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Pagetoid Dyskeratosis and Pachydermodactyly-Mechanically induced Dermatoses ?

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Introduction: Pachydermodactyly is a rare benign form of digital fibromatosis, preferrably seen in adolescent males. The disease is characterized by a painless soft tissue swelling of the proximal interphalangeal (PIP) joints of the second to the fifth finger.
Pagetoid dyskeratosis is an incidental finding in a variety of lesions of the skin and squamous mucosa, presenting as a brownish maculae, typically found in areas of the skin, which are mechanically stressed such as intertriginous areas, buttocks and genitals. We herein report the first patient, who simultaneously presented these two rare diseases in the area of the hands.
Patient/methods: We report a case of a 14-year-old boy, who was sent to our department because of a brownish pigmentation of digit I-III and IV of the left hand and digit IV of the right hand, which were evolving four months. Additionally he presented a swelling of the lateral and dorsal regions of the metacarpophalangeal joints of digit III and IV of both hands since six months. He did not show any inflammatory signs or symptoms. The patient's history did not reveal any repetitive trauma, contact with chemicals and dyes or substance abuse.
Results: Laboratory studies showed no abnormalities, namely antinuclear antibodies, anti-doubled-stranded DNA, and rheumatic factor were found negative. X-ray exams only revealed augmentation of soft parts in the PIP.
Dermoscopic examination suggested that the lesions could be pigment lesions. A biopsy specimen of the fingertip showed no melanocytes, pigment- or hemosiderin deposits and no tumorous alterations. Pale pagetoid cells within the epidermis, some of which were dyskeratotic, led to the diagnosis of pagetoid dyskeratosis.
Discussion: Some authors postulate that pagetoid dyskeratosis is caused by trauma and friction. It is often present in areas which are subject to friction or moisture from occlusion. Pachydermodactyly is oft en associated with mechanical stress due to repetitive movements.
The observation of the two rare conditions in one patient, support the idea that both are induced by repetitive microtraumatic actions. To our knowledge we herein present a new case of pagetoid dyskeratosis of the hand and first in combination with pachydermodactyly.

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Increased expression of heat shock protein 90 in keratinocytes and mast cells in psoriasis

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Psoriasis is a chronic inflammatory skin disease and various stress factors mediate initiation and perpetuation of skin inflammation. Heat shock protein (HSP) 90 is a protein that acts as chaperone to protect cells from environmental stress signals and also has many roles in cell survival, cytokine signaling, such as IL-17 receptor signaling, as well as adaptive and innate immune responses. To elucidate the role of HSP90 in psoriasis, we assessed HSP90 expression and found that HSP90α, the inducible isoform of HSP90, was significantly upregulated in epidermal keratinocytes and mast cells of lesional psoriatic skin and downregulated following ustekinumab therapy. In vitro heat stress induced HSP90α mRNA expression and protein secretion in both human keratinocyte cell line HaCaT and human cord blood-derived mast cells (CB-MCs). HSP90α mRNA was upregulated by IL-6, IL-17, and IL-22 and HSP90α protein secretion was increased by TNF-α in HaCaT cells. HSP90α mRNA upregulat ion and protein production were induced by stem cell factor in CB-MCs. Finally, inhibition of HSP90 reduces proliferation in HaCaT cells and survival in CB-MCs. Our findings suggest that HSP90 from keratinocytes and mast cells is a key regulator in both the induction and maintenance of psoratic inflammation and provide a rationale for a novel therapeutic approach with HSP90 inhibitors.

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Expression of IL-17 in acute generalized exanthematous pustulosis and generalized pustular psoriasis

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Introduction & objectives: Acute generalized exanthematous pustulosis (AGEP) is a rare pustular disorder, mostly attributed to drugs, and characterized by sterile pustules, fever, and neutrophilia. CXCL8 producing drug-specific T lymphocytes are involved in the pathogenesis of AGEP. IL-17 is a pro-inflammatory cytokine and involved in various neutrophil-related autoimmune diseases. However, little is known about the role of IL-17 in AGEP. In this study we investigated the expression of IL-17 in AGEP in comparison to generalized pustular psoriasis (GPP) and normal skin.
Materials & Methods: Skin biopsy specimens were obtained from patients with AGEP (n=8), GPP (n=7) and from normal controls (n=8). IL-17 mRNA and protein expression were evaluated by in situ hybridization and immunohistochemistry. IL-17 positive cells were counted in the epidermis, upper dermis,