


Posters

# FULMINANT SJS/TEN-LIKE PEMPHIGUS VULGARIS WITH HIGH ANTI-DSG1 TITERS AND CMV REACTIVATION: AN IMMUNOLOGICAL CHALLENGE

**R. Daher[2], A. Verdelli[1], L. Sanna[2], V. Corti[2], A. Magnatta[2], S. Landini[2], M. Donati[3], I. Bonanni[1], A. Corrà[4], V. Ruffo Di Calabria[5], E. Mariotti[6], C. Della Bella[7], L. Giovannoni[8], M. D'Elios[7], A. Moggi Pignone[9], M. Caproni[1]**

*(1)Rare Dermatological Diseases Unit, Department of Health Sciences, University of Florence, Florence, Italy; (2)Section of Dermatology, Department of Health Sciences, University of Florence, Florence, Italy; (3)Dermatology Department, University of Modena and Reggio Emilia, Modena, Italy; (4)-Dermatology Unit, Ospedale San Bortolo, Vicenza, Italy; (5)SSD Dermatologia, Cardinal Massaia Hospital, Asti, Italy; (6)Unit of Dermatology, Azienda USL Toscana Nord Ovest, Lucca, Italy; (7)Department of Molecular and Developmental Medicine, University of Siena, Italy; (8)UOC Ricerca e Sviluppo Clinical Practice, AOU Careggi, Florence, Italy; (9)Department of Experimental and Clinical Medicine, Division of Internal Medicine, University of Florence, Italy*

***Keywords:*** *Pemphigus-Vulgaris, Anti-Desmoglein-1.*

**Introduction.** Pemphigus vulgaris (PV) is an autoimmune blistering disease driven by IgG autoantibodies against desmoglein (Dsg) 1 and 3. Rare fulminant variants may mimic Stevens-Johnson syndrome/toxic epidermal necrolysis (SJS/TEN). We report an exceptionally severe SJS/TEN-like PV in a previously healthy woman, focusing on its immunological basis and management.

**Materials and Methods.** Case report of a 55-year-old woman with an unremarkable history. The work-up included histology, direct (DIF) and indirect immunofluorescence (IIF), anti-Dsg1/Dsg3 ELISA, a viral panel and malignancy screening; disease activity was scored by PDAI.

**Results.** The patient developed rapidly progressive mucocutaneous erosions evolving into extensive epidermal detachment (PDAI activity 25), without drug trigger or fever. Histology showed suprabasal acantholysis; DIF revealed intercellular IgG1 and C3; IIF was positive (1:80). ELISA demonstrated markedly elevated anti-Dsg1 (107.5 IU/mL) with only modest anti-Dsg3 (9.6 IU/mL). Anti-envoplakin was negative and malignancy screening unremarkable, while rat-bladder IIF was unavailable. CMV reactivation (IgM positive) was detected and treated with ganciclovir. The Dsg1-dominant, discordant profile together with complement-fixing IgG1/C3 deposition supports antibody- and complement-mediated acantholysis as drivers of the SJS/TEN-like phenotype. Combined rituximab and intravenous immunoglobulin achieved rapid re-epithelialization. At 12-month follow-up the patient was in complete clinical and serological remission (PDAI 0; anti-Dsg negative), with no malignancy.

**Conclusions.** This case expands the clinical spectrum of PV, showing that high anti-Dsg1 titers, complement activation and CMV-related immune dysregulation can converge to produce a life-threatening SJS/TEN-like phenotype. Early B-cell-directed therapy combined with immunoglobulin can be life-saving, and prolonged oncological surveillance remains warranted.

**References**

1. Schmidt E, Kasperkiewicz M, Joly P. Pemphigus. Lancet. 2019;394:882-894.2. Harman KE, Seed PT, Gratian MJ, Bhogal BS, Challacombe SJ, Black MM. The severity of cutaneous and oral pemphigus is related to desmoglein 1 and 3 antibody levels. Br J Dermatol. 2001;144:775-780.3. Ahmed AR, Spigelman Z, Cavacini LA, Posner MR. Treatment of pemphigus vulgaris with rituximab and intravenous immune globulin. N Engl J Med. 2006;355:1772-1779.