal infiltration of multiple large round and ovoid cells with a foamy cytoplasm corresponded to xanthomatous histiocytes (blue arrows), and occasional Touton giant cells (yellow arrow) (Vivascope 3000 Handheld; Mavig GmbH, Munich, Germany) (F-J) Well-demarcated dome-shaped dermal infiltrate consisting of foamy histiocytes without any pigment and atypia, occasional Touton giant cells, and scattered mixed inflammatory cells. Peripheral CMN components consisting of deep and periadnexial infiltration of melanocytes, and within the lesion focal pigmentation and deeply located clustered nevus cells (H&E x40, 100, 100, 100, 100 respectively). On immunohistochemistry, the foamy cells were CD1a (-), S100 (-) (K), CD68 (+) (L), and HMB45 (-) (M), whereas nevus cells were S100 (+) (K) and CD68 (-) (L) (x200).

References

1. Syed HA, Fillman EP. Juvenile Xanthogranuloma. In: *StatPearls*. Treasure Island (FL): StatPearls Publishing; February 15, 2026. PMID: 30252359.
2. Hernández-San Martín MJ, Vargas-Mora P, Aranibar L. Juvenile Xanthogranuloma: An Entity With a Wide Clinical Spectrum. Xantogranuloma juvenil: una entidad con amplio espectro clínico. *Actas Dermosifiliogr (Engl Ed)*. 2020;111(9):725–733. DOI:10.1016/j.ad.2020.07.004