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Severe Generalized Pustular Psoriasis of von Zumbusch in an elderly patient: a case of remarkable response to IL-36 Inhibition

Iranzu Barandika Urrutia[1], Zita Álvarez Bobillo[1], Beatriz Clemente Hernández[1], Itziar Muelas Rives[1], Marcial Álvarez Salafranca[1], Ana María Morales Callaghan[1], Ignacio Hernández Aragües[1], Tamara Gracia Cazaña[1], Yolanda Gilaberte Calzada[1]

[1]Miguel Servet University Hospital, Zaragoza, Spain

Introduction & Objectives:

Generalized pustular psoriasis (GPP) is a rare, severe autoinflammatory skin disease characterized by widespread sterile pustules on erythematous skin, often with systemic involvement. The von Zumbusch subtype is the most acute and potentially life-threatening form. GPP may occur even in the absence of plaque psoriasis, posing diagnostic and therapeutic challenge. Dysregulation of the IL-36 signaling pathway plays a key pathogenic role in GPP flares.

Case Presentation:

An 81-year-old woman with no personal or family history of psoriasis presented to the emergency department with a 3-month history of pustular skin lesions. Initially treated with systemic corticosteroids (1.5 mg/kg/day), she experienced rapid worsening after abrupt discontinuation. She was admitted with systemic symptoms: fever, urinary tract infection, syncope, and atrial fibrillation decompensation. Examination revealed confluent pustules (2–3 mm) on erythematous plaques with peripheral scaling, affecting 38% of body surface area. GPPGA scores were 4 for pustules and 3 overall.

Blood tests showed leukocytosis (13.7 x10^3/µL) and elevated CRP (64.7 mg/L). Skin biopsy confirmed GPP (spongiform pustules of Kogoj, papillary edema, lymphocytic infiltrate). Due to the severity and comorbidities, a single intravenous dose of spesolimab was administered. The clinical response was rapid and significant: pustules resolved by day 3 (GPPGA pustules: 0), and only mild erythema remained at day 7 (GPPGA total: 1). No adverse effects were observed.

Conclusion:

This case underscores the importance of recognizing GPP in elderly patients without a history of psoriasis and highlights abrupt corticosteroid withdrawal as a key flare trigger. Spesolimab, a first-in-class IL-36 receptor antagonist, targets the key inflammatory pathway in the GPP pathogenesis. It provides rapid pustular clearance and systemic symptom relief, with a favorable safety profile and convenient single-dose administration. In this case, spesolimab enabled fast disease control and avoided escalation to broader immunosuppression. Its efficacy and tolerability support its role as a frontline therapy for acute GPP flares, especially in complex or high-risk patients.