Dermatology Practical & Conceptual

Pseudoxanthomatous Mastocytosis: A Rare Variant in a Hispanic Male

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Case Presentation

A 34-year-old Hispanic man presented to the dermatology department with multiple itchy yellowish nodules on his trunk and extremities (Figures 1A and B), which had appeared at six months of age. No abnormalities were found on general laboratory tests, and serum tryptase was requested. Dermoscopy revealed a pigment network and brown streaks on a yellowish pink background (Figure 1C). During the physical examination, rubbing the lesions induced only erythema, indicating a negative Darier sign. Histopathology showed a diffuse infiltration of mast cells in the dermis (Figure 1D), confirmed by Giemsa staining (Figure 1E) and CD117 immunohistochemistry (Figure 1F), which demonstrated a density of 70 mast cells per mm². The diagnosis of pseudoxanthomatous mastocytosis was established, and treatment was initiated with oral antihistamines.

Teaching Point

Pseudoxanthomatous mastocytosis (PXM) is an extremely rare variant of mastocytosis, first described in 1875 as "xanthelasmoid mastocytosis." It is a form of diffuse cutaneous mastocytosis characterized by multiple yellowish papules and flat nodules resembling xanthomas [1]. Notably, rubbing frequently triggers erythema without wheals (negative Darier sign) in this variant [1]. Histopathologically, PXM shows dermal mast cell infiltration, similar to other forms, but it is distinguished by a deeper infiltration, potentially contributing to the yellowish color of the lesions [2]. Clinical features and histopathological findings confirm the diagnosis. Treatment options include antihistamines, avoidance of triggers, topical or systemic steroids, phototherapy, and interferon [1].

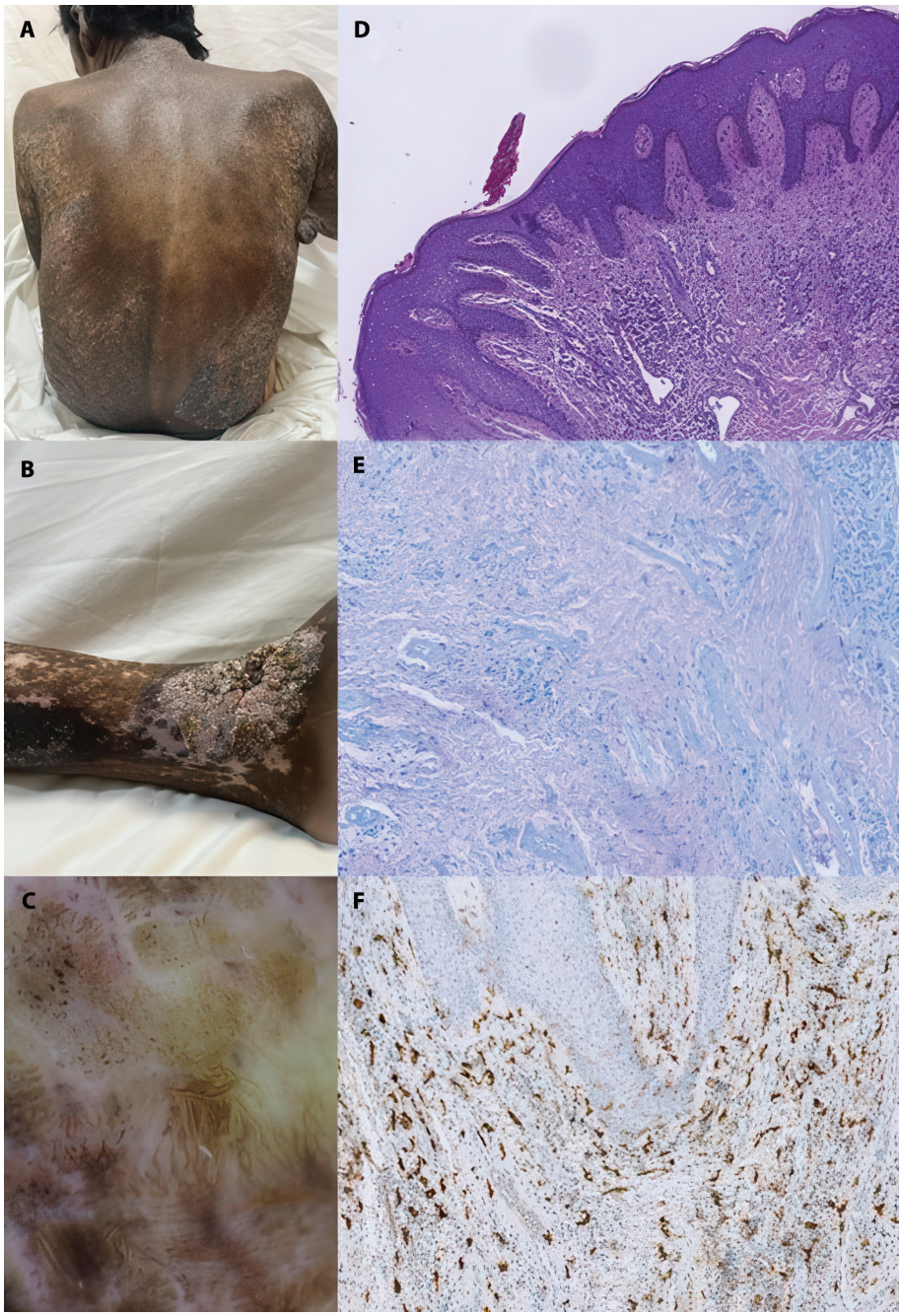

Figure 1. Male patient presenting with multiple yellowish and pink confluent nodules predominantly located on the (A) lower back and (B) extremities. (C) Dermoscopy of the nodules revealed a pigment network and brown streaks on a yellowish pink background. (D) Skin biopsy stained with H&E shows infiltration of mast cells in the dermis (120x magnification). (E) Giemsa stain demonstrating the metachromatic granules of mast cells (200x magnification). (F) Immunohistochemical staining revealed CD117 positive cells (200x magnification), with a mast cell count of 70 per mm².

This case is interesting because it represents a highly uncommon variant, with only a few cases documented in the literature. Moreover, while certain forms of cutaneous mastocytosis often resolve spontaneously, PXM typically persists into adulthood, as observed in our patient. No additional systemic involvement has been reported [2].

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